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Recurrent primary extracranial scalp meningioma: a case report

B. Ulhaq[1], S. John[2], S. Ziaulhaque[3], Siraj Ulhaq[4]

[1] Spine Surgery, Combined Military Hospital, Rawalpindi, PAKISTAN
[2] Neurosurgery, United Medical and Dental College, Karachi, PAKISTAN
[3] Neurosurgery, Akhtar Saeed Medical & Dental College, Lahore, PAKISTAN
[4] Neurosurgery, Asfendiyarov Kazakh National University, Almaty, KAZAKHSTAN

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Extracranial meningioma,
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En bloc excision

ABSTRACT

Background: Primary extracranial meningiomas are rare neoplasms that may arise without intracranial or dural involvement and are frequently misdiagnosed as benign scalp lesions. A lack of routine histopathological evaluation of excised scalp masses may delay the diagnosis of rare tumors.

Case Presentation: We report a 28-year-old woman with a recurrent left parieto-occipital scalp mass associated with positional headache and intermittent vertigo. She had undergone prior excision of a similar lesion five months earlier without histopathological examination and was lost to follow-up. Examination revealed a firm, mobile subcutaneous mass. Contrast-enhanced magnetic resonance imaging demonstrated a well-circumscribed extracranial lesion without intracranial extension or dural attachment.

The patient underwent complete en bloc excision. Intraoperatively, the lesion was highly vascular. Histopathology confirmed a World Health Organization Grade I meningothelial meningioma with epithelial membrane antigen and progesterone receptor positivity and a low Ki-67 index (~2%). At one-year follow-up, the patient remained recurrence-free with complete symptom resolution.

Conclusion: This case highlights a diagnostic pitfall in which the omission of histopathological evaluation following initial excision led to the delayed diagnosis of a primary extracranial scalp meningioma. It emphasizes that imaging alone is insufficient to exclude rare neoplasms and underscores the need for routine pathological examination of all excised scalp lesions to prevent misdiagnosis and recurrence.

BACKGROUND

Meningiomas are typically benign neoplasms arising from arachnoid cap cells of the meninges and account for approximately 30-37% of all primary central nervous system tumors [1]. Although

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Corresponding author:
Simon John

Neurosurgery, United Medical and Dental College, Karachi, Pakistan

sims4.jpanthers@example.com

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most are intracranial, primary extracranial meningiomas are exceedingly rare, representing less than 2% of all meningiomas [2].

Primary extracranial meningiomas occur independently of intracranial lesions and lack dural attachment, distinguishing them from secondary extracranial extensions of intracranial tumors. Fewer than 400-500 cases have been reported in the literature across all anatomical sites, with the scalp among the most frequently involved extracranial locations.

The pathogenesis remains uncertain, with proposed mechanisms including ectopic arachnoid cell rests, migration along cranial nerve sheaths, or meningothelial differentiation of pluripotent mesenchymal cells [3]. Clinically and radiologically, these lesions often mimic more common benign scalp tumors, posing a diagnostic challenge [4].

We report a rare case of recurrent primary extracranial scalp meningioma without dural attachment in a young female, initially excised without histopathological evaluation, resulting in early recurrence. This case highlights critical diagnostic pitfalls and underscores the importance of complete surgical excision and routine pathological assessment of all scalp lesions.

To our knowledge, recurrent primary extracranial scalp meningioma without dural attachment following excision without histopathological evaluation is rarely reported, highlighting a critical diagnostic and management gap.

Case Presentation

A 28-year-old woman with no significant prior medical history presented with a recurrent swelling located in the left parieto-occipital region of the scalp near the vertex. This was accompanied by severe unilateral headaches that worsened with changes in position, along with intermittent episodes of vertigo, collectively causing marked impairment in her daily activities over the preceding five months.

She had undergone prior excision of a similar lesion at an outside facility five months earlier; however, histopathological evaluation was not performed, and she was subsequently lost to follow-up. The swelling recurred nearly 2 cm away from the same site and progressively increased in size over the following year.

There was no history of head trauma, prior epidural hematoma or intracranial surgery, radiation exposure, or family history of neoplastic disease. Additionally, there were no clinical features suggestive of neurofibromatosis or other tumor predisposition syndromes.

On examination, the lesion was firm, mobile, and subcutaneous, located near the previous surgical scar, without overlying skin changes or tenderness. The lesion measured approximately 2.0 × 2.3 × 2.5 cm. No regional lymphadenopathy was noted. Neurological examination was normal.

She underwent a non-contrast CT scan of the brain, which demonstrated a scalp swelling associated with scalloping of the outer table of the skull, with extension into the calvarial bone.

Contrast-enhanced MRI revealed a well-circumscribed extracranial mass without intracranial extension, dural tail sign, or pachymeningeal enhancement, favoring a benign scalp tumor of uncertain origin (Figure 1). These findings effectively excluded secondary extracranial extension of an intracranial meningioma.

