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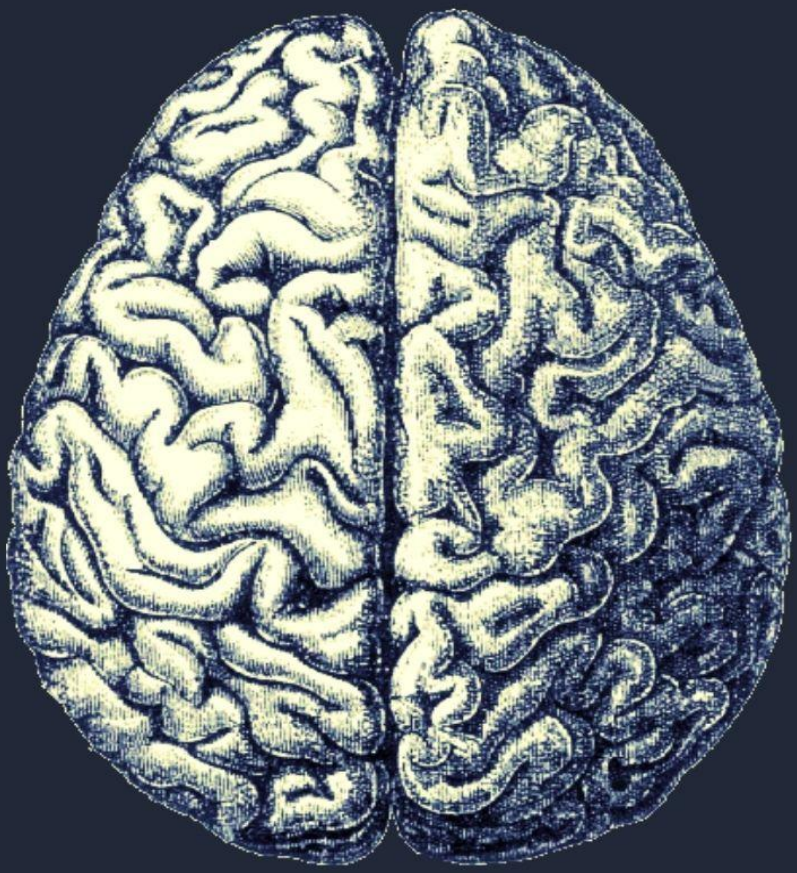

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Contents

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Cost-effective 3D reconstruction of skull defects

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Keywords
cranioplasty, 3D printing, titanium mesh, decompressive craniectomy, skull reconstruction

ABSTRACT

Cranial defects following decompressive craniectomy or tumour resection can result in major cosmetic, protective, and neurological deficits, including the "syndrome of the trephined". While patient-specific PEEK and titanium implants offer excellent anatomical and functional results, their high-cost limits accessibility in many healthcare systems. This study describes a cost-effective cranioplasty technique that uses in-house 3D-printed skull models or templates to pre-contour standard titanium mesh implants. We report three representative cases of large cranial defects secondary to tumour resection, complex infection with decompressive craniectomy, and severe traumatic brain injury. In each case, preoperative CT data were used to generate a patient-specific 3D model, which guided precise shaping of the implant and restoration of the native cranial curvature. This workflow combines the affordability and availability of conventional materials with the accuracy of computer-aided planning, minimises intraoperative modelling time, and reduces the need for expensive industrial patient-specific implants. Our early experience suggests that 3D-assisted contouring of standard implants is a robust and accessible strategy for restoring cranial integrity and improving cosmetic outcomes in resource-constrained settings.

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1. INTRODUCTION

Cranioplasty is one of the oldest neurosurgical procedures, dating back to antiquity. Historically performed mainly to protect the underlying brain, modern cranioplasty serves three critical functions: cosmetic restoration, mechanical protection, and the restoration of physiological hydrodynamics and intracranial pressure [1]. The latter is particularly relevant in the "syndrome of the trephined" (sinking skin flap syndrome), in which atmospheric pressure acting on an unshielded brain leads to neurological deterioration, headaches, and focal deficits [2]. Once cerebral edema has subsided and the patient survives the initial insult, the resulting cranial defect frequently necessitates secondary reconstruction to restore both appearance

and neurophysiological function [3]. Beyond these structural and hemodynamic effects, restoring cranial integrity may also facilitate recovery of higher-order cognitive and language functions, in line with contemporary neurolinguistic and cognitive perspectives on brain plasticity [14].

The choice of material for cranioplasty remains a matter of debate, balancing biocompatibility, mechanical strength, aesthetic outcome, radiological behavior, and cost [4]. Autologous bone has long been considered the gold standard, but its use is limited by high resorption rates and infection risk, often necessitating flap disposal [5]. Synthetic alternatives such as titanium mesh, PEEK, and PMMA have therefore become indispensable. Titanium mesh is widely used due to its high strength, biocompatibility, and low infection rates compared to autologous grafts [6]. However, conventional intraoperative contouring of titanium mesh for large or complex defects is technically demanding and may result in suboptimal cosmetic results or sharp edges that risk scalp breakdown. Patient-specific implants (PSI) manufactured from PEEK or titanium provide excellent anatomical accuracy but are expensive and frequently inaccessible in low- and middle-income settings [7,8].

Three-dimensional (3D) printing has emerged as a promising strategy to bridge this gap. Using preoperative CT imaging, a virtual model of the skull and defect can be generated; from this, 3D-printed anatomical models or templates enable preoperative or extracorporeal shaping of standard materials (titanium mesh or PMMA), thereby reducing cost while preserving the advantages of patient-specific reconstruction [9–11]. Moreover, casting or contouring the implant away from the brain avoids the exothermic polymerization risks associated with intraoperative PMMA molding [10].

Infection risk and optimal timing of cranioplasty remain crucial considerations. Many authors advocate reconstruction 3–6 months after craniectomy to allow for resolution of brain edema and scalp healing, although earlier cranioplasty (within 12 weeks) may help mitigate the syndrome of the trephined in selected patients [12].

In this context, the aim of our study is to describe an in-house, cost-effective 3D-assisted cranioplasty workflow using 3D-printed skull models or templates to contour standard titanium mesh implants, and to illustrate its application through three representative cases of large cranial defects of different etiologies.

2. Methods / 3D-assisted cranioplasty workflow

Preoperative thin-slice skull CT scans were exported in DICOM format, processed in dedicated 3D reconstruction software ([software name]) for segmentation of the cranial vault and defect, and mirrored from the intact side to restore native curvature. The resulting 3D model was exported as an STL file and printed on a desktop FDM 3D printer using PLA filament, either as a full-scale skull or local template depending on defect morphology and surgeon preference. After sterilization according to institutional protocol, the models were used pre- or intraoperatively to contour standard titanium mesh on the back table, ensuring close adaptation to the cranial curvature and smooth edges. All procedures were performed by the same neurosurgical team, and demographic, radiological, operative, and early clinical and cosmetic outcomes were prospectively recorded.

3. Case reports

3.1. Case 1 – Infectious etiology with decompressive craniectomy

A 20-year-old male presented to the Emergency Department with intense headache and left-sided hemiparesis. He had a known history of frontal sinusitis. Preoperative imaging identified a life-threatening intracranial infectious process consisting of a right frontal subdural empyema, a parasagittal subdural empyema, and a left epidural abscess. Due to the severity of the infection and intractable intracranial hypertension, the patient underwent a life-saving decompressive craniectomy with evacuation of the empyemas and cranialization of the frontal sinus. The craniectomy was extended to allow adequate decompression of the edematous brain.

After complete resolution of the infection and clearance for reconstruction, restoration of the cranial vault was prioritized. Given the patient's young age and the extensive fronto-temporo-parietal defect, achieving an optimal cosmetic result was essential. To avoid the prohibitive costs of PEEK PSIs, a cost-effective 3D-assisted reconstruction was chosen. A patient-specific 3D anatomical model of the skull defect was generated from CT data and printed in PLA. This model served as a template for

shaping a titanium mesh implant preoperatively on the back table. This hybrid technique combined the affordability of standard titanium mesh with the precision of computer-assisted design and eliminated many of the irregularities associated with free-hand bending.

Postoperative CT demonstrated restoration of the cranial contour, with the implant perfectly adapted to the bone margins and providing rigid protection of the underlying brain. The herniation seen on preoperative imaging was reversed. The patient was discharged with improved neurological status, and his hemiparesis resolved over the following weeks. At 6-month follow-up, there were no implant-related complications or infections.

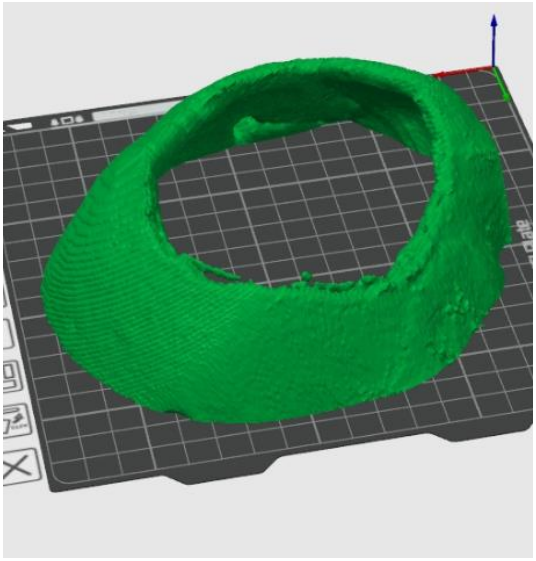

Figure 1. 3D-printed mold on which the titanium mesh was shaped to match the patient-specific cranial curvature.

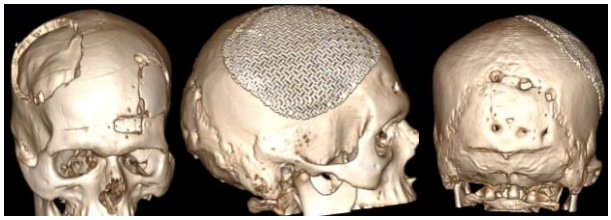

Figure 2. Preoperative and postoperative CT images demonstrating cranial reconstruction with a titanium mesh implant.

3.2 Case 2 – Tumor-related cranial defect

A 71-year-old male was admitted to the Neurosurgery Department following an episode of right-sided focal motor seizures that secondarily generalized. On admission, the patient was conscious and cooperative (GCS 14) but reported headache and vertigo. Native and contrast-enhanced CT scans revealed a large, mixed-density parasagittal mass in the left fronto-parietal region. Imaging showed an extra-axial lesion with nodular components and a fluid element, exerting mass effect with compression of the left lateral ventricle. The tumor infiltrated the calvarial bone and the superior sagittal sinus, necessitating removal of the involved bone flap during surgery. [13]

To address the resulting cranial defect, a titanium mesh implant was used. The mesh was contoured intraoperatively to replicate the natural curvature of the fronto-parietal region and to achieve a precise fit with the surrounding bone margins. Postoperative CT confirmed appropriate implant positioning, restoration of the cranial vault, and a symmetrical aesthetic profile. The postoperative course was uneventful. The patient was discharged in good general condition, without new neurological deficits, and with a highly satisfactory cosmetic result.

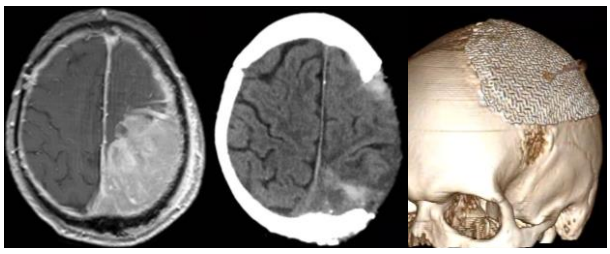

Figure 3. (left) Preoperative contrast-enhanced MRI demonstrating an intracranial mass with intraosseous extension; (center) postoperative CT scan following tumor ablation and craniectomy; and (right) 3D reconstruction of the cranial defect with a contoured titanium mesh implant.

3.3 Case 3 – Traumatic brain injury with extensive cranial defect

A 41-year-old male was admitted to the Emergency Department after a scooter accident with polytrauma. On arrival, he was intubated and sedated. Initial CT imaging revealed a devastating craniocerebral injury: a right parieto-temporo-occipital epidural hematoma exerting significant mass effect, an acute fronto-temporo-parieto-occipital subdural hematoma, and a midline shift of approximately 13 mm. Multiple skull fractures involved the parietal and temporal bones.

Emergency surgery was performed with evacuation of the hematomas and a large right-sided fronto-temporo-parietal decompressive craniectomy. Although the life-saving procedure was successful, follow-up CT scans in the subacute phase showed herniation of brain parenchyma through the craniectomy site, progression of edema, and distortion of cerebral architecture. Given the magnitude and irregular morphology of the defect, free-hand intraoperative contouring of a reconstruction implant would have been difficult and prone to cosmetic asymmetry.

Using the patient's CT data, a 3D model of the skull was generated and printed. This model was used preoperatively to precisely pre-contour a titanium mesh implant, matching the patient's individual cranial curvature. Intraoperatively, the pre-shaped mesh required only minimal adjustments and was fixed flush with the bone margins. This approach reduced operative time and provided rigid protection against atmospheric pressure, effectively reversing the sinking skin flap and parenchymal herniation. The reconstruction restored the symmetry of the skull and stabilized the previously herniated brain tissue.

The patient was discharged in good general condition, and his subsequent course was favorable, with progressive neurological improvement and complete recovery at 6-month follow-up, without implant-related complications.

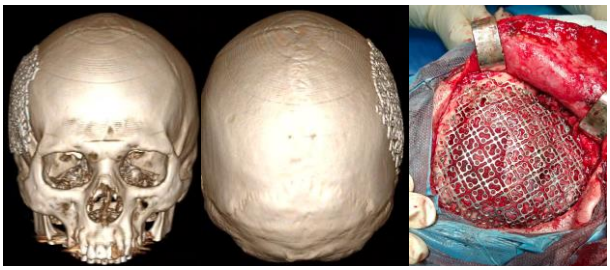

Figure 4. Postoperative CT 3D reconstructions (left and center) and intraoperative aspect (right).

4. Discussions

Reconstruction of large cranial defects is a multidimensional challenge that requires balancing functional restoration, cosmetic outcome, and economic feasibility. In many healthcare systems, particularly those with constrained resources, the high cost of industrial patient-specific implants represents a major barrier to optimal reconstruction. Our case series shows that combining in-house 3D printing with standard titanium mesh allows patient-specific restoration of cranial anatomy while maintaining affordability.

Titanium mesh remains an attractive material due to its mechanical strength, biocompatibility, and relatively low infection rates compared to autologous bone [6]. However, free-hand intraoperative contouring of mesh for large and complex defects is technically demanding and can result in irregular contours, "temporal hollowing," or palpable step-offs. By using 3D-printed models or templates derived from preoperative CT data, we were able to restore the native cranial curvature more reliably. In the presented cases, postoperative imaging confirmed a close anatomical fit and satisfactory cranial symmetry, with results comparable to those reported for high-end PSIs [7–11].

From a cost perspective, 3D-assisted contouring of standard implants offers a substantial economic advantage. The material cost of a 3D-printed PLA model and titanium mesh is typically only a fraction of that of a custom-made PEEK or titanium PSI, while still providing individualized reconstruction. This is particularly relevant in high-risk situations such as infectious etiologies or patients with a history of empyema, where implant removal may be required. In such scenarios, discarding an expensive PSI represents a significant financial loss, whereas replacing a standard titanium mesh contoured on a new 3D model is more acceptable from a health-economic standpoint.

Infection remains one of the most feared complications of cranioplasty, with reported rates between 4% and 12% [7,12]. In our small series, no implant-related infections occurred, including in the patient with a history of intracranial empyema. Although our follow-up is limited, this suggests that

3D-assisted use of standard materials does not carry an inherently higher infectious risk compared with more expensive alternatives, provided appropriate patient selection, timing, and perioperative management are observed.

The present work has several limitations. It is a small, single-center case series without a control group of conventional cranioplasties or industrial PSIs. Follow-up is relatively short, and functional outcomes were not quantified using standardized scales. Furthermore, we did not perform a formal cost-effectiveness analysis, which would be valuable to objectively compare this approach with commercial patient-specific solutions. Nevertheless, our experience supports the concept that in-house 3D printing can meaningfully expand access to individualized cranial reconstruction, particularly in centers where resources are limited but basic 3D printing infrastructure is available.

5. Conclusion

Cost-effective 3D-assisted cranioplasty using in-house 3D-printed models or templates to contour standard titanium mesh implants represents a robust and accessible alternative to expensive industrial patient-specific implants. In our small case series, this approach enabled accurate restoration of cranial anatomy, satisfactory cosmetic results, and adequate protection of the brain, without early implant-related complications. From a risk–benefit perspective, it is particularly appealing in resource-constrained settings and in patients at increased risk of infection, where the potential need for implant removal must be weighed against cost. We advocate the development and integration of simple in-house 3D printing workflows in neurosurgical centers, so that "cost-effective" reconstruction can remain synonymous with "clinically effective" rather than "low quality".

Table 1. Overview of the three 3D-assisted cranioplasty cases.

Parameter	Case 2	Case 1	Case 3
Age / Sex	71 / M	20 / M	41 / M
Etiology	Parasagittal extra-axial tumor with bone and sinus invasion	Intracranial infection (subdural empyema, epidural abscess)	Severe TBI with acute EDH + SDH and skull fractures
Defect location	Left fronto-parietal	Fronto-temporo-parietal	Right fronto-temporo-parietal
Decompressive craniectomy	No	Yes	Yes
3D printing use	Anatomical model	3D skull model for pre-contouring	3D skull model for pre-contouring
Implant material	Titanium mesh	Titanium mesh	Titanium mesh
Early outcome	Good cosmetic result, no new deficits	Hemiparesis resolved, no reinfection	Restored cranial symmetry, no early complications
Follow-up	3 months, no complications	6 months, no complications	6 months, no complications

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Incidence and severity of complications in postoperative spondylodiscitis after lumbar disc herniation surgery

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ABSTRACT

Background: Postoperative spondylodiscitis is a rare but serious complication after lumbar disc herniation surgery, with incidence varying between 0.2–4%. It is mainly related to intraoperative contamination and host-related risk factors. This study aimed to evaluate demographic variables, lifestyle-related factors, and clinical outcomes in patients diagnosed with postoperative spondylodiscitis.

Methods: We conducted a retrospective observational study on 85 patients treated between 2018 and 2024 in a tertiary care centre. Clinical and paraclinical data were collected, including demographic characteristics, residence, lifestyle-related risk factors, discectomy level, complications, treatment type, motor deficit, and hospitalisation duration. Statistical analyses were performed using χ^2 and Student's t-tests, with significance set at $p < 0.05$.

Results: Complications were observed in 24 patients (28.2%), predominantly at the lumbar level. Age, sex, and residence showed no significant associations with complication status ($p > 0.05$). In contrast, lifestyle-related factors such as smoking, alcohol consumption, and poor hygiene were significantly correlated with complications (χ^2 test, $p < 0.05$). Patients with complications had longer hospital stays (23.25 vs. 12.66 days, $p < 0.05$). Abscesses were the most frequent complication, followed by epiduritis. The presence of complications was significantly associated with motor deficits ($p < 0.05$).

Conclusion: While demographic factors did not influence outcomes, lifestyle-related risk factors and surgical treatment were associated with increased complications and prolonged hospitalisation. Early identification of high-risk patients and aggressive management of abscess formation remain essential to reduce morbidity. Keywords: postoperative spondylodiscitis, lumbar disc herniation, spinal infection, complications, lifestyle risk factors, hospitalisation, motor deficit.

Abbreviations: N = number of patients; p = p-value; r = Pearson correlation factor; SD = standard deviation; t-test = Student's t-test; χ^2 = Chi-square test.

Keywords
postoperative spondylodiscitis, lumbar disc herniation, spinal infection, lifestyle-related risk factors

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Introduction

Postoperative spondylodiscitis is a rare but serious complication following lumbar disc herniation surgery, with an incidence reported between 0.2% and 4%, depending on surgical technique and patient-related risk factors [1]. The condition is characterized by infection of the intervertebral disc and adjacent vertebral bodies, often leading to prolonged hospitalization, need for reoperation, and in severe cases, long-term disability [2,3]. Early diagnosis remains difficult due to the non-specific clinical presentation, which includes back pain, fever, and elevated inflammatory markers that may overlap with expected postoperative changes [2,4,5]. Several studies have indicated that the presence of comorbidities such as diabetes, smoking, and immunosuppression increases susceptibility to postoperative infections, while intraoperative factors including prolonged surgical time and intraoperative bleeding may also contribute [4-6]. Furthermore, lifestyle-related factors such as poor hygiene, alcohol consumption, and tobacco use have been implicated in exacerbating the risk of postoperative complications, although evidence remains heterogeneous [7,8]. The clinical course of postoperative spondylodiscitis varies significantly; some patients respond well to conservative treatment with antibiotics, while others require revision surgery due to abscess formation, epidural involvement, or persistent neurological deficits [9,10]. Given its impact on patient morbidity and healthcare burden, it is essential to better understand the risk factors, complication patterns, and outcomes of this condition in order to develop improved strategies for early recognition, prevention, and management.

Materials and methods

Study design and population

We conducted a retrospective observational study including 85 patients diagnosed with postoperative spondylodiscitis following lumbar disc herniation surgery. Patients were identified from the institutional database and included if they met clinical, radiological, and/or microbiological criteria for postoperative spondylodiscitis.

The cohort consisted of 24 patients with complications and 61 patients without complications. Patients were further subgrouped according to age, sex, place of residence (urban vs rural), lifestyle-related risk factors (smoking, alcohol consumption, poor hygiene), treatment modality (conservative vs surgical), and discectomy level (thoracolumbar, lumbar, lumbosacral).

Variables and data collection

The following variables were collected: age, sex, place of residence, presence of lifestyle-related risk factors, type of treatment received, duration of hospitalization, presence and type of complications, and presence of neurological motor deficit.

Complications were categorized as: abscess, epiduritis, fluid collection, myositis, or empyema.

Statistical analysis

Statistical analysis was performed using descriptive and inferential methods. Continuous variables were expressed as mean ± standard deviation (SD), while categorical variables were reported as frequencies and percentages.

Normality of continuous variables was assessed using the Shapiro–Wilk test.

Outliers were checked with standardized Z-scores (no extreme values identified, $-3 < Z < 3$).

Homogeneity of variances was verified using Levene's test.

The relationship between age and hospitalization duration was tested with the Pearson correlation coefficient ($r = 0.007$, $p \geq 0.05$, no significant correlation).

Comparisons of mean age between patients with and without complications were performed with the Student's t-test.

Associations between categorical variables (presence of complications vs sex, residence, lifestyle risk factors, treatment type, and motor deficit) were tested with the Chi-square (χ^2) test of independence.

All analyses were two-tailed, with statistical significance set at $p < 0.05$.

Results

When analyzing the relationship between age and the presence of complications, patients with complications had a slightly lower mean age (61.29 years, SD±11.611) compared to those without complications (63.75 years, SD±13.608). Although complications were more frequently observed in patients aged between 55 and 79 years, the overall difference in age between the two groups was not statistically significant. The distribution of age did not

differ substantially between subgroups, and no extreme values were identified (Table 1).

Table 1. Patient age distribution according to the presence of complications

Grouping Criteria	Cohort Subgroups	No. of patients	Median Age
Presence of complications	Yes	24	61.29 (SD±11.611)
	No	61	63.75 (SD±13.608)
Total		**85**	**63.06 (SD±13.055)**

Regarding the relationship between age and length of hospitalization, the two variables are not correlated (r = 0.007, p-value ≥ 0.05); therefore, hospitalization duration does not increase with patient age.

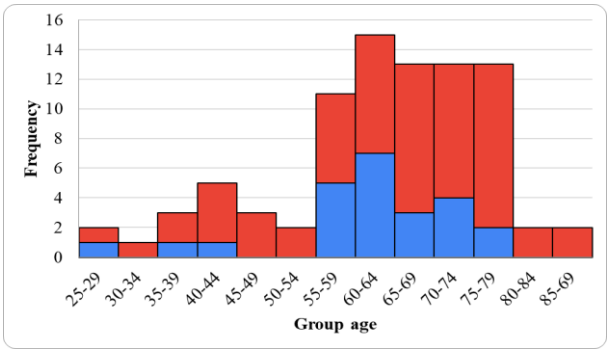

Figure 1. Age group distribution by complication status.

Red bars represent patients without complications, while blue bars represent those with complications. The number of patients with complications is lower than that of patients without complications in the older age groups. Most patients with complications are concentrated between ages 55 and 79. Statistical analysis confirmed that age distributions did not differ significantly between groups (Levene's test p ≥ 0.05; Student's t-test p ≥ 0.05), although normality was not met in either subgroup (Shapiro–Wilk p < 0.05) (Figure 1).

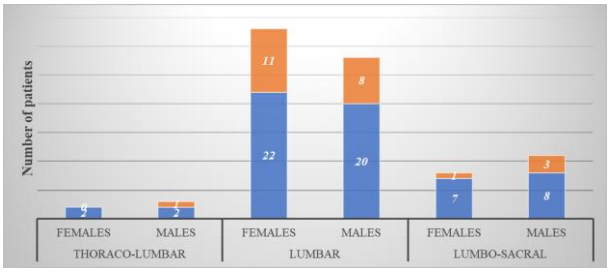

Figure 2. Distribution of patients with and without complications by spinal level and sex.

Blue bars represent patients without complications, while orange bars represent patients with complications. Out of the 85 patients included in the study, 24 developed complications. The association between complication occurrence and sex was analyzed only for the lumbar region, as the minimum number of observations per category was not met in the other regions. No statistically significant association was found between sex and the presence of complications in lumbar spondylodiscitis (χ^2 test, $p > 0.05$) (Figure 2).

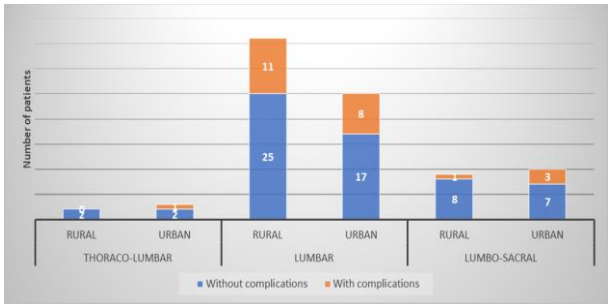

Figure 3. Distribution of patients with and without complications by spinal level and residence.

No statistically significant association was found between residence (urban/rural) and complications in lumbar spondylodiscitis (χ^2 test, $p > 0.05$) (Figure 3).

Table 2 presents the presence or absence of complications in patients with lifestyle-related risk factors. Among the total participants included in the study, only 18 individuals exhibited lifestyle-related risk factors, some of whom had two or even three risk factors. The presence of at least one risk factor was significantly associated with the occurrence of complications (χ^2 test, $p < 0.05$).

Level of discecomy	Lifestyle factors	Without complications	With complications
Thoraco-lumbar	*Smoking*	1	1
	Alcohol	0	1
	Poor hygiene	1	0
Lumbar	*Smoking*	**3**	**4**

	Alcohol	2	1
	Poor hygiene	1	**4**
Lumbo-sacral	*Smoking*	**4**	0
	Alcohol	1	2
	Poor hygiene	1	0

Table 2. Presence of complications in patients with lifestyle-related risk factors by level of discectomy

83 of 85 patients received conservative treatment, consisting of antibiotic therapy, either a single antibiotic or combination regimens involving two or three antibiotics from different classes. Surgical treatment was significantly associated with the presence of complications (χ^2 test, $p < 0.05$). Table 3 illustrates the average length of hospital stay for the total patient cohort and for subgroups (patients with complications versus those without complications, and by level of discectomy). The mean hospital stay was significantly longer for patients with complications compared to those without complications (Student's t-test, $p < 0.05$).

Level of discectomy	Complications	Number of patients	Median hospital stay
Thoraco-lumbar	Yes	1	10
	No	4	1.5 (SD±0.577)
Lumbar	Yes	19	23.74 (SD±17.944)
	No	42	10.57 (SD±13.182)
Lumbo-sacral	Yes	4	24.25 (SD±20.855)
	No	15	21.47 (SD±36.975)
Subtotal	**Yes**	**24**	**23.25 (SD±17.797)**
Subtotal	**No**	**61**	**12.66 (SD±21.645)**
Total		**85**	**15.65 ((SD±21.080)**

Table 3. Average length of hospital stay by complication status and level of discectomy.

In the studied cohort, complications were most frequently observed at the lumbar level, with abscesses being the most common type. Thoraco-lumbar and lumbo-sacral levels showed fewer complications overall. Abscess formation predominantly affected the lumbar region, while other complications such as epiduritis, fluid accumulation, myositis, and empyema were less frequent across all levels (Table 4).

Level of discecomy	Complication type	Number of patients
Thoraco-lumbar	No complications	4
	Abcess	0
	Epiduritis	0
	Fluid build-up	1
	Miozitis	0
	Empiema	0
Lumbar	No complications	**42**
	Abcess	11
	Epiduritis	4
	Fluid build-up	2
	Miozitis	1
	Empiema	1
Lumbo-sacral	No complications	15
	Abcess	3
	Epiduritis	1
	Fluid build-up	0
	Miozitis	0
	Empiema	0
Total	**No complications**	**61**
Total	**Complications**	**24**

Table 4. Frequency of complications and diagnostic approaches in the study cohort

Figure 4 shows the frequency of patients according to the presence of complications and motor deficit. The presence of complications was significantly associated with the occurrence of motor deficit (χ^2 test, $p < 0.05$).

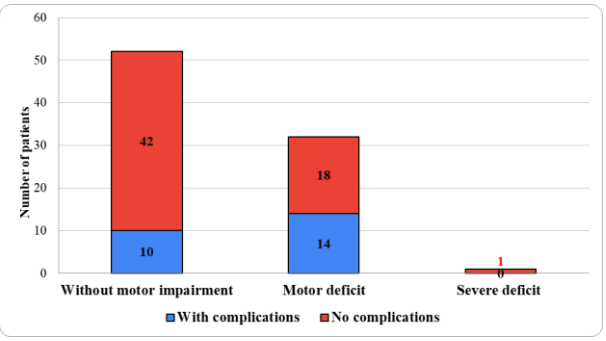

Figure 4. Frequency of patients by complication status and presence of motor deficit

Discussions

In our study, the incidence of postoperative spondylodiscitis was associated with lifestyle-related risk factors such as smoking, alcohol consumption, and poor hygiene, while demographic factors including age, sex, and place of residence did not show significant associations with the presence of complications. These findings are consistent with previous reports suggesting that lifestyle-related and comorbidity factors play an important role in susceptibility to postoperative spinal infections (11,12).

Similar to our results, other studies have shown that age alone is not a predictor of postoperative complications, although most cases cluster in the middle to older age groups (13). In our cohort, patients aged between 55 and 79 years were most affected, which aligns with findings indicating that comorbidities and immunological decline in these age groups may predispose to infections rather than age itself being an independent risk factor (14).

We also observed that sex was not significantly associated with the occurrence of complications in lumbar cases. This is in agreement with several reports in the literature showing no consistent gender-related predisposition to postoperative disc infections (15).

Importantly, we found that patients with complications had a significantly longer hospital stay compared to those without complications, which reflects the increased burden of postoperative spondylodiscitis. This observation is supported by prior studies highlighting prolonged hospitalization and healthcare costs as major consequences of these infections (16,17).

The most frequent complication in our study was abscess formation, followed by epiduritis, consistent with previous findings where abscesses are described as a common driver for the need of surgical intervention (18). Our results reinforce the need for early recognition and aggressive management of abscesses in order to reduce morbidity and prevent neurological deficits.

Conclusions

This retrospective study on 85 patients with postoperative spondylodiscitis following lumbar disc herniation surgery highlights several key findings. Complications were present in 24 patients (28.2%), most frequently in the lumbar region, with abscess formation as the predominant type.

Age did not significantly influence the occurrence of complications (Student's t-test, $p \geq 0.05$), nor was there a correlation between age and hospitalization duration ($r = 0.007$, $p \geq 0.05$). Most complications were concentrated in the 55–79 years age group, but age itself was not a predictor. Similarly, sex and place of residence showed no significant associations with the occurrence of complications (χ^2 test, $p > 0.05$).

In contrast, lifestyle-related factors such as smoking, alcohol consumption, and poor hygiene were significantly associated with complications (χ^2 test, $p < 0.05$). Surgical treatment was also strongly correlated with the presence of complications (χ^2 test, $p < 0.05$), underlining the higher risk in this subgroup.

Hospitalization was significantly prolonged in patients with complications compared to those without (23.25 vs. 12.66 days, Student's t-test, $p < 0.05$), demonstrating the additional healthcare burden. Moreover, the presence of complications was significantly associated with motor deficits (χ^2 test, $p < 0.05$), reflecting the clinical severity of these cases.

Overall, our results emphasize that while demographic variables such as age, sex, and residence did not influence outcomes, lifestyle-related factors, type of treatment, and the development of abscesses play a central role in the evolution and prognosis of postoperative spondylodiscitis. Early recognition of complications and targeted interventions are essential to limit prolonged hospitalizations and neurological deterioration.

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Single vs multiple pathogens in brain abscesses: two cases with distinct aetiologies

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ABSTRACT

Brain abscesses represent life-threatening focal infections with diverse aetiologies. In the current era of escalating antimicrobial resistance, medical management is increasingly challenged and empiric regimens may be insufficient. We report two young immunocompetent male patients with brain abscesses due to distinct aetiologies: a monomicrobial subdural and epidural collection caused by Streptococcus constellatus and a polymicrobial post-traumatic orbitofrontal abscess associated with Enterococcus faecalis and Escherichia coli. Both patients underwent urgent surgical drainage combined with broad-spectrum intravenous antibiotics, later tailored to culture results. The first case illustrates the risk of suboptimal oral therapy and poor adherence, with subsequent relapse and development of antimicrobial resistance, whereas the second case highlights infectious complications following penetrating orbito-cranial trauma. These contrasting cases emphasise the need for timely neurosurgical intervention, prolonged targeted antimicrobial therapy, and strict avoidance of unsupervised antibiotic use in the management of brain abscesses.

Keywords
brain abcess, subdural empyema, epidural abcess, polimicrobial infection, streptococcus constellatus, antibiotic resistance

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1. INTRODUCTION

A brain abscess is defined as a localized accumulation of pus within the brain parenchyma or meninges, enclosed by a distinct capsule. This pathology typically arises as a secondary complication stemming from adjacent infections or from systemic sources. [1] Brain abscesses account for a small proportion of intracranial space-occupying lesions, but they carry substantial morbidity and mortality, especially when diagnosis or treatment is delayed. Most cases arise by contiguous spread from otitis, mastoiditis, or sinusitis, or via hematogenous dissemination from distant infective foci such as endocarditis or lung abscesses. Penetrating head trauma and neurosurgical procedures represent less frequent but clinically important causes, often associated with polymicrobial infections and more resistant organisms. [2] In contemporary series, brain abscess accounts for approximately 1–2% of intracranial space-occupying

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lesions, with reported mortality rates of about 5–15% despite advances in imaging and antimicrobial therapy.

Microbiological patterns typically differ according to the route of infection. Contiguous and odontogenic abscesses usually involve streptococci of the Streptococcus anginosus group, anaerobes, and mixed oral flora, whereas hematogenous abscesses tend to be monomicrobial. Post-traumatic and post-neurosurgical abscesses are more often polymicrobial and may include staphylococci, enterococci, and aerobic Gram-negative bacilli. [3]

In this context, we present two contrasting cases: a large epidural and subdural collection due to a single Streptococcus constellatus strain and a post-traumatic orbitofrontal abscess associated with multiple pathogens. By comparing their clinical course and management, we aim to highlight the practical implications of single versus polymicrobial infection for neurosurgeons. [4]

2. Methods

We conducted a descriptive retrospective case report including two male patients diagnosed with intracranial abscess/empyema and treated in the Neurosurgery Department of Sibiu County Emergency Hospital. Clinical data, imaging findings, operative notes, microbiological results and follow-up information were obtained from institutional medical records. Microbiological diagnosis was established by culture of material collected intraoperatively from the abscess cavity and/or associated soft-tissue collections. Demographic characteristics, presumed source of infection, anatomical location, number and type of pathogens, surgical procedures, antimicrobial regimens and clinical outcomes were extracted and are summarised in Table 1. Written informed consent was obtained from both patients for publication of this case report and the accompanying images.

