ISSN 1220-8841 (Print)
ISSN 2344-4959 (Online)

ROMANIAN
NEUROSURGERY

Vol. XXXX | No. 2 June 2026

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DOI: 10.33962/roneuro-2026-035

Romanian Neurosurgery (2026) XXXX (2): pp. 182-185
DOI: 10.33962/roneuro-2026-035
www.journals.lapub.co.uk/index.php/roneurosurgery

Primary malignant transformation of an intradiploic epidermoid cyst to invasive squamous cell carcinoma: a case report

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Keywords
Squamous cell carcinoma, Epidermoid cyst, Skull neoplasms, Occipital bone, Neoplastic cell transformation

ABSTRACT

Malignant transformation of an intradiploic epidermoid cyst into an invasive squamous cell carcinoma (SCC) is an exceptionally rare and highly aggressive clinical entity. We present the case of a 72-year-old male with a three-year history of worsening headaches. Neuroimaging revealed an extra-axial cystic lesion arising from the right occipital diploic space, associated with a distinct calvarial bone defect and significant mass effect on the posterior fossa structures.

The patient successfully underwent surgical resection under a tailored neuroanesthetic regimen. Histopathological examination confirmed a primary intradiploic epidermoid cyst with malignant transformation to invasive SCC. This report highlights the clinical presentation, complex intraoperative neuroanesthetic and surgical challenges, radiological markers of malignancy, and critical pathological findings of this rare condition.

INTRODUCTION

Primary intradiploic epidermoid cysts are rare, benign, slow-growing congenital lesions resulting from ectodermal inclusions trapped within the diploic space of the skull during neural tube closure [1]. While typically indolent and asymptomatic for decades, these cysts can grow large enough to cause calvarial swelling, headaches, or focal neurological deficits.

Malignant transformation of a primary intraosseous epidermoid cyst into invasive squamous cell carcinoma (SCC) is an exceedingly rare phenomenon, with only six cases reported in the medical literature [2,3]. Malignant transformation converts a benign, easily plane-dissectable lesion into an infiltrative, highly aggressive tumor that carries a poor prognosis. This case report details the successful surgical management and multi-disciplinary lessons learned from a patient presenting with this rare pathology.

CASE DESCRIPTION

This case report meets the CARE (CAse REport) guidelines. A 72-year-old male presented with a 3-year history of progressive, worsening headache. On clinical evaluation, there was an insidious

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ISSN 2344-4959 (online)
ISSN 1220-8841 (print)

First published
June 2026 by
London Academic Publishing
www.london-ap.uk

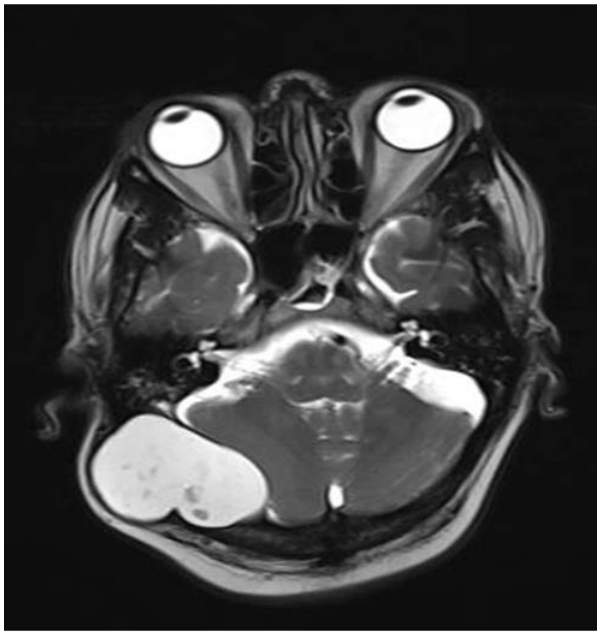
onset of symptoms without focal motor deficits, though signs of localized skull tenderness were noted.

Magnetic Resonance Imaging (MRI) demonstrated a large extra-axial cystic lesion arising directly from the right occipital diploic bone. The lesion was associated with a prominent calvarial bone defect and exerted a marked mass effect on the adjacent posterior fossa structures, including the cerebellum and brainstem (Figure 1). While the presentation primarily mimicked a benign intradiploic epidermoid cyst, the irregular bone destruction and areas of heterogeneous enhancement raised suspicion for a more aggressive process.

Given the posterior fossa localization and mass effect, a meticulous neuroanesthetic plan was executed to optimize cerebral perfusion and mitigate an intracranial pressure (ICP) surge. Smooth anesthetic induction was achieved using propofol, rocuronium, and fentanyl to prevent abrupt hemodynamic fluctuations. Following endotracheal intubation, an ultrasound-guided scalp block was performed to minimize the surgical stress response and manage perioperative pain. Anesthesia was maintained with an oxygen-air-sevoflurane mixture, titrated alongside a continuous infusion of dexmedetomidine at 0.7 mcg/kg/hr. End-tidal carbon dioxide monitoring was utilized to avoid hypercarbia and secondary increases in ICP.