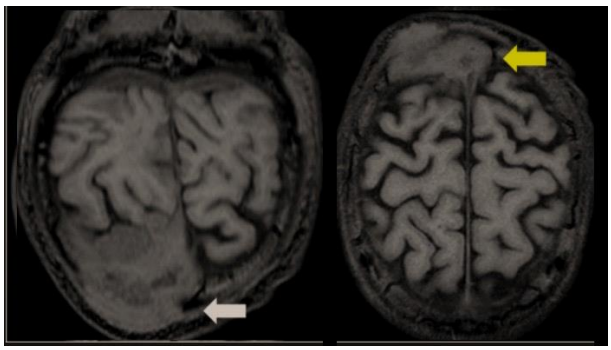

Figure 1. Contrast-enhanced MRI brain (axial views), demonstrating a primary extracranial scalp meningioma in the left parieto-occipital region. (A) Lower axial cut showing a well-defined extracranial soft tissue mass (white arrow) without evidence of intracranial extension, dural tail sign, or perilesional edema. (B) Higher axial cut at the level of the vertex demonstrating the scalp-based location of the lesion (yellow arrow) with preservation of the underlying cortical architecture and no pachymeningeal enhancement, confirming the extracranial nature of the tumor.

The patient underwent en bloc surgical excision under general anesthesia. Intraoperatively, the tumor was highly vascular, and bleeding necessitated transfusion of one unit of packed red blood cells.

Histopathological examination confirmed a WHO Grade I meningothelial meningioma, with immunohistochemistry positive for epithelial

membrane antigen (EMA) and progesterone receptor (PR), and a low Ki-67 index (~2%).

Additionally, CT angiography and CT venography of the cerebral vasculature were performed to exclude any vascular communication with the intracranial compartment, and the findings were unremarkable.

At the 1-year follow-up, the patient remained recurrence-free, with complete resolution of headaches and a significant improvement in quality of life.

Discussion

Primary extracranial meningiomas pose significant diagnostic challenges due to their atypical location and absence of dural attachment, often mimicking common benign scalp masses. Unlike secondary extracranial meningiomas arising from direct intracranial extension, primary lesions are entirely extracranial. In the largest reported series by Rushing et al., comprising 146 cases, the scalp was the most common site of involvement; however, recurrent cases without dural attachment remain exceptionally uncommon [4].

Table 1. Selected reported cases of primary extracranial scalp meningioma without dural attachment. WHO, World Health Organization; NR, no recurrence; M, male; F, female.

Author(s)	Year	Location	Age / Sex	WHO Grade	Dural Attachment	Recurrence (months)
Rushing et al. [4]	2009	Scalp (series, n=146)	0.3–88 yrs; M>F	I (88%)	Absent	Yes (26/110 at ≥120 months)
Tong et al. [3]	2020	Posterior scalp, near the vertex	30s / M	I (meningothelial)	Absent	Yes (no histology at index)
Kieu et al. [5]	2021	Left frontoparietal scalp	38 / M	I (meningothelial)	Absent	No (NR)
Kim et al. [6]	2018	Right frontotemporal scalp	Adult / F	I (meningothelial)	Absent	No (48 mo)
Present Case	**2025**	**Left parieto-occipital scalp**	**28 / F**	**I (meningothelial)**	**Absent**	**Yes (5 months; no histology at index)**

Our case is notable for early recurrence following excision without histopathological evaluation, a critical and underreported issue, likely resulting in incomplete resection and failure to recognize the lesion's neoplastic nature [4,5]. Radiologically, the absence of a dural tail sign, intracranial extension, or perilesional edema contributed to diagnostic uncertainty, underscoring that extracranial meningioma should remain a differential diagnosis for any atypical or recurrent scalp mass. Histologically, extracranial meningiomas are indistinguishable from intracranial counterparts, with characteristic EMA and PR positivity and low proliferative indices, as seen in our patient [4,7].

The pathogenesis remains debated; proximity to the prior surgical site raises the possibility of ectopic meningothelial proliferation or implantation, though a definitive mechanism cannot be established. Complete en bloc resection is the treatment of choice and is critical to prevent recurrence, as illustrated by our case [4,5]. This report reinforces the importance of routine histopathological evaluation of all excised scalp lesions and highlights that preoperative imaging alone may be insufficient for definitive diagnosis.

Conclusions

Recurrent primary extracranial scalp meningioma without dural attachment is an exceptionally rare entity that can mimic benign scalp lesions, leading to misdiagnosis and inadequate initial treatment. This case highlights the critical importance of routine histopathological evaluation, accurate preoperative assessment, and complete surgical excision. Early recognition and meticulous management are essential to prevent recurrence and ensure optimal outcomes.

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