3. Case 1

A 21-year-old male presented to our emergency department with a severe headache with onset approximately 3 weeks prior and a left-sided motor deficit. The patient was conscious but uncooperative and confused, with a Glasgow Coma Scale (GCS) score of 14.

A multidetector computed tomography (MDCT) scan was performed, revealing a right frontal subdural fluid collection, a thin right parasagittal subdural fluid layer, and an extensive anterior high-frontal and left parasagittal fluid collection. Additionally, spontaneous hyperdensity of the falx cerebri and tentorium, marked diffuse cerebral edema of the right hemisphere, and fluid-density accumulation occupying the paranasal sinuses were observed. (Figure A)

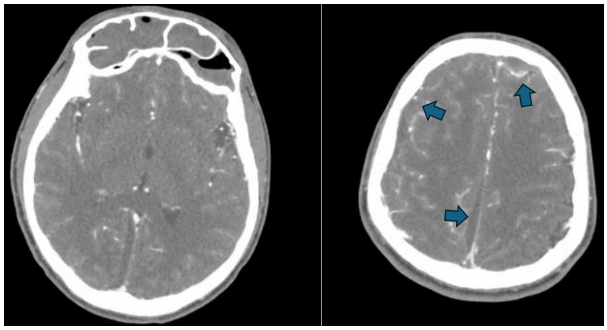

Figure A. Initial MDCT scans showing right frontal and parasagittal subdural collections with significant right hemispheric edema

Based on imaging investigations and patient history, the diagnosis of left frontal epidural abscess, and right parasagittal and fronto-temporo-parietal (FTP) subdural empyema was established.

A right FTP decompressive craniotomy was performed, with lavage and drainage of the right frontal and left parasagittal subdural empyema, and of the left epidural abscess.

The following day, a reintervention was performed, consisting of the enlargement of the decompressive craniectomy to allow expansion of the edematous brain.

Microbiological analysis revealed the presence of *Streptococcus constellatus*, susceptible to the majority of antibiotics. Intravenous empirical therapy with a third-generation cephalosporin plus metronidazole was initiated and later tailored to the susceptibility profile of S. constellatus. The patient remained hospitalised, with serial CT/MRI showing progressive reduction of the collections and mass effect.

The patient was discharged with instructions to avoid physical exertion, exposure to cold and damp environments, and local trauma. The prescribed home treatment regimen included antibiotics, antiepileptics, analgesics, antipyretics, and vitamins.

Despite medical advice, the patient continued unsupervised oral levofloxacin (750 mg/day) for several months after discharge. At

readmission, cultures from the recurrent abscess demonstrated a markedly reduced susceptibility profile, with sensitivity retained only to teicoplanin. This evolution suggests that prolonged, suboptimal oral therapy and poor adherence may have contributed to the selection of a resistant S. constellatus strain.

Over the following months, the patient was readmitted twice. The first readmission, prompted by complaints of headache, involved the evacuation of the right occipital abscess contents and excision of the capsule (Figure B). The second admission was for cranioplasty (Figure C)

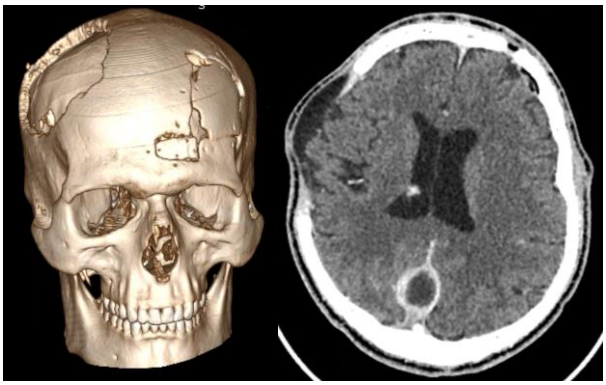

Figure B. Left: Postoperative imaging appearance; Right: Well-encapsulated occipital interhemispheric abscess identified after 1 month of antimicrobial therapy.

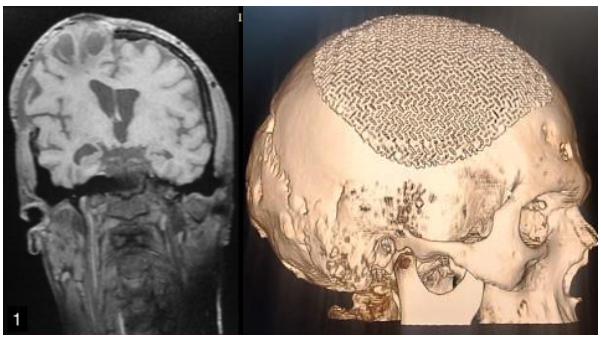

Figure C. 1 – Brain MRI revealing fungus cerebri (transcalvarial herniation of the cerebral parenchyma) through the craniectomy defect. 2 – CT scan status post right frontoparietal cranioplasty (6 months later).

At 6-month follow-up, he remained seizure-free under levetiracetam, with mild residual left-sided weakness and no radiological evidence of recurrent infection.

4. Case 2

We present the case of a 33-year-old male admitted for a right upper eyelid abscess three days following a penetrating cranio-facial trauma with a metallic object. CT imaging revealed a complex orbital fracture, pneumocephalus, and an intraparenchymal bone fragment (Figure C), which were initially managed conservatively. However, despite the resolution of the local infection, the patient developed delayed post-traumatic seizures, necessitating urgent transfer to the Neurosurgery department for the management of the penetrating orbito-cranial injury. He had no history of immunosuppression or major chronic disease.

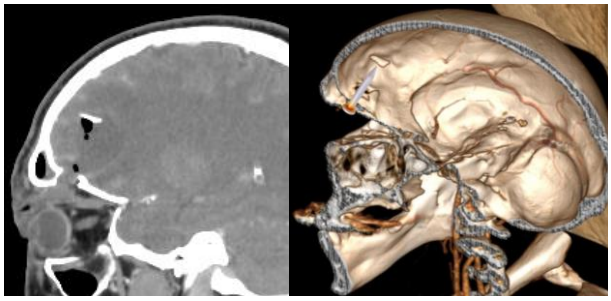

Figure D. Left – Cranial CT scan demonstrating a comminuted and displaced fracture of the right orbital roof. Associated findings include a small right frontal intraparenchymal pneumocephalus and limited subarachnoid hemorrhage. Right – 3D reconstruction demonstrating the trajectory of the metallic object and the migrated bone fragment secondary to the fracture.

The patient received an initial antibiotic regimen consisting of Ceftriaxone, Vancomycin, and Metronidazole. However, due to the lack of clinical improvement and the onset of an epileptic seizure 4 days after starting antibiotic treatment, the decision was made to proceed with neurosurgical intervention.

Repeat CT revealed an enlarging right frontal ring-enhancing lesion with surrounding edema and mass effect, despite 4 days of antibiotic therapy. Given the lesion size (>2 cm) and the new-onset seizures, surgical evacuation was indicated.

Neurosurgical intervention was performed under general anaesthesia, consisting of a right frontal craniotomy with evacuation of the right frontal abscess and removal of the intraparenchymal bone fragment.

Bacteriological examination isolated *Enterococcus faecalis* from the cerebral abscess and *Escherichia coli*

from the eyelid abscess. Both pathogens were found to be susceptible to antibiotics.

Based on susceptibility testing, ceftriaxone was de-escalated and the regimen was switched to high-dose ampicillin plus gentamicin to provide synergistic bactericidal activity against E. faecalis, while anaerobic coverage was maintained with metronidazole. The E. coli isolate from the eyelid abscess was fully susceptible to third-generation cephalosporins and fluoroquinolones and did not produce extended-spectrum β-lactamases.

At the time of discharge, the patient was conscious, cooperative, and temporo-spatially oriented. He was in good general condition, remained seizure-free, and showed complete resolution of neurological symptoms. A home treatment regimen consisting of antibiotics, anticonvulsant therapy, and corticosteroids for cerebral edema management was prescribed.

At 12-month follow-up, the patient had no recurrent seizures under anticonvulsant prophylaxis and CT showed complete resolution of the lesion.

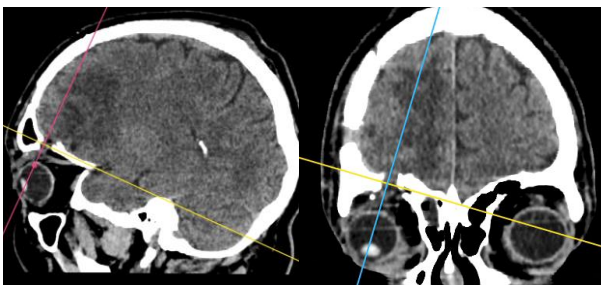

Figure E. Postoperative CT demonstrating regression of cerebral edema and reduction of the abscess cavity.

5. Discussion:

Streptococcus constellatus is a gram-positive, catalase-negative, facultatively anaerobic coccus that typically arranges in chains. The organism is characterized by its propensity to cause purulent infections and abscesses, a trait that distinguishes it from many other viridans streptococci. [2] In the Streptococcus family, antibiotics penetrate abscesses poorly and bacteria exposed to sub-inhibitory concentrations may develop mutations in penicillin-binding proteins (PBPs). These mutations alter the antibiotic target site, preventing penicillin from binding effectively. [3],[4]

Escherichia coli (E. coli) is a gram-negative, rod-shaped bacterium. It is the most dominant facultative anaerobe in the non-pathogenic human intestinal flora. [5] Resistance is primarily mediated by the secretion of beta-lactamases, which destroy the antibiotic molecule prior to its action. Isolates producing Extended-Spectrum Beta-Lactamases (ESBLs) are capable of degrading cephalosporins, whereas carbapenemase-producing stains inactivate carbapenems. [6][7]

Enterococcus faecalis is a gram-positive facultatively anaerobic coccus. While it morphologically resembles streptococci, it is distinguished by its extreme environmental durability. [8] E. faecalis exhibits intrinsic resistance to cephalosporins. It possesses specific proteins (PBP4 and PBP5) that have a very low affinity for these antibiotics, allowing the bacterium to synthesize its cell wall even in their presence. [9]

In our first patient, the infection was caused by a single S. constellatus strain, likely originating from paranasal sinus disease, as suggested by the CT findings. This pattern is consistent with previous reports, in which S. anginosus group streptococci are frequently isolated from brain abscesses arising from sinusitis, otitis, or odontogenic infections. By contrast, post-traumatic abscesses and post-neurosurgical infections are more often polymicrobial and may involve environmental Gram-negative bacilli, staphylococci, and enterococci.

The second case reflects this pattern: the penetrating orbitofrontal injury led to a polymicrobial infection involving E. faecalis and E. coli. Polymicrobial abscesses are associated with a higher risk of antimicrobial resistance and typically require broader empiric coverage, often combining a third-generation cephalosporin with metronidazole and, in selected settings, vancomycin or a carbapenem until culture results are available.

Penetrating orbito-cranial injuries are uncommon but carry a high risk of intracranial infection, particularly when associated with retained foreign bodies and contamination by skin, sinus, or environmental flora. Small orbital wounds may initially appear benign and the intracranial trajectory can be easily overlooked, leading to delayed diagnosis of cranial injuries and brain abscesses.

Awareness of these mechanisms is crucial, as early imaging and prophylactic broad-spectrum antibiotics may reduce the risk of severe infectious complications.

Antibiotic resistance is escalating. The contributing factors are multifactorial,

encompassing the use of antibiotics in the agro-industry, non-adherence to treatment regimens and self-medication, with the latter being the most prevalent. [10][11]

Current guidelines emphasise that successful treatment of brain abscess almost always requires a combination of surgical drainage and prolonged high-dose antimicrobial therapy. Empiric treatment is guided by the presumed source and is later narrowed based on cultures, with a recommended total duration of 6–8 weeks of intravenous therapy, followed by careful imaging and clinical follow-up. The need for repeated aspirations or craniotomy is dictated by lesion size, location, mass effect, and response to therapy; abscesses larger than 2–2.5 cm or those causing significant mass effect are usually managed surgically. [12]

Both of our patients required surgical evacuation due to mass effect and neurological symptoms. In the first case, early decompressive craniectomy was necessary to control malignant cerebral edema, while in the second case craniotomy allowed removal of both the abscess and the intraparenchymal bone fragment along the transorbital trajectory. [13]

The clinical course of the first patient illustrates the dangers of unsupervised oral antibiotic use in the setting of intracranial infection. Prolonged home-based levofloxacin therapy, outside of specialist supervision and without appropriate intravenous induction, may have contributed to selection of a less susceptible S. constellatus strain and delayed definitive management. This observation aligns with broader data showing that self-medication, poor adherence, and inappropriate antibiotic choice are major drivers of antimicrobial resistance at the population level.

Prognosis in brain abscess depends on multiple factors, including preoperative neurological status, level of consciousness, presence of multiple lesions, intraventricular rupture, and comorbidities such as immunosuppression. Both of our patients were young and immunocompetent, presented with relatively preserved GCS scores, and received timely neurosurgical and antimicrobial treatment, which likely contributed to their favourable functional outcomes despite the severity of their intracranial infections.

Limitations

A limitation of this report is the small number of patients and the absence of systematic reporting of laboratory parameters and long-term neurocognitive outcomes. Nevertheless, the contrasting aetiologies and microbiological patterns provide useful illustrative scenarios for everyday neurosurgical practice.

6. Conclusion:

These cases highlight the central role of neurosurgical drainage combined with prolonged, carefully supervised antimicrobial therapy in the management of brain abscesses with different etiologies. While the first patient presented with an extensive monomicrobial collection due to S. constellatus, the second developed a polymicrobial post-traumatic abscess involving E. faecalis and E. coli. In both scenarios, timely surgical intervention, culture-guided antibiotic therapy, and close radiological follow-up were essential for a favourable outcome. The relapse and resistance pattern observed in the first case further underline the risks of unsupervised, prolonged oral antibiotic use in such severe infections and reinforce the need for strict antibiotic stewardship and patient education.

From a practical perspective, these cases suggest that monomicrobial abscesses arising from contiguous sinus disease and polymicrobial post-traumatic orbitofrontal abscesses may differ in their microbiological spectrum and empiric coverage requirements, but they share a common need for timely neurosurgical drainage and prolonged, supervised antimicrobial therapy.

Table 1. Key differences between the two brain abscess cases.

Parameter	Case 1 – Non-traumatic empyema (single pathogen)	Case 2 – Penetrating orbitofrontal abscess (multiple pathogens)
Patient / etiology	21-year-old male; likely contiguous spread from sinus infection	33-year-old male; penetrating trans-orbital trauma with metallic object
Location / imaging	Left frontal epidural + right parasagittal/FTP subdural empyema; marked right hemispheric edema	Right orbitofrontal intraparenchymal abscess near orbital roof fracture; limited SAH/pneumocephalus

Parameter	Case 1 – Non-traumatic empyema (single pathogen)	Case 2 – Penetrating orbitofrontal abscess (multiple pathogens)
Microbiology	**Single pathogen:** *Streptococcus constellatus*	**Polymicrobial:** *Enterococcus faecalis* (brain) + *Escherichia coli* (eyelid)
Main treatment	FTP decompressive craniotomy with drainage; later occipital abscess evacuation and cranioplasty; IV antibiotics + prolonged unsupervised oral levofloxacin at home	Right frontal craniotomy, abscess evacuation and bone fragment removal; IV ceftriaxone + vancomycin + metronidazole, then targeted therapy
Complications	Fungus cerebri, recurrent abscess; selection of fluoroquinolone-resistant *S. constellatus*	Early post-traumatic seizures; no documented infectious recurrence
Outcome at discharge	Conscious, oriented, afebrile; improved motor status	Conscious, oriented, seizure-free on anticonvulsants; good general condition

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Psychiatric and neurobehavioral effects of posterior fossa surgery. A literature review

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ABSTRACT

The posterior fossa, housing the brainstem and cerebellum, has traditionally been viewed as exclusively responsible for motor functions. However, recent literature has highlighted the profound cognitive and psychiatric implications of lesions and surgery in this area. This literature review analyses case studies, clinical series, and systematic reviews regarding post-surgical neuropsychiatric sequelae in the posterior fossa. Relevant studies on cerebellar mutism, cerebellar cognitive affective syndrome, and emotional lability were included. Posterior fossa pathology is associated with a wide spectrum of disorders. Posterior Fossa Syndrome (PFS) is a common pediatric complication, characterised by mutism and irritability. In adults, although rarer, subtle personality changes, executive dysfunction, and severe emotional lability caused by brainstem compression can occur. Pathophysiological mechanisms involve the disruption of cerebello-cerebral circuits (diaschisis). Recognising postoperative psychiatric complications is essential. Neuropsychological assessment should be routine in posterior fossa surgery to optimise patient recovery and quality of life. This review may serve as a structured framework for future prospective and statistical studies that systematically evaluate psychiatric and cognitive outcomes after posterior fossa surgery.

Keywords
posterior fossa, cerebellum, emotional lability, cerebellar mutism, posterior fossa syndrome

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1. INTRODUCTION

The posterior fossa syndrome (PFS) is marked by a range of linguistic, cognitive, and behavioural-affective symptoms that may arise following cerebellar lesions of various causes [1]. Surgeries within the posterior fossa represent a major challenge in neurosurgery. Although historically the cerebellum was primarily associated with motor coordination, the "cerebellar revolution" of recent decades has demonstrated an intrinsic connection between posterior fossa lesions and neuropsychiatric manifestations. Studies have shown that the cerebellum modulates cognitive and affective processes through reciprocal connections with the cerebral cortex and limbic system [2].

The anatomical basis of psychiatric disorders in posterior fossa pathology lies in the connections of the cerebellum with supratentorial structures. Disruption of the balance of dopaminergic

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and serotonergic networks plays a central role. Cerebellar lesions can lead to a "deafferentation" of thalamo-limbic circuits or the interruption of cerebellar output to midbrain areas (locus coeruleus and raphe nuclei) [3]. Wickenhauser et al. emphasize the role of the dentato-thalamo-cortical tract; its injury causes cerebello-cerebral diaschisis, reducing metabolism in the contralateral frontal lobes, which explains the executive deficit and mutism [4]. This paper aims to synthesize the psychiatric effects of posterior fossa surgery, ranging from mutism and executive dysfunction to psychotic states and emotional lability.

In the pediatric population, PFS is a well-documented complication, affecting a significant percentage of children operated on for vermian tumors (especially medulloblastoma) [5]. Pollack (2001) describes this syndrome as a neurobehavioral complex including: transient mutism: Delayed onset (1-6 days postoperatively); severe emotional lability: uncontrolled crying, extreme irritability; personality changes: social withdrawal or bizarre behavior [5]. A systematic review conducted by Hanzlik et al. demonstrated that survivors present long-term neuropsychological deficits, including lowered IQ and attention problems, exacerbated by adjuvant treatment [6].

Emotional lability (pathological laughing or crying) is a disabling symptom associated with brainstem compression. In a recent case-control study, Prakash et al. (2023) showed that extra-axial tumors compressing the pons can trigger these involuntary manifestations [7]. Surgical decompression led to complete remission of symptoms in all study patients and improved quality of life [7].

Although less frequently reported than in children, posterior fossa syndrome also affects adults. Marien et al. (2013) and Wibroe et al. (2018) documented cases of transient mutism and affective disorders in adults following tumor surgery [1, 8]. Symptoms include: executive dysfunction (planning, working memory); personality changes (disinhibition or affective flattening); linguistic difficulties (agrammatism, dysprosody). Omar et al. (2014) draw attention to the difficulty of diagnosing CCAS in adults due to confounding factors such as anxiety and fatigue [9].

There is evidence linking structural posterior fossa abnormalities (e.g. Dandy-Walker cysts, megacicterna magna, tumors) to the onset of psychoses, bipolar disorders, and personality disorders, suggesting that the cerebellum is an essential modulator of mental state [3].

2. Methods

This article is a narrative literature review. We searched PubMed and Google Scholar using combinations of the terms "posterior fossa", "cerebellum", "cerebellar mutism", "posterior fossa syndrome", "cerebellar cognitive affective syndrome", "emotional lability", "pathological laughing and crying", and "brainstem" up to March 2025. We prioritised clinical case reports, case series, prospective cohorts, and systematic reviews that described psychiatric, cognitive, or behavioural outcomes after posterior fossa lesions or surgery in paediatric and adult populations. Reference lists of key articles were screened to identify additional relevant publications. Non-English papers without an English abstract and studies without clear neuropsychiatric data were excluded.

3. Psychiatric and neurobehavioral manifestations according to lesion location in the posterior fossa

For the neurosurgeon, it is clinically helpful to relate psychiatric and cognitive sequelae to the exact location of the posterior fossa lesion. The available literature indicates that midline vermian lesions, lateral hemispheric lesions, involvement of the cerebellar peduncles and dentato-thalamo-cortical pathways, intrinsic brainstem lesions, and developmental malformations of the posterior fossa are each associated with relatively characteristic neuropsychiatric profiles [1–7].

Cerebellar vermis and midline lesions

The cerebellar vermis is crucial for affect modulation and social behaviour and is the key structure implicated in posterior fossa syndrome. In children, midline tumours (especially vermian medulloblastoma) frequently lead to delayed-onset mutism, severe emotional lability and marked personality change in the early postoperative period [4,5]. Pollack describes a triad of transient mutism, severe irritability or pathological crying, and pronounced personality change or behavioural regression [5]. Adults with midline lesions may not develop a full PFS, but subtle combinations of reduced speech output, emotional instability and

personality change have been described [1,8,11]. These manifestations represent the affective–behavioural expression of cerebellar cognitive affective syndrome and can persist as long-term deficits in attention, executive function and social cognition [1,5,6].

Cerebellar hemispheric lesions

Lesions restricted to the cerebellar hemispheres, particularly the posterior lobules, more often present with the "cognitive" aspects of CCAS. Patients may show impaired planning and set-shifting, reduced verbal fluency, visuospatial disorganisation and slowed information processing, with only modest motor signs [1,2,9]. Right hemispheric lesions tend to predominantly affect language and verbal executive functions, whereas left hemispheric lesions more often impair visuospatial abilities. Personality changes are usually milder than in vermian damage, but apathy and irritability are not uncommon, and these deficits are easily overlooked without targeted questioning or formal neuropsychological testing.

Cerebellar peduncles and dentato-thalamo-cortical pathways

The cerebellar peduncles, especially the superior cerebellar peduncle and the dentato-thalamo-cortical tract, are critical for cerebello-cerebral communication. Wickenhauser et al. and other authors have shown that injury to these outflow pathways produces cerebello-cerebral diaschisis with reduced metabolism in contralateral frontal regions, explaining postoperative mutism and frontal-type executive dysfunction [4]. In paediatric series, involvement of the inferior cerebellar peduncle has been consistently associated with more severe and complex PFS phenotypes, including mutism, sleep-disordered breathing, cognitive regression and language disturbance [4,5]. Thus, peduncular damage can generate a full CCAS/PFS picture even when the cerebellar cortex appears relatively preserved.

Brainstem compression and intrinsic brainstem lesions

The brainstem, particularly the pons, plays a central role in emotional expression and arousal. Extra-axial posterior fossa tumours compressing the pons may cause pathological laughing or crying and emotional incontinence; in the series by Prakash et al., decompression led to complete remission of these symptoms and improved quality of life [7]. Intrinsic brainstem lesions can similarly produce emotional lability, rapid mood shifts and reduced initiative through disruption of corticobulbar and monoaminergic pathways [3,7]. In extensive lesions, disorders of consciousness and respiration dominate, but apparently "purely neurological" brainstem compression can be the principal driver of severe postoperative emotional dysregulation.

Developmental malformations and CSF-space abnormalities

Developmental anomalies such as Dandy-Walker malformation, megacisterna magna or large posterior fossa arachnoid cysts have been linked with major psychiatric syndromes. Pollak et al. described patients with such malformations who presented with psychotic disorders, bipolar illness or marked personality change in the absence of supratentorial lesions [3]. It is likely that longstanding distortion or hypoplasia of the vermis and cerebellar outflow pathways alters the maturation of cerebello-limbic circuits, predisposing to psychosis and mood instability. In some cases, psychiatric symptoms are the presenting complaint and posterior fossa pathology is discovered only after brain imaging.

Pediatric versus adult profiles

Although the underlying anatomy is the same, the psychiatric and cognitive sequelae of posterior fossa lesions differ in emphasis between children and adults. In children, vermian tumours and peduncular involvement most often produce the full PFS picture with mutism, striking emotional lability, behavioural regression and long-term reductions in IQ and attention [4–6]. Adjuvant treatments (radiotherapy, chemotherapy) further compound these deficits [6]. From a neurolinguistic perspective, early language acquisition depends on widely distributed cortico-subcortical networks in which the cerebellum has a modulatory role. In a recent review, Cîndea I.E. and Cîndea C. integrated neuropsychological and functional imaging data to show that language development is closely linked to the maturation of fronto-temporo-cerebellar circuits [10]. This framework helps explain why posterior fossa lesions in childhood can lead not only to transient mutism but also to persistent language and communication

disorders. In adults, posterior fossa surgery more commonly results in partial or attenuated syndromes: transient mutism or speech disturbance, subtle personality change, executive dysfunction and mood symptoms. Full-blown PFS is rare but well documented [1,8,9]. Omar et al. underline that in adults, fatigue, anxiety and premorbid psychiatric conditions can mask or mimic CCAS, making systematic cognitive and affective assessment essential [9].

Rare and atypical psychiatric presentations

Beyond the more common patterns of mutism, emotional lability and executive dysfunction, the literature also describes rarer psychiatric phenomena associated with posterior fossa pathology. Pollak et al. documented cases in which posterior fossa lesions were associated with psychotic syndromes, bipolar disorder and severe personality change, sometimes leading to an initial misdiagnosis of primary psychiatric illness [3]. Case reports outside the core series have also described manic episodes, catatonia-like states and vivid hallucinosis in the context of cerebellar or brainstem lesions. Although these presentations are uncommon, they are clinically important because psychiatric onset in mid-life with atypical features or neurological signs should prompt neuroimaging including the posterior fossa; and in neurosurgical patients, new-onset psychosis, mania or catatonia in the postoperative period may represent an extreme form of cerebellar cognitive affective dysfunction rather than a de novo primary psychiatric illness.

4. Clinical implications for neurosurgical practice

Posterior fossa surgery carries significant risks not only for motor function but also for neuropsychiatric integrity. The evidence reviewed confirms that the cerebellum and brainstem participate in distributed networks subserving cognition, affect and behaviour, and that disruption of these networks can produce a recognisable cerebellar cognitive affective syndrome in both children and adults [1–3,9]. In children, this syndrome frequently overlaps with posterior fossa syndrome, characterised by delayed mutism, severe emotional lability and behavioural regression after midline tumour resection [4–6]. In adults, similar circuits can be affected, but the clinical picture is usually more subtle, with partial mutism, personality change and executive dysfunction that are easily misattributed to nonspecific postoperative factors [1,8,9]. In line with previous analysis of surgical versus conservative management in supratentorial intracerebral hemorrhage, where neurosurgical decisions were closely tied to prognostic factors and long-term outcomes [11], similar structured decision frameworks are needed for posterior fossa lesions, explicitly integrating the risk of psychiatric and cognitive sequelae.

Anatomically, midline vermian lesions and developmental malformations of the posterior fossa are most strongly associated with affective dysregulation, personality change and, in some cases, psychotic or bipolar syndromes [3–5]. Lateral hemispheric lesions predominantly produce cognitive deficits—executive dysfunction, visuospatial impairment and language disturbance—with relatively modest motor signs [1,2,9]. Involvement of the cerebellar peduncles and dentato-thalamo-cortical pathways is a key determinant of severity, because disruption of these outflow tracts leads to cerebello-cerebral diaschisis with mutism and frontal-type deficits [3,4]. Compression or intrinsic involvement of the brainstem, especially the pons, may manifest as pathological laughing or crying and other forms of emotional incontinence, often reversible after decompression [3,7].

These findings have direct implications for neurosurgical practice. Pre-operative counselling should include specific information about potential cognitive and psychiatric sequelae, especially in paediatric patients with vermian tumours and in adults undergoing extensive midline or peduncular resections. Post-operative follow-up should incorporate simple, standardised screening for mutism, emotional lability, personality change and executive deficits. Where abnormalities are identified, referral to psychiatry, neuropsychology and rehabilitation services is essential. Paediatric series demonstrate that early recognition and structured cognitive and behavioural interventions can improve educational and adaptive outcomes in posterior fossa tumour survivors [5,6].

5. Conclusions

The cerebellum is not only a coordinator of movement but an essential modulator of the mind. Subtle postoperative changes in personality or executive function and severe emotional lability

should not be dismissed as transient or purely reactive phenomena; they often represent the clinical expression of a well-defined cerebellar syndrome. The literature reviewed, including studies on posterior fossa surgery and posterior fossa syndrome, strengthens the evidence that involvement of the cerebellar peduncles—particularly the inferior cerebellar peduncle—is consistently associated with mutism, cognitive and psychiatric disorders, sleep-disordered breathing and language impairment [3–5]. Systematically integrating lesion-location–based risk assessment, routine neuropsychiatric screening and early cognitive–behavioural rehabilitation into posterior fossa care pathways is therefore crucial to optimise not only survival but also long-term quality of life and social reintegration in these patients.

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Visual improvement following the treatment of internal carotid complex aneurysms through internal carotid ligation combined with STA-MCA bypass

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Keywords
internal carotid complex aneurysms, ophthalmic artery, carotid ligation, superficial temporal artery to middle cerebral artery anastomosis

ABSTRACT

Objective: The combination of internal carotid artery (ICA) ligation with superficial temporal artery (STA) to middle cerebral artery (MCA) bypass has proven effective in the treatment of internal complex carotid aneurysms (ICCAs). Indirect exclusion through thrombosis is anticipated to alleviate the mass effect on adjacent neurovascular structures. However, visual impairment may occur secondary to thrombosis of the ICA and ophthalmic artery. The purpose of this study is to emphasise the paradoxical stabilisation and improvement of vision observed in certain cases.

Methods: A retrospective study was conducted from 2016 to 2020, analysing data from a cohort of patients with ICCAs, among whom nine patients underwent ICA ligation in combination with STA-MCA anastomosis. Preoperative and postoperative visual findings were analysed and compared.

Results: All patients underwent STA-MCA bypass in addition to complete ICA ligation. Postoperative clinical outcomes remained unchanged in 8 out of 9 patients. Complete aneurysm exclusion through thrombosis was achieved at 12 months. The STA-MCA anastomosis remained patent in 6 out of 9 patients, with no clinical deterioration observed, except in one case. Notably, irrespective of graft patency, this patient experienced a minor parietal stroke, from which full recovery was achieved after six months. Furthermore, an improvement in visual acuity was observed in two patients at three months.

Conclusion: The ICCA treatment, which involves ICA ligation combined with STA-MCA bypass, alleviates the mass effect on adjacent neurovascular structures. Despite the occurrence of ICA thrombosis, an enhancement in visual acuity has been documented, which can be attributed to the patency of the ophthalmic artery via the meningeal anastomotic circuits.

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Introduction

Visual impairment caused by Internal Carotid Complex Aneurysms (ICCAs) stems from various factors. The ipsilateral optic nerve, oculomotor, trochlear, and abducens nerves are affected along their course by either the mass effect or the Water Hammer phenomenon of large aneurysms. Additionally, the substantial volume of blood within giant aneurysms generates a whirling and swirling stream, leading to a steal effect (1). Consequently, hypoperfusion of the ophthalmic artery results in dysfunction of the optic nerve.The only efficacious treatment involves an indirect exclusion through thrombosis, achieved via a combination of internal carotid artery (ICA) ligation and superficial temporal artery-middle cerebral artery (STA-MCA) bypass. Theoretically, management and exclusion of ICCAs alleviate the compression on the involved nerves.

Gradual thrombus retraction induces the involution of giant aneurysms through progressive fibrosis and reduction of mass effect (2). Carotid ligation facilitates thrombosis of all segments of the internal carotid artery, from cervical ligation to the bifurcation. Moreover, the ophthalmic artery may also undergo complete thrombosis.

In this report, we present cases of visual improvement observed in 2 patients out of 9 who were operated on for ICCAs at our institution between 2016 and 2019. The management involved carotid ligation combined with STA-MCA bypass, with comparative imaging assessments of the aneurysm, internal carotid artery, and ophthalmic artery pre- and post-operatively. The visual outcomes are documented and analyzed within this publication.

Materials and Methods

Patients

We conducted a retrospective analysis of clinical and imaging data from September 2016 to December 2019 for patients treated at our institution. The study included only those patients with intracranial cavernous carotid aneurysms (ICCAs) presenting with symptoms such as cranial nerve palsy or persistent headaches unrelieved by conventional analgesics. Asymptomatic patients or those with episodic headaches were not immediate candidates for this intervention; instead, they were referred for endovascular assessment and treatment abroad. Furthermore, patients in a coma with ruptured ICCAs in the intensive care unit are not managed using the proposed approach procedure.

Preoperative Evaluation

A clinical examination, encompassing neurologic and ophthalmologic assessments, is conducted during the initial visit. The modified Rankin scale is also evaluated**.** Epidemiologic data and imaging findings—including side, size, and location on the internal carotid artery—are documented in **Table 1**.

Table 1. The Baseline Data of Patients with Complex Internal Carotid Aneurysms

Patient Number	Age (years)	Sex	Side	Size (mm)	Symptoms	Location
1	36	Male	Right	37,81	Headaches, seizure, hallucination, visual field deficit	Paraclinoi
2	20	Female	Right	28,56	Headaches, homolateral visual acuity decrease	Paraclinoi
3	19	Female	Right	45,83	Headaches, Bilateral visual loss	Paraclinoi
4	68	Female	Left	18.63	Headaches, blurred vision, decrease in visual acuity	Intracaverno
5	30	Male	Left	25,70	Headaches, homolateral visual loss, 3rd nerve palsy	Paraclinoi
6	65	Female	Right	44,20	Cavernous sinus syndrome	Transitiona
7	45	Female	Right	22.10	Headaches, 3rd nerve palsy,	Intracaverno
8	54	Female	Left	16.10	Headaches, left eye blurred vision.	Paraclinoi
9	48	Male	Left	23,52	Headaches, homolateral visual loss	Transitiona

Decision Making

In the absence of balloon test occlusion within the country, we evaluated the patients based on their Willis Polygon Score (WPS) as described by L. Houari et al. (3). The decision-making organizational chart is outlined in Image 1, where WPS 7 indicates the number of missing segments in the Willis polygons.

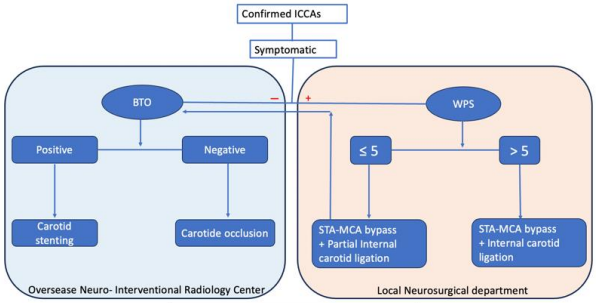

Image 1: Diagram showing the decision making.

Operative Procedure

The patient is prepared and positioned in accordance with standard procedures, with access established to the cervical region on the side of the aneurysm. The carotid bifurcation is meticulously dissected, and the ligation at the origin of the internal carotid artery (ICA) is performed using either a secure double knot spaced one centimeter apart or a single knot occluding approximately three-quarters of the carotid lumen (a knot designed to permit passage of the hook's tip), employing a non-resorbable suture.

A superficial temporal artery is dissected, harvested, and clipped distally to a length of 10 cm. The extremity is prepared for the anastomosis, a temporo-parietal bone flap is removed, the dura mater is opened, and an M3 artery is selected after the dissection of the outer part of the sylvian fissure. A temporal clip is applied, and a termino-lateral anastomosis is performed with silk sutures.

In the absence of indocyanine green, the anastomosis is assessed using surgical techniques that involve gently occluding the proximal and distal segments of the M3 artery while observing arterial pulsation.

The entire procedure is conducted under normotensive conditions, with a slight elevation observed during the temporary clipping phase of the anastomosis.

The dural closure initially begins at the frontal aspect and then progresses to the temporal region, using a pedicle layer of temporal muscle to ensure a secure entry of the STA without undue compression.