Surgical exploration revealed a highly adherent, infiltrative tumor capsule that violated normal tissue planes, directly invading the occipital bone and underlying dura mater. Despite the loss of the typical benign dissection planes and increased vascularity from tumor neovascularization, an uncomplicated near-total surgical resection was successfully achieved. Copious saline irrigation and perioperative steroids were administered to prevent chemical meningitis from keratinaceous cyst spillage. The patient emerged stable from anesthesia without immediate lower cranial nerve deficits or respiratory compromise.

Figure 1. Axial T2-weighted magnetic resonance image demonstrating a large hyperintense cystic lesion in the posterior fossa, with compression of the cerebellar hemisphere and mass effect on adjacent posterior fossa structures, consistent with an intradiploic epidermoid cyst with malignant transformation. [The manuscript's case narrative states the lesion was right-sided, while this figure caption as submitted states “left posterior fossa”; author to confirm the correct laterality before publication.]



Pathological Findings

Grossly, the resected tissue consisted of fragmented bony elements and soft, friable, keratin-flecked cyst walls. Microscopic evaluation confirmed the diagnosis of malignant transformation arising directly within a pre-existing epidermoid cyst.

Portions of the cyst wall retained an orderly lining of well-differentiated, stratified squamous epithelium, lacking cytological atypia. Abrupt zones of transition were observed, demonstrating severe dysplasia progressing to squamous cell carcinoma in situ (Bowenoid changes) and frankly invasive squamous cell carcinoma. The malignant squamous cells demonstrated marked pleomorphism, high mitotic activity, and hyperchromatic nuclei; sheets of them were seen directly infiltrating the dense fibrotic cyst wall and adjacent skull bone trabeculae. Distinct keratin pearl formation was observed (Figure 2).

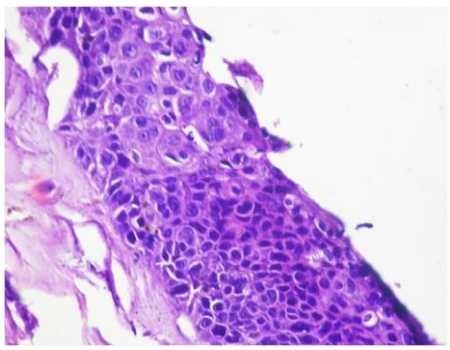


Figure 2. (A) High-power histopathological section (H&E stain) detailing a segment of the cyst lining undergoing malignant transformation. There is a distinct loss of normal cellular architecture, characterized by marked nuclear pleomorphism, hyperchromasia, high nuclear-to-cytoplasmic (N:C) ratios, and mitotic figures spanning the thickness of the squamous epithelium, consistent with high-grade dysplasia / carcinoma in situ. (B) Histopathological section showing invasive squamous cell carcinoma arising from an epidermoid cyst wall. The specimen demonstrates malignant squamous epithelial proliferation with keratinization and stromal invasion adjacent to the cystic epithelial lining.

Discussion

Benign epidermoids typically feature a distinct pearly capsule that is easily dissected from surrounding neural structures [4]. However, upon malignant transformation, the tumor aggressively infiltrates the dura, bone, venous sinuses, and adjacent parenchyma. Gross total resection is frequently hindered by these compromised tissue planes [5]. Rapid symptom progression, severe localized pain, or new-onset cranial neuropathies in a patient with a known stable cyst should immediately raise suspicion for malignant transformation.

While standard epidermoid cysts appear homogeneous with clear margin definitions, features that favor malignant change include irregular or poorly defined margins, direct osteolytic bone destruction, structural heterogeneity or loss of uniform diffusion restriction on DWI, vivid contrast enhancement along the cyst capsule or within soft-tissue nodules, and significant perilesional vasogenic edema [6,7].

Posterior fossa tumors necessitate advanced hemodynamic and airway planning. The potential for sudden brainstem manipulation or venous air embolism, particularly if the patient is positioned semi-sitting, demands continuous EtCO2 tracking and invasive monitoring. Utilizing multimodal regimens like intraoperative dexmedetomidine infusion combined with ultrasound-guided scalp block facilitates smooth emergence, enabling immediate postoperative neurological evaluation while safeguarding against abrupt spikes in intracranial pressure [8,9]. There is always disruption of normal intracerebral blood flow in any kind of brain tumor pathology that compresses the ventricles [10]. Therefore, for the need of optimal prediction of emergence, even exhaled-gas assessment of propofol should be incorporated to generate accurate prediction models [11,12,13].

Accurate diagnosis requires comprehensive sampling of the specimen to capture the focal transitional zones. Histopathology must demonstrate a clear continuum from benign stratified squamous epithelium to high-grade dysplastic changes and invasive cords of SCC [14]. Immunohistochemical staining with p63 or p40 clarifies the lineage, while elevated proliferation markers like Ki-67 define the high-grade components [15]. However, due to institutional constraints, we were not able to perform these tests for the present case. Nevertheless, a systemic workup is mandatory to rule out osteolytic skull metastasis from a distant primary carcinoma.

Conclusion

Primary intradiploic SCC remains a highly lethal malignancy with a tendency for early local recurrence and leptomeningeal carcinomatosis. Even after successful surgical cytoreduction, the overall prognosis remains poor. Aggressive management using adjuvant radiotherapy, with or without chemotherapy, is generally indicated to target microscopic residual disease. Early surgical intervention for symptomatic intradiploic cysts before malignant transformation occurs offers the best opportunity for a definitive cure.

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