CALVARIAL MENINGIOMA: CASE REPORT OF AN OSTEOLYTIC LESION MIMICKING METASTASIS

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ABSTRACT
Meningiomas are the most common benign intracranial neoplasm which are predominantly dura based. Primary intraosseous calvarial meningiomas are rare. Although majority of them are benign, atypical and malignant variants also occur and its awareness is important to prevent any misdiagnosis. The authors take this opportunity to report a case of primary calvarial meningioma along with review of literature, occurring in a 47 year old female, for its rare occurrence.

Keywords: meningioma, intraosseous, calvarial, extradural


INTRODUCTION
Primary intraosseous calvarial meningioma is a type of primary extradural meningioma. Primary extradural meningioma is a rare entity of meningioma occurring at varied sites like cranial bones, nose, paranasal sinus, lungs and mediastinum; primary calvarial meningioma is even rarer1. As the name indicates, primary intraosseous calvarial meningioma is limited to the skull bone with no epidural or subcutaneous component. These lesions behave mainly in a benign fashion but occasional atypical and malignant cases have been reported2.

CASE REPORT
A 47 year old Saudi female presented with the complaint of a painful swelling in the posterior aspect of head since 6 months. It was not associated with any history of seizures, vomiting or any neurological deficit. There was no history of trauma or weight loss. On examination, the swelling was bony hard, fixed, non tender, non pulsatile and non compressible, measuring 3x3 cm with no skin changes. The swelling had an irregular surface, ill defined margin, and was continuous with the calvarium. General as well as systemic examinations and basic routine investigations were within normal limits.

CT scan showed an osteolytic lesion in the left posterior parietal calvarial bone with no gross intracranial soft tissue component (Figure 1). The lesion measured 2.5 cm in diameter. Ventricular system was normal with no midline shift or deformity. Extra axial CSF spaces were normal with no posterior fossa abnormality. Considering the possibility of metastatic bone tumor, whole body scintigram was performed, which did not show any abnormality.

Figure 1: CT Head showing an osteolytic lesion in the posterior parietal calvarial bone.

Patient underwent posterior craniotomy and cranioplasty. Peroperatively posterior parietal bone was diffusely expanded in size of 3x2.5 cm with smooth outer and inner table, which was excised. The excised tissue was sent for histopathological examination.

Gross examination showed a single bony tissue, measuring 3x2.8x0.5 cm, with focal darker areas. The bony tissue was processed for decalcification,
cut and entirely processed. Microscopic examination revealed a tumor consisting of nests of oval cells and masses of spindle cells in between the normal bony trabeculae (Figure 2,3). The cells were arranged in a syncytium with indistinct cytologic borders. At few places, rudimentary meningothelial whorls and nuclear clearing were identified (Figure 4). No evidence of necrosis, significant mitoses or nuclear pleomorphism was seen. Immunohistochemical examination revealed tumor cells positive for epithelial membrane antigen (EMA) (focally) (Figure.5A) and vimentin (Figure.5B). Differential diagnosis of metastatic carcinoma was also considered but tumor was negative for CK5/6, CEA and CKAE1/AE3.

Figure 2: section showing nests of oval & spindle cells interspersed between bony trabeculae (H&E; 40x).

Figure 3: section showing tumor cells comprising of oval and spindle cells, along with normal bony trabeculae (H&E; 200x).

Figure 4: section showing nests of oval cells with few showing nuclear clearing (↑) (H&E; 400x)

Figure 5: (A) section with tumor cells showing positivity to EMA (40x) & (B) section with tumor cells showing positivity to vimentin (400x)
DISCUSSION

Meningiomas are the most common benign intracranial neoplasm. They account for 15-20% of all intracranial tumors, being second only to gliomas in prevalence. The cell of origin for meningioma is meningocyte, found in the arachnoid layer of the meninges, which usually accumulate in clusters known as “arachnoid cap cells”. The meningiomas are usually subdural and intracranial/spinal in location but occasional epidural meningiomas also occur. The meningioma arising outside the intracranial/spinal location are considered primary extradural/ectopic.

Although reported sporadically, primary extradural meningioma is a well accepted entity, accounting for 1–2% of meningiomas. The first case of primary extradural meningioma was reported by Winkler in 1904; a case of paravertebral subcutaneous primary extradural meningioma in a 10-year-old girl. Paranasal sinuses and nasal cavity are the most common extradural locations. Meningiomas rarely originate within the skull bone, referred to as calvarial meningioma or primary intraosseous meningioma.

One hypothesis states that primary intraosseous meningioma arise from arachnoid cell rests which are caught within extradural tissues at the time of embryologic development. These arachnoidal cells may accompany the vessels, exiting nerves or the periosteam attached to the sutures. Trauma is another factor being postulated in the etiology of primary intraosseous meningiomas; however due to the uncertain association of meningioma with the head injury, its role in the development of meningiomas is controversial.

Primary intraosseous meningiomas are usually reported at fronto-temporal region, orbits and anterior cranial fossa, having more predilection for areas near the suture lines, especially the coronal or pterional sutures. Males and females are equally affected. Primary intraosseous meningiomas have a bimodal age distribution with a peak during the second decade and a second peak at fifth to seventh decade.

Radiologically, intraosseous meningiomas show hyperostosis in majority of the cases, however, their appearance can vary from the common osteoblastic appearance to the rare osteolytic form, as has been in our case. The osteolytic lesions generally show a more aggressive course and represent an advanced stage, unlike our case which showed no features of atypia or malignancy. Majority of these primary intraosseous meningiomas are histologically benign, however atypical and malignant cases (upto 11-33%) have also been reported.

An important differential diagnosis is meningioma en plaque, which shows tumor in the form of a sheet, along the dural surface. This growth is accompanied with marked hyperostosis, which is often disproportionate to the actual tumor size. Meningioma en plaque characteristically involves the sphenoidal wing in contrast to primary intraosseous meningioma, which has a predilection for areas near the suture lines.

The other differential diagnosis is metastatic deposits, which on microscopic examination reveal cells of primary origin and can be further confirmed on immunohistochemistry.

CONCLUSION

In spite of primary intraosseous meningioma being rarer lesion, their possibility should always be kept in the differential diagnosis specially if the lesion is exclusively in cranial bones and near the suture.

REFERENCES