In addition to the preoperative measures, saline-diluted heparin and calcium inhibitors are administered at the anastomosis site. In the intensive care unit, the patient receives an intravenous salicylic acid following a three-hour computed tomography scan.

Results

A total of nine patients underwent surgical intervention for ICCAs during the study period. All patients received internal carotid artery (ICA) ligation combined with superficial temporal artery (STA)-middle cerebral artery (MCA) anastomosis. The cases are sequentially numbered from 1 to 9, each exhibiting distinct characteristics detailed in Table 1. Epidemiological data analysis indicated a predominance of female patients, with an average age of 43 years. Preoperative visual deficits were observed in seven out of nine patients, third nerve palsy in three patients, and one patient presented with seizures.

The aneurysm sizes ranged from 16.10 mm to 45.83 mm, with an average size of 29.16 mm. Five of the nine aneurysms were classified as giant and were primarily located in the paraclinoid segment of the internal carotid artery.

In addition to the STA-MCA anastomosis, all patients underwent ICA ligation at its cervical origin.

1. The surgical outcomes:

The evaluation of surgical outcomes employing Angio CT and Angio MR across multiple sequences is documented in Table 2, providing a comprehensive assessment of aneurysm morphometry, including thrombosed regions. Both the aneurysm and the internal carotid artery (ICA) were examined at 6 and 12 months using steady-state magnetic resonance imaging.

Table 2: Surgical outcomes

Patient Number	Anastomosis Patency (Angio CT)			Aneurysm Thrombosis (Angio	
	3 hours after surgery	1 month	3 months	6 months	12 mor
1	Yes	Yes	Yes	80%	100%
2	Yes	Yes	Yes	100%	100%
3	No	No	No	80%	100%
4	Yes	Yes	Yes	100%	100%
5	No	No	No	100%	100%
6	Yes	Yes	Yes	100%	100%
7	Yes	Yes	Yes	100%	100%
8	Yes	Yes	Yes	100%	100%
9	Yes	No	No	100%	100%

The aneurysm exclusion via thrombosis is achieved in all cases, with 7 out of 9 patients attaining 100% exclusion and the remaining 2 patients achieving 80% at six months; by twelve months, the exclusion rate reaches 100%. Postoperative assessment of the STA-MCA anastomosis was conducted at three hours, one month, and three months following surgery. The anastomosis remained patent in 7 out of 9 patients and was non-patent in 2 after three hours. Notably, in one patient, the anastomosis transitioned from being patent to thrombosed at one-month post-surgery.

2. Clinical outcomes:

The assessment of clinical outcomes in comparison to the preoperative status is documented in **Table 3**. The modified Rankin scale indicates stabilization across all patients. Ophthalmologic evaluations remained stable, with no alterations observed in the visual field; furthermore, an improvement in visual acuity was noted in two cases at the three-month follow-up.

Table 3: Clinical preoperative / post-operative comparison

Patient Number	Modified Rankin scale			Ophthalmic evaluation				
	Admission	Discharge	3 Months	Visual filed		Acuity at the side of the aneurysm		
				Preop	3 months	preop	3 months	12 months
1	1	1	1	HH	HH	18/20	18/20	18/20
2	1	1	1	N	N	6/20	10/20	10/20
3	3	3	3	BB	BB	0/20	0/20	0/20
4	1	1	1	N	N	10/20	16/20	16/20
5	1	1	1	MB	MB	0/20	0/20	0/20
6	1	1	1	MB	MB	0/20	0/20	0/20
7	1	1	1	N	N	6/20	6/20	6/20
8	1	1	1	N	N	16/20	16/20	16/20
9	1	1	1	N	N	4/20	4/20	4/20

N: normal, **HH:** homonymous Hemianopsia, **BB:** Binocular Blindness, **MB:** Monocular Blindness.

A patient is reported to have a lateral homonymous hemianopsia with preserved visual acuity. This clinical condition is attributed to ipsilateral optic tract compression caused by the aneurysm (see Images 2, 3, 4). Heme MRI sequences reveal hemosiderin deposition and parenchymal destruction of the optic tract, indicated by an arrow in Image 3. The patient's visual impairment persists unchanged following surgical intervention.

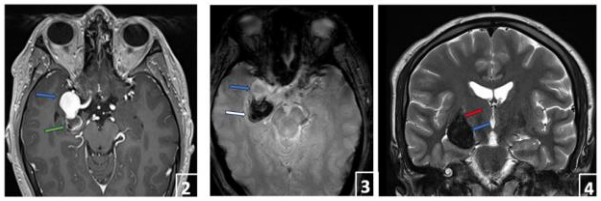

Image 2: Axial T1-weighted enhanced brain MRI showing a prebifurcaion right internal carotid aneurysm (blue arrow) and a posterior pseudo-aneurysmal sac partially thrombosed, compressing the optic tract (green arrow).

Image 3: axial T2* weighted gradient echo MRI, shows the aneurysm (blue arrow) and the hemosiderin deposition in the parenchyma corresponding to the optic tract (white arrow), which is a sign of previous aneurysm bleeding.

Image 4: Coronal T2-weighted MRI showing an aneurysm in the ambient cistern measuring 25 mm on this axis (blue arrow). The superior part of the aneurysm compresses the optic tract (red arrow).

Binocular blindness was observed in a single patient, attributable to the mass effect exerted by the large aneurysms on the sellar and suprasellar regions (see blue arrows in images 5, 6, and 7).

The optic chiasm is displaced anteriorly and appears laminated vertically as a thin layer by the anterior segment of the aneurysm (indicated by the yellow arrow in image 5). Notably, this patient's visual impairment persisted postoperatively.

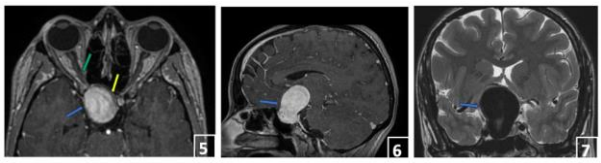

Image 4: Axial T1-weighted enhanced brain MRI showing a giant right internal carotid aneurysm (blue arrow) measuring 45,83 mm at its greatest axis, the optic nerves are normal, the chiasm is stretched anteriorly (yellow arrow) by the aneurysm.

Image 5: Sagittal T1-weighted enhanced brain MRI showing a giant right internal carotid aneurysm (blue arrow) enlarging and eroding the anterior wall and the floor of the sella turcica.

Image 6: Coronal T2-weighted MRI showing a giant internal carotid aneurysm (blue arrow). The superior part of the aneurysm raises the floor of the third ventricle.

3. Illustrative case

A 45-year-old woman with a history of embolization of a paraclinoid internal carotid aneurysm five years prior presents with persistent headaches and a left eye visual acuity of 10/20. Cerebral angiography revealed revascularization of a left paraclinoid aneurysm, characterized by coiling material at the base of an enlarged fusiform aneurysm (purple arrow, Image A), measuring 16 x 11 mm. The patient underwent a homolateral Hunterian internal carotid artery ligation and superficial temporal artery middle cerebral artery (STA-MCA) bypass. Postoperative assessment indicated stability, with a reduction in headache severity and improvement in visual acuity from 10/20 to 16/20 at three months. Follow-up imaging, including Angio MRI at three months, confirmed aneurysm thrombosis and patency of the anastomosis (red arrow, Image B). The internal carotid artery appears completely thrombosed, while the ophthalmic artery remains patent (blue arrow, Image C), as indicated by the yellow arrow.

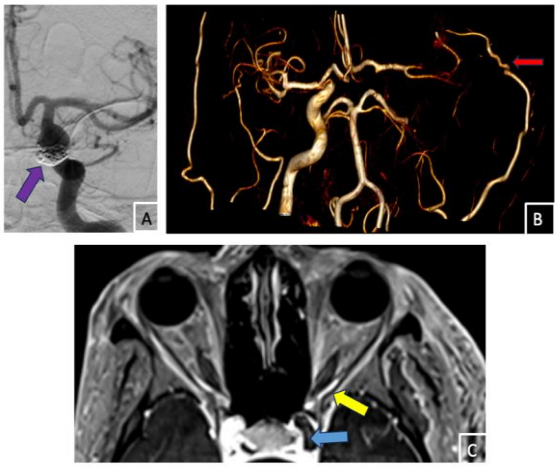

Image A: Cerebral angiography illustrates the revascularization of a left paraclinoid aneurysm, with coiling material positioned at the base (indicated by the purple arrow) of an enlarged fusiform aneurysm.

Image B: The anteroinferior three-dimensional reconstruction view derived from the Angio MR reveals the exclusion of the aneurysm and the internal carotid artery from the circulatory system. Additionally, the patency of the anastomosis and the dilation of the recipient artery are evident (indicated by the red arrow) at the three-month follow-up.

Image C: Axial enhanced T1 MRI, illustrating complete carotid thrombosis (blue arrow) with the ophthalmic artery remaining patent (yellow arrow).

Discussion

The morphology and hemodynamic alterations associated with intracranial carotid cavernous aneurysms (ICCAs) exert varying effects on the adjacent neurovascular structures, resulting in visual impairment. The reported recovery rates for visual disturbances caused by aneurysm compression range from 50% to 65% (4,5), whereas the recovery rate for external ophthalmoplegia is approximately 70% (6). Limited literature addresses these differential recovery rates. Matano et al. (7) elucidated that the disparity is attributable to differences in embryological origin and development between the optic nerve and other cranial nerves, leading to distinct responses to compression.

These discrepancies can be elucidated by considering two types of visual impairment. The first pertains to intra- and extraocular motility deficits, predominantly caused by the mass effect exerted on the oculomotor, trochlear, and abducens nerves, which are primarily situated within the cavernous sinus. The primary manifestation is ophthalmoplegia.

The second type involves impairment of the optic nerve itself. It is posited that, in addition to the mass effect, hemodynamic alterations—such as hypoperfusion resulting from a steal phenomenon observed during angiographic dye injection and the water hammer effect transmitting shock waves to the optic nerve during each systole—contribute to the pathology.

From a theoretical standpoint, the surgical exclusion of ICCAs diminishes their influence on neural structures. Practically, indirect exclusion through thrombosis, achieved via internal carotid artery (ICA) ligation combined with superficial temporal artery-middle cerebral artery (STA-MCA) bypass, is regarded as the most effective intervention. Carotid ligation promotes thrombus formation, thereby reducing the risk of further neural compromise and restoring vascular stability, particularly along the internal carotid artery, from the cervical ligature to the bifurcation, including the origin of the ophthalmic artery. Strother et al. (2) observed that gradual thrombus retraction causes aneurysm involution through progressive fibrosis and reduction of the mass effect. Matano et al. (7) reported that visual acuity correlates with a poorer prognosis following treatment of giant paraclinoid aneurysms than with ophthalmoplegia.

In our cohort of nine patients undergoing surgical treatment for ICCAs via ICA ligation and STA-MCA bypass, we observed stabilization of visual status. Notably, an improvement in visual acuity was documented in two patients despite complete ICA thrombosis and a paradoxically patent ophthalmic artery (see image C of the illustrative case).

This outcome can be attributed to the chronic hypoperfusion induced by the steal effect, which facilitated the effectiveness of the anastomotic circuits. Specifically, there are three known anastomoses connecting the ophthalmic artery (OA) and the middle meningeal artery (MMA) (7,8): the recurrent meningeal branch, the anterior falx artery, and the anastomotic branch with MMA, all of which sustain flow in the ophthalmic artery in the event of ICA thrombosis. In such cases, these anastomoses enlarge and become the primary sources of blood supply to the orbital region and optic nerve.

A limitation of our study is the absence of angiographic investigation into the anatomical variations of the ophthalmic artery, its branches, and the associated anastomotic circuits. Furthermore, due to the stabilization of visual fields at three months post-treatment, no repeat angiographic assessments were performed at the twelve-month mark.

Conclusions

ICA ligation with STA MCA bypass is an effective treatment of ICCAs; the exclusion of the aneurysms by thrombosis relieves the extra and intraocular motor nerves as well as the optic nerve. Despite ICA thrombosis, ophthalmic artery flow, according to anatomic variation, is secured by anastomotic circuits.

Abbreviations

ICCAs: Internal Carotid Complex Aneurysms, **ICA**: Internal Carotid Artery, **STA**: Superficial Temporal Artery, **MCA**: Middle Cerebral Artery, **ICU**: Intensive Care Unit, **WPS**: Willis Polygon Score, **BTO**: Balloon test Occlusion, **MRI**: Magnetic Resonance Imaging, **CT**: Computerized Tomography, **3D**: Tridimensional, **IV**: Intravenous, **mRS**: modified Rankin Scale, **A1**: Precommunicant segment of the anterior cerebral artery, **M3**: Branches of the middle cerebral artery located in the sylvian fissure. **N:** normal, **HH:** homonymous Hemianopsia, **BB:**

Binocular Blindness, **MB:** Monocular Blindness, **Preop:** preoperative status, **OA:** Ophthalmic artery, **MMA:** Middle meningeal artery.

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Hybrid embolization and radiosurgery for glossopharyngeal glomus tumour. A successful case report

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Keywords
glomus, neurosurgery, embolization, radiosurgery, glossopharyngeal nerve

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Abstract

Introduction: Tumours of the glossopharyngeal nerve are extremely rare, posing a challenge in diagnosis and treatment. These tumours can cause a variety of neurological symptoms, including sore throat, numbness of the tongue, decreased sensitivity, and changes in taste, difficulty swallowing, and dizziness. Due to their rarity, there is no standardised treatment protocol, and the approach is often tailored to the individual patient.

Aim: To present a rare case of glomus tumour of the glossopharyngeal nerve, and that the hybrid method of embolisation and radiosurgery proved to be a successful treatment.

Case description: We present the case of a 31-year-old female patient who came to the Emergency Centre due to excruciating headaches, vomiting and loss of taste and

tingling in the tongue. After a detailed neurological examination and MRI diagnostics, a glossopharyngeal nerve tumour was discovered in the jugular foramen region. Due to the tumour's proximity to vital structures, surgical resection would have been high-risk. Therefore, it was decided to apply a hybrid approach, combining embolisation and radiosurgery. MRI scans showed a decrease in the size of the tumour. The patient was monitored regularly, and the results remained stable.

Conclusion: Hybrid treatment with embolisation and radiosurgery represents an effective option for the treatment of rare tumours of the glossopharyngeal nerve. This combination allows for the reduction of symptoms, reduction of tumour size, and improvement in patients' quality of life. Further research is needed to determine the optimal treatment protocols for these rare tumours.

Introduction

Tumors of the glossopharyngeal nerve are extremely rare, posing a challenge in diagnosis and treatment. These tumors can cause a variety of neurological symptoms, including sore throat, numbness of the tongue, decreased sensitivity, and changes in taste, difficulty swallowing, and dizziness. Due to their rarity, there is no standardized treatment protocol, and the approach is often tailored to the individual patient.

Case report

A 31-year-old female patient presented to the Emergency Center in March 2024 due to complaints of left-sided pulsating headache, nausea, and vomiting. Regarding previous headache episodes, the patient stated that she had experienced them before, but they were mostly correlated with her menstrual cycle. However, the headache she felt this time was extremely severe and unbearable. The patient mentioned that she had taken analgesics, but vomited them up. During the patient's examination, a blood pressure of 133/80mmHg was recorded, with a heart frequency rate of 90/min. Based on the neurological examination, it was noted that she complained of numbness of the tongue on the left side and decreased sensitivity.

The patient was then subjected to a CT diagnosis of the endocranium (Figure 1). It was found that in the left lateral cerebellomedullary cistern and cerebellopontine cistern, an irregular, lobulated, heterodense expansive lesion of approximate dimensions 25x16x29 mm was observed, propagating from the left jugular foramen, remodeling (eroding) its walls, with heterogeneous post-contrast opacification. Differential diagnosis suggested either a centrally cystic degenerated lesion or a necrotic lesion with accompanying compressive effect on the ipsilateral aspect of the medulla and cerebellum, in contact with the medial side of the left internal jugular vein. The radiological finding raised suspicion of a jugular paraganglioma, schwannoma, or meningioma. Also, at the level and partly below the left aspect of the foramen magnum, along the dura mater, a smaller expansive lesion was seen, encasing 3/4 of the circumference of the left vertebral artery like a "muff," measuring 11x14 mm, appearing separate without clear continuity with the previously described lesion, which could differentially represent either a meningioma or an exophytic component of the previously described lesion. Also, on Figure 2., the 3D VR reconstruction is displaying vascular structures of the skull base. Note the two masses located left and superior of the foramen magnum, connected by a thin vascular bridge.

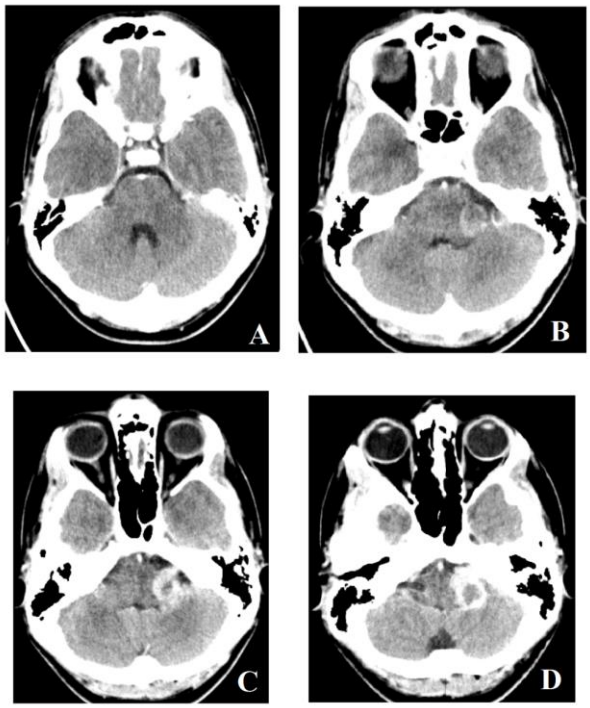

Figure 1.

After complete diagnosis and preparation, the patient was referred from the Emergency Center for further in-hospital treatment to the Clinic of Neurosurgery. During that time, from March 28, 2024. until April 12, 2024., an MRI of the

endocranium was performed (Figure 3.), which showed a tumor in the area of the jugular foramen on the left with an associated minor change below a mixed (solid-cystic) change in the left lateral cerebellomedullary and cerebellopontine cisterns, with areas of necrosis/cystic degeneration and in contact with the medial side of the left v. jugularis internae. The described strand propagates caudally under the foramen magnum, anterolaterally to the left in relation to the medulla, with which it is in contact, and that both changes are connected by an isthmus. On Figure 3, high-resolution, contiguous, thin-section coronal and sagittal MRI reconstructions are shown, displaying both masses and the bridge connecting them. Contrast-enhanced MP-RAGE MRI reconstruction shows enhancement patterns of the described masses.

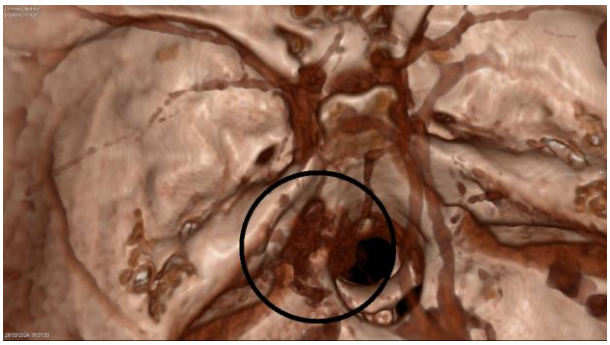

Figure 2.

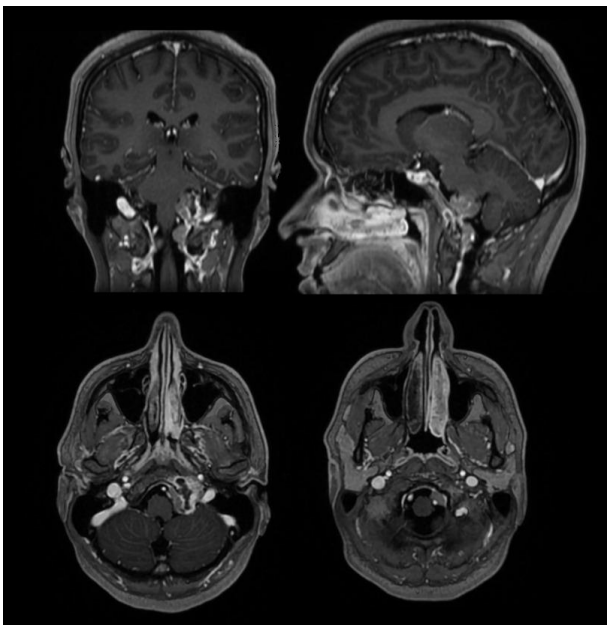

Figure 3.

As part of the preoperative preparation, the patient was examined by an endocrinologist. Normal findings were established for chromogranin A, and urinary catecholamines; there were no signs of secretory activity of the observed tumor lesion indicative of a glomus jugulare tumor. She was then examined by an otorhinolaryngologist who, based on audiometric diagnostics, found mild sensorineural hearing loss on the right side at 250, 4000, and 8000Hz, and mild to moderate sensorineural hearing loss on the left side at 4 and 8kHz.

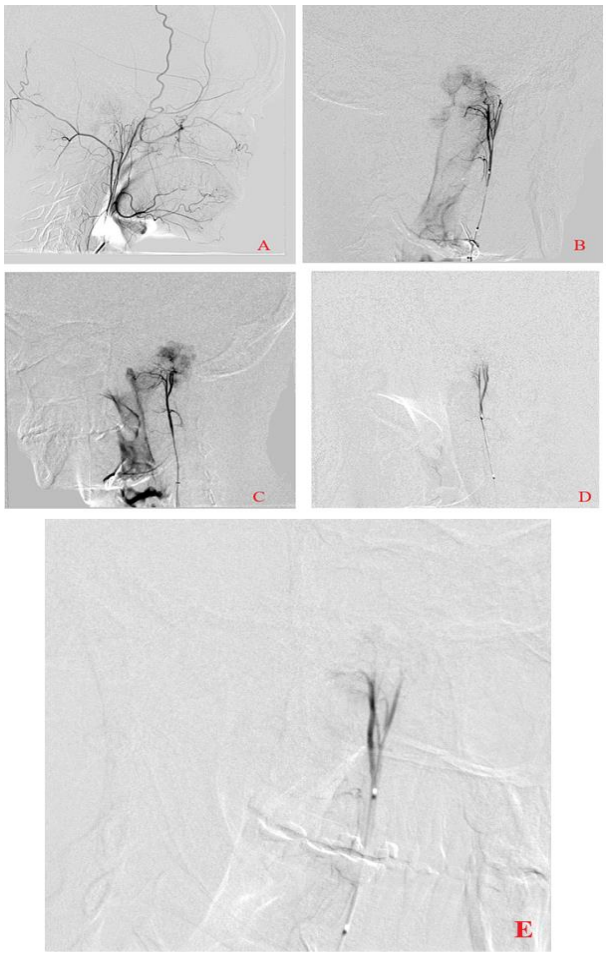

Figure 4.

The angiography of the blood vessels of the head and neck (Figure 4) - on the angiograms of both carotid and posterior basins from both vertebral arteries in the projection of the left pontocerebellar angle, a zone of pathological opacification is shown, which is vascularized from the branches of the ascending pharyngeal artery on the left side. After that, endovascular treatment was performed - the

pre-procedural angiograms from the left external carotid artery (ACE) show a zone of pathological opacification vascularized from the branches of the left AphA, then a microcatheter is used to access the feeding arteries and the particle embolization agent PVA Contour (150-250 μm) is applied, which devascularizes the tumor. She was then examined by a neurosurgeon in this department and presented to the Gamma Knife Council. The conclusion was that the patient needs to do an endocrinological examination in terms of additional evidence of a glomus jugular tumor.

Figure 4 shows a profile image of the carotid artery before the intervention (A), then the working position of the eastern pharyngeal artery (AphA) before the intervention, where predominantly marginal tumor opacification is visible (B, C), then the working position of the eastern pharyngeal artery (AphA) after the intervention (D, E). Figure 5. shows a distal profile image of the left external carotid artery (ACE) after embolization.

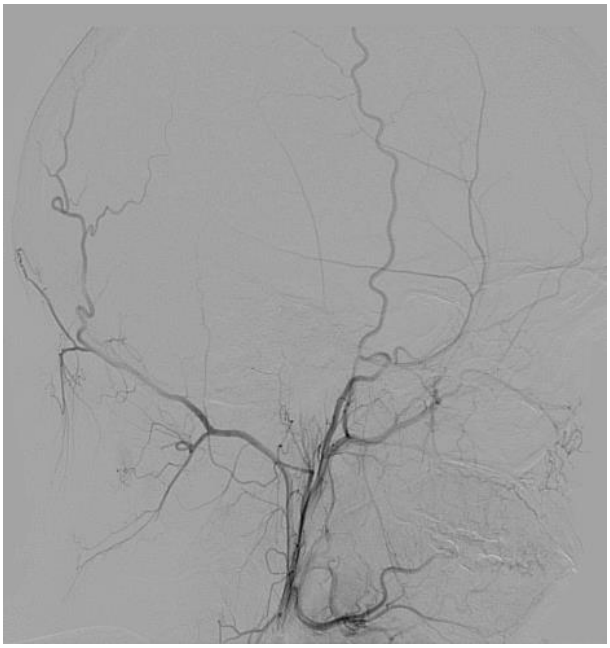

Figure 5.

After standard preparation, the patient underwent radiosurgical intervention in May 2024. A Leksell stereotactic frame was placed under local anesthesia. A pre-procedural MRI of the endocranium was performed according to protocol. Multiple radiation "shots" were delivered using different collimators. A dose of 14Gy at the 50% isodose was applied to the (T1) embolized tumor in the region of the left jugular foramen. The patient tolerated the procedure well.

Following the procedure, the patient was discharged for further home care in unchanged general and neurological condition compared to admission. The patient was regularly monitored and controlled, and no recurrence of the pathological change was recorded.

Discussion

Glomus tumors (paragangliomas) are highly vascular neuroendocrine neoplasms arising from the paraganglionic system. These tumors most commonly originate from the carotid body, jugular bulb, vagus nerve, and tympanic plexus. However, primary involvement of the ascending pharyngeal artery (APhA) is exceptionally rare (1). Even more uncommon is a tumor spanning multiple cisternal compartments while extending inferiorly beyond the cranium, and are considered complex cases (2).

Morphology

MRI reveals an expansile lesion in the left cerebellomedullary and cerebellopontine cisterns, with a second lesion of similar imaging characteristics just below the foramen magnum, anterolateral left to the medulla oblongata. The two lesions appear connected via an isthmus passing through the foramen magnum.

Both lesions exert a moderate mass effect, compressing the anterolateral part of the pons and left cerebellar peduncles while slightly remodeling adjacent bone. The superior lesion borders the medial border of the left internal jugular vein, whereas the inferior lesion encases approximately 75% of the left vertebral artery circumference.

MRI Characteristics

On T1-weighted imaging, both lesions are slightly hypointense relative to normal parenchyma, with punctate hyperintensities suggestive of flow voids, indicative of hypervascularity. Marked susceptibility on SWI and intense enhancement on post-contrast T1-weighted images further emphasize their vascular nature. On T2W/FLAIR, the lesions are moderately hyperintense. DWI demonstrates focal high signal areas, but the ADC map reveals no significant diffusion restriction—consistent with

either high cellularity or rich vascularity, both characteristic of paragangliomas. (3, 6)

Special Characteristics

The isthmus connecting the two lesions raises the possibility of either a single, extensive tumor following neurovascular structures or multiple lesions bridged by tumor tissue. A multifocal paraganglioma may appear contiguous, particularly in patients with hereditary paraganglioma syndromes (10). Additionally, a vascularized fibrous band rather than tumor tissue cannot be entirely excluded.

Given the hypervascular nature of these tumors, it is plausible that a single, expansive glomus tumor could arise from the APhA and extend superiorly. As a key feeder of skull base structures, the APhA may serve as a vascular conduit for tumor infiltration into adjacent compartments.

Interventional Radiology Findings

The patient underwent cerebral digital subtraction angiography (DSA), which demonstrated pathological opacification in the left cerebellopontine (CP) angle. The lesion was found to be vascularized by the ascending pharyngeal artery (APhA), confirming the primary feeder for the tumor. Following this, the patient underwent successful embolization, which effectively devascularized the tumor, reducing its vascularity and improving the management outlook. (7)

Gamma Knife Radiosurgery and Endocrine Consultation

In light of the clinical and radiologic findings, including the hypervascularity and the anatomical location, Gamma Knife radiosurgery was suggested as a possible treatment option. This approach is particularly beneficial for tumors located in difficult-to-access areas like the skull base, where surgical resection might carry significant risks. The decision to consider radiosurgery underscores the high vascularity of the lesion and the potential for targeted, minimally invasive treatment. (8)

Additionally, an endocrine consultation was requested due to the suspicion that this could represent a glomus jugulare tumor, a well-known form of paraganglioma that often presents in similar anatomical locations. Endocrinologists may assess for hereditary syndromes or metabolic abnormalities associated with paragangliomas, which could influence long-term management and follow-up care. (9)

Conclusion

The lesion's intense vascularity, flow voids, and enhancement pattern, combined with its anatomical location and bone remodeling rather than destruction, make an atypical paraganglioma the most plausible diagnosis. The isthmus-like structure linking the two lesions further supports a contiguous growth pattern, distinguishing it from other neoplasms. The tumor's likely origin from the ascending pharyngeal artery, a known vascular feeder of skull base structures, aligns with the expected behavior of a paraganglioma rather than any of the alternative diagnoses discussed. The findings from cerebral DSA, which demonstrated vascularization from the APhA, and the successful embolization to devascularize the tumor further confirm the diagnosis. The consideration of Gamma Knife radiosurgery, alongside endocrine consultation, underscores the complex nature of this case and the need for a multidisciplinary approach in managing such a rare and highly vascular tumor.

Schwannoma (e.g., Jugular Foramen or Vagal Schwannoma)	Schwannomas may arise along cranial nerves and exhibit moderate enhancement but typically follow a nerve's trajectory and do not demonstrate significant vascularity. In contrast, this lesion displays marked hypervascularity with flow voids on MRI and susceptibility artifacts on SWI, features strongly suggestive of paraganglioma. Additionally, schwannomas are well-circumscribed and lack the infiltrative growth pattern or isthmus-like connection seen in this case.
Meningioma (Skull Base Meningioma)	The absence of dural attachment, calcifications, or a "dural tail" sign argues against meningioma. Meningiomas typically cause hyperostosis rather than bone remodeling and lack the characteristic flow voids and intense vascularity observed here. The lesion's involvement of neurovascular structures and its continuity across the foramen magnum further support a diagnosis of paraganglioma over meningioma.
Metastatic Disease	Hypervascular metastases (e.g., from renal cell carcinoma or thyroid carcinoma) could theoretically mimic a paraganglioma's imaging characteristics. However, metastases often present as multiple, poorly circumscribed masses with aggressive bone destruction rather than the remodeling seen here. Additionally, the lack of a known primary malignancy and the tumor's contiguous growth through the foramen magnum favor a primary skull base neoplasm over metastasis.
Hemangioblastoma	Hemangioblastomas are highly vascular and may demonstrate flow voids but are more commonly found in the cerebellum and often present with a cystic component and an enhancing mural nodule, features absent in this case. Furthermore, the lack of association with von Hippel-Lindau disease and the lesion's extra-axial location make hemangioblastoma less likely.
Chordoma	Chordomas typically arise from the clivus or sacrum, presenting as destructive, lobulated masses with high T2 signal intensity. However, they generally do not exhibit the extensive flow voids and intense enhancement characteristic of paragangliomas. Unlike chordomas, the present lesion demonstrates vascular remodeling rather than aggressive bone invasion.
Aneurysm or Vascular Malformation	Aneurysms and vascular malformations can mimic hypervascular tumors, but they lack a solid parenchymal component on post-contrast MRI. Additionally, the isthmus-like connection and progressive expansion into adjacent spaces are inconsistent with a purely vascular anomaly. While MRA/CTA would be necessary to definitively exclude a vascular malformation, the imaging characteristics strongly favor a neoplasm.

Figure 6. Differential Diagnosis and Comparison with Present Case (4, 5).

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Gross total resection of a rare epidermoid cyst in the supplementary motor area without postoperative SMA syndrome. A case report and literature review

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Keywords
epidermoid cyst, supplementary motor area, microsurgery, SMA syndrome, neuronavigation

Abstract

Epidermoid cysts rarely occur in the supplementary motor area (SMA), where resection carries a particular risk of postoperative SMA syndrome. We present the case of a 25-year-old woman who developed persistent headache and intermittent dizziness. Preoperative MRI revealed a right medial frontal lesion with low T1, high T2, and restricted diffusion, consistent with an epidermoid cyst. Microsurgical resection was performed via a right interhemispheric approach under neuronavigation. Internal decompression was followed by capsule dissection along the tumor–brain interface. Particular care was taken to preserve bridging veins, avoid fixed retractors, and minimise traction on the SMA cortex. A superficial cortical vein with focal wall irregularity was reinforced with hemostatic material instead of being sacrificed. Gross total removal was achieved with minimal blood loss.

The patient recovered without neurological deficits, was mobilised on the third postoperative day, and was discharged home. Histopathology confirmed the diagnosis. Follow-up MRI at six months showed no recurrence, and her modified Rankin Scale score was 0.

This case highlights that, although SMA epidermoid cysts are exceedingly rare, complete resection with full functional preservation is achievable. Careful surgical planning, venous preservation, and low-traction microsurgical techniques are crucial to avoid SMA syndrome in this eloquent cortical region.

Introduction

Epidermoid cysts are rare congenital lesions of ectodermal origin, representing only 0.2-1.8% of all intracranial tumors[1].They arise from ectodermal inclusions during neural tube closure[2] and are usually located in the cerebellopontine angle, sellar region, or fourth ventricle. In contrast, their occurrence in the supplementary motor area (SMA) is exceptionally unusual[1,3].

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The SMA occupies the medial aspect of the superior frontal gyrus (Brodmann area 6) and plays an important role in motor planning, initiation of voluntary movement, and speech activation within the dominant hemisphere[4,5].Surgical intervention in this particular region is occasionally faced with the specific risk of SMA syndrome, characterized by transient contralateral weakness, mutism, urinary incontinence, and apathy. While recovery usually occurs within days to weeks, these deficits can be distressing for both patients and surgeons[6,7]. Thus, resection in this eloquent area requires careful balancing of tumor clearance with functional preservation.

Here, we describe the management of a young woman with an epidermoid cyst in the SMA.

Using a right interhemispheric approach guided by neuronavigation, gross total resection was achieved without postoperative SMA syndrome.We also review the relevant literature on SMA and parasagittal lesions to place our surgical strategy and outcome in a broader clinical context.

Description

History and Examination

A 25-year-old right-handed female surgical nurse presented with a 2-week history of persistent headache accompanied by intermittent dizziness. She denied nausea, vomiting, motor weakness, dysarthria, or sphincter disturbances. On admission, her neurological examination was normal. Her past medical history was notable only for a previous cesarean section.

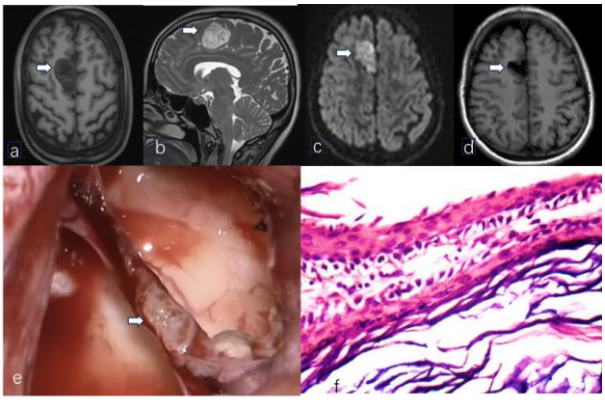

Figure 1 .a. Axial T1-weighted MRI showing hypointensity. **b.** Sagital T2-weighted MRI showing hyperintensity **c.** Diffusion-weighted imaging (DWI) demonstrating marked hyperintensity, consistent with an epidermoid cyst. **d.** Axial postoperative MRI confirming gross total resection of the lesion. **e.** After tumor removal, superficial cortical vein showed thinning and focal irregularity of the wall. **f.** Histopathological examination (hematoxylin–eosin stain, original magnification ×200) showing keratinized stratified squamous epithelium, consistent with an epidermoid cyst

Imaging

Magnetic resonance imaging (MRI) demonstrated a well-circumscribed lesion in the right supplementary motor area (SMA). The mass appeared hypointense on T1-weighted sequences, hyperintense on T2, and showed marked restricted diffusion on diffusion-weighted imaging (DWI). Computed tomography (CT) revealed a hypodense lesion without contrast enhancement. These radiological features were consistent with an epidermoid cyst. (Figure a.b.c)

Surgical Technique

Surgery was performed under general anesthesia, patient was placed positioned supine and the head secured using Mayfield clamp. Neuronavigation was used to localize the lesion and guide the craniotomy. A small right frontal parasagittal bone flap was used to expose the superior sagittal sinus. After the opening of the dural, the interhemispheric fissure was carefully dissected under high magnification, then particular attention was paid to preserve the bridging veins. Sharp dissection of arachnoid adhesions provided access to the lesion.

The removal of the tumor was started with internal decompression, followed by capsule dissection along the tumor-brain interface. Sharp dissection was used to minimize traction on the SMA cortex. Bridging veins and the superior sagittal sinus were preserved carefully during the process. No fixed retractors were applied; instead, minimal dynamic retraction was achieved using gentle suction. Intraoperatively, superficial cortical vein showed thinning and focal irregularity of the wall (Figure e); which was finally wrapped around with hemostatic material rather than sacrifice. The resection cavity was irrigated with saline containing dexamethasone. Gross total resection was obtained, with an estimated blood loss of 50 mL.

Postoperative Course

The patient awoke without motor or language deficits. She was mobilized on postoperative day 3 and discharged home, with normal motor strength and intact cranial nerves. Histopathology confirmed

an epidermoid cyst (Figure d). At 6 months, MRI confirmed no recurrence (Figure d) and her mRS score remained 0.

Discussion

Although intracranial epidermoid cysts are benign and slow-growing, their management in eloquent areas such as the supplementary motor area (SMA) is particularly challenging. The SMA is responsible for motor planning, initiation, and speech activation, and injury to this region or its connections can produce a transient SMA syndrome with contralateral akinesia or mutism[4–6].

Most patients recover spontaneously, reflecting a reversible functional disconnection rather than permanent tissue damage[7,8].

Functional imaging and electrophysiological studies suggest that postoperative SMA syndrome is associated with temporary disturbance of the inter-hemispheric and intra-hemispheric motor network. Preservation of the cingulum and callosal fibers and maintenance of venous drainage are crucial for rapid functional recovery[7,8]. After SMA surgery, plastic recruitment of the contralateral SMA and adjacent premotor areas contributes to restoration of motor function[9,10].

Compared with intra-axial lesions such as gliomas or meningiomas, epidermoid cysts offer distinct surgical advantages.They are extra-axial, relatively avascular, and possess a well-defined tumor-brain interface, allowing for internal decompression and gentle capsule dissection without traction on the eloquent cortex[1-3].Several series have demonstrated that gross or near-total resection achieves excellent long-term control with low recurrence rates[5,6].However, when bridging veins or cortical venous complexes are involved, the goal should be complete resection while preserving venous outflow.

In this case, a small parasagittal craniotomy and neuronavigation-guided interhemispheric approach were used to expose and resect the tumor. Cerebrospinal fluid release enabled adequate relaxation, and no fixed retractor was needed. A superficial cortical vein with focal irregularity was reinforced rather than sacrificed, preventing postoperative venous infarction and SMA syndrome. Similar microsurgical strategies—minimal retraction, precise capsule dissection, and venous preservation—have been shown to reduce neurological deficits after SMA and parasagittal lesion surgery[9,10].

Table 1. Summary of Reported SMA or Parasagittal Lesion Cases and Postoperative SMA Syndrome

No	Author (Year)	Study type	Lesion type	Surgical approach	Intraoperative adjuncts	SMA syndr.	Rec. time	Key findings / comments
1	Yamakawa et al., 1989[1]	Retrospective series (n=33)	Intracranial epidermoid (incl. frontal)	Various	None	Not reported	—	Early large series; total removal key to long-term control.
2	Hasegawa et al., 2022[5]	Large cohort (n=63)	Intracranial epidermoid cysts	Various	Neuronavigation, microsurgery	—	—	Extent of resection determines recurrence; GTR safest.
3	Schembri & Grech, 2015[7]	Case report	Interhemispheric epidermoid cyst	Interhemispheric	Neuronavigation	None	—	Demonstrates rare parasagittal location; full recovery.
4	Pinson et al., 2022[9]	Review	SMA lesions (various tumors)	Various	Mapping, MEP	33–75%	2–8 weeks	Comprehensive SMA-syndrome review; deficits

								usually transient.
5	Palmisciano et al., 2022[10]	Systematic review	SMA tumors	Various	Mapping, MEP	30–70%	2–12 weeks	Recovery correlates with preserved SMA–cingulate network.
6	Potgieser et al., 2014[11]	Review/fMRI analysis	SMA syndrome (functional study)	—	—	Yes (transient)	—	SMA syndrome due to reversible network dysfunction.
7	Acioly et al., 2015[15]	fMRI case study	Medial frontal tumor	Interhemispheric	fMRI-guided mapping	Present	3 weeks	Contralateral SMA recruitment supports recovery.
8	Berg-Johnsen et al., 2018[16]	Case series	Parasagittal meningioma	Interhemispheric	Mapping, MEP	43%	2–6 weeks	SMA syndrome frequent but transient; low morbidity.
9	Tsai et al., 2022[17]	Case report + review	Parasagittal meningioma	Interhemispheric	Navigation, MEP	Present	4 weeks	Low-traction microsurgery ensures full recovery.

SMA, supplementary motor area; CPA, cerebellopontine angle; MEP, motor evoked potential; GTR, gross total resection; fMRI, functional magnetic resonance imaging.

A summary of reported SMA and parasagittal lesions, including lesion type, approach, and functional outcome, is presented in Table 1. Across published series, the incidence of SMA syndrome after tumor surgery ranges from 30% to 75%, but symptoms are almost always transient.

Our experience confirms that, with careful microsurgical planning and functional awareness, gross total resection of an epidermoid cyst in the SMA region can be achieved without postoperative SMA syndrome.

Conclusion

SMA epidermoid cysts are exceptionally rare. Combining precise preoperative planning, neuronavigation, low-traction microsurgical techniques, and venous preservation can enable gross total resection without SMA syndrome. This case reinforces that eloquent cortex tumors can be resected safely while preserving neurological function.

Abbreviations

SMA = Supplementary Motor Area;
MEP = Motor Evoked Potential;
SEP = Somatosensory Evoked Potential;
MRI = Magnetic Resonance Imaging;
DWI = Diffusion Weighted Imaging;
CT = Computed Tomography.

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Paediatric intradural cerebellopontine angle chordoma mimicking meningioma

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ABSTRACT

Chordomas are rare, notochord-derived neoplasms, most commonly affecting the sacrum and clivus, and exceedingly rare in the cerebellopontine angle (CPA), especially in children. This report describes a 10-year-old male presenting with hearing loss, giddiness, vomiting, and left-sided facial palsy, who was found to have a CPA chordoma that was SMARCB1-deficient, confirmed via histopathology. The clinical presentation and management are discussed, along with a review of the scant global literature.

Keywords
chordoma,
paediatric,
intradural

INTRODUCTION

Chordomas are rare malignant tumors arising from ectopic remnants of notochord. They account for 1–3% of primary bone tumors, mostly extradural with sometimes trans-dural extension. They have a predilection for the midline particularly the sacrum and spheno occipital synchondrosis of clivus (1). Primarily intradural chordomas are rarely reported in literature, less than 10 case for our knowledge. Most of these are seen in adult patients. Paediatric intradural chordomas only 4 cases have been reported till now. Cerebello pontine angle (CPA) chordomas are very rare, with only a handful of cases reported globally, and even fewer in paediatric patients. Intradural chordomas are usually benign lesions with no bony involvement. As there was no bony involvement in our case, we suspected tentorial meningioma as primary diagnosis. (1,2)

CASE PRESENTATION

A 10-year-old male child presented to us with gradually progressive hearing loss, giddiness for 3 months, intermittent episodes of vomiting. On neurological examination child had left-sided Grade 3 facial palsy and sensorineural hearing loss. Fundoscopy was suggestive of Grade 2 papilledema. On radiological examination Contrast-enhanced CT/MRI demonstrated a large heterogenous solid-cystic lesion in the left CPA with hydrocephalus. The patient underwent surgical resection of left CPA lesion with CSF diversion in form of ventriculo- peritoneal shunt.

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Intraoperatively lesion was based on tentorium cerebelli, reddish grey in colour, highly vascular and suckable mimicking a meningioma. Complete excision of lesion as achieved with coagulation of dural attachment. All cranial nerves were preserved. Postoperatively, clinical recovery was achieved, and the patient was eventually discharged after suture removal. (Fig 1,2,3)

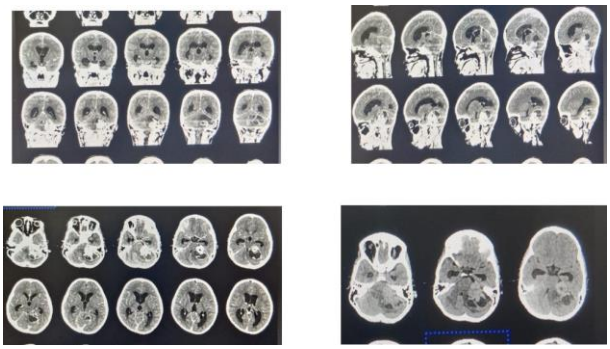

Figure 1. Preop CT scan of patient showing the left CP angle lesion

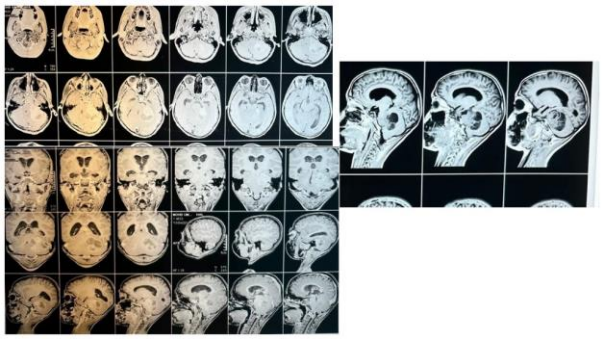

Figure 2. Pre op MRI with contrast

Histopathological evaluation of the resected tissue revealed poorly differentiated chordoma, with loss of SMARCB1/INI1 nuclear expression, confirming the diagnosis of a SMARCB1-deficient chordoma. The tumor cells were strongly positive for brachyury, CK pan, and EMA, consistent with chordoma. (4)

Discussion

Chordomas develop from remnants of notochord throughout the axial skeleton. Chordomas are rare slow growing primarily bony tumors with potential for trans-dural extension. They have malignant potential; symptoms are primarily due to compression of surrounding structures as they are slow growing. Rarely they can show acute progression of symptoms due to intertumoral haemorrhage.

Chordomas are usually extradural lesions with transdural extension in some cases. Purely intradural chordomas are rare, less than 4% of chordomas. Purely intradural chordoma in paediatric patients is even more rare as only 4 cases have been reported in literature till today. Purely intradural chordomas are usually well-defined lesions compared to extradural chordomas, they have no bony invasion, less adherent to surrounding structures. That is the reason these lesions can be resected completely causing less recurrence. (1,2)

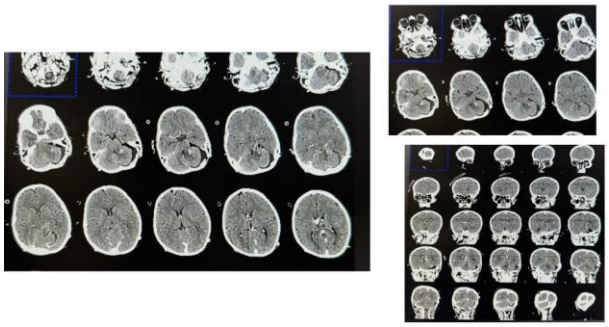

Figure 3. Post Op CT scan showing complete excision

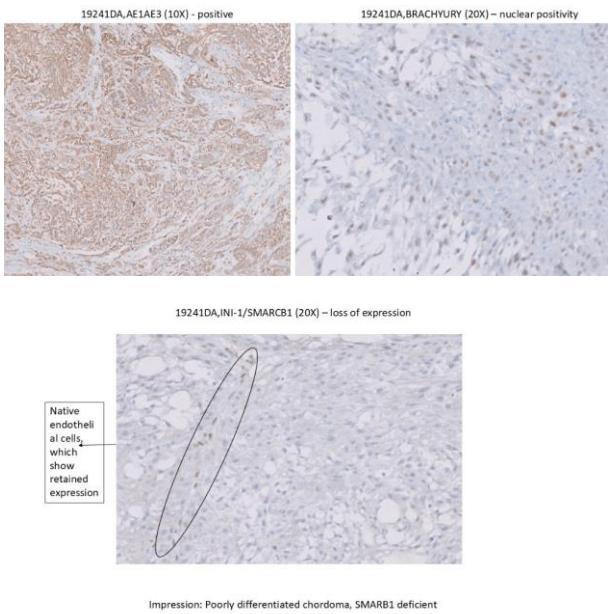

Figure 4. Histopathological image

Pathogenesis of intradural chordomas is not clear, various theories have been proposed. According of "migration" theory, some remnants of embryological

notochord are displaced intradural, as in cases of head trauma, from which intradural chordomas can originate. Other theory suggests malignant transformation of ecchordosis physaliphora (EP), a benign developmental ectopic remnant of intradural notochord tissue attached to the clivus. Even though EP and intradural chordomas are similar histologically, EP are smaller lesion(<2cm) which are usually asymptomatic. The differential diagnosis of these lesions includes dermoid, neurenteric and arachnoid cysts. (2,4)

CPA chordomas are notably rare, with only 2–3 cases described worldwide to date, and this case is among the first reported in Asia. The mainstay of treatment is surgical excision aiming for maximal safe resection. Role of adjuvant radiotherapy is debatable due to local recurrence rate shown even after subtotal resection. Ito et al reported zero recurrences in 18intradural chordomas among 17 patients with 5 to 144 months of follow-up while Vellutini et al reported a case of tumor recurrence two years after the initial diagnosis. So, role of adjuvant chemotherapy is still debatable. But considering the potential side effects of radiation on paediatric brain we propose careful observation rather than radiotherapy in these patients. (1,2,3)

Conclusion

Paediatric CPA intradural chordomas are rarely reported. Our case demonstrates the need to consider these lesions in the differential diagnosis of lesions of the CPA. As these lesions are slow growing and surgically completely resectable, role of adjuvant radiotherapy is debatable. Case pooling and further research are required to better understand and manage this entity.

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Surgical strategy and predicator of insular glioma in a tertiary centre

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ABSTRACT

Introduction: The insular glioma is a rare condition in neurosurgical practice. The treatment of insular glioma lacks a distinct approach. This study aims to review the outcomes of insular glioma surgery and discuss strategies to minimise the risk in adults who have undergone initial or repeat resection of insular gliomas of all grades.
Methods: An observational study was done among 50 patients with insular gliomas who were admitted to the Department of Neurosurgery of a tertiary care centre during January 2015-August 2025. The treatment decisions and neurosurgical outcomes of the patients with insular glioma were analysed. All patients were assessed with either computed tomography, magnetic resonance imaging, or both.
Results: Among the admitted patients, the youngest patient was 25 years, and the oldest was 76 years, while there were 30 males and 20 females. Most of the patients, 42 (84%), presented with headache and 32 (64%) presented with seizures of sudden onset. Right-sided insular gliomas were observed in 27 (54%) patients, but 23 (46%) patients had left-sided tumours, and 27 (57%) patients were operated on with the Transylvanian approach and 20 (42%) patients with the transcortical approach. Extent of resection above 90% or above was achieved in 52% of cases, and EOR of 70-90% in 48% of cases.
Conclusion: The surgical approach for insular gliomas requires technical mastery of intraoperative technologies to minimise postoperative morbidity.

Keywords
craniotomy,
extent of resection,
glioma,
insular,
Transylvanian,
transcortical

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INTRODUCTION

Insula is a deep and complex anatomical structure (1). The term insular cortex was first labelled by German neurologist, J. C. Reil in 1809.Insular gliomas are not uncommon, accounting for over 25% of all low-grade gliomas (LGGs) and 10% of all high-grade gliomas (HGGs) (4,13). Gliomas within the insular region have historically been difficult locations for the aggressive resection of LGGs or HGGs due to the complicated shape, organization of the insular cortex, its functional significance and its close relationship with the internal cerebral artery, the middle cerebral artery, and lenticulostriate vessels (1).Due to the significant surgical risks, some researchers have indicated that insular tumors are inoperable, opting instead for stereotactic biopsies for diagnosis, followed by chemotherapy and/or radiotherapy as alternative treatments. In contrast, other researchers have reported positive outcomes with insular tumors resections by utilizing specialized

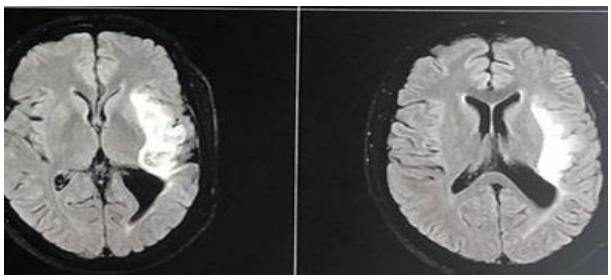
microsurgical techniques and a thorough understanding of insular anatomy. To maximize resection and reduce morbidities, the surgical strategy for insular glioma excision has necessitated a thorough grasp of surgical anatomy, glioma biology, subcortical white matter neuroanatomy, precise microsurgical technique, and intraoperative mapping (10,12,17). The aim of this study was to review the outcomes of insular glioma surgery.

Materials And Methods

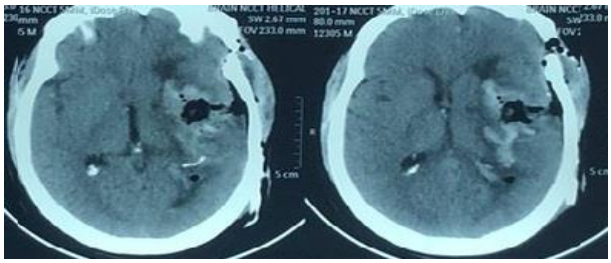
An observational study was done among 50 patients with insular gliomas who were admitted to the Department of Neurosurgery, National Academy of Medical Sciences, Bir Hospital from during January 2015-August 2025. All the insular gliomas were admitted to the neurosurgical intensive care during the study period and were included in the study. Those who received special preoperative treatment, such as radiotherapy and chemotherapy were excluded. All patients were evaluated with either a computed tomography (CT) scan, magnetic resonance imaging (MRI), or both. MRI brain is the ideal diagnosis tool for insular gliomas. Patient characteristics like age, history of hypertension/ Diabetes, Glasgow Coma Scale (GCS) on admission, routine laboratory tests, CT scan and MRI results were evaluated. We reviewed insular glioma presentation, radiological findings, and biological, anatomical, and clinical factors associated with outcomes after resecting tumors in this region. Outcome assessment was based on the data from the medical records of patients during their hospitalization, Glasgow Outcome Score (GOS) and 6-month period after discharge. Data were entered and statistically analyzed using SPSS Software.

Results

There were 30 males and 20 females with ages ranging from 25 to 76 years. Right-sided insular gliomas were observed in 27 (54%) patients but 23 (46%) patients had left-sided tumors. Most of the patients, 42 (84%) presented with headache and 32 (64%) presented with seizures of sudden onset. All patients underwent craniotomy with excision tumors except 3 (%) had undergone biopsy procedure after burr hole. Biopsy cases were old with radiologically high-grade tumors, and 27 (57%) patients were operated on with a Transylvanian approach and 20 (42%) patients with a transcortical approach. Among 6 (12%) patients who had undergone a previous resection, 4 (8%) presented with tumors recurrence and 2 (4%) had a residual tumor.

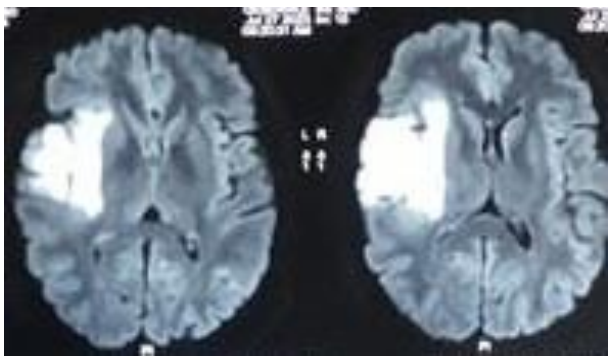


A.



B.

Figure 1. Preoperative axial T1-weighted, magnetic resonance imaging MRI, (A) revealed expansible left insular mass lesion and immediate post-operative computed tomography (CT) scans (B) showing excision of lesion.



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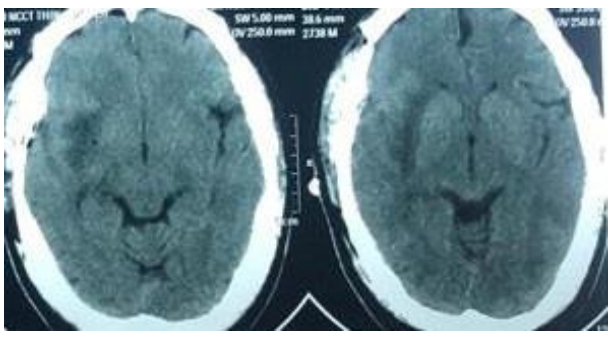

D.

Figure 2. Preoperative axial flare MRI (C) indicated right insular mass lesion and CT scans (D) showing post-operative excision of lesion.

Among them, 5 (9%) patients had severe brain swelling post operatively who had undergone transylvian approach. All of them went to an emergency craniotomy but they developed hemiparesis and expressive aphasia. Among them, there was one mortality, even a decompressive craniectomy. Radiologically, contrast enhancement on T1 imaging and cystic insular glioma had good prognostic factors post operatively, no matter either transylvian or transcortical approach. Extent of resection 90% or above was achieved in 52% of cases, and extent of resection of 70-90% in 48% of cases. According to histological grade, there were WHO Grade II (n=33, 66%), WHO Grade III (n=14, 28%) and WHO Grade IV(n=3,6%). All patients had postoperative chemotherapy and radiotherapy as oncological protocol. All of these cases were kept in regular follow up in the outpatient department.

Discussion

Surgical resection of insular tumors was established in the 1990s by Yaşargil and Reeves (21). The insula has a pyramidal shape with an apex that was found and protected within the folds of the sylvian fissure, under the triangularis of inferior frontal gyrus and is a hidden lobe situated in the depth of sylvian fissure(16,17,18,19). A huge network of afferent connections has allowed the insula to be dedicated to a wide range of motor, sensory, affective, and cognitive tasks (9,11). The main objective of surgery is to remove as many insular gliomas as possible while preserving the patient's speech and motor function and increasing their chances of survival. Combination of microsurgical technique, cortical mapping, and insular anatomy are needed for the resection of insular glioma. Both the transylvian and transcortical corridors to the insula are associated with low morbidity profiles (1, 6,8,12,15, 16)

MRI plays a significant part in the diagnosis and further characterization of insular glioma. In order to reach a maximal safe resection of these invasive tumors, advanced imaging systems, such as perfusion imaging, diffusion imaging, spectroscopy, and positron emission tomography (PET) imaging, have become progressively valuable surgical tools, particularly, when used in combination with intraoperative brain mapping (3,8,9,11, 16). In our series, contrast enhancement on T1 imaging and cystic lesions had good prognostic factors in insular glioma.

The insular cortex receives a rich blood supply from the internal cerebral and middle cerebral arteries. The middle cerebral artery bifurcates at the limen insula, giving rise to between 1 and 6 insular M2 branches that cover the surface of the insula. The M2 branch located over the central insular sulcus terminates as the rolandic artery, and understanding this can help prevent damage to the vessel. The lenticulostriate arteries traverse the anterior perforated substance and can be challenging to identify and preserve during surgery for insular gliomas (1,9,21). Surgical intervention of glial tumors located in the insula is particularly difficult because of the close proximity to the internal capsule. Around 95% of patients had fewer neurological deficits, allowing them to function independently and eventually return to work overtime (2 ,4,5,7,8). These results demonstrated that a pterional craniotomy with microsurgical approach to insular tumors could be considered. In the English literature, surgeons meticulously dissected anatomical structures and described surgical techniques to maximize safety (2,6). There are some recommendations: 1) Broad splitting of the sylvian fissure, 2) awake craniotomy with cortical and subcortical mapping to identify the overlying motor cortex tract and the internal capsule subcortically, and 3) meticulous suprasylvian dissection to avoid coagulation of the long perforating M2 segment and lateral lenticulostriate arteries during insular tumor resection. and 4) tumor removal piecemeal manner through windows created between vascular structures (3,6,9,10). The transcortical approach emphasizes a subpial resection of the insula lesion which has expanded popularity with the evolution of intraoperative mapping techniques and awake craniotomies (1). Berger et al. reported transcortical approach to insular tumors series and divided the insula into four zones to establish surgically relevant insular function and to correlate with outcome surgical resection (1,5,6). Ultimately, the approach is a multifaceted decision and involves the knowledge of the individual surgeon as well as patient presentation, anatomy and neurological function.

In this study the incidences of insular gliomas in males were 60% and 40% in females which is in contrast to another study, where male and female

incidence was 42% and 62% respectively (17,18). In another study, seizure was found to be the most common presentation in 70.6% patients and in similar another study found in 55.1% patients 2,3 but in our study most common symptom was headache in 42 (84%) followed by seizure 16 (32%). Left side of tumor was more common (55.7%), whereas in our study right side tumors were more common 54% and left side tumors were seen in 46% patients (17,18). In our study, WHO grade 2 tumor was most common accounting to 66%, whereas in another study, WHO grade 2 tumor was most common in 65.7% patients and 34.3% patients had WHO Grade 3 tumor. In a similar study, EOR above 90% was achieved in 42% of cases, and EOR of 70-90% in 51% of cases, which was similar to our findings in which extent of resection 90% or above was achieved in 52% of cases, and extent of resection of 70-90% in 48% of cases (6,10, 14,15,18,20).

Tumor resection improves survival, but increasing extent of resection poses a risk of neurological compromise. This study is the first in the literature to analyze predictors of insular glioma. Predictors of poor outcome involved WHO grade IV glioblastomas, advanced age, diabetes and transylvian approach. Predictors of favorable outcome comprised younger age at diagnosis (< 40 years); contrast enhancement on T1 imaging and cystic insular glioma radiologically, EOR > 90%, WHO grade I, II, and III histologically; and transcortical surgical approach. This study was an observational study of a single institution and included a small sample size. So, this study cannot be generalized among the whole population of Nepal. We recommended further prospective study to know better prognostic factors of insular glioma.

Conclusion

Insular glioma surgery is challenging surgery due to its peculiar anatomical characteristics and carries substantial morbidity and complications. Younger age; radiologically contrast enhancement on T1 imaging and cystic lesion, transcortical surgical approach, EOR > 90%, and histologically WHO grade I, II, and III ad predicated as favorable outcome in insular glioma surgery.

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A poignant odyssey of migrating distal end of ventriculoperitoneal shunt from inguinal canal to anal canal. An arduous situation

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ABSTRACT

Ventriculoperitoneal (VP) shunt procedure is the most commonly performed and widely accepted treatment method for hydrocephalus of any aetiology. Unfortunately, VP shunt is associated with varying complications beginning right from its ventricular end to the peritoneal end [1]. Here we present a challenging case of migrating distal end of VP shunt with its extrusion through the inguinal region, followed by per anal protrusion in the same patient. Scarcity of literature on sequential dual protrusion of the distal end of VP shunt in the same patient makes it a very uncommon case attracting our attention.

Keywords
hydrocephalous, ventriculoperitoneal (VP) shunt, inguinal shunt protrusion, per anal shunt protrusion, silicon allergy

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INTRODUCTION

Ventriculoperitoneal shunt is the mainstay of treatment for hydrocephalous but the complications rate is very high. After shunt placement, approximately 11-25% patients develop shunt failure within first year [2-5]. The number of shunt revisions and replacements is higher in pediatric patients than the adults [4, 5]. Most common causes of shunt failure are shunt obstructions followed by infection [5, 6] with infections causing early and obstruction/occlusion amounting to late shunt failures [7]. Other common complications include abdominal pseudocyst formation, spontaneous bowel perforation, inguinal hernias, liver abscess [8-10]. Rarer complications are extrusion of peritoneal end into the stomach, gall bladder, vagina, liver, urinary bladder, bowel, colon, diaphragm and scrotum [11, 12]

CASE PRESENTATION

Here is a case of an infant boy who presented to our department of Neurosurgery, super specialty block's outdoor with a large sized head and abnormal body movements. Radiology of head done which was suggestive of hypoxic-ischemic encephalopathy with dilated ventricles and multiple cysts in the brain. He underwent right VP shunt surgery at our center at the age of 2 months. After one month of the surgery he

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developed skin erosions of the abdominal wall over the catheter tract with obstruction of the distal end. Second surgery was done and distal end was revised with placement into the peritoneum via a fresh incision on the abdomen [Fig.1]. Two months later he presented with the distal end catheter tip extruding via the left inguinal region with a clear CSF coming through it (FIG.2a).

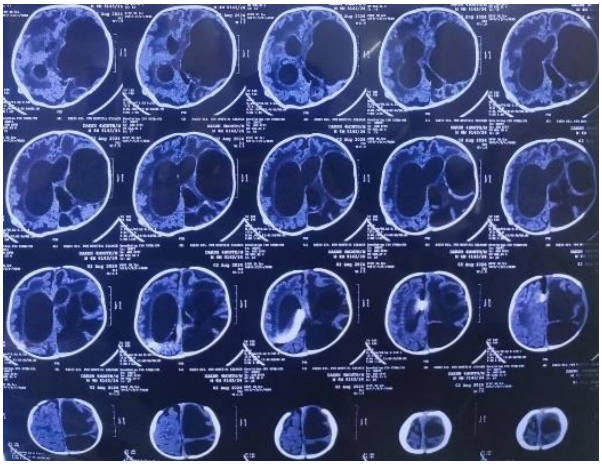

Figure 1. CT scan showing multiple cysts with right VP shunt catheter in right lateral ventricle.

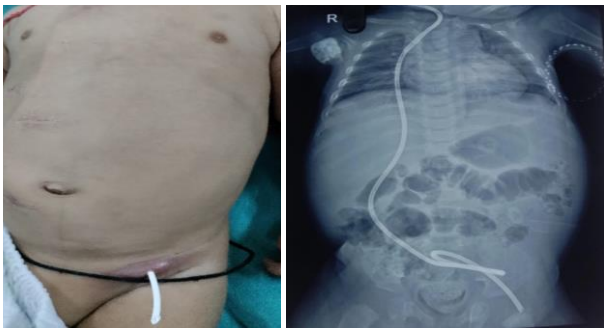

Figure 2. a) Clinical photograph of the same patient with distal end extruding out of the inguinal canal and dribbling of clear CSF. (b) X- ray showing shunt catheter in abdomen with distal end in left inguinal region.

Child was fully conscious but febrile. USG abdomen was done which showed catheter tip passing through left inguinal region with surrounding inflammation and sub-centemetric inguinal lymph nodes suggestive of cellulitis. Erect abdominal X-ray showed shunt tube in the abdominal cavity with distal end in left inguinal region (FIG 2b). CSF study was done and after ruling out the shunt infection third surgery was planned. Clavicular incision was given and distal end (extruded tip from the inguinal region) was pulled down. Left sided VP shunt procedure was performed. Whole assembly of the shunt was changed (Fig.3). Post op event was uneventful and child was discharged in a stable condition. Later after 3 months child presented with fever and extrusion of distal catheter tip per anal (Fig.4).

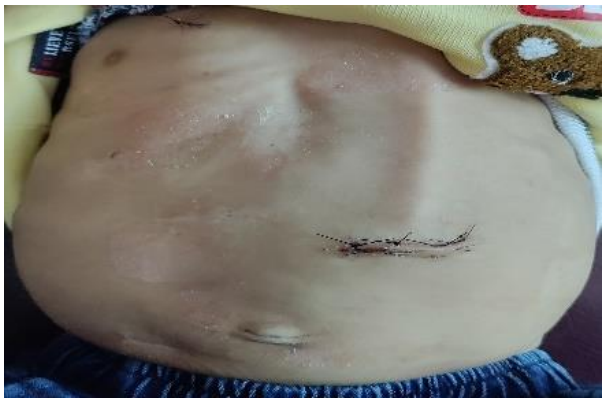

Figure 3. Clinical photograph with multiple healed scars of previous surgery and left abdominal incision of third surgery.

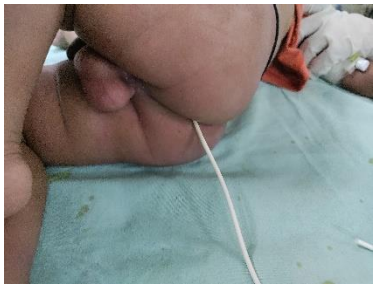

Figure 4. Photograph showing extrusion of distal end of the catheter through anus.

On examination child was conscious but irritable with dribbling of CSF from the extruded part of the VP shunt. There were no signs of meningeal irritation or peritonitis. Child was put on antipyretics and antiepileptics. Plain CT head showed ventricular end in the left lateral ventricle and the associated cyst with no ventriculomegaly on the right side (Fig.5). Child underwent fourth surgery. Skin incision given at the clavicle and shunt tube was divided. Extruded part was gently pulled out through the anus. Ventricular end was exteriorized and CSF was sent for culture and sensitivity. Empirical antibiotics were started. Child improved and after three sterile cultures, exteriorized part was clamped. There were no signs of raised intracranial pressure and anterior

fontanelle was lax. Ventricular end was removed and child was discharged in stable condition with regular follow-up advice.

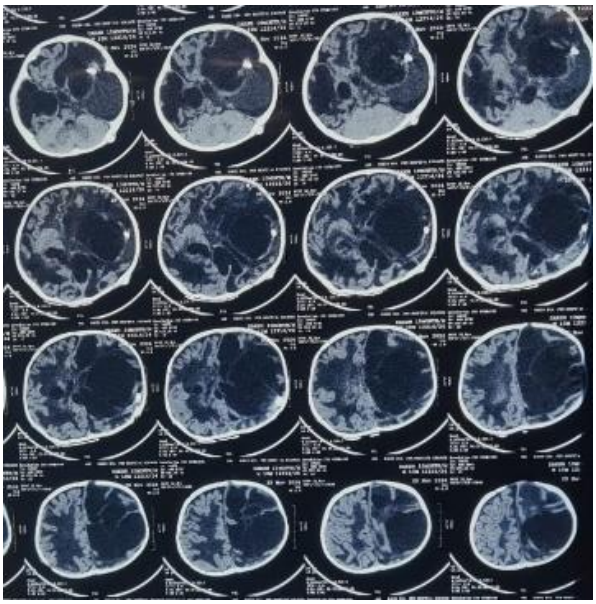

Figure 5. Computed tomography showing ventricular end in left lateral ventricle with no ventriculomegaly on right side.

Discussion

VP shunt surgery just like other operative procedures, is not free from complications. Extrusion of distal end of the catheter is a rare complication. To our knowledge no such case of dual extrusion of the shunt catheter in single patient has been reported till date making this a very unusual presentation. Majority of the cases present months after the surgery and most of the patients are asymptomatic until visualization of the extruded catheter from the skin or the natural orifice. Although the exact pathophysiology behind shunt extrusion is not well-established, but few mechanisms have been proposed [13]. Thin intestinal wall of children, stiff and pointed catheter end, use of trocars by the surgeon, infection, previous surgery and silicon allergy are the factors apart from others that could affect the incidence of shunt extrusion along with other abdominal complications [14, 15].

Wu et al. studied the Office of Statewide Health Planning and Development Database and found highest rate of shunt malfunction in neonates followed by children and then adults [4]. Children with early shunt complication had a greater odds of a subsequent complication[4]. Type of hydrocephalous was found to be the strongest independent risk factor for VPS complication with congenital and obstructive type having more risk than non-communicating type[4] . Low socioeconomic status and male gender were associated with increased risk of complications [4] .

Bowel perforation by the distal catheter tip is rare with a reported incidence of 0.1- 0.7% [16-18] although higher incidence have been reported by some series [19,20]. Majority of the cases of bowel perforation occur in children [17] which may be due to relatively thin bowel wall in infants, especially in children with myelomeningocele in whom bowel innervation may be insufficient [17]. Colon is the most common site of bowel perforation with per anal extrusion of the shunt catheter being the most prevalent symptom [21]. Different hypotheses have been proposed to explain this complication. Catheter stiffness in one of the factors causing perforation. G Muthoni et al. [20] compared the stiffness of three commonly used types of catheter, including Medtronic, Codman bactiseal and Chabra shunt catheters. They studied 197 shunted pediatric patients, out of them 6 patients developed perforation and all of them had Chabra shunts. This led to the conclusion that Chabra shunts to be the most stiff. Later on it was discovered through experiments that Chabra shunt catheters are the least stiff among the three but have the highest frictional force [20]. Distal catheter tends to be entangled in omentum at the site of perforation[22,23]. Increased frictional force of the catheter may promote its entanglement in abdominal tissues predisposing to bowel perforation [20] .

Silicon allergy may cause perforation through bowel as well as abdominal wall and other skin erosions around the catheter [22-24]. Patients with silicon allergy have history of recurrent shunt malfunction due to catheter obstruction. Symptoms resolve completely with replacement of distal catheter with the one made of polyurethane material [22, 24].

The reported incidence of inguinoscrotal complications is around 10-20% [25, 26, 27-29]. Increased intra-abdominal pressure is often implicated as a causative factor for inguinal hernias in patients with hydrocephalous treated with VP shunt [25, 26]. Abnormal neuromuscular function may also be factor responsible for these complications [27].

Conclusion

To summarize, extrusion of distal end of VP shunt is rare. Dual extrusion in single patient is further very unusual and to the best of our knowledge, there is no such case reported in the past. The odyssey of the migrating shunt is very painful both for the patient and the family. Out of the various proposed theories, we couldn't find the exact mechanism for shunt extrusion and its variable presentations. Complications associated with shunt extrusion like meningitis should be addressed promptly to reduce the morbidity and mortality associated with them. More research work is required to find out the factors causing extrusion and explain their unusual and unpredictable presentation among patients. We emphasize on lifelong follow-up and education of the patients/guardians who take care of them regarding the complications. Proper guidelines should be framed to deal with these complications promptly and adequately thereby reducing the associated morbidity and mortality.

Abbreviations

VPS: ventriculoperitoneal shunt,
NCCT: non contrast computed tomography.

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Descending spinal vascular axis in contact with the artery of Adamkiewicz. Anterior spinal artery or descending branch of the artery of Adamkiewicz?

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ABSTRACT

Objective: This study aims to determine whether the descending vascular axis distal to the Adamkiewicz loop is a continuation of the AKA or the ASA, using both morphological and statistical analyses.

Method: A cadaveric study was conducted on 15 adult male specimens. Following posterior access and removal of the spinal cord en bloc, low-pressure injections were performed into the ASA to visualize spinal vasculature. In several cases, the AKA was identifiable without injection due to its prominent calibre. Using digital callipers, morphometric measurements were obtained for the ascending branch, arch, and descending segment of the AKA, as well as the ASA segment proximal to the AKA loop.

Results: The mean diameter of the ASA was 1.0 mm (range: 0.5–1.86 mm), with 80% of specimens exhibiting a calibre under 1 mm. The ASA occupied less than 10% of the spinal cord's ventral surface. The AKA had a mean diameter of 1.14 mm, while the descending vascular axis measured 1.69 mm on average (range: 0.8–2.1 mm). In all specimens, the AKA presented as a continuous single trunk with ascending, arching, and descending components. The descending vascular axis showed greater morphological similarity in calibre to the AKA than to the ASA.

Conclusion: Morphometric evidence suggests that the descending spinal axis beyond the AKA loop more closely resembles a continuation of the Adamkiewicz artery rather than the anterior spinal artery.

Keywords
Adamkiewicz artery, spinal cord, vascular anatomy

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INTRODUCTION

In the initial description of spinal cord vascularization, Lazorthes et al, reported that the ventral side of the spinal cord is vascularized by the anterior spinal artery (ASA) (5). This artery travels along the median ventral fissure from top to bottom and is reinforced along its course by various radiculomedullary arteries. At the level of the conus medullaris, the ASA anastomoses with the two dorsolateral spinal arteries, forming the anastomotic loop of the conus medullaris (3,5,11). According to this initial description, the ASA runs along the entire length of the spinal cord.

The Adamkiewicz artery (AA) is described as an ascending artery that joins the ASA, bending to form its characteristic hairpin loop. Most authors agree that the vascular segment beyond the junction of these two arteries represents the continuation of the ASA (6,12). In fact, the Adamkiewicz artery is the main radiculomedullary artery supplying the ASA at the level of the lumbar enlargement (10,11). It is also considered the largest intradural blood supply to the anterior spinal cord (2).

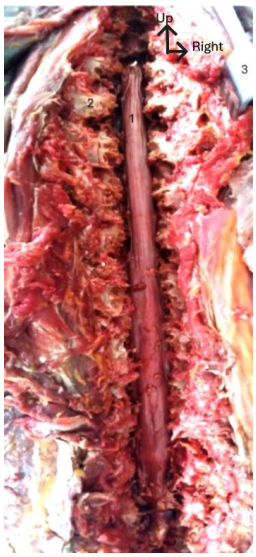
We previously published an anatomical study on the ASA–AA junction and defined three types of junctions based on the diameters of the ASA, AA, and the descending branch of the ASA, which were correlated with neurological risk (7). Morphologically, the Adamkiewicz artery appears as a single trunk with an ascending branch, a loop, and a descending branch. Based on purely morphological observations, we noted trivial but consistent similarities in caliber between the Adamkiewicz artery and the descending spinal axis, more so than with the portion of the ASA proximal to the junction.

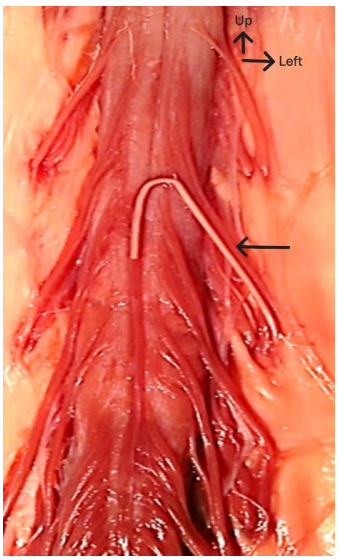
From these findings, we currently hypothesize that the descending spinal axis beyond the Adamkiewicz loop may be more accurately described as the continuation of the Adamkiewicz artery rather than the ASA. The present study aims to determine the true origin of the descending spinal axis following the Adamkiewicz loop through both morphological and statistical analysis.

Material and method

A cadaveric study was conducted on a series of 15 adult male specimens. The vertebral column was accessed via a posterior approach (Fig. 1), and the spinal cords were meticulously excised en bloc following bilateral resection of the nerve root attachments. The anterior dura was then incised to allow for low-pressure injection into the anterior spinal artery (ASA), which was readily identifiable within the median ventral fissure. This injection enabled clear visualization of the radiculomedullary arteries, including the artery of Adamkiewicz. Notably, in several specimens, the Adamkiewicz artery could be identified without ASA injection due to its distinctive caliber (Fig. 2).

Using a digital caliper, precise morphometric measurements were obtained for critical segments of the Adamkiewicz artery. These included the diameter of the ascending branch, the curvature of its characteristic hairpin loop, the diameter of the anterior spinal artery (ASA) segment proximal to the loop, and the vertical arterial segment extending distally from the arch (Fig. 3).





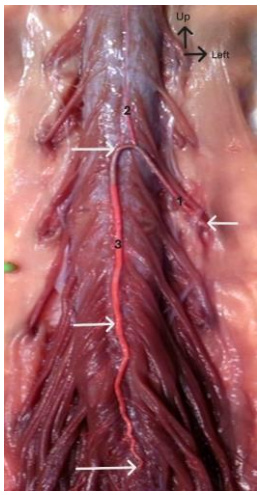


Figure 1. Thoracolumbar posterior arch resection for dura exposition: 1: Dura mater; 2: posterior arch resected; 3: Beckman retractor.
Figure 2. Adamkiewicz artery (arrow) with big caliber reveled without colorant injection.
Figure 3: Anatomical preparation showing lower spinal cord vascularization: 1: Adamkiewicz artery; 2: anterior spinal artery; 3: descending spinal axis. White arrows show the entire AA as a single trunk in the present description.

Results

The average diameter of the anterior spinal artery (ASA) was 1.0 mm, with a range from 0.5 mm to 1.86 mm. In 80% of specimens, the ASA caliber was less than 1 mm. The artery occupied less than 10% of the ventral surface of the spinal cord, which measured an average width of 9.4 mm (ranging from 8 mm to 14 mm). Table 1 summarizes the ASA caliber, AA caliber and the vertical axis after AA loop caliber spinal cord surface, and the percentage of ventral surface covered by the ASA and the AA.

Morphometric analysis of the Adamkiewicz artery (AA) revealed a mean diameter of 1.14 mm at its middle third, with the majority of specimens

exhibiting a diameter greater than 1 mm. The descending vascular axis distal to the Adamkiewicz arch measured an average of 1.69 mm, with observed extremes ranging from 0.8 mm to 2.1 mm. In all specimens, the Adamkiewicz artery consistently appeared as a single-trunk vessel, comprising an ascending branch, a transverse arch, and a descending axis.

Discussion

The anterior spinal artery, the Adamkiewicz artery, and the radiculomedullary arteries share a closely related embryological origin. The ASA consistently exhibits a relatively small diameter, averaging around 1 mm, which aligns with the findings of Ko et al. in their anatomical study of 15 specimens (4). Suh et al., in a separate study involving 26 specimens, reported a mean ASA diameter of approximately 0.57 mm (9).

In our study, the intradural portion of the Adamkiewicz artery demonstrated an average diameter of 1.14 mm, consistent with measurements reported by Beaza et al (8). The descending vascular axis distal to the AA's arch measured 1.69 mm in diameter. In a meta-analysis of 375 patients, Taterra et al. found that in 71.3% of cases, the AA formed a continuous conduit from the aorta to the ASA (12).

To address our initial hypothesis, a morphological comparison was undertaken. The aim was to evaluate the diameter of the descending vascular axis in relation to both the ASA and the AA. It was observed that the descending vascular axis was, on average, 1.84 times larger in diameter than the ASA, and 1.5 times larger than the AA. In a previous investigation, we noted that the diameter of the descending segment closely matched that of the AA at the level of its arch; however, no definitive hypothesis was proposed regarding its origin (7).

A detailed statistical analysis using the Wilcoxon test for paired samples ($\alpha = 0.05$) revealed a statistically significant morphological difference between the descending vascular axis and the ASA ($p = 0.003$), as well as between the descending vascular axis and the AA ($p = 0.001$). Notably, the caliber of the descending axis appeared to diverge from that of the ASA, while approximating the caliber of the AA.

Subsequent bivariate analysis indicated that the difference in caliber between the descending axis and the AA ($p = 0.300$), as well as between the descending axis and the ASA ($p = 0.407$), was not statistically significant. However, correlation analysis revealed a stronger association between the descending vascular axis and the AA ($r = 0.287$) compared to the ASA ($r = 0.208$). Based on these morphological and statistical findings, the descending vascular axis appears to share greater similarity with the AA than with the ASA.

Furthermore, all anatomical specimens consistently displayed the Adamkiewicz artery as a single, uninterrupted trunk of large caliber extending from its dural penetration to its distal termination. These preparations also revealed that the ASA terminated at the apex of the AA loop centrally within the ventral median fissure in 10% of cases and laterally in the remainder. This finding supports the notion that the ASA may represent a chain of anastomoses terminating at the apex of the AA loop.

In a cadaveric study, Alleyne et al. described the intradural segment of the AA referred to as the "medial trunk" as a single continuous vessel following its junction with the ASA (1). The concept of the AA as a singular, dominant trunk challenges the traditional depiction of spinal cord vascular anatomy. However, it offers a compelling explanation for the profound neurological deficits associated with injury to the AA, which would compromise vascular supply to the entire lower spinal cord.

Table 1. Morphometric features of AA, ASA and vertical vascular axis.

Spec	AA	ASA	Vert Ax	Cord surf	AAs (%)	ASAs (%)	Vert Axs (%)
1	0.80	1	0.80	9	8.89	11.11	8.89
2	1.12	1.12	1.50	9	12.44	12.44	16.66
3	1.26	0.60	1.20	9	14	6.67	13.33
4	0.61	1.20	1.53	8	7.62	15	19.12
5	1.77	1.86	1.86	14	12.64	13.29	12.64
6	1.90	1.50	2.10	10	19	15	21
7	0.90	1	2	10	9	10	20
8	1.20	0.8	1.25	9	13.33	8.89	13.88
9	0.80	0.5	2	9.5	8.42	5.26	21.05
10	0.90	1	1.90	9	9	11.11	21.11
11	1.20	0.75	1.70	9	13.33	8.33	18.88
12	1.10	1	1.85	8.5	12.94	11.76	21.76
13	1.15	0.95	1.80	9	12.77	10.56	20
14	1.20	1	2	8	15	12.50	25
15	1.12	0.75	1.90	10	11.2	7.50	19

AA= Adamkiewicz artery, ASA= anterior spinal artery, Vert AX= vertical axis, Cord surf= spinal cord surface,

AAs= Adamkiewicz artery projection on spinal cord surface, ASAs= anterior spinal artery projection on spinal cord surface, Vert Axs= Vertical axis projection on spinal cord surface.

Conclusion

In light of this descriptive anatomical study, the Adamkiewicz artery (AA) consistently appeared as a single, uninterrupted trunk. After perforating the dura mater, it exhibited an initial extramedullary ascending segment, ultimately giving rise to a descending branch closely adherent to the median ventral fissure of the spinal cord. Morphologically, the descending vascular axis appears to represent a continuation of the AA rather than a segment of the ASA. These findings suggest that the AA serves as a critical conduit connecting the anterior and posterior medullary vascular territories.

This unique arterial configuration may explain the profound neurological deficits frequently observed following injury to the AA, as such damage compromises blood flow to a substantial portion of the spinal cord. Nevertheless, this conclusion remains a preliminary hypothesis and warrants validation through studies involving larger sample sizes.

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Mass lesion in cases of cerebral arteriovenous malformations post gamma knife radiosurgery or embolization. Pathophysiology and management algorithm

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ABSTRACT

Background: Cerebral Arteriovenous Malformations (AVM) have been conventionally treated with surgery, embolization and/or Gamma Knife Radiosurgery (GKRS). This article is to present a rare complication after Embolization/ Gamma Knife Radio Surgery for Cerebral AVMs.

Method: 05 patients with cerebral AVMs presenting with an unusual complication of a mass lesion at the site of the treated lesion were treated. Two modes of index treatment were used: endovascular embolization and/or GKRS. These patients developed new-onset neurological deficits at varying intervals after index treatment. They were investigated radiologically, revealing a mass lesion at the site of the treated AVM. The pathophysiology of this complication, along with the management algorithm, has been studied and is presented.

Results: All the patients in the series responded well to surgery. The histopathological examination revealed vascular elements in all cases without any evidence of neoplasm.

Conclusion: Delayed presentation as a mass lesion of a treated AVM is unusual. The mass lesion in cerebral arteriovenous malformations with suspicion of malignant transformation or with unresponsive raised intracranial pressure may mandate craniotomy and excision.

Keywords
cerebral AVM,
gamma knife radiosurgery,
radiation necrosis,
mass lesion

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INTRODUCTION

Cerebral arteriovenous malformations (AVMs) are abnormal connections of arteries and veins of the brain, resulting in arteriovenous shunting of blood. The population prevalence of brain AVM is estimated to be 10–18 per 100,000 adults with a new detection rate (i.e., incidence) of approximately 1 per 100,000 person-years[1]. Overall mortality rates in AVM patients range from 0.7%–2.9% per year[2]. The commonest presentation of the cerebral AVMs is intracranial haemorrhage. Seizure is the second most common presentation of AVM, which occur in 20%–45% of patients[3].

The various modalities conventionally used to treat cerebral AVMs are embolization, Gamma Knife RadioSurgery (GKRS) and surgical excision4. The choice of treatment depends upon the size of the lesion, location of the lesion and venous drainage of AVM, consolidated and summarized as Spetzler Martin Grading.

Physiology of AVM treatment with embolization and Gamma Knife Radiosurgery

With the cyno-acrylate glue is deposited in the feeders of the AVM, the venous channels collapse, vascular channels undergo constriction and the nidus shrinks[5, 6]

Gamma Knife Radiation induces intimal necrosis with resultant obliteration of the abnormal vascular channels and shrinkage of the AVM nidus. Immediate post procedure perinidus oedema after Gamma Knife Radiation is not uncommon and can be bothersome, especially in cases, where the nidus size is big and maximum radiation dosage is used.

Adverse Effect Profile of AVM embolization and Gamma Knife RadioSurgery

Adverse effects after embolization

Some minor side effects may be observed shortly after embolization of an AVM by some patients. The side effects are usually temporary and should subside within a few days to weeks[7].

Headaches are not infrequently reported. Other possible complication is stroke like symptoms such as weakness in one arm or leg, numbness, tingling, speech disturbances and visual problems. The risk of permanent stroke or death after embolization is low.

The risk of bleeding reduces, but doesn't become zero after embolization of AVM cases. Residual risk of bleeding is directly proportional to the initial size of the AVM and the size of residual nidus post embolization.

Adverse effects after Gamma Knife RadioSurgery

The common short term complications, which can happen are:

a) Rebleeding
b) Incomplete obliteration
c) Encephalomalacia/encephalocele
d) Fresh neurological and cranial nerve deficits
e) Seizures
f) Persistent headaches
g) Alopecia at the local irradiated site

Majority of these events occurs within 3 years of radio-surgical treatment[9] The most common long term complication of Gamma Knife Radiosurgery are Radiation necrosis, cyst formation, Haemorrhage (CEIH) and increased seizure frequency. Radiation necrosis effects almost 33% of all patients, however clinical manifestations owing to Radiation Necrosis effect about 1.7 – 7.6% of all patients treated with GKRS.

We here, present an unusual complication which was noticed in few of our patients following embolisation/ GKRS, which has not been widely reported so far and has not been studied. We here present a series of five patients who underwent transarterial embolization for cerebral arteriovenous malformation, were asymptomatic in the initial post procedure period. On follow up, they presented with a space occupying lesion with surrounding oedema at the site of initial nidus, resulting in local and hemispheric mass effect.

All these patients presented with clinical features of raised Intracranial pressure, which didn't respond to decongestants and needed to be treated with decompressive craniectomy, to which they promptly responded with improvement in sensorium. The CEMRI Brain did not show any bleeding from the AVM or any evidence of a fresh acute event except the finding of fresh onset perilesional oedema with mass effect. The Histopathological examination of the mass revealed only vascular tangles and was devoid of any tumour tissue, even when each thin section of the excised mass was deliberately examined. The consent of the patients has been obtained for publishing this data.

Case No. 1

Ms IS

13yrs old girl presented with history of fall from treadmill with LOC for about 15 min followed by severe headache and vomiting in August 2012. MRI with MRA revealed Left sylvian fissure SAH due to left frontal AVM with intranidal aneurysm (Fig 1). Endovascular coiling of the intranidal aneurysm was done on 03 Oct 2012 (Fig 2) followed by GKRS for the left frontal AVM on 02 Feb 2013. DSA, 3yrs Post GKRS in Dec 2016 showed complete obliteration of AVM and no residual sac of intranidal aneurysm (Fig 3). She was asymptomatic for 9yrs Post GKRS. In Sep 2021, she presented with complaints of headache, vomiting and nuchal pain. NCCT Head + CT

Angiography followed by CEMRI Brain showed mass lesion in left sylvian fissure region with no aneurysm (Fig 4a). She was initially managed with Mannitol, Lasix and Steroid therapy. She responded favorably with reduction in headache and vomitings. She was given steroids for 6 weeks and gradually tapered off. Within one week of stopping steroids, she presented again with features of raised intracranial pressure. She was readmitted and underwent excision of the mass lesion in Oct 2020 in view of recurrent features of mass effect and diagnostic dilemma between radiation necrosis and malignant transformation. Intraoperatively, it was found to be a well-defined, firm, vascular mass lesion localized in the left sylvian fissure with well-defined planes abutting the coiled aneurysm.

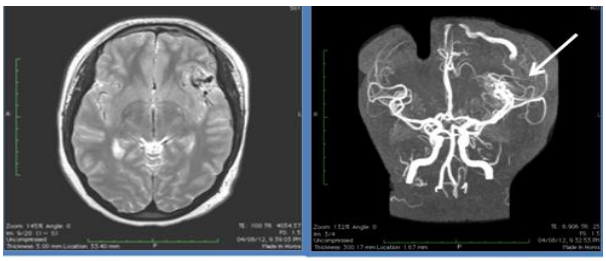


Fig 1 : MRI Brain and MR Angio at presentation (Aug 2012)

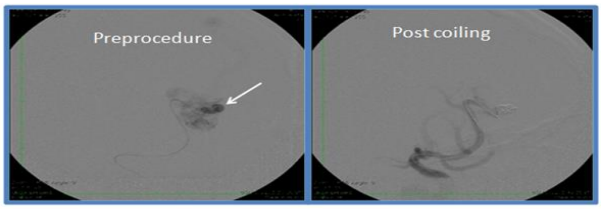
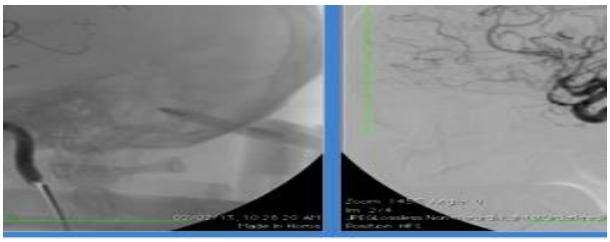


Fig 2 : DSA showing AVM with intranidal aneurysm and post coiling



entation **Fig 3B** Interval

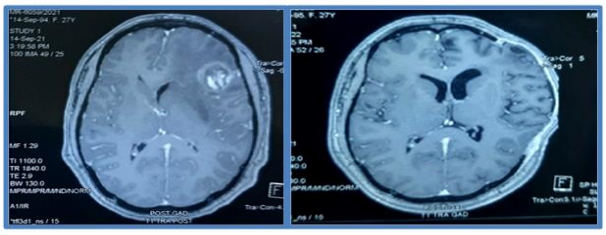
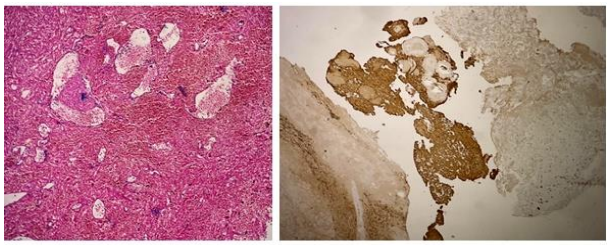


Fig 4 : Preop and postop MRI (Sep 2021)



5A : Large areas of haemorrhage, d necrosis, ectatically dilated blood **Fig 5B** GFAP IHC highlights gliotic brain tissue vessels between the hyalinised blood vessels

HPE – vascular elements interspersed with gliotic brain (Fig 5A & B)

Case No. 2

Ms YK

This 39yrs old lady patient presented with left sided hemiparaesis in Apr 2013. Investigations revealed Right Parietal AVM sized 46 x 38 x 49mm. The patient underwent Gamma Knife Radiosurgery for this AVM at Army Hospital R & R on Jun 2013. Post procedure angiogram runs revealed 80% reduction in the AVM nidus size (Nidus size 10 x 8mm on MRI Angiogram in Dec 2016 and 6.2mm diameter in MRI with Angiogram in Nov 2017). The patient remained asymptomatic for about 7yrs post treatment (till Jan 2020). In Jan 2020, patient presented with progressive left hemiparaesis. Investigations revealed an ill defined 16 x 12 x 14mm SOL in right parietal region with perilesional oedema and adjacent cystic lesions (tumefactive cysts) with toal conglomerulate measuring 44 x 41 x 55mm. CT angiogram and DSA revealed no patency of vessels at the location of the nidus. The patient was managed with decongestants and conservative measures, and the hemiparaesis improved (and so did the oedema radiologically). She presented again in Apr 2020 to a nearby hospital in Dehradun (due to COVID restrictions on inter-state travel) with left hemiplegia and altered sensorium. NCCT Head and MRI Brain revealed 30 x 20 x 10mm mass lesion in the right parietal region and surrounding oedema with adjacent tumefactive necrotic cysts (total volume 7 x 5 x 5cm) with mass effect and midline shift to the left side. She was initially given a trial of elective ventilation and decongestants, however there was no improvement in the neurological status and hence, Right Parietal Craniotomy and excision of the lesion was done.

Histopathology - blood clots, areas of infarction, multiple variable sized vessels and gliosed brain tissue with no features of any neoplasm (Fig 6A).

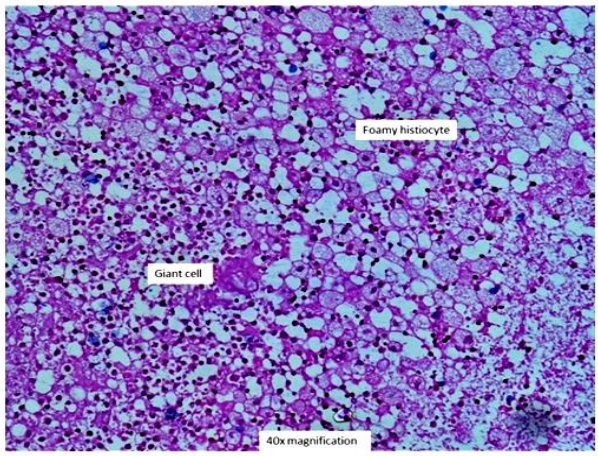


Fig 6A : HPE of case 2 Sheets of foamy histiocytes, inflammatory cells and an occasional giant cell

Case No. 3

Mr NK

This 43yrs old male patient was treated for AVM in Left Parietal region. Patient first presented with throbbing headache in Mar 2017. He was detected to be having superior sagittal sinus thrombosis with no abnormality on MR Angiogram. On follow up MRI with MR Angiogram and Venogram, he was found to be having partial recanalisation of superior sagittal sinus with Dural based AVM in right parietal region. He underwent glue embolisation in Dec 2018. Follow up MRI in Aug 2019 revealed an ill-defined mass lesion in the left parietal region (21 x 33 x 22mm) with mild perilesional edema. Follow up imaging (Aug 2020) revealed an increase in the size of this lesion (now 37 x 37 x26mm).

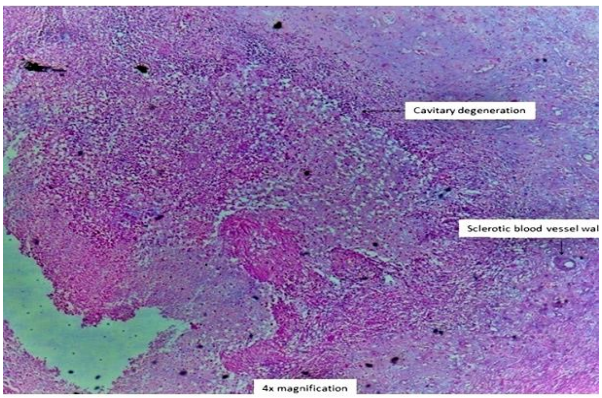


Fig 6B HPE of case 3 :- Cavitary degeneration containing foamy histiocytes and inflammatory cells with surrounding thickened sclerotic blood vessel walls

CT angiogram and DSA Cerebral vessels did not show any recanalisation of vascular components of the nidus. In view of progressive left hemiparaesis, radiological progression of the lesion and doubt in diagnosis, the patient was subjected to craniotomy and excision of the lesion on 26 Aug 2020.

Histopathology- glial brain tissue with foam cells and vascular elements consistent with AV malformation (post embolisation) (Fig 6B).

Case No 4

Mr SK

This 29yrs old male patient had an AVM in Lt Temporo-parietal region. Patient presented with seizures in Feb 2007. Investigations revealed AVM sized 65 x 63 x 66mm in Lt Temporo-parietal region. The patient underwent coil embolization for this AVM at Army Hospital R & R in 2007. Post procedure angiogram runs revealed 80-90% reduction in the AVM nidus size. He subsequently underwent Gamma Knife Radiosurgery in staged fashion on May 2017 and Feb 2018. The patient remained asymptomatic for 2yrs post treatment. In Jul 2019, patient presented with Right Hemiparaesis and dysphasia. NCCT Head revealed an ill defined SOL in the left Temporo-parietal region with perilesionaloedema, with adjacent tumefactive necrotic cysts and midline shift to Right side of 12mm. CT angiogram and DSA revealed 65 x 63 x56mm vascular lesion with minimal revascularisation. The patient was managed with decongestants and conservative measures, however the symptoms intensified and to relieve the raised Intracranial pressure the patient was subjected to Left frontotemporoparietal decompressive craniectomy and excision of the lesion on 27 Jul 2019.

Histopathology revealed a tangle of vessels and no features of any neoplastic transformation (Fig 6C).

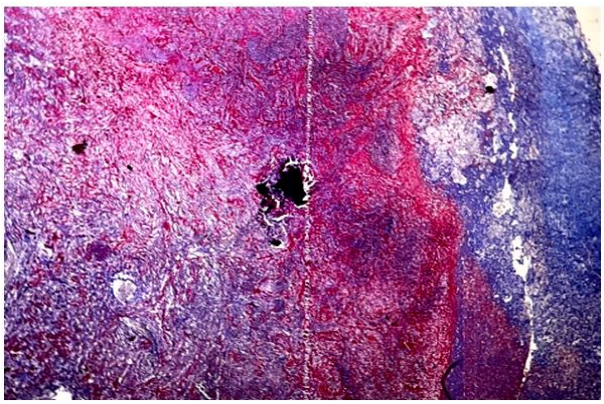
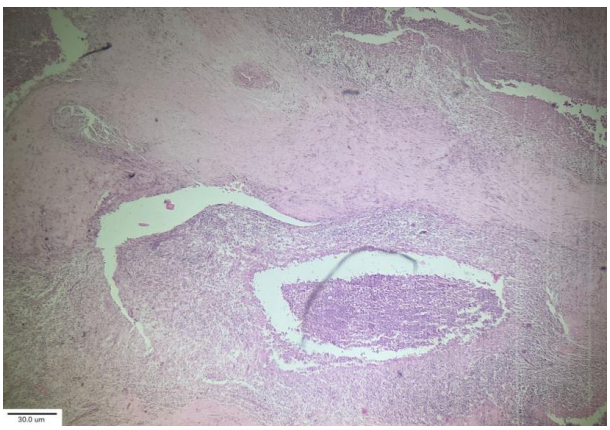


Fig 6C : HPE slide of Case 4, Masson Trichrome stain highlights Fibrin (reddish areas) and areas of fibrosis (Blue)

Case no 5

Mr HD

This 29yrs male presented with sudden onset right hemiparaesis on 04 Aug 24. His NCCT Head revelaed large bleed on left frontal region extending towards left basal ganglia. His DSA cerebral vessels revealed Lt frontal AVM 9x9.1x11mm in size with feeders from Lt MCA and draining vein draining to SSS. He was managed conservatively for the bleed and was taken up for endovascular embolisation of the AVM on 29 Aug 2024. He recovered symptomatically and his post procedure angiographic runs showed 80% obliteration of the nidus. He was discharged with follow up advice. He remained well till 16 Feb 2025 (~ 6months from initial treatment), when he developed right hemiparaesis and was admitted. His MRI Brain revealed a large 38 x 34 x 30mm mass lesion at the site of treated nidus. He was managed with decongestants and had slow, but appreciable symptomatic recovery. He was discharged and remained well for 3months, however in May 2025, he again had similar symptoms and was admitted. NCCT Head revealed mass lesion with massive surrounding oedema. He was given a trial of decongestants, however, he didn't show much improvement and was ultimately taken up for decompressive craniectomy with excision of the lesion on 19 Jun 2025. Histopathological examination of the sample revealed chronic inflammatory changes within congested blood vessels. (Fig 6d)



Result

We have described a clinical entity which effected five patients of cerebral AVM treated at our centre with embolisation or GKRS or both (in a sequential fashion) (**Table 2**), (out of total 106 cerebral AVM patients treated in the given time frame). At varying intervals from the index treatment, they presented with features of raised intracranial pressure and their management algorithm has been elucidated. The pathophysiology of the said complication has also been studied. In view of unresponsive cerebral oedema, they needed decompressive craniectomy and excision of the nidus. The histopathological examination of the excised lesion revealed vascular elements, glial brain tissue and no neoplastic transformation. They all responded well to the treatment.

Discussion

The ideal goal of cerebral AVM treatment is to completely obliterate the AVM nidus and normalization of cerebral hemodynamics[8]. This is to emphasise the possibility of delayed radiation necrosis at the site of treated AVM, presenting as a mass lesion with mass effect. Earlier, there have been isolated case reports for delayed presentation with increased intracranial pressure after Radiosurgery[10], however similar entity following embolisation has not been reported. The management of such an entity should be initially with decongestants, however in non-responsive cases decompression of the lesion with or without decompressive craniectomy may be needed.

The preoperative evaluation of such cases is aimed at:

a) Detecting whether there is a residual or recanalised AVM nidus with or without rebleed
b) If there is no AVM nidus or rebleed identified, then to differentiate between radiation necrosis and post radiation malignant transformation

Various modalities which can be instituted to differentiate between radiation necrosis and malignant tranformation are:

a) NCCT Head and CEMRI Brain – indirect evidences like cystic degenration more common with radiation necrosis, enhancing lesions and distant non contiguous lesions more suggestive of malignant transformation
b) MR Perfusion Imaging – increase in local perfusion is indicative of malignant transformation
   (i) PET Scan - FDG PET has a sensitivity of 65-81% and specificity of 40-94% to differentiate between radiation necrosis and tumour recurrence.

Other PET modalities:

(ii) F-F DOPA: 3,4 dihydroxy 6,18F-fluoro-L-Phenylalanine has a specificity of 86% and sensitivity- 98% to differentiate between radiation necrosis and tumour recurrence.

(iii) F-FET: O-2-18F-fluoroethyl-L-tyrosine has specificity of 100% and sensitivity- 93%

(iv) C-METL-methyl-11C-methionine has specificity- 83% and sensitivity- 88%

However, it is important to note that these statistics of the PET Scan modalities have been validated for known cases of neoplastic conditions, which underwent radiation therapy and are on follow up and may not hold true for the Post GKRS cases of cerebral AVMs.

Suggested Pathophysiology of this complication after embolisation is likely to be related to the normal perfusion pressure breakthrough (NPPB), impaired venous drainage, and embolization-induced angiogenesis. This in turn means formation of collaterals around the embolised arterial elements. These may be vessels with abnormal wall and permeability, thus causing gradual extravasation around the nidus, ultimately leading to cumulative oedema and presentation as a mass lesion. Pathophysiology following GKRS is more elusive and may be related partially to radiation necrosis and partially to neoangiogenesis.

Even after a deliberate investigatory protocol, it may not be possible, at least in few cases, to conclusively differentiate between post radiation necrosis and malignant transformation, and diagnostic dilemma may form an independent indication to operate, especially in younger individuals. Increasing, non-responsive oedema with raised intracranial pressure and consequent neurological deficits may warrant surgical decompression with removal of the conglomerate at the AVM site, in its own merit.

This is to generate familiarization about this entity, which may either be under treated considering it to be a post treatment effect or over treated considering it to be malignant transformation of a benign lesion. However, when there is a persistent diagnostic dilemma, compounded with the non-responsive clinical condition of the patient, surgical excision should be readily offered. The epidemiological profile of the patient may also play a role in decision making.

This highlights the need of cautious follow up of these patients for a longer duration as two of these five patients developed the complication with raised intracranial pressure more than 5years after the embolisation procedure. To gain insight into the pathophysiology of such delayed cerebral reaction to a procedure, more cases of similar kind need to be studied with inclusion and exclusion of all scientifically important information.

Table 1. Spetzler Martin Grading

Spetzler- Martin Grading	Points	Supplementary Grading
Size, cm		**Age (in yrs)**
<3cm	1	<20
3-6cm	2	20-40
>6cm	3	>40
Venous Drainage		**Bleeding**
Superficial	0	Yes
Deep	1	No
Eloquence		**Compactness**
No	0	Yes
Yes	1	No
Total	**5**	

Conclusion

The follow up of a group of AVM patients treated with embolization/ GKRS revealed that, after a duration ranging from 1.5-12yrs, these patients presented with a mass lesion with significant perilesional oedema at the site of preexisting AVM. CT Angiogram and DSA revealed no significant flow in the vascular malformation. They were subjected to decompressive craniectomy and excision of the nidus. The histopathological examination of the excised lesion revealed vascular elements, glial brain tissue and no neoplastic transformation. This description is to notify a rare, serious and hitherto unexplained complication of cerebral AVM patients undergoing embolisation/ GKRS. Further studies with larger patient subsets are required to establish the pathophysiology of such a complication and also predictive factors at the time of index treatment.

Table 2. Summary of cases with course of illness and treatment rendered

S No	Age	Sex	Location of lesion	Size of lesion	Time of Treat-ment	Details of treatment rendered	Time lag for Symptom appearance after embolization	Deficits	Resolution of symptoms after craniotomy
1	22y	F	Left Temporal	6.5cc	2012	GKRS	9yrs	Headache, vomiting nuchal pain	Recovered
2	39y	F	Rt Parietal region	46 x 38 x 49mm	Jun 2013	Gamma Knife Radiosurgery	7yrs	Left hemiparaesis, LOC	Consious, oriented, hemiparaesis improving
3	43y	M	Lt parietal region	21 x 12 x 18mm	Dec 2018	Embolisation	1.5yrs	Right hemiparaesis	hemiparaesis improving
4	30y	M	Lt temporo-parietal region	65 x 63 x 66mm	2007 Feb 2018	Embolisation Gamma Knife Radiosurgery	12yrs post embolisation 1.5yrs post GKRS	Right hemiparaesis, Dysphasia	Partial resolution
5	29y	M	Lt Frontal		Oct 2024	Embolisation (onyx)	6months	Rt hemiparaesis	Weakness completely recovered

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Neurofibromatosis type 1 associated with multiple internal and external anterior abdominal wall defects. A case report

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Keywords
congenital hydrocele, diastasis recti, neurofibromatosis, umbilical hernia

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ABSTRACT

Neurofibromatosis type 1 is a relatively common autosomal dominantly inherited genetic disorder. It is characterised by variable clinical manifestations, including café-au-lait spots, axillary or inguinal freckling, cutaneous neurofibromas, plexiform neurofibroma, bony lesions, optic glioma, and iris Lisch nodules, which constitute the clinical diagnostic criteria. In addition, a wide range of systemic abnormalities, including skeletal deformities, cardiovascular anomalies, neurocognitive deficits, as well as nervous system and non-nervous system tumours, have been described in patients with NF1. We present a previously unreported systemic association in NF 1 in an adolescent male: the presence of bilateral congenital hydrocele, divarication of rectus abdominis muscles and umbilical hernia, all external and internal defects of the anterior abdominal wall.

INTRODUCTION

Neurofibromatoses (NF) are a relatively common genetic disorders with autosomal dominant inheritance[7]. They are a heterogenous group of diseases with high predisposition to the development of benign and malignant tumours affecting different parts of the body[9, 21]. Three types of NF, neurofibromatosis type 1 (NF1), neurofibromatosis type 2 (NF2), and shwannomatosis, have been described[15]. NF1 is the most common type accounting for about 96% of the cases[14, 17]. It occurs in 1 in 3500 live births and is characterized by variable expression of clinical manifestations including café-au-lait spots, axillary or inguinal freckling,

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cutaneous neurofibromas, plexiform neurofibromas, bony lesions, optic glioma, and iris Lisch nodules[20]. A number of other abnormalities including skeletal deformities, cardiovascular anormalies, neurocognitive deficits, and several nervous and non-nervous system tumours have been described in patients with NF1[9]. However, to the best of our knowledge, co-existence of bilateral congenital hydrocele, divarication of rectus abdominis muscles (DRAM) and umbilical hernia in a patient with NF1 has not been previously reported in the English literature. One other example of such rare associations, albeit intracranial, reported from Africa not too long ago was that of pleomorphic xanthoastrocytoma in another boy with NF1[2].

In this report we present an 11 year old boy with NF1 and a novel spectrum of systemic associations involving both internal and external anterior abdominal wall defects.

Case Description

An 11 year old boy first presented at our hospital aged 2 years with painless bilateral scrotal swellings. The swellings were first noticed about 18 months earlier and had been increasing in size progressively. He had been a product of term pregnancy given birth to at gestational age of 39 weeks and 2 days. The birth weight was 3.2kg. Examination at this first hospital presentation in childhood revealed non-tender, brilliantly transilluminating bilateral scrotal masses. In addition, there were a 2-cm separation of the rectus abdominis muscles, a 1-cm umbilical defect, and a 2cm right sided occipital mass. A clinical diagnosis of bilateral congenital hydrocele with divarication of the recti abdominis and umbilical hernia was made. He underwent surgical procedures of bilateral herniotomy and hydrocelectomy; while the divarication of the abdominal recti and the umbilical hernia were managed non-operatively.

The boy presented in our hospital 9 years later with a progressively enlarging occipital swelling (Fig. 1). The swelling was first observed at birth as a < 2cm painless occipital skin mass, and was the same one that had been documented at the first hospital presentation for the abdominal wall defects. The mass gradually increased in size over time till review, necessitating presentation because of the cosmetic blemish it had become. There was a paternal family history of dark pigmented skin patches and neurofibromas. Clinical examination showed a soft 10cm by 8cm by 3cm largely right sided, painless, occipital mass with overlying dark skin patch; it had no attachment to the underlying bone. He also had multiple cafe-au-lait spots over the skin of the limbs and the trunk (Fig. 2), as well as iris harmatomas. There were bilateral hairline inguinal scars (Fig 3). The DRAM had resolved spontaneously while the umbilical defect has reduced significantly (about 3mm residual defect).

A clinical diagnosis of plexiform neurofibroma in a patient with neurofibromatosis type 1 was made. He subsequently underwent excisional biopsy of the occipital mass which was histologically confirmed to be a plexiform neurofibroma (Fig 4).

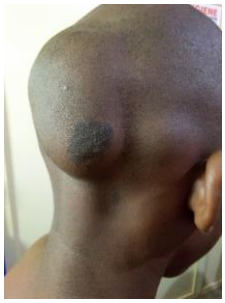

Figure 1. 10cm by 8cm occipital plexiform neurofibroma in our patient.

Figure 2. Photograph of the patient showing multiple cafe-au-lait spots.

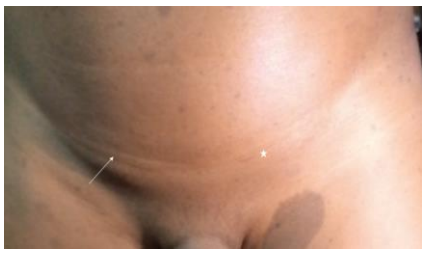

Figure 3. Photograph of the patient showing the well-healed scars of the previous bilateral herniotomy (arrow on the right, star on the left).

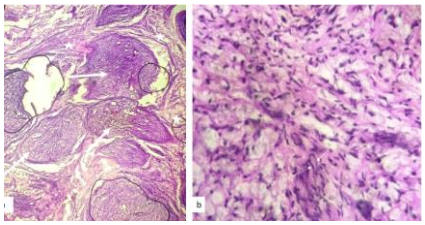

Figure 4. (a) low power (x4) photomicrograph of the lesion's histology specimen showing spindle cells (arrows) separated by fibrocollagenous stroma (stars). (b) high power (x40)

photomicrograph showing elongated spindle cells with wavy nuclei (arrows).

Discussion

Neurofibromatosis type 1 (NF1), the most frequent of the neurofibromatoses, is a relatively common genetic disorder[7, 15]. It is largely diagnosed clinically using well defined criteria[9, 12]. A wide range of abnormalities including cutaneous lesions, skeletal deformities, cardiovascular defects, neurocognitive deficits, as well as nervous system and non-nervous system tumours have been described in patients with NF1[9]. Not previously reported however, to the best of our knowledge, is the occurrence of multiple anterior abdominal wall defects both external and internal in a patient with NF 1. We present one case of such, an 11-year-old boy who was managed at our hospital at the age of 2 years for divarication of rectus abdominis muscles, umbilical hernia and bilateral congenital hydrocele but presented 9 years later with an occipital mass that was increasing in size progressively; surgical pathology of this lesion post-excision was reported as plexiform neurofibroma.

Neurofibromatosis type 1 results from a germline mutation in the NF1 gene, a tumour suppressor gene located on the long arm of chromosome 17 (17q11.2). It encodes neurofibromin which is a negative regulator of the Ras proto-oncogene, a key signalling molecule in cell growth and differentiation[8, 10, 18]. Divarication of rectus abdominis muscles (DRAM) is very rare and is usually congenital when present[11]. Congenital DRAM can occur as autosomal dominant transmission without associated syndrome or sequence; or as a part of some clinical syndromes like the prune-belly, Beckwith-Wiedemann, Simpson-Golabi-Behmel, and some other midline-defect syndromes[3, 4, 5, 16].

In contrast, the exact cause of umbilical hernias is unknown, although higher incidence has been reported in African, and Africa-American infants compared to Caucasians[1]. Greater risks of umbilical hernia has also been documented in premature and low-birth-weight infants, children with metabolic disorders like hypothyroidism; autosomal trisomies like trisomy 18 and 21; as well as syndromes like Beckwith-Wiedemann, Marfan and Simpson-Golabi-Behmel[22, 23]. As for congenital hydroceles, they result from failure of closure of the Processus Vaginalis, the exact pathogenesis of which is still a subject of controversy[6]. Familial inheritance[6, 13], and association of hydrocele with the FG syndrome (Opitz-Kaveggia syndrome)[19], have however been documented.

Apart from the clinically documented family history of NF1 in the patient presented in this report, we are not in the position to provide any other genetic / familial explanation for the associations with the internal and external anterior abdominal wall defects seen in him. Perhaps it is a mere coincidence or maybe there is an as yet unidentified genetic or familial predispositions. Most non-syndromic DRAM, paediatric umbilical hernia, and congenital hydroceles resolve spontaneously, surgery being indicated in the latter if the hydrocele persists beyond the age of 2 years. Our patient had surgery for the hydrocele at age 2 years and was managed non-operatively for the DRAM and umbilical hernia to a good outcome.

Plexiform neurofibromas develop in about 30-50% of patients with NF1 and carry a lifetime risk of malignant transformation[9]. Indication for surgery in plexiform neurofibroma include pain referrable to the lesion, functional compromise, and cosmetic disfigurement[20]. Our patient presented and had excision of his occipital lesion because of the latter reason.

Conclusion

NF1 is a relatively common inherited disorder with a wide range of associated anomalies. We have presented what must be a novel clinical spectrum of associations of NF 1 in an 11-year-old adolescent boy from Nigeria: a rare case of divarication of rectus abdominis muscles, umbilical hernia and bilateral congenital hydrocele in a child who fulfilled the clinical criteria for NF1. He was managed earlier in childhood for the associated abdominal wall lesions but presented 9 years later for the management of an occipital plexiform neurofibroma because of its cosmetic disfigurement.

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Calcified chronic subdural hematoma with subacute presentation. A case report

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ABSTRACT

Chronic subdural hematomas are a very common disease occurring spontaneously, following trauma, ventriculoperitoneal shunt and postcranial surgeries, with an incidence of 1.72-20.6 per 100,000 persons per year. But sometimes calcification may occur over the outer layer of membrane or within the collection, which may occur from 6 months to many years [1]. The exact aetiology and pathogenesis of calcification remains unknown; it was first reported by a Bohemian pathologist carl von Rokitansky, in 1884[2]. Due to its appearance sometimes, it's calledan armoured brain [3] or matryoshka head after Russian nesting dolls [4]. Though limited cases have been reported majority of cases are asymptomatic, and surgical management is advocated for those having mass effect or neurological deficit. Inan article published in 2020 by Turgut et al total of 114 cases have been reported [5]. The total incidence of calcification in chronic subdural hematoma is about 02-2.7% [6]. We report a case that presented with a neurological deficit and was treated surgically with craniectomy and complete excision.

Keywords
calcified chronic subdural haematoma, calcification

CASE REPORT

A 63-year male patient presented with progressive weakness of right side of upper limb followed by lower limb for the last 18- months along with altered sensorium and headache for past 7 days. Upon examination patient was having GCS- 13 with both pupils reacting to light equally, vitals were normal and motor examination revealed power of 4/5 on right upper and lower limb with reduced touch sensation and two point discrimination. Patient was not having any history of trauma, fever, hypertension, diabetes mellitus, medications or any cranial surgery.

Patient Underwent CT-scan which showed large well defined extra-axial hypo dense collection with calcification on left front-temporo-parietal region with thickness of 4.5 cm and mass effect of 1.5 cm. [Fig.1-A]. Upon MRI examination it revealed large subdural sub acute hematoma over left cerebral convexity and brain parenchymal compression. [fig. 1-b] patient also underwent digital subtraction angiography (dsa) 4 vessel which was unremarkable.

Patient was planned for surgical intervention and left aided fronto tempura parietal craniectomy was performed and well defined subdural collection was found upon bone removal. [fig.2-a] Which

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was not adherent to underlying arachnoid and It was easily dissected and stripped off without any damage to underlying arachnoid or pia. [Fig2-B] The overlying Dural defect was covered with pericranium, the excised mass was showing altered blood products with soft to firm consistency and foul smell. Patient had not complications post-surgery and post operative CT Image showed complete removal of mass without any residual or recent changes. Patient was Discharged on 5th postoperative day and with GCS- of 15 with right sided motor weakness improved on 3 months follow up visit.

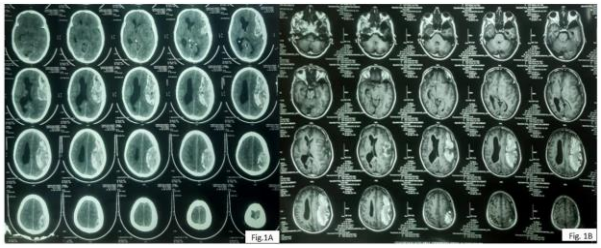

Figure 1-a. Ct scan showing large extra axial collection with calcification on left fronto-temporo-parietal region.

Figure 1-b. T1 mri image showing large hyperdense area over left fronto-temporo-parietal region subdural space.

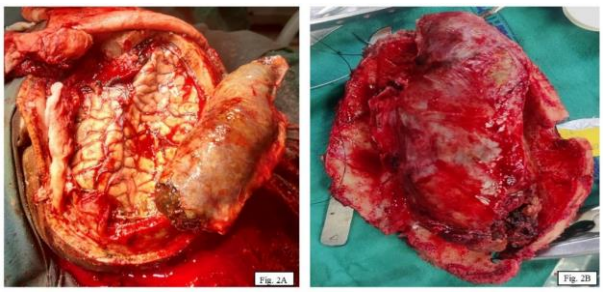

Figure 2-A. Shows craniectomy and large organized subdural collection that was easily seperated from underlying arachnoid with compressed brain parenchyma, foul smelling collection was there.

Figure 2-b. Complete excision of collection en-mass with both layers and bone specimen, collection is mixed old and altered blood.

Discussion

Subdural hematoma usually progress to chronic form over period of 7 to 21 days with cause being trauma, infection, ventricular shunt over drainage, post-surgical. Anticoagulant & antiplatelet use and coagulation disorders. The pathogenesis of subdural hematoma from acute to chronic stage have been well studied and local and systemic factors have roles with neomembrane formation with collagen deposition and liquefaction of the collection. But why some chronic subdural hematomas progress with calcifications stays unknown and some vascular thrombosis and calcification of the membrane have been thought to be responsible which usually presents as calcification of inner, outer or both the membranes of chronic subdural hematomas [7].

The process of calcification has been found to be occurring over the period from 6 months to many years and is more common in children's than young adults or old age persons [1]. Many times, repeated bleeding into the cavity have also been found as a causative factor for calcification. Upon radiological investigations it appears like helmet inside skull and given name armored brain [3] or matryoshka head [4] appearance after similarities with russian nesting dolls. When the calcification progresses to final stage of ossification it can be called ossified chronic subdural hematomas.[8]

The treatment of these entities is unanimous and ranges from conservative in old age patients to twist drill & burr hole craniostomy to complete craniotomy/craniectomy and complete removal depending upon location, thickness and neurological status of patient [9]. Satisfactory results have been found with and without membrane removal without damaging the underlying brain parenchyma without any re-accumulation of blood [10]. Our patient was having very thick collection with calcification, so he underwent craniectomy and complete excision of hematoma and inner membrane.

Conclusion

Calcified chronic subdural hematomas are rare entity with unknown pathogenesis and not guidelines about the management. Most cases present with headache, altered consciousness or neurological deficit and all cases can be easily diagnosed with ct scan and mri. The management should be tailored according to the individual hematoma characteristics and patient neurological and overall status from

conservative follow-up, burr hole drainage to craniotomy and removal.

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Retained knife fragment in thoracic spine following stab injury. A rare case with full neurological recovery

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ABSTRACT

This case report presents a rare instance of a retained knife fragment within the thoracic spine following a stab injury in a 24-year-old male. The patient presented with stable vitals and no neurological deficit despite the knife penetrating near the spinal cord at the T9–T10 level. Prompt imaging, surgical intervention, and multidisciplinary coordination resulted in complete recovery without a lasting neurological deficit. This case highlights the importance of rapid assessment and careful surgical management in preventing secondary injury.

Keywords
penetrating spinal injury, retained knife, thoracic spine, neurological recovery, neurotrauma

INTRODUCTION

Penetrating spinal injuries constitute a relatively uncommon subset of spinal trauma, accounting for a small percentage of all spinal cord injuries worldwide. Their epidemiology and mechanisms vary significantly depending on geographic, social, and economic factors. According to Wyndaele and Wyndaele (2006), the global incidence of spinal cord injuries ranges from 10 to 83 cases per million population annually, with stab and knife injuries forming a minor but clinically significant proportion [1]. These injuries most frequently involve the thoracic region, owing to its relative immobility and narrow canal diameter [2].

The neurological presentation in such cases is diverse and depends on the depth and trajectory of penetration. Waters et al. (1995) reported that stab-induced spinal injuries often present with incomplete lesions and demonstrate better recovery potential than blunt trauma, given the localized nature of cord damage [2]. However, retained knife fragments within the spinal canal are particularly rare, posing both diagnostic and therapeutic challenges. Early radiological assessment and multidisciplinary coordination are critical for preventing secondary cord injury. The present case adds to the limited literature describing thoracic spinal stab wounds with retained metallic fragments but preserved neurological function, emphasizing the importance of timely surgical management.

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CASE REPORT

A 24-year-old male presented to the emergency casualty after being stabbed during a physical assault. On examination, two linear stab wounds measuring approximately 3–5 cm × 1–2 cm and 4-5 cm × 1-2 cm were noted in the left sub-scapular paraspinal region, with active bleeding at presentation. The patient was hemodynamically stable (GCS 15/15) and demonstrated full motor power (5/5) in all four limbs with no neurological deficit. An initial X-ray suggested possible thoracic involvement (Figure 1).

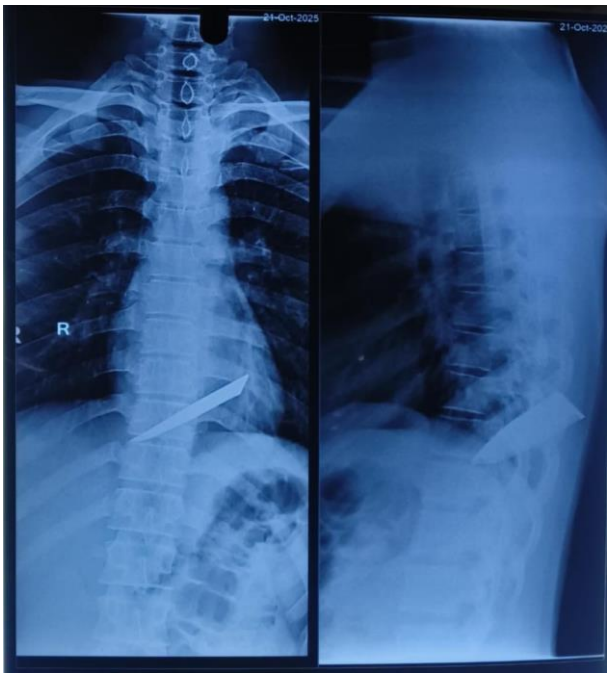

Figure 1. Chest X-ray showing the retained knife fragment.

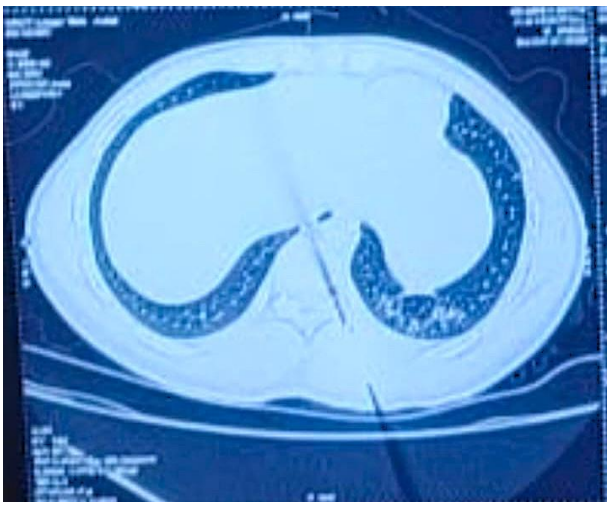

Figure 2. Knife blade around the vertebral body in HRCT.

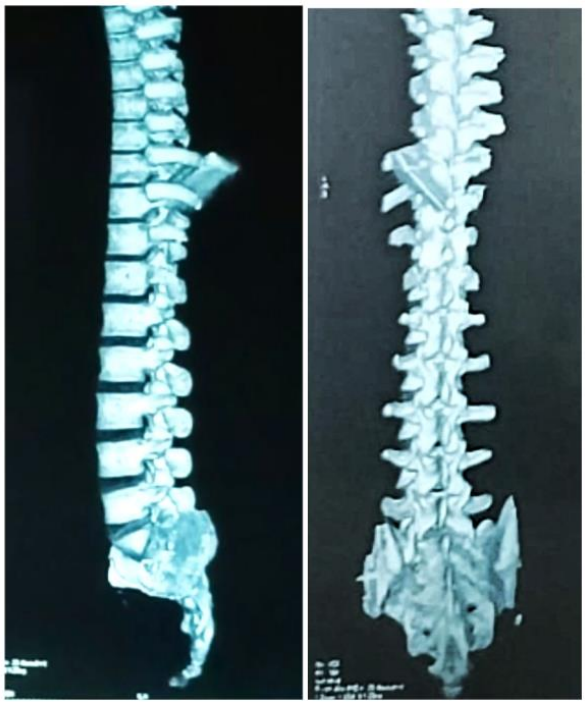

Figure 3. 3DCT spine image showing the knife in the vertebra at the T9-T10 level.

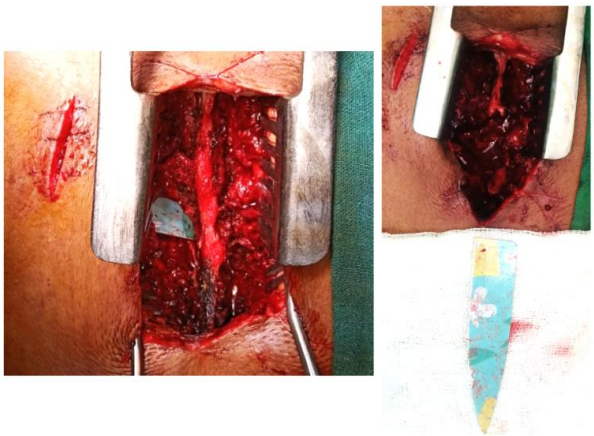

Figure 4. Retained knife in the vertebra and its comparison to the incision wound and nearby stab wound.

After wound closure and stabilization, HRCT thorax and 3D spine reconstruction were performed, revealing a retained metallic knife fragment tracking towards the vertebral body at T9–T10 (Figures 2 and 3). The patient underwent surgery after consent and preparation. A midline posterior incision was made, and the muscle planes were separated under X-ray guidance. Partial laminectomy was performed, revealing the knife transecting the posterior spinal cord and dura (Figure 4). The knife was carefully extracted, dura repaired, hemostasis ensured, and

closure done with drain placement. Postoperatively, the patient developed mild bilateral lower limb weakness (power 4/5). MRI findings suggested postoperative edema, and the patient was managed conservatively with physiotherapy, leading to full neurological recovery. Postoperative X-rays confirmed successful removal (Figure 5). The patient was discharged without any residual deficit.

Figure 5. Postoperative MRI suggesting postoperative edema.

Discussion

Penetrating spinal injuries from sharp objects such as knives are most often the result of interpersonal violence, and their management requires a precise understanding of spinal anatomy and the mechanism of injury [1]. While gunshot wounds dominate spinal trauma in Western literature, knife-induced injuries remain more common in developing countries, often affecting young males in assault-related scenarios [3].

The absence of neurological deficit in the presence of a retained intramedullary or intradural knife fragment, as seen in this patient, is exceptionally rare. Li et al. (2012) described a similar thoracic stab injury with a retained knife blade and no neurological deficit, underscoring that the final outcome depends largely on the injury trajectory and avoidance of vascular or neural structures [4]. Early and accurate imaging, including CT and 3D reconstruction, remains essential for localization and surgical planning, minimizing intraoperative risk.

Surgical extraction should be performed under direct visualization to prevent further cord damage. In our case, careful laminectomy and controlled removal of the blade, followed by dural repair and hemostasis, resulted in complete neurological recovery—consistent with the favorable outcomes reported by Ndoumbe et al. (2015), where early intervention and absence of preoperative deficits predicted better prognosis [3].

Overall, literature supports that patients with stab-related spinal trauma, particularly those with incomplete or no initial neurological deficits, exhibit a high potential for functional recovery [2, 3]. This reinforces the critical role of multidisciplinary management involving emergency, radiology, and neurosurgical teams in optimizing outcomes for such rare and high-risk injuries.

Conclusion

Early imaging, surgical precision, and coordinated multidisciplinary management led to full recovery without neurological deficit in this rare case of retained knife fragment within the thoracic spine. Such cases highlight the potential for excellent outcomes even in penetrating spinal trauma when managed promptly and appropriately.

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Management of Moyamoya disease. A systematic review and meta-analysis on surgical revascularization, outcomes and clinical manifestations

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Keywords
Moyamoya disease, internal carotid arteries (ICA), direct bypass, indirect bypass, artery temporal superficial, middle cerebral artery, revascularisation

ABSTRACT

Background: Moyamoya disease (MDD), defined by a chronic, progressive stenosis of the terminal portion of the internal carotid arteries (ICA) on both sides, carries the anomalous vascular information network, which functions as a collateral pathway to the brain. The aim is to understand the management of moyamoya disease (MMD) in terms of the approaches and different types of arterial revascularisations (direct, indirect, and combined), regardless of the pathological mechanism of origin to be investigated.

Materials and methods: A review was conducted following the PRISMA (Preferred Reporting Items for Systematic Reviews and Meta-Analyses) guidelines. The review focused on moyamoya disease, outcomes before and after revascularisation, and related treatment modalities (STA-MCA) in various databases, including ScienceDirect and PubMed/MEDLINE, using the PRISMA guidelines, R, and Excel. Only studies published in English up to August 2025 were included.

Results: Our systematic review included 4800 patients, comprising (N=2866, 60%), patients, (N=2024,42%), surgical interventions, and (N=2112,44%,), revascularisations. See Table 1. (N=1199, 25%), patients included (N=977, 20%), surgical approaches (N=615, 13%), and revascularisations.

Conclusion: This study has shown that moyamoya disease (MMD) is a vascular concern that, regardless of ethnicity, is very rare; cases have been reported in Europe and the United States, as well as in the Hispanic population, but none have been reported in African regions. Therefore, encountering this pathology cannot be ruled out, and one should be up-to-date on the types of revascularisations, whether STA-MCA, ACA-PCA.

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Introduction

Moyamoya disease (MDD), defined by a chronic, progressive stenosis of the terminal portion of the internal carotid arteries (ICA) on both sides, carries the anomalous vascular information network, which functions as a collateral pathway to the brain stem. is understood from its appearance as a smoke-like vascular network on cerebral angiography, which was termed "moyamoya." Angiography for this disease was described in mid-1957, for both carotid arteries or bilaterally, and designated as a pathological entity in the 1960s. [1]. MMD, we proceed with a bypass of the superficial artery to the middle cerebral artery. (STA-MCA); this is the surgical method of choice for moyamoya disease (MMD); at the time of ischemic onset, it will improve cerebral blood flow. This method, by increasing flow, is also essential in preventing rebleeding in patients with moyamoya without early hemorrhagic disease. When they undergo bypass surgery, perioperative catheterization will be included, which will be essential in assessing postoperative complications. [2].

New studies indicate that RNF213 can modify the E3 ubiquitin ligase associated with the East Asian gene. Another variant of this gene is the non-Arg4810Lys variant of RNF213, which is associated with moyamoya disease in European countries, etc. This Arg4810Lys variant is said to be the most strongly genetically associated and is generally associated with moyamoya disease. [3]. MMD: There are factors that influence stenosis, its vascularization, and angiogenesis. The pathophysiology includes surgical and nonsurgical approaches that influence the pathogenesis so much that they tend to halt and slow its progression. Therefore, the intervention is surgical revascularization through direct and indirect bypass techniques, leading to the restoration of cerebral hypoperfusion. [4]. Moyamoya disease can develop in both children and adults, although its characteristics manifest differently. In children, ischemia is a common manifestation, presenting with an irreversible and progressive deterioration of nerve function with an unknown pathogenesis, unlike in adults, who begin with bleeding and require early intervention. It should precede treatment of the disease, which can be classified as direct and indirect bypass surgeries with orifice trepanation with anastomosis of the superficial temporal artery (STA), encephalomyosynangiosis (EMS), and encephaloduroarteriosynangiosis (EDAS), direct surgery tends to reconstruct intra-extracranial vessels with anastomosis of the superficial temporal artery and/or the middle cerebral artery (STA-MCA). Indirect surgery in children with moyamoya has been shown to be effective. Although both direct and indirect surgery are recommended, they tend to have a better response in children. [5].

Material and Methods

A review was conducted following the PRISMA (Preferred Reporting Items for Systematic Reviews and Meta-Analyses) guidelines. The review focused on moyamoya disease, outcomes before and after revascularization, and related treatment modalities (STA-MCA). A comprehensive search was conducted in various databases, including ScienceDirect and PubMed/MEDLINE, using the PRISMA guidelines, R, and Excel. Search terms included "Moyamoya disease, MMD" in children and adults, along with terms specifying surgical therapies and techniques, different rehabilitation methods, and pathologies associated with the disease. Only studies published in English up to August 2025 were included.

The PICO (Population, Intervention, Comparison, Outcome) framework was used to define the study population, focusing on patients aged 1 to 75 years with moyamoya disease (Figure 1). Search Strategy and MeSH Terms

The search strategy incorporated MeSH terms (Medical Subject Headings) related to revascularization, both direct and indirect, in Moyamoya disease (MMD), with a focus on disease management.

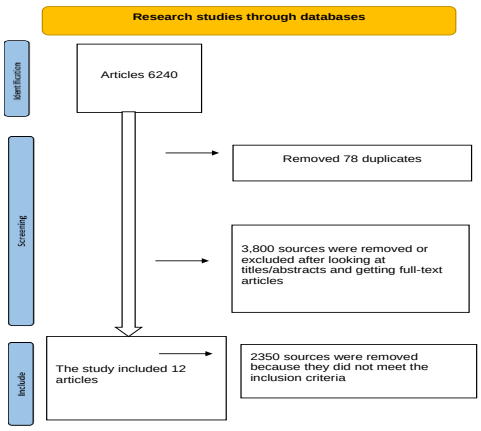

Figure 1. Flowchart of the articles selected according to the PRISMA protocol base of Moyamoya disease, (MMD).

Comprehensive Search Strategy Keywords

The comprehensive search strategy incorporated the following MeSH terms:

Moyamoya disease/inmmunotherapy"[Mesh]
Moyamoya disease/nanotechnology"[Mesh]
Moyamoya disease/cerebrospinal fluid"[Mesh]
Moyamoya disease/chemically induced"[Mesh]
Moyamoya disease/complications"[Mesh]
Moyamoya disease/congenital"[Mesh]
Moyamoya diseasediagnosis"[Mesh]
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Moyamoya disease/genetics"[Mesh]
Moyamoya disease/history"[Mesh]
Moyamoya disease/immunology"[Mesh]
Moyamoya disease/metabolism"[Mesh]
Moyamoya disease/microbiology"[Mesh]
Moyamoya disease/mortality"[Mesh]
Moyamoya disease/pathology"[Mesh]
Moyamoya disease/physiopathology"[Mesh]
Moyamoya disease/prevention and control"[Mesh]
Moyamoya disease/psychology"[Mesh]
Moyamoya disease/radiotherapy"[Mesh]
Moyamoya disease/rehabilitation"[Mesh]
Moyamoya disease/surgery"[Mesh]
Moyamoya disease/therapy"[Mesh]

Keywords

Additional keywords included "Revascularization of Moyamoya disease," "anastomosis" "Carotid arteries" "Direct bypass," "Indirect bypass," "Artery temporal superficial," "Middle cerebral artery" and "Surgical Approaches.

Data Collection

We collected data from the included studies covering various aspects of the manifestations of Moyamoya disease (MMD). We also included diagnosis, clinical and surgical management, and treatment. Factors influencing the genetics of the RNF213 Arg4810Lys variant were examined, along with details on surgical or conservative techniques, as well as revascularization as the treatment for patients with MMD. Novel therapies for anastomosis, which induce recovery and regeneration of arterial circulation, were also reviewed.

Data Extraction

Data extraction from studies that met the inclusion criteria was performed using a standard, generalized search for the study of Moyamoya disease (MMD). Relevant information related to therapy or immunotherapy for MMD was extracted from manuscripts that met the search methods. We focused on relevant studies on MDD, population demographics, details of the intervention and control, and the methodology used in each study.

Risk of Bias in Individual Studies

In each study, we assessed risk of bias and applicability issues using different Kaplan survival methodologies, along with revascularization of the affected arteries, both the superficial temporal arteries and the middle cerebral artery, restoring their flow and anatomy in MMD. The datasets analyzed focused on various current and modern treatments for Moyamoya disease in children and adults, as we searched and focused on MMD and its genetic, molecular, and cellular factors. This review included standard protocols for conventional and surgical therapeutic removal and treatment.

Statistical Analysis

For each data point and relevant event, the odds ratio (OR) with the mean difference as the summary statistic was used to analyze MMD. The outcomes of interest were defined using the 95% confidence interval (CI) and the weighted mean difference (OR). A random-effects model was used to estimate outcome measures using data from MMD patients from the included studies to date. R software 5.3 and Excel were used for all primary and subgroup analyses, and a P value of 0.05 was considered statistically significant.

RESULTS

Our systematic review included 4800 patients, comprising (N=2866, 60%) patients, (N=2024,42%), surgical interventions, and (N=2112,44%,) revascularizations. See Table 1. (N=1199, 25%), patients included (N=977, 20%), surgical approaches, and (N=615, 13%), patients included revascularizations. See Table 2. (N=735, 15%), patients from three studies were also included.

(N=297, 6%), direct bypass procedures are the most common, (N=1702, 35%), and indirect bypass studies. See Table 3. A 2023 analysis of a randomized controlled trial in adult moyamoya patients that included bilateral direct revascularization versus therapy for hemorrhagic MMD demonstrated a reduction in revascularization with surgery (2.7%/year vs. 7.6%/year; P = .04). This study demonstrated that hemorrhages originating posteriorly from the cerebral or posterior choroidal arteries could rebleed and would therefore benefit from reintervention. Nearly all studies strongly support revascularization as beneficial for patients with MMD symptoms. Therefore, the positive findings were limited to patients with hemorrhagic MMD who underwent surgery. While in patients with ischemic MMD it has been seen in class 2a, its limitation was found with type C evidence, according to the AHA/American Stroke Association GUIDE, 2021, established as beneficial revascularization for the same prevention of ischemic stroke and/or transient ischemic stroke, TIA [6].

A 2024 retrospective, multicenter cohort comparative investigation aimed to determine the differences in stroke onset between moyamoya disease and moyamoya syndrome. Both surgical and conservative therapies were assessed by multivariate Cox regression analysis. Out of 2,565 patients, 2,349 diagnosed with MMD were included. No significant difference was observed between the two groups; nevertheless, it was demonstrated that individuals undergoing surgical treatment had a reduced incidence compared to those receiving conservative treatment. Hazard ratio (HR): 0.487; 95% confidence interval (CI): 0.334-0.711; $p < 0.001$. The modified Rankin scale prior to admission was > 2 (HR: 3.139; 95% CI: 1.254-7.857; $p = 0.015$), and perioperative problems were noted (HR: 8.666; 95% CI: 3.476-21.604; $p < 0.001$). The difference is that stroke implications rose in patients with moyamoya syndrome, although both groups benefited from revascularization, therefore it was indicated to urge and minimize perioperative problems as a way of preventive [40].

A supplementary retrospective investigation with 65 individuals aimed to elucidate the distinctions in clinical symptoms between moyamoya illness and moyamoya syndrome. Eighteen percent of patients were classified as asymptomatic with an underlying illness, compared with 82% of patients with moyamoya disease. Evaluating neurological signs, 66% of both groups had cerebrovascular manifestations, with ischemic stroke present or not, and 32%–42% without hemorrhagic strokes. Headache was prevalent in 18% of patients, and only 26% had phenotypes. Patients with ≥2 presented with stroke, TIA, headache, and seizures. Imaging studies demonstrated ≥1 ischemic lesion, while posterior circulation lesions were impacted in 51%. Forty-seven patients underwent surgical procedures, and 45 patients were administered aspirin. Subsequently, following diagnosis, 12 patients suffered additional strokes. From symptom onset and stroke incidence to follow-up, only 5 instances were seen per year, 26%; 19 patients presented with intellectual disability, 8 of whom had epilepsy, while 7 had behavioral difficulties. Therefore, in patients with MMD vs. MMS, the difference was more complex in patients with external circumstances for an incMMS ($p = 0.021$). It should be mentioned that studies of stroke patients detected within 4 years had a lower incidence of intellectual or cognitive damage. [41].

Monitoring has two intraoperative approaches: first, fluorescence phenomena, and second, oscillatory neural activity. Some unique techniques have been developed under real-time blood flow conditions employing indocyanine green videography, while others focused on electrophysiological characteristics of high gamma activity (HGA). Indocyanine green videography (ICG-VG) was employed in four patients with moyamoya plus two patients with AVM, while DEHGA was used in four patients with brain tumors via craniotomy techniques in the conscious patients. Perfusion imaging parameters were also used, as was the case with perfusion imaging techniques that revealed shortening and increased hyperfusion when connected to the surgical microscope after moyamoya revascularization and flow visualization techniques, providing key anatomical and functional information for the removal of AVMs under the microscope. [42, 43, 44]

Discussion

MMD: Its origins, pathogenesis, and relationship between diagnosis and clinical symptoms are still under controversy, especially in other ethnic groups than in Asians. Susceptibility to the RNF213 p.R4810K mutation is regarded uncommon in Asians than in

Caucasian Europeans. Therefore, MMD should be treated in various ethnic groups [45].

Etiology

We are reminded that the genetics of moyamoya disease is of interest. However, chromosome 17 has been discovered as a carrier of the gene that may be sensitive to MMD in East Asian individuals. However, it was discovered that women were more likely to be impacted by familial MMD. Another study suggested that it was due to autosomal dominant inheritance with incomplete familial penetrance. Additionally, another study studying the genome and the specific locus discovered that the RNF213 gene was the first susceptible gene, but the biggest number of carriers were of the p.R4859K gene for MMD in Japan, which would explain its frequency compared to Western countries. p.R4859K is associated to autoimmune quasi-MMD and arteriosclerotic MMD [46].

Epidemiology of Moyamoya Disease (MMD)

Its incidence is relatively high geographically in East Asia, and according to some research, the prevalence of MMD is 10 per 100,000 individuals, while the incidence rate is 1 per 100,000 in Japan. Moving to South Korea, the incidence is 2 per 100,000, and its prevalence is 16 per 100,000, whereas in the West it is less than 1 per 100,000. In regions of North America, but in the United States, the trend is increasing. In the Chinese city of Nanjing, the prevalence of moyamoya was 3 per 100,000 as of 2000. Thus, in the latest investigations, 2,430 cases of moyamoya have been reported since 1976. A peak rate worldwide was identified in young people with moyamoya disease (MMD), between 20 and 30 years of age. The difference has increased according to geography and sex, and in foreign cases, the growth has been more prevalent in women than in men. [47].

Pathology of Moyamoya Disease (MMD)

Based on spontaneous progressive occlusions with bilateral inclinations to the terminal branches of the internal carotid arteries, the pathological vasculature of the intracranial vessels in individuals with MMD has been examined. Thickening of the intimal layer of fibrocellular origin, an increase in smooth muscle cells, irregular undulation of the internal elastic laminae, and attenuation of the medial layer without atherosclerotic or inflammatory alterations have been reported. Previous findings have indicated caspase-3 apoptosis in the middle cerebral arteries, which is related with histological abnormalities. The collateral branches in MMD are the so-called dilated perforating arteries, a constant mix of vessels and their growth. According to pathological investigation, the elastic laminae are weakened with the creation of microaneurysms and previously collapsed or thrombotic arteries. Additionally, a light microscope examination of the superficial temporal artery revealed enlarged inner membranes, with disturbed and absent elastic layers. This study indicated that the smooth muscle cells in the inner membrane of the superficial temporal artery were filled with a basement membrane material. Therefore, it was considered that these histological abnormalities must have been a consequence of ischemic and hemorrhagic strokes in patients with moyamoya disease [48].

Moyamoya disease with Down syndrome

The relation between Moyamoya disease with Down syndrome has been noticed as a higher-than-expected frequency, but it is co-occurring in patients with Down syndrome. Existing data in the US national database suggested that 3.8% of patients admitted with MMD were affected. These patients, among Hispanics and Caucasians, represented 15% with ischemic strokes. Although Down syndrome has not been specifically represented in Asian countries, its clinical presentation is more widespread, and although most treated patients presented with symptoms or local deficits with bilateral involvement, the pathological mechanism of MMD in patients with Down syndrome is also not fully understood. Cardiac problems have been documented in 40% to 50% of patients with Down syndrome, impacting the body's circulatory system. There is also an increase in retinal veins flowing into the optic disc in Down syndrome. There are further symptoms from diabetic retinopathy through diabetes mellitus, such as the low incidence of malignancy in these patients. Therefore, reduced levels of systemic angiogenesis and high amounts of endostatin minimize diabetic retinopathy in people with Down syndrome. Although this angiogenesis potential may be linked to Down syndrome with moyamoya, people with moyamoya tend to have thyroid dysfunction, and the immunological dysregulation in Down syndrome could lead to MMD in some cases [49].

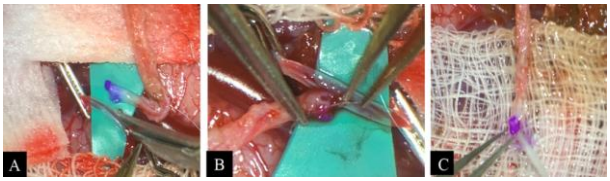

Figure 5. Moyamoya MMD procedure a,b,c) Revascularization STA-MCA.

Conservative Treatment of MMD

According to a review of a global survey, just 31% explored long-term aspirin use. Antiplatelet therapy has been utilized for stroke prevention in people with MMD. According to some research, antiplatelet medication is sufficient, however its usefulness in preventing stroke is currently being investigated. However, it cannot prevent recurrent cerebral infarction in people with MMD. Ischemic damage in persons with MMD is not considered embolic damage, but rather a hemodynamic infarction. Pathological changes in the vasculature of individuals with MMD, toward the ICA bifurcation, are not considered endothelial damage, which is more prone to platelet adhesion. In this circumstance, ischemic stroke prevention will not be helpful in people with MMD. However, persons getting antiplatelet therapy likely to develop issues with bleeding; the therapy is not connected with cerebral hemorrhage. In symptomatic MMD, its alternative use should not be considered. Ischemic stroke can impact posterior circulation, which is considered a risk factor for ischemic stroke [50].

Direct Revascularization

This is a proven effective procedure, the first employed between the superficial temporal artery and middle cerebral artery (SMA-MCA) bypass. The first surgical report was performed by Yasargil in 1967. Considered one of the most exact surgical treatments to date, however, modern technology breakthroughs have modified it over time. The STA is employed as a primary donor channel for the anastomosis of a distal branch to the middle cerebral artery. STA has both parietal and frontal branches, which can both be donors, and is followed using surface Doppler and neuronavigation, citing an angiographic diameter of 1 mm or more. Although the angiographic STA may be reduced, it can be enhanced during dissection with surgical measurement and appropriate flow following the anastomosis. When dissecting the STA, care must be given to avoid any form of vasospasm or damage by leaving any soft tissue remnants of the galea. When irrigating with heparinized solution after clipping the proximal STA to make the anastomosis, care must be given to protect the STA branches to avoid scalp necrosis. Remember that combined bypasses should be performed with a bigger incision than direct bypasses, which can be associated with 3-15% skin scarring. The linear incision will be even more essential, with less healing issues (1.6%), while curved incisions will be made in 3.8% and Y-type incisions in 17%. STA-MCA is conducted less frequently than those performed in the anterior cerebral artery and posterior cerebral artery. [51]. For ACA revascularization, a substantial piece of the STA, no less than 10 cm, must be dissected, with the craniotomy directed into the anterior cerebral artery. If revascularization affects the ACA and MCA, then both STAs are dissected bilaterally.

Therefore, two distinct craniotomies must be performed, one toward the Sylvian fissure and the other toward the ACA section. [52]. After passing the donor vessel of the STA, more toward the bone bridge between both craniotomies, thereby producing the medial anastomosis. Therefore, both STAs must be removed to minimize any damage of the scalp. We can also use the occipital artery for revascularization of the MCA or PCA area. However, direct revascularization of the posterior quadrant is more complex, particularly for the smaller recipient arteries if the occipital artery is employed. The average diameter of the recipient vessels should be 1 mm, although 0.8–0.9 mm can be considered. after performing a normal craniotomy, the middle meningeal branches should be preserved after exposing the dura, and after opening the dura, its collaterals should be preserved around it for a distal cortical anastomosis, which can be confirmed by preoperative angiography [53, 54].

Indirect revascularization

This is a technique for developing new vessels through the blood flow of vascularized tissue, stimulating the formation of new vessels. Unlike direct bypass, indirect bypass does not usually use vascular anastomosis. Other revascularization types, such as encephaloduroarteriosynangiosis (EDAS) and encephalomyosynangiosis, are considered, depending on the presence of several burr holes, such as synangiosis, followed by

encephaloduromyoarteriopericranial and omental transplantation. This method of indirect revascularization is advised for patients with recipient vessels of inadequate size for direct anastomosis, in pediatric and adult cases of MMD, which is regarded rare. The postoperative time will be shorter, and hypoperfusion syndrome does not develop postoperatively. If we employ the collateral branches of the distal pulmonary artery, we will confine direct bypass to the posterior temporoparietal-occipital regions, making direct bypass a good alternative for stroke revascularization in the PCA areas. The occipital artery can be used for indirect bypass compared to EDAS. Indirect bypass has shown beneficial in youngsters compared to adults. The increased indirect rate in children is attributable to cerebrovascular plasticity at very early ages. [55].

Combined revascularization

This is simply the combined use of direct and indirect revascularization for the treatment of MMD. Its objective is to obtain the cerebrovascular hemodynamic improvement of direct bypass, together with the benefits of indirect bypass. Direct bypass assists with revascularization as a backup method to direct anastomosis. Quantitative results vary depending on the patient. Although some suggest that direct bypass has advantages over indirect bypass in patients with MMD, adult patients with symptomatic MCA and ACA territories should be treated with combined revascularization instead of direct bypass, and combined bypass will be more effective or beneficial in pediatric patients. One restriction of combined revascularization is its increased use in dissection and surgical exposure. [56, 57].

Moyamoya disease (MMD) in pediatric patients usually presents with ischemic events, with intracranial hemorrhage occurring in only 10% of cases, compared to 60% in adults. [58, 59]. Spontaneous acute subdural hematoma in MMD is very rare; 1 to 6 cases have been reported. Acute subdural hematoma presents as a complication of MMD, as it is due to the rupture of transdural vessels. [60]. Some patients temporarily lose consciousness and develop lower limb weakness. For example, an SDH can be identified on a brain CT scan by noting a sudden increase in intracranial pressure, which is the cause of the symptoms. [61]. However, the location of the SDH, around the parasagittal area, could explain the lower limb weakness. An acute subdural hematoma results from a tear of the bridging veins due to head trauma. [62]. In contrast, spontaneous acute subdural hemorrhage can be caused by bleeding from the cortical artery, arteriovenous malformations, coagulopathies, or neoplasms like meningiomas, metastases, spontaneous intracranial hypotension, cocaine use, and arachnoid cysts. [63, 64].

Patients with MMD may experience complications such as hemorrhage, hydrocephalus, and cerebral palsy. [65]. Patients with MMD may be instructed to undergo encephaloduro-arteriosynangiosis (EDAS) to improve blood perfusion, with a risk of stroke. The outcome can be unpredictable due to the MMD condition. Intraventricular hemorrhage with hydrocephalus—this could progress to a complicated neurological prognosis. [66]. Also, the duration or persistence of these complications. An external ventricular drain (EVD) should be placed; therefore, a peritoneal shunt. VP shunt. Imaging studies will reveal the accuracy of placement versus a decrease in ventricular size, as well as a reduction in IVH and hydrocephalus; therefore, drainage is a positive indication. [67]. and effective optimization of CSF decompression. Verbal impairment may limit baseline functions, leading to failure to notice decompensation. A critical sign present in MMD in a patient with cerebral palsy may be seizure activity, which can be detected through urgent neuroimaging. [68]. Moyamoya disease (MMD) and multiple sclerosis have distinct pathogenesis. The clinical presentation and radiological presentations are similar, which has led to misdiagnosis of MMD. [69].

Limitations and future directions

Several investigations have raised the disadvantage of MMD in the context of stroke: it is more prevalent, and bleeding assessment is class C evidence. According to the AHA/American Stroke Association (2021) guidelines, revascularization was identified as effective for the prevention of ischemic stroke and/or transient ischemic attack (TIA). The difference is that the incidence of stroke rose in patients with moyamoya syndrome, while both groups benefited from revascularization, so urging and minimizing perioperative problems was indicated as a

preventive approach. When dissecting the superficial temporal artery (STA), care must be taken to avoid any vasospasm or damage, leaving any soft tissue remnants of the galea. When irrigating with heparinized solution after cutting the proximal STA to conduct the anastomosis, care must be given to protect the STA branches to avoid scalp necrosis. However, direct revascularization of the posterior quadrant is more complex, particularly for smaller recipient arteries if the occipital artery is employed. A mixture of combined revascularization may be employed in the future, although laboratory samples should be gathered for further evaluation. [70].

Conclusion

This study has proven that moyamoya disease (MMD) is a vascular problem that, regardless of ethnicity, cannot be ignored, as very rare cases have been documented in Europe and the United States, as well as in the Hispanic population, but none have been identified in African locations. Therefore, encountering this pathology cannot be ruled out, and one should be up-to-date on the types of revascularizations, whether STA-MCA, ACA-PCA, since their effectiveness in blood return after performing the procedure has been demonstrated, taking into account direct revascularization or direct bypass and indirect revascularization and indirect bypass as advantages and disadvantages that help us further with this type of pathology.

Table 1. Patients with Moyamoya disease treated with anatomical revascularization of both the superficial temporal artery and the middle meningeal artery and some variants.

Author	Kind of study	Patients No.	Range of age	Surgical intervention	Revascularization	Bypass	Follow up	Mortality	P= value
Yoshida Y. et al. 1999[7].	Retrospective	28	7 to 69 years (mean 39.2 years)	19	10	EDA, EMS, STA-MCA	14.2 years	5	P>0.05
Kawaguchi S. et al.2000 [8].	CT	22	43 years	11	6	STA-MCA bypass	0.8 to 15.1 years	N/A	p<0.05
Duan L. et al. 2012[9].	Retrospective	802	28 (range, 0.5-77) years.	500	773	EDAS	26.3 months (range, 6.0-101.9 months).	3	*P*<0.01)
Choi et al. 2013[10].	Retrospective	44	44.9 years (range: 17-65 years)	29	35	STA-MCA	12 to 105 months (mean of 55.4 months)	N/A	*p* > 0.05
Liu X. et al. 2013[11].	Retrospective	97	(mean age 31±10 years; range 5–56 years)	54	97	STA-MCA/EDAS	N/A	6	p<0.001
Miyamoto S. et al. 2014[12].	RCT	160	+18	80	42	STA-MCA Direct+indirect	5 years		*P*=0.048)
Han C. et al. 2015[13].	Retrospective	6	(range, 1-23 years)	6	5	N/A	18-108 month	N/A	N/A
Huang Z. et al. 2015[14].	Retrospective	154	33.95 years.	126	124	EDAS	36.12 months	N/A	P < 0.001
Jang DK. et al. 2017[15].	Retrospective /Comparative study	249	N/A	158	91	STA-MCA Direct+indirect	6-year	N/A	p = 0.014
Liu X. et al. 2015[16].	Case series	528	2-67 years	406	406	EMAS	39.5 months)	N/A	p<0.001

Lee SB. et al. 2012[17].	Retors pective	142	N/A	126	124	STA-MCA/EDAS	10 years	N/A	P < 0.05
Kang K. et al. 1996[18].	Retros pective	312	(39.4 ± 9.1 years old)	133	186	STA-MCA	48 (IQR 32–67) months	17	*P* = 0.006
Mizoi K. et al. 1996[19].	TC	23	(mean age, 35; range, 20-59)	23	16	STA-MC	6 months	N/A	p = 0.056
Zhang M. et al. 2020[20].	Retros pective	219	N/A	219	157	STA-MC	18 months (3-69 months	N/A	P = 0.004
Lukshin VA. et al. 2021[21].	Cohort study	80	N/A	134	40	N/A	N/A	N/A	N/A

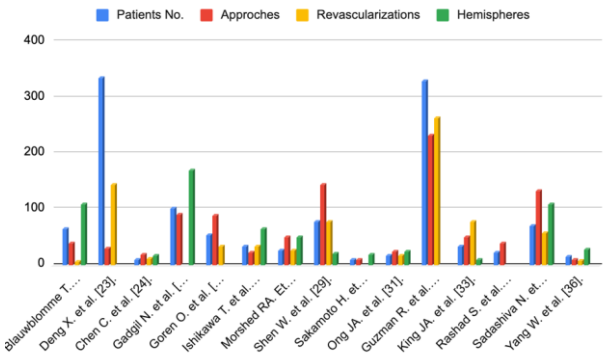

Figure 2. Graphics of patients with Moyamoya disease treated with revascularization.

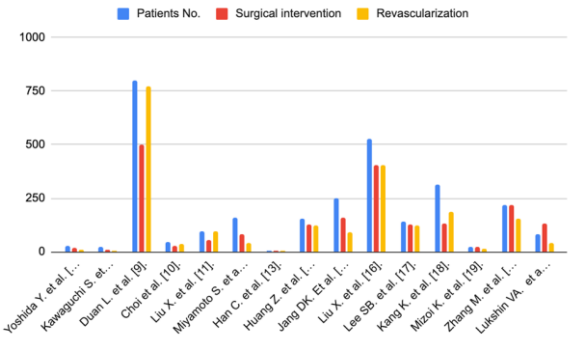

Figure 3. Graphs Patients with Moyamoya disease with revascularization approaches and intervention of the cerebral hemispheres.

Table 2. Patients with Moyamoya disease with different surgical approaches.

Authors	Kind of study	Year	Patients No.	Approaches	Revascularizations	Hemispheres	follow-up
Blauwblomme T. et al. [22].	Retrospective	2017	64	39	5	108	4.2 years
Deng X. et al. [23].	Retrospective	2021	336	30	144	N/A	N/A
Chen C. et al. [24].	Retrospective	2018	10	18	12	16	63.4 months ± 36.0.
Gadgil N. et al. [25].	Retrospetive	2018	102	90	N/A	169	4.3 years
Goren O. et al. [26].	Retrospective	2021	54	88	34	N/A	3-6 years
Ishikawa T. et al. [27].	Cohort study	1997	34	23	34	64	> 5 postoperative years
Morshed RA. Et al. [28].	Retrospective	2020	26	49	26	49	2.6 years
Shen W. et al. [29].	Retrospective	2017	77	143	77	20	28.43 ± 15.31 months
Sakamoto H. et al. [30].	Cohort study	1997	10	10	N/A	19	4 years,
Ong JA. et al. [31].	Retrospective	2020	16	24	16	24	3 months to 12 year
Guzman R. et al. [32].	Cohort study	2009	329	233	264	N/A	4.9 years
King JA. et al. [33].	Cohort study	2010	33	50	78	10	4.1 years.
Rashad S. et al. [34].	retrospectivey	2016	23	38	N/A	N/A	3 and 131 months
Sadashiva N. et al. [35].	Retrospective	2016	70	133	58	108	15.9 months (range 3-62 months)

Yang W. et al. [36].	Retrospective/case reports	2017	15	9	7	28	11.6 years

Table 3. Patients with Moyamoya disease were approached by direct and indirect revascularization.

Author	Kind of study	Year	Patients No.	Direct Vascularization or bypass	%	Indirect Vascularization or bypass	%
Deng X. et al. [23]	Retrospective	2021	336	70	21	447	75
Ha EJ. Et al. [37].	Longitudinal and crossectional analysis	2019	627	N/A	N/A	773	81
Ishikawa T. et al. [27].	Cohort study	1997	34	48	70	16	47
Morshed RA. Et al. [28].	retrospectivo	2020	23	33	69	16	69
Guzman R. et al. [32].	Cohort study	2009	329	96	29	264	80
Sadashiva N. et al. [35].	Retrospective	2016	70	17	21	50	71
Wang Y. et al. [36].	Retrospective/case reports	2022	144	N/A	N/A	37	25
Mizoi K. et al. [19]	TC	1996	23	16	69	7	30
Chou SC. et al. [38].	Retrospective	2022	50	17	34	50	100
Zheng EY. et al [39].	retrospective	2023	58	N/A	N/A	58	100

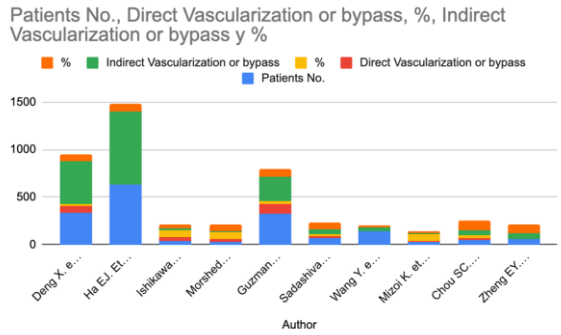

Figure 4. Graphs of patients with Moyamoya disease undergoing direct and indirect bypass.

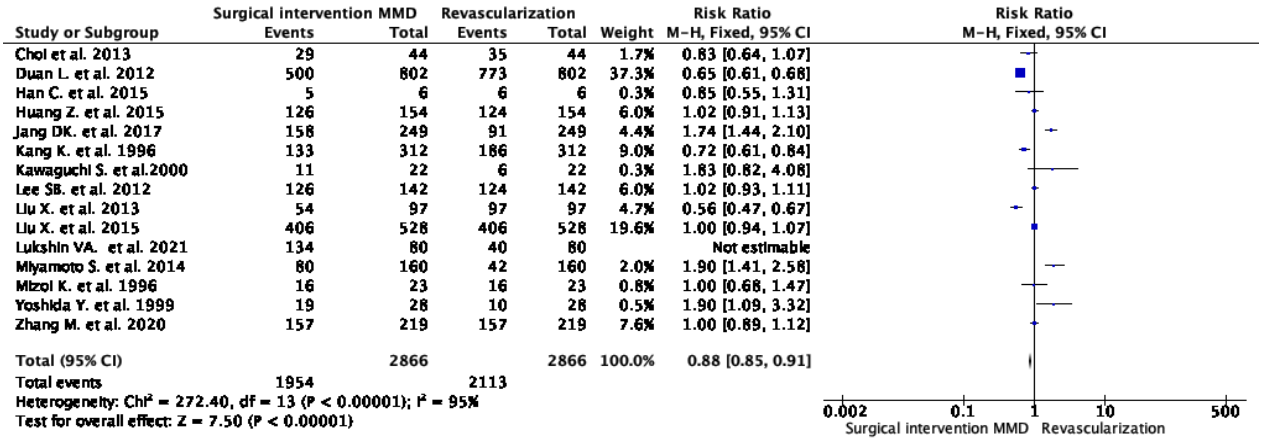

Figure 6. Forest plot Risk-ratio of patients with moyamoya disease MMD.

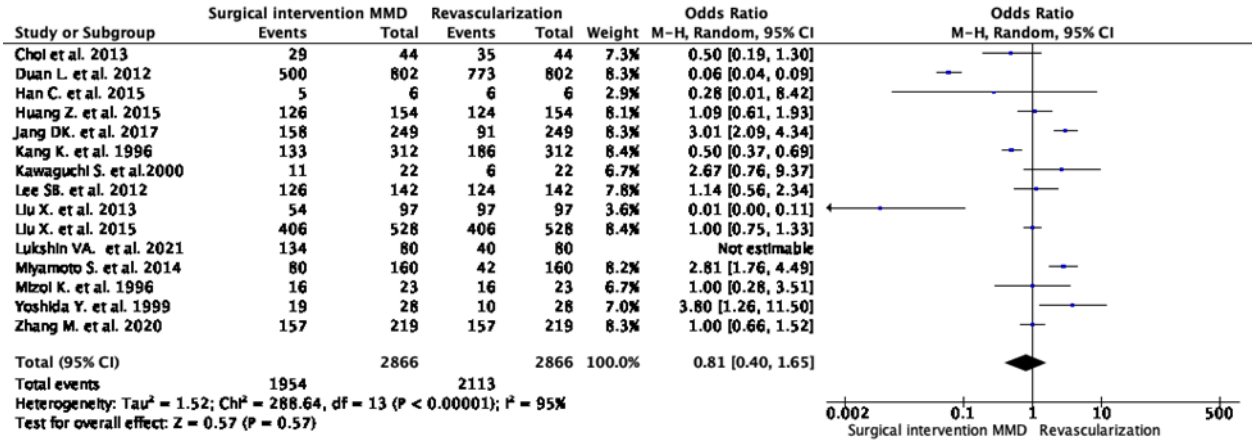

Figure 7. Forest plot Odds ratio of patients with moyamoya disease MMD, surgical intervention vs. revascularization.

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Lumbar spinal stenosis associated with alkaptonuria. Case report

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ABSTRACT

Alkaptonuria is a rare autosomal recessive metabolic disorder caused by a deficiency of the homogentisic acid oxidase enzyme. In this disease, degenerative changes occur in the intervertebral discs and connective tissue due to pigment accumulation. This report presents a 45-year-old female patient who developed multi-level lumbar spinal stenosis due to alkaptonuria and underwent surgical treatment. Clinical findings, radiological imaging, and characteristic pigment accumulation observed during surgery were discussed. In this patient with severe spinal stenosis unresponsive to conservative treatment, surgical decompression provided significant improvement in symptoms, while intraoperative black disc material and pigmented ligamentum flavum provided important clues for diagnosis. As spinal degeneration associated with alkaptonuria can be aggressive, these cases should be followed up regularly over the long term.

Keywords
Alkaptonuria, lumbar spinal stenosis, spinal degeneration

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INTRODUCTION

Alkaptonuria is an autosomal recessive metabolic disorder. These cases involve a deficiency of the enzyme homogentisic acid oxidase, which plays a role in phenylalanine and tyrosine metabolism (1, 2). It manifests itself primarily as dark pigment accumulation (ochronosis) in cartilage and connective tissue (3). Over time, progressive degeneration also occurs in the spine and joints of these patients (4, 5).

The degeneration occurring in connective and cartilage tissue presents itself in patients mainly as widespread joint pain. Over time, back and neck pain accompany the patients' extremity complaints. Spinal instability and/or limited mobility may develop in relation to the severity of degeneration (6). Disc herniations, stenosis, and spondylolisthesis are spinal problems that can be seen in these patients (7, 8). This report presents a case of alkaptonuria in which the patient had previously undergone surgery for cervical and lumbar disc herniations and subsequently developed multi-level lumbar stenosis. The case was examined in terms of clinical, radiological, and surgical treatment, and the results were discussed.

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CASE PRESENTATION

A 45-year-old female patient who underwent surgery for lumbar disc herniation in 2013 and cervical disc herniation in 2017 was evaluated for progressive neurogenic claudication. On examination, no findings were observed except for hypoesthesia below the left knee. In addition to complaints of pain in her back and legs, the patient had neurogenic claudication that worsened after approximately 50 meters. Her medical history revealed that during her 2013 surgery for an extruded lumbar disc herniation at the L5-S1 level, dark pigment accumulation was observed in the disc material, leading to further investigation and a diagnosis of alkaptonuria. Similarly, dark pigment accumulation in the disc material was reported in the 2017 surgery performed for cervical disc herniation (Figure 1).

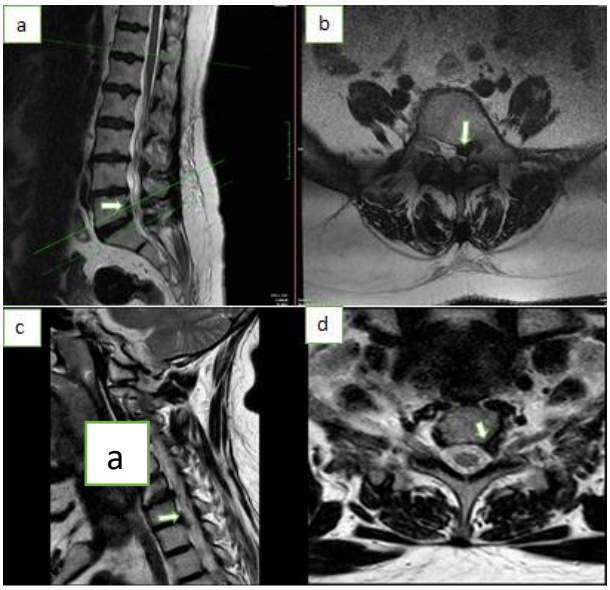

Figure 1. (a) Sagittal T2-weighted lumbar MRI showing an upwardly extruded lumbar disc herniation (*white arrow*), (b) axial T2-weighted lumbar MRI with a disc herniation compressing the left S1 nerve root (*white arrow*), (c) sagittal T2-weighted cervical MRI revealing an inferiorly migrated cervical disc herniation (*white arrow*), and (d) axial T2-weighted cervical MRI demonstrating a disc herniation impinging on the left cervical nerve root (*white arrow*).

The lumbar Magnetic Resonance Imaging (MRI) scan performed due to neurogenic claudication revealed significant lumbar degeneration and the presence of multi-level spinal stenosis (Figure 2). When compared to the previous lumbar MRI, it was observed that lumbar degeneration had significantly increased and progressed to severe spinal stenosis (Figures 1, 2).

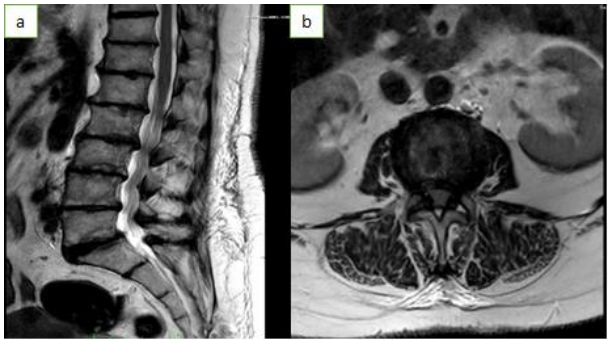

Figure 2. (a) Sagittal T2-weighted lumbar MRI showing narrowing of intervertebral disc spaces, multilevel osteophytic degeneration, spondylopathy, and grade 1 spondylolisthesis at the L5–S1 level, (b) axial T2-weighted lumbar MRI demonstrating spinal canal stenosis at the L2–L3 level.

Lumbar Computed Tomography (CT) revealed narrowing of the intervertebral disc spaces with a vacuum phenomenon, Schmorl's nodes, osteophytic spurs along the vertebral body margins, facet joint sclerosis, a defect in the pars interarticularis of L5, and Grade I spondylolisthesis at the L5-S1 level (Figure 3).

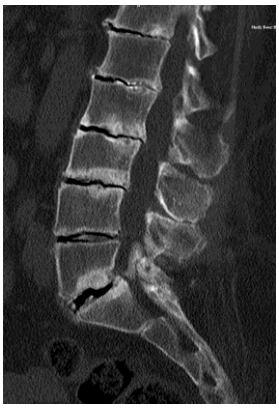

Figure 3. Sagittal lumbar CT; the calcification and narrowing of intervertebral disc spaces, Schmorl's node formations, osteophytic degeneration, spinal sclerosis, and grade 1 spondylolisthesis at the L5–S1 level are seen.

The patient, who had severe spinal stenosis and did not respond to conservative treatment, underwent surgery. During the surgery, bilateral partial hemilaminectomy and foraminotomy were performed at the levels where stenosis was observed. Spinal instrumentation was not performed because the patient did not want it. At these levels, dark pigment accumulation in the ligamentum flavum was seen accompanying degeneration of the flavum. Postoperatively, the

patient's pain and neurogenic claudication improved significantly. Pathological examination of tissue samples obtained during surgery reported the presence of brown pigment deposition and degeneration (Figure 4).

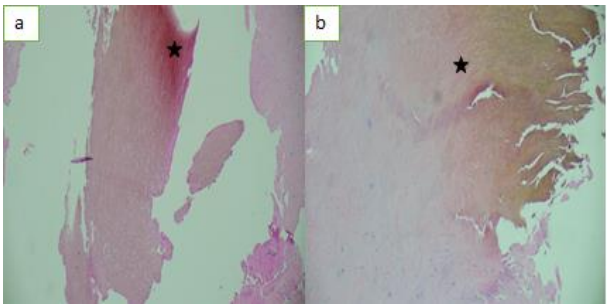

Figure 4. Microscopic examination of the ligamentum flavum shows brownish pigment deposition consistent with ochronotic degeneration due to homogentisic acid accumulation (stars). (a) H&E ×4, (b) H&E ×20.

Discussion

Alkaptonuria is an autosomal recessive metabolic disorder caused by a deficiency of the homogentisate 1,2-dioxygenase enzyme. This enzyme deficiency leads to the accumulation of homogentisic acid (HGA) in the body. Over time, HGA oxidizes, particularly in structures containing type 2 collagen, such as cartilage, connective tissue, and intervertebral discs, and converts into pigmented benzoquinone derivatives, which irreversibly bind to the tissue. Ochronotic pigment accumulation increases inter-fibrillar cross-linking and reduces tissue elasticity. Consequently, connective tissues become stiffened, brittle, and structurally compromised. (9, 10). Over time, this degenerative process in connective tissue leads to serious structural problems in the joints, and the clinical symptoms of patients are related to the severity of the degeneration (11). HGA accumulation has also been reported in the spinal region, causing similar degeneration (1, 12).

This degenerative process is particularly pronounced in the lower lumbar segments where axial loading is excessive. The structural deterioration in the connective tissue eventually leads to inadequate stabilization of the spine and increased loading on the posterior structures. This situation particularly increases the mechanical stress on the ligamentum flavum. In response to increased load and ochronotic pigment accumulation, elevated secretion of tissue inhibitors of metalloproteinase-2 (TIMP-2) results in hypertrophy of the ligamentum flavum (10, 13). Furthermore, the process associated with alkaptonuria in the intervertebral discs gradually disrupts the structure of the disc, facilitating the development of disc herniation (14). The resulting disc herniation further exacerbates the severity of spinal stenosis. In our case, there was a history of surgery for disc herniation at three levels: L5-S1, C6-C7, and C7-T1.

Although there were no specific findings related to alkaptonuria in the radiological diagnosis, findings indicative of segmental degeneration, such as narrowing of the disc space, calcification, vacuum phenomenon, osteophyte formation, endplate sclerosis, ligamentum flavum, and facet joint hypertrophy, can be observed on MRI and tomography (15, 16). These findings indicating segmental degeneration were also present in our case.

In most cases, degenerative changes associated with alkaptonuria become clinically apparent after the third or fourth decade of life. In most cases reported in the literature, the lumbar region is affected. Thoracic and cervical involvement has been reported more rarely (17, 18). Clinical symptoms and findings vary depending on the location and severity of the involvement. However, pain and limited mobility, as in our case, are the most common complaints in patients. In our case, both the lumbar and cervical regions were affected, and surgery for multi-level disc herniation and spinal stenosis had been performed at different times. With this feature, our report presents a unique example of how spinal degeneration related to alkaptonuria may pursue a particularly aggressive pattern.

Surgically, the distinctive dark blue-black coloration of degenerative disc material in patients with alkaptonuria is the most striking finding of the “black disc phenomenon.” In many cases, it has been reported that the nucleus pulposus fragments removed during discectomy are blackish in color and lighten in color upon contact with air (8, 19, 20). A similar appearance is also seen in the ligamentum flavum (9, 10). In our case, similar pigment accumulation in the ligamentum flavum was observed macroscopically and microscopically.

Since there is no definitive method to eliminate the underlying cause of alkaptonuria, treatment is limited to measures that support the control of

degenerative changes. Although protein (tyrosine-phenylalanine) restriction and vitamin C supplementation are recommended, nitisinone therapy, which reduces homogentisic acid production, has been reported to be beneficial in experimental studies (21, 22, 23). In cases with spinal involvement, surgical treatment can significantly improve patients' quality of life if there is severe pain and progressive neurological findings (18).

Conclusion

Although alkaptonuria is a rare disease, it should be kept in mind in the differential diagnosis of young patients with rapidly progressive, widespread spinal degeneration. Encountering black disc material and/or pigment accumulation in the ligamentum flavum during surgery is an important clue for alkaptonuria. In cases with advanced spinal stenosis that do not respond to conservative treatment, surgical intervention can significantly improve symptoms. Furthermore, it should be remembered that spinal degeneration associated with alkaptonuria, as in our case, can have an aggressive course, and such cases require periodic evaluation for spinal complications.

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Clinical and radiological assessment of diffuse axonal injury in traumatic brain injury patients. A retrospective study

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ABSTRACT

Background: Diffuse Axonal Injury (DAI) is a severe form of traumatic brain injury (TBI), characterised by widespread damage to white matter tracts due to shearing forces. It is often associated with high morbidity and mortality, especially when diagnosis and management are delayed.

Objective: This study aims to evaluate the clinical, demographic, and radiological features of DAI patients and assess their treatment outcomes. Additionally, it investigates the correlation between MRI-based Adams Grading and prognosis using the Glasgow Outcome Scale (GOS).

Methods: A retrospective analysis was conducted on 30 patients diagnosed with DAI and admitted to the Department of Neurosurgery, SVP Hospital, Ahmedabad, from June 2022 to June 2024. Data collected included age, gender, mechanism of injury, initial Glasgow Coma Scale (GCS) score, MRI findings (Adams Grade), treatment modalities, and GOS scores at discharge or follow-up.

Results: DAI was most commonly observed in young adults aged 11–30 years, with road traffic accidents being the leading cause. MRI demonstrated superior diagnostic accuracy compared to CT. Adams Grading showed a strong correlation with clinical outcomes: higher grades (II and III) were associated with poorer GOS scores (1–3), while lower grades (I) had better outcomes (4–5). Early neurocritical care and timely imaging significantly influenced recovery.

Conclusion: DAI predominantly affects young individuals, primarily due to preventable trauma. MRI plays a crucial role in early diagnosis and prognostication. A multidisciplinary approach, including prompt neuro-intensive care and rehabilitation, is essential for improving outcomes in DAI patients.

Keywords
diffuse axonal injury,
traumatic brain injury,
MRI grading,
Glasgow coma scale,
Glasgow outcome scale

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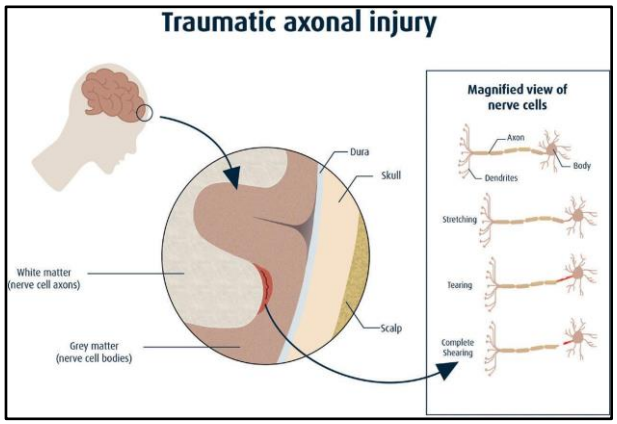
INTRODUCTION

TBI is divided into three groups; mild, moderate, and severe. This division is based on the Glasgow Coma Scale (GCS) in the (sub)acute phase after trauma[1]. Axonal injury is mostly seen in patients with moderate to severe TBI, but also occurs in patients with mild TBI[2,3,4]. Axonal injury is mostly seen after high-energy level trauma, such as road traffic accidents and falls from height[2]. In these types of trauma, acceleration and deceleration forces can cause shear injury of axons[5-

7]. In the brain, the cell body of the neuron lies in the more dense grey matter and the axon in the white matter with a lower density. Due to the difference in density between grey and white matter, acceleration deceleration forces result in a difference in speed between grey and white matter. This results in the stretching of the axons (figure 1)[6]. The shear injury can cause primary lesion to the axon (axonotomy) and secondary injury as a result of an overstretched axon. This secondary or delayed injury is a result of axonal stretch followed by a progressive cascade of pathologic cellular mechanisms disrupting the structural integrity of the axon and neuron [8].

Figure 1. Illustrates the mechanism of traumatic axonal injury due to acceleration-deceleration forces.



Schematic representation of acceleration-deceleration forces leading to shear injury at the grey-white matter interface. These forces cause stretching, tearing, or complete shearing of axons, resulting in primary and secondary axonal damage.

Axonal injury is referred to as diffuse axonal injury (DAI) or traumatic axonal injury (TAI). In radiologic imaging of TBI the definition of DAI is multiple axonal lesions in multiple brain regions, when lesions are confined to only one region it should be named TAI[9]. In clinical practice DAI and TAI are being used almost interchangeably. Most patients with axonal injury have sustained moderate or severe TBI, but after mild TBI axonal injury can also be present[4].

In patients with axonal injury a Computed Tomography (CT) scan of the brain can appear normal, but sometimes microbleeds can be visible indicating the presence of axonal injury. Magnetic Resonance Imaging (MRI) is more sensitive for axonal injury. It can be visible as microbleeds on a T2 star gradient echo (T2*GRE), a fast field echo (FFE), and/or susceptibility weighted image (SWI)[10-13].

The condition is often associated with prolonged unconsciousness and poor neurological outcomes. This study provides a comprehensive understanding of DAI to improve early diagnosis and patient care.

Aim and Objectives

This study aims to:

- Assess the demographic profile of DAI patients
- Evaluate clinical presentations and Glasgow Coma Scale (GCS) scores
- Correlate MRI-based Adams Grading with clinical outcomes
- Analyze treatment modalities and patient prognosis

Methodology

This study consists of analysis of 30 cases of Diffuse axonal injury admitted and treated in the Department of Neurosurgery, SVP Hospital, Ahmedabad from June 2022 to June 2024. Patient`s data were documented in case record form and were followed up till the end of the study. Written and informed consents were taken of all patients' relatives before participation in the study.

Inclusion Criteria

1. Patient`s relatives who are willing to participate in the study.
2. Patients of any age.
3. Patients who have Diffuse axonal injury

Exclusion Criteria

- Patient`s relatives who are not willing to participate in the study.
- Patients who do not have diffuse axonal injury.

Total 30 patients were included in the study of which 24 were male and 6 were female. Patients were categorized into mild, moderate, and severe DAI based on MRI grading. Outcomes were analyzed statistically.

Results

Key findings from the study include:

- Age Group Most Affected: 11-30 years (highest incidence, 50%), followed by 31-50 years (30%).

- Common Causes: Road traffic accidents (70%) were the leading cause, followed by falls (20%) and assault-related injuries (10%).

Table 1. Age distribution of patients with diffuse axonal injury (DAI)

Age Group	Number of Patients	Percentage (%)
≤10	2	6.67
11–30	15	50.00
31–50	6	20.00
51–70	5	16.67
≥71	2	6.67

Maximum incidence is in the young population due to higher incidence of motor vehicle accidents in young people [Table 1].

Table 2. Association between DAI Grade III and Admission GCS Scores

GCS Score on Admission	Number of Patients	Percentage (%)
<8	11	100

As shown in Table 2, all patients with DAI Grade III had a GCS score <8 on admission.

Table 3. Association between DAI Grades I/II and Admission GCS Scores

GCS Score on Admission	Number of Patients	Percentage (%)
≥8	11	57
<8	8	43

More than half of patients with DAI Grades I/II had GCS scores ≥8 at admission [Table 3].

Table 4. Distribution of Diffuse Axonal Injury (DAI) grades among study cohort

DAI Grade	Number of Patients	Percentage (%)
I	8	26.66
II	11	36.66
III	11	36.66

There was no significant predilection for any particular DAI grade among the cohort [Table 4].

Table 5. Correlation between dai grade and duration of ICU stay (survivors only)

Hospital ICU Stay	DAI Grade III	DAI Grade II	DAI Grade I
>10 days	2/2 (100%)	7/10 (70%)	2/8 (25%)
<10 days	0	3/10 (30%)	6/8 (75%)

Higher DAI grades correlated with longer ICU stays [Table 5].

Table 6. Final outcomes based on Glasgow Outcome Scale (GOS) in DAI patients

Final Outcome (GOS)	Number of Patients	Percentage (%)
GOS 1 (Death)	10	33.33
GOS 2 (Vegetative)	1	3.33
GOS 3 (Severe Disability)	3	10.00
GOS 4 (Moderate Disability)	9	30.00

Final Outcome (GOS)	Number of Patients	Percentage (%)
GOS 5 (Good Recovery)	7	23.33
Total Favorable Outcomes (GOS 4–5)	16	53.33%

Final outcome analysis showed that 53.33% of patients had favorable outcomes (GOS 4–5), while 33.33% died [Table 6].

Table 7. Discharge and mortality outcomes in DAI patients

Outcome Type	Number of Patients	Percentage (%)
Discharged	20	66.66
Died	10	33.33

Out of 30 patients, 20 were discharged, and 10 met with mortality [Table 7].

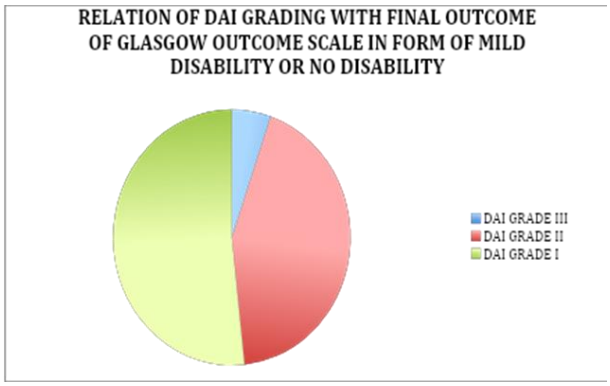


Figure 2. Relationship between MRI-Based Adams Grading and Glasgow Outcome Scale (GOS) at discharge

Bar chart showing the distribution of patient outcomes (GOS scores) stratified by DAI grade. Grade III injuries show a higher proportion of poor outcomes (GOS 1–3), while Grade I injuries are associated with better recovery (GOS 4–5).

Discussion

Maximum incidence of Traumatic brain injury and hence DAI is in the young population due to higher incidence of motor vehicle accidents in young people. In the age group of 11-40 there are 17 patients (57%) in our study. Males are frequently involved in Road traffic accidents. In our study, 24 patients (80%) are male. Previous studies have established that MRI is the gold standard for diagnosing DAI, with Adams Grading (Grade I-III) predicting severity. However, limited research has focused on patient outcomes based on MRI findings and Glasgow Outcome Scale (GOS) scores in an Indian setting. This study aims to bridge this gap by correlating MRI grades with clinical outcomes.

The study findings align with existing literature that young adults are the most affected by DAI due to high-speed vehicle accidents. Peeters W, et al. in their study 'Epidemiology of traumatic brain injury in Europe' and Maas AI, et al in their study 'Moderate and severe traumatic brain injury in adults' published in The Lancet Neurology. 2008;7(8) also had similar findings[15,16]. MRI grading was found to strongly correlate with neurological outcomes, emphasizing the importance of early imaging. Patients with Grade I DAI had better recovery, while Grade III cases showed high mortality and severe disability rates. Fiersching et al. reported mortality in his study in which relation was established between Mortality in DAI patients and MRI grading[14]. Farukh javeed, Lal Rehman, Ali afzal published an article in Surgical Neurology International about DAI which also showed similar findings[17].

The use of neurocritical care (mechanical ventilation, sedation, and supportive therapy) plays a key role in improving survival rates. However, long-term rehabilitation and physiotherapy remain essential for neurological recovery.

Conclusion

There is a critical role of MRI-based grading in predicting DAI outcomes. Early diagnosis and intensive neurocritical care significantly improve prognosis. Given the high incidence of DAI in young adults due to road traffic accidents, preventive measures such as helmet use, traffic regulation enforcement, and public awareness are necessary. Future research should focus on long-term functional recovery and rehabilitation strategies for DAI survivors.

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Impact of coagulopathy on the management and outcome of chronic subdural hematoma

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chronic subdural hematoma,
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ABSTRACT

Background: Chronic subdural hematoma (CSDH) is one of the commonest conditions encountered in neurosurgical practice. It is a disease more commonly seen in the elderly, and its incidence is expected to rise as the population of persons above 65 years increases. With improving survival and an increasing ageing population, the use of antithrombotic medications to prevent and treat cardiovascular diseases appears to be on the rise. Though trauma is the leading cause of CSDH, these pharmacologic agents alter coagulation and have been associated with the rising recurrence of CSDH. There appears to be a complex relationship between CSDH and coagulopathy, which can lead to rebleeding or recurrence after surgical evacuation of the hematoma.

Aim/Objectives: To determine the relationship between the presence of coagulopathy and outcome in patients who underwent burr hole drainage of CSDH.

Methodology: This was a retrospective cohort study on all patients who underwent drainage of CSDH at the University of Benin Teaching Hospital over a 19-year period from June 2006 to May 2025. Clinical data were obtained from a computerised log of patients' records and analysed using STATA software version 12.

Results: One hundred and forty patients were studied. The male-to-female ratio was 4:1, and most patients were above 60 years (55.5%). Fifty-one per cent (51%) had coagulopathy - 10.07% were on antiplatelet medications and 0.71% were on warfarin. Seventy-two per cent (72%) of patients with coagulopathy had a history of trauma. Bilateral CSDH was found in thirty-six per cent (36%) of patients with coagulopathy. The patients who had coagulopathy had lower mean hematoma volumes, longer hospital stay, and accounted for all the patients (4) who required ICU admission. The presence of coagulopathy did not alter the rate of recurrence, reoperation, and GOS at discharge.

Conclusion: There is a high incidence of coagulopathy in patients who have CSDH, and most of them would require prompt evacuation irrespective of haematoma volume. Pre- and post-operative substitution of coagulation factors is associated with very good outcomes despite poor neurological status at presentation.

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Introduction

Chronic subdural hematoma (CSDH) is one of the commonest conditions encountered in neurosurgical practice. It is the slow-growing encapsulated fluid collection of blood and blood degradation products in the potential space between the dura mater and arachnoid coverings on the brain surface.[1,2] Since its first description by Wepfer in 1657, our understanding of the pathophysiology and clinical features has evolved rapidly.[3] The propagation of CSDH and its recurrence is known to result from fibrinolytic hyperactivity, which occurs within the hematoma capsule and systemic blood, amongst other mechanisms.[4] This ensuing coagulopathy contributes significantly to outcome following surgical evacuation.[5]

Globally, it is a disease more commonly seen in the elderly, with an incidence of about 8-58 patients per 100,000 population.[6] The prevalence of CSDH is expected to rise as the elderly population increases.[7]

There appears to be a complex relationship between CSDH and coagulopathy. On the one hand, CSDH can lead to coagulopathy, and on the other hand, patients with coagulopathy are at higher risk for developing CSDH, i.e., coagulopathy is a known risk factor for CSDH.[8,9] There are several recognized causes of coagulopathy, which include anticoagulant or antiplatelet therapy, liver disease, chronic kidney disease with dialysis, renal failure, or hematologic conditions that may impair coagulation, such as hemophilia. Thus, even mild trauma can lead to significant bleeding in such individuals.

In CSDH patients in whom no recognized causes for coagulopathy exist, the presence of CSDH itself is also known to cause coagulopathy.[10] Though less commonly discussed, large or recurrent CSDHs may consume clotting factors or activate fibrinolysis, leading to a secondary coagulopathy. Several studies suggest fibrinolytic activity within the hematoma membranes contributes to ongoing bleeding and expansion of the hematoma.[1] Several mechanisms have been proposed in the development of secondary coagulopathy in CSDH. These include hyperfibrinolysis in chronic subdural membranes, which produce tissue plasminogen activator (tPA), promoting fibrinolysis and preventing clot stabilization.[4] In CSDH, there is local consumption of coagulation factors within the hematoma cavity as well as inflammatory processes in the membranes, both of which may alter coagulation profiles. A secondary disseminated intravascular coagulopathy (DIC) - like picture may present in severe or recurrent cases, though it is rare. There is excessive activation of both coagulation and fibrinolysis, which are important in the progressive enlargement of CSDH.[10] The excessive activation in coagulation is mainly through the extrinsic pathway.[10] Higher levels of plasminogen at initial operation have been identified in patients subsequently experiencing CSDH recurrence. This suggests that markers of hyperfibrinolysis may be able to predict those at highest risk of recurrence and therefore help guide treatment and follow-up.[1]

The age-long treatment for CSDH, irrespective of the institution, has been evacuation through burrhole craniostomy, besides other surgical procedures.[8,11] However, surgical evacuation alone will not suffice in patients who have coagulopathy-associated CSDH, whether or not there was antecedent trauma. Thus, treatment of coagulopathy and surgical evacuation of CSDH have to be addressed in tandem in this group of patients, either preoperatively, postoperatively, or both.[12] In patients with substitution of coagulation factors, outcome has been found to be worse if patients only had post - operative correction.[4]This suggests that for safe and effective treatment of patients with coagulopathy - associated CSDH, there should be both pre-operative and post-operative substitution of coagulation factors to avert complications such as recurrence, reoperations, prolonged hospital stay, and even increased mortality following surgical evacuation.

We investigated the association between coagulopathy and outcome in CSDH patients in the University of Benin Teaching Hospital, Nigeria, a referral centre for most neurosurgical cases in our subregion.

Materials and methods

This was a retrospective cohort study on all patients who underwent surgery for CSDH at the Neurosurgical unit of the University of Benin Teaching Hospital over a 19-year period from June 2006 to May 2025. All patients were admitted via the Accident and Emergency Department, outpatient clinics, or were referred from other units within the hospital. They all underwent full neurosurgical evaluation before admission, and diagnosis was

confirmed by cranial computerized tomographic (CT) scans or Magnetic Resonance Imaging (MRI).

All the patients had burr-hole drainage of the haematoma. Single or double burr holes were sited based on the discretion of the attending surgeon. Standard frontal and parietal burr holes were made using the Hudson brace and perforator. Copious irrigation of the subdural space was done with warm antibiotic-impregnated saline until the effluent was clear. They were nursed flat post-operatively on our neurosurgical wards until mobilized, except for very few ill patients who went to the intensive care unit (ICU). No surgical drains were used postoperatively.

Out of 511 patients operated on within the study period, complete records were found in only 140 patients, with a data retrieval ratio of 27.4%. Data obtained from case notes was entered into a predesigned proforma, which was then uploaded into a computer database. These included demographic, clinical, radiological, and laboratory profiles. The history of trauma, anticoagulation therapy, diabetes, cardiac, renal, and hematologic conditions was particularly noted. We determined whether anticoagulation had been used, and all patients had a clotting profile done on admission.

Coagulopathy was defined as a derangement in clotting profile, low platelet count, use of antithrombotic medications, hematological disorder with an increased risk of bleeding, hepatic failure, or hemodialysis. Platelet counts <100,000 per microliter (normal 100,000-400,000 per microliter), Prothrombin time >15 sec (11-15 seconds), Partial Thromboplastin Time with Kaolin >45 seconds (20-45 seconds), and International Normalized Ratio (INR) >1.2 were taken as abnormal. Antithrombotic medications recorded included low-dose aspirin, clopidogrel, and warfarin. All patients who had coagulopathy were given fresh frozen plasma and tranexamic acid peri-operatively.

The patients were divided into two groups: those with and without coagulopathy. We compared, in the two groups, the patients' age, pre-operative Glasgow Coma Score (GCS), history of trauma, tobacco and alcohol use, and radiologic findings. Intraoperative hematoma volumes, reoperation, recurrence rates, need for ICU admission, duration of hospitalization, and Glasgow outcome score at discharge were also studied. Routine post-operative CT in CSDH was not performed unless indicated by neurologic deterioration or failure to improve neurologically following surgery.

Data were analyzed using STATA software version 12. Continuous variables expressed in means (± SD) were tested using Student's t-test for the difference between group means of CSDH patients without versus with coagulopathy. Categorical variables were compared using the Chi-squared test or Fisher's exact test (if any cell in a category was < 5). P-value < 0.05 was considered statistically significant.

Results

The case file records for one hundred and seventy-five patients were obtained; however, only one hundred and forty had complete data. The mean age was 60.34 years. Most patients were above 60 years (55.5%). There were one hundred and thirteen males and twenty-seven females (M: F = 4:1). The male-to-female ratio for patients without coagulopathy was 7:1, while for those with coagulopathy, it was 3:1. (See Table 1)

Table 1. Laboratory variables and the use of anti-thrombotic medications in CSDH patients with coagulopathy

Variables (n)	Frequency	Percentage
Platelet count, n= 115		
<100,000/μL	8	6.96
Prothrombin time, n= 136	37	27.21
>15 seconds		
PTTK, n= 135		
>45 seconds	23	17.04
INR, n=126		
>1.20	40	31.75
Anticoagulant, n=140		
Warfarin	1	0.71
Antiplatelet, n=139		
Clopidogrel	4	2.88
Low dose aspirin	9	6.47
Clopidogrel and low dose aspirin	1	0.72

μL: microlitre; PTTK: Partial Thromboplastin Time with Kaolin; INR: International Normalized Ratio

Only 14 patients had a history of antithrombotic use: warfarin, 1(0.71%); clopidogrel, 4(2.88%); low-dose aspirin, 9(6.47%); and both clopidogrel and low-dose aspirin, 1(0.72%). (See Table 2).

In the two groups of patients, trauma appeared to be the commonest aetiologic factor for the CSDH (71.77%). Approximately 50 % of patients took

alcohol, and 14.39% used tobacco, but there was no observable difference in the presence or absence of coagulopathy. Patients who had coagulopathy were more likely to present with lower GCS scores, though this was not statistically significant (p=0.079). Most patients, however, presented with mild deterioration in the level of consciousness with a GCS of 13-15. The presence of coagulopathy did not affect the appearance of bilateral lesions(p=0.189).

Table 2. CSDH patients without versus with coagulopathy by demographic, aetiology, lifestyle, clinical and radiologic variables

Variables	n (%) Total= 140	No Coagulopathy n= 69 (49.29%)	Coagulopathy n=71 (50.71%)	P-value
Age (years), n=137	2(1.46)	0	2	0.879
0-10	1(0.73)	0	1	π
11-20	3(2.19)	2	1	
21-30	13(9.49)	7	6	
31-40	16(11.68)	8	8	
41-50	26(18.98)	12	14	
51-60	76(55.47)	37	39	
Above 60	60.34(17.16)	60.27(15.11)	60.40(18.98)	
Age, Mean (SD), years				0.9633μ
Gender, n= 140	27(19.29)	9	18	0.065
Female	113(80.71)	60	53	α
Male				
Aetiology, n= 124	89(71.77)	43	46	0.615
Trauma	35(28.23)	17	18	α
Others				1.000
				π
Alcohol, n= 140	69(49.29)	33	36	0.733
No	71(50.71)	36	35	α
Yes				
Smoking, n=139	119(85.61)	59	60	0.705
No	20(14.39)	9	11	α
Yes				
GCS at presentation, n=128	7(5.47)	2	5	0.079
3-8	31(24.22)	10	21	π
9-12	90(70.31)	48	42	
13-15				
Bilateral haematoma, n= 135	93(68.89)	49	44	0.189
No	42(31.11)	17	25	α
Yes				
Side on CT, n= 134	50(37.31)	21	29	**0.047**
Left	42(31.34)	27	15	**α**
Right	42(31.34)	17	25	
Bilateral				

CSDH: Chronic subdural Haematoma; π: Fisher's exact test; SD: Standard Deviation; μ: Student's t-test between group means; α: Chi-squared test

In patients with coagulopathy, more left-sided lesions (42%) were observed compared to the right side (21%), while in patients without coagulopathy, more right-sided lesions (41%) were seen compared to the left (32%). The difference in sidedness was statistically significant (p=0.047). (See Table 3)

Table 3. CSDH patients without versus with coagulopathy by intraoperative findings and outcomes

Variable	n (%) Total= 140	No Coagulopathy n= 69 (49.29%)	Coagulopathy n= 71 (50.71%)	P-value
Haematoma volume, ml (n=122)	8(6.56)	6	8	0.600
0-50	62(50.82)	28	34	π
51-100	22(18.03)	13	9	
101-150	17(13.93)	10	7	
151-200	8(6.56)	3	5	
201-250	2(1.64)	1	1	
251-300	3(2.46)	2	1	
Above 300	132.77(85.74)	137.30(95.99)	127.93(73.76)	0.5486
Mean (SD) ml				
Recurrence, n= 139	131(94.24)	65	66	0.983
No	8(5.76)	4	4	α
Yes				1.000
				π
Re-operation, n= 135	1(0.74)	0	1	0.549
Craniectomy	5(3.70)	2	3	α
Old burr hole	129(95.56)	65	64	1.000
No				π
ICU Care, n= 140	136(97.14)	69	67	**0.045**
No	4(2.86)	0	4	**α**
Yes				0.120
				π
GOS at discharge, n= 139	1(0.72)	0	1	0.620
	0(0)	0	0	π
Death	1(0.72)	1	0	
Persistent vegetative state	4(2.88)	1	3	
Severe disability	133(95.68)	66	67	

Moderate disability				
Good outcome				
Duration of hospitalization, n=114	15.45(9.00)	14.13(6.64)	16.52(10.46)	0.1603
Mean (SD) days				

CSDH: Chronic subdural haematoma; π: Fisher's exact test; SD: Standard Deviation; μ: Student's t-test between group means; α: Chi-squared test; ICU: Intensive Care Unit; GOS: Glasgow Outcome Score; α: range.

Table 3 compares the mean hematoma volumes, recurrence, reoperation rate, ICU admissions, GOS at discharge, and duration of hospitalization in patients with and without coagulopathy. The mean hematoma volumes measured intraoperatively were 137.30ml for patients without coagulopathy and 127.93ml for patients with coagulopathy. In the no-coagulopathy group, there were 2 outliers with 500 and 600 ml of intraoperative haematoma volume. These two patients had CSDH complicating the insertion of ventriculoperitoneal shunts. When these outliers were removed and data reanalyzed, the no-coagulopathy group mean (n=61) was 123.77 (SD 59.80) ml and the Coagulopathy group mean (n=59) was 127.93 (SD 73.76) ml. The combined mean (n=120) was 125.81 (SD 66.78) ml with no statistically significant difference observed between the two groups (p= 0.7345).

Overall, the recurrence rate was 5.76%. The presence of coagulopathy did not affect the rate of recurrence(p=0.983). Not all recurrent cases were operated. In coagulopathy-associated CSDH, all recurrences were significant enough to require reoperation. In one patient, a limited craniectomy was done to evacuate the clot.

Only patients with coagulopathy required ICU admission, and this accounted for 2.86% of the total number of patients seen (p=0.045). One (0.72%) death was recorded in a patient with coagulopathy. GOS at discharge was a good outcome in 95.6% with no difference observed in the presence or absence of coagulopathy.

The mean duration of hospital stay was 16.52 days in patients with coagulopathy and 14.13 days in patients without coagulopathy. The difference was not statistically significant (p=0.1603)

Discussion

Defining coagulopathy can be challenging due to the several hematological parameters involved.[13] In the study of coagulopathy associated with CSDH, common investigations include simple tests ranging from clotting profile and complete blood counts to complex ones such as clotting factor assay and thromboelastography.[10]

The incidence of chronic subdural hematoma rises sharply with advancing age, irrespective of aetiology.[14, 9,15,16,17] Our study shows that coagulopathy associated with CSDH was also common in the 6th decade, and the male-to-female ratio was 3:1. There was no statistically significant difference in the mean ages of patients with and without coagulopathy (p=0.963). This likely meant that the pattern of age-related hemostatic changes, comorbidities, liver/kidney dysfunction, and local hematoma-driven fibrinolysis in the hematoma membrane was similar. [18,19,20,21]

In this study, it was found, surprisingly, that only 10.71% of patients operated for CSDH were on antithrombotic agents: more patients were on antiplatelets compared to warfarin. It was expected that a higher number of elderly patients would be on cardio- and neuroprotective antithrombotic medications. This strongly contrasted previous studies, which observed a higher incidence of antithrombotic use, which ranged between 29 and 43%.[22,23,24].Thus, with more than fifty percent (50%) of patients having coagulopathy, the strong association between coagulopathy and CSDH is thus underscored.

In large studies, approximately two-thirds of patients had a history of trauma.[25] Stroobandt et al found a trauma rate of 80%.[26] This was similar to our finding in which 71.8% were ascertained to have a traumatic origin. This was in contrast to Konig et al, who found trauma in only 48%.[12] This rate depends, to a large extent, on the number of patients in whom a history of trauma could be recalled.

The presence of coagulopathy appeared to play a role in the level of consciousness at presentation. More patients with coagulopathy in our study tended to present with lower GCS scores. As expected, this should relate to larger hematoma volumes. This correlated well with our study in which we found that patients with coagulopathy had higher mean hematoma volumes only after correcting for outliers (127.93 ml vs 123.77 ml ; p=0.7345). Is it possible that other factors besides the volumetric effect are responsible for the lower GCS at presentation?

The incidence of bilateral hematoma in patients with coagulopathy in our study was 36.2%. This was much higher compared to previous reports, which ranged from 17.4% to 25.52%.[12,27,28]. Our findings, however, appear to be congruent with those of Oyama et al, who postulated that bilateral CSDHs tended to occur more frequently in patients with coagulation abnormalities.[29] There is, however, no clear-cut consensus on the role of anticoagulation in the bilateral occurrence of CSDH. Garba Sunday et al[30] in their review article, noted that anticoagulation contributed to bilateral CSDH, while Yu-Hua Huang[28] found no association. In our environment, in which routine brain scans are not done and late presentation is common, this may have contributed to the increased frequency of bilateral lesions seen.

In our study, we found that left-sided lesions were more common. Patients with coagulopathy had twice as many left-sided lesions as right-sided lesions. In patients without coagulopathy, right-sided lesions were more common. This difference was statistically significant (p=0.047). MacFarlane et al had previously noted that left-sided lesions were more common.[31] With the higher occurrence of left-sided lesions, it is important to ask if this is a result of a biological or anatomical process. Right-sided (non-dominant) lesions are generally not associated with communication difficulties or impairment of dominant hand function but rather more subtle symptoms and signs, such as inattention and geographical dyspraxia. This may therefore lead to their underdiagnosis rather than a true increase in left-sided lesions. Coagulopathy resulting in larger hematomas and more significant neurologic deterioration may explain why left-sided lesions were more commonly diagnosed.

With burr hole evacuation of CSDH, there is usually a rapid improvement in the clinical status of patients. Out of seven patients who presented with a GCS<8, only four eventually required ICU admission. Only patients with coagulopathy were admitted into the ICU, and for this reason, they tended to have a longer duration of hospitalization. The longer duration of hospitalization, including ICU care, means that patients with coagulopathy had higher costs of treatment, which has a significant impact in a low-resource setting.[32]

Recurrence of CSDH after the first burr hole craniostomy is not rare. The rate of clinically significant recurrence that required surgical evacuation ranged from 2-37% after initial burr hole evacuation with or without postoperative drain.[33,34] Its causes range from patient-specific risk factors, radiologic risk factors, surgical risk factors, and subdural fluid characteristics.[33] In our study, the recurrence rate was 5.54% and there was no statistically significant difference between the presence or absence of coagulopathy (p=0.983). This is similar to findings by Vinredai et al in their review paper, where they observed that the use of anticoagulants or antiplatelet did not lead to an increase in the rate of recurrence of CSDH.[33] Toshiyuki et al also found that there were no significant differences in the incidence of radiographic deterioration or reoperation of ipsilateral or contralateral hematomas between patients with and without antithrombotic therapy after surgical treatment of unilateral CSDH.[24] Forster et al, however, observed that perioperative antithrombotic therapy led to higher recurrence and worse outcomes.[35] Our study also showed that the presence of coagulopathy did not lead to a higher reoperation rate (p=0.549). This is probably because patients with laboratory documented coagulopathy associated with CSDH had pre- and post-operative substitution of coagulation factors.

Coagulopathy has been identified as an independent risk factor for mortality.[5]Mortality rates in CSDH patients have been reported to be as high as 16.7%.[15] In this study, only one mortality (0.72%) was observed, and it was in a patient with coagulopathy. The perioperative correction of coagulopathy in patients undergoing surgery may have significantly reduced its impact. This makes it extremely important that all patients with CSDH should have coagulation screening done. When derangements are seen, these should be corrected as early as possible before surgery. The overall outcome following surgery for CSDH is, however, favourable.[25]

The presence of coagulopathy in chronic subdural haematoma (CSH) requires correction of coagulation to facilitate surgery.[12] In virtually all our patients who had derangements in clotting profile, tranexamic acid was commenced, and fresh frozen plasma (FFP) was given before and after surgery. Fresh whole blood was also given when FFP was not available or the patient was noted to have low hematocrit. In hospitals where facilities for advanced coagulation studies, including clotting factor assays,

could be done, substitution of the deficient factor pre- and postoperatively has been found to improve outcome.[12] Fresh frozen plasma is rich in factors II, V, VIII, IX, X, XI, and antithrombin III and is readily available in our environment. Since there is a high prevalence of coagulopathy in CSDH patients, it is recommended that, in emergency cases, FFP be administered if there is not enough time to carry out laboratory tests. This is especially important in low-resource settings where delayed referral and late presentation are common.

Limitations of the Study

Due to challenges with diagnostic laboratory capability, the definition of coagulopathy was narrow.[36] It is thus possible that some patients with subclinical coagulopathy would have been missed, leading to a lower incidence of coagulopathy. The relatively small sample size may have impacted the power of the study.

Conclusion

There is a high incidence of coagulopathy in patients who have CSDH requiring surgical evacuation, irrespective of haematoma volume. Our results suggest that patients with coagulopathy are more likely to present on the left side, have lower admission GCS scores, require ICU admission after surgery, and have longer hospital stays. The presence of coagulopathy did not increase the frequency of bilateral lesions, clinical recurrence after surgery, or need for reoperation. This therefore highlights the need for pre- and post-operative substitution of coagulation factors, which improves outcomes despite poor neurological status at presentation.

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