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INTRA-OSSEOUS MENINGIOMA – A RARE ENTITY

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ABSTRACT

Meningioma are the most common and benign intracranial neoplasms, accounting for 20% of all primary intracranial tumors. The subset of extradural meningioma arising from the calvarium is called Intraosseous Meningioma. Primary intra-osseous meningioma of calvarial origin is a rare occurrence confined to the calvarial bone. This report outlines the clinical and radiological features of a frontal intraosseous Meningioma, along with its surgical management.

KEYWORDS: Calvarial lesion, Intra-osseous meningioma, Hyperostosis, Surgical excision.

INTRODUCTION

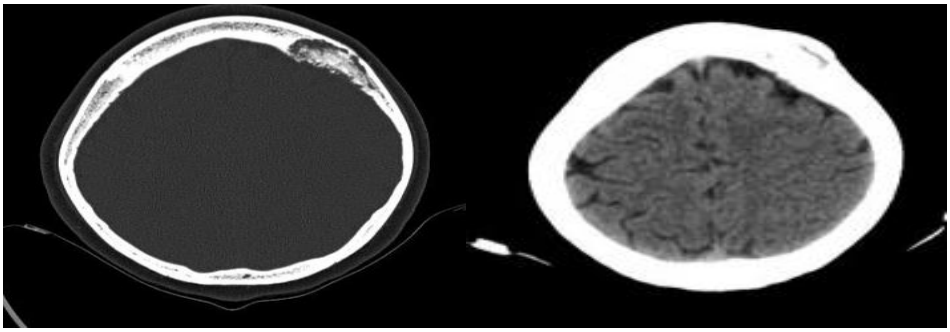
There are various causes for common scalp swellings, notably involving the calvarium. The differential diagnosis of a bony calvarial swelling includes Osteoma, Epidermoid tumor, Eosinophilic granuloma and rarely Metastasis or Intra-osseous Meningioma. The importance of clinico-radiological correlation along with surgical management of a rare entity is described in this report.

CASE REPORT

A 44 years old lady, presented with a swelling over the left frontal scalp since a year, and of late she was complaining of pain over the swelling. She had no history of prior head trauma or fever. There was no history suggestive of raised intracranial tension or any focal neurological symptom.

Clinically, there was about 4x3cm well defined hard swelling over the left frontal region, with no discoloration of the overlying scalp. There was no evidence of discharge or audible bruit from the swelling. There were no other swellings over the scalp.

CT scan brain with bone windows showed an expansile osteolytic lesion with central hyperostosis involving frontal bone on the left side abutting the coronal suture; in addition there was erosion of both inner and outer tables of frontal bone (Fig:1). There was no evidence of intra-cerebral extension of the lytic frontal bone lesion. The possibilities of Calvarial intraosseous venous malformation (Hemangioma), Solitary Plasmacytoma or Metastatic tumor were considered. The evaluation for primary source of Metastasis was done which did not reveal any evidence of systemic malignancy.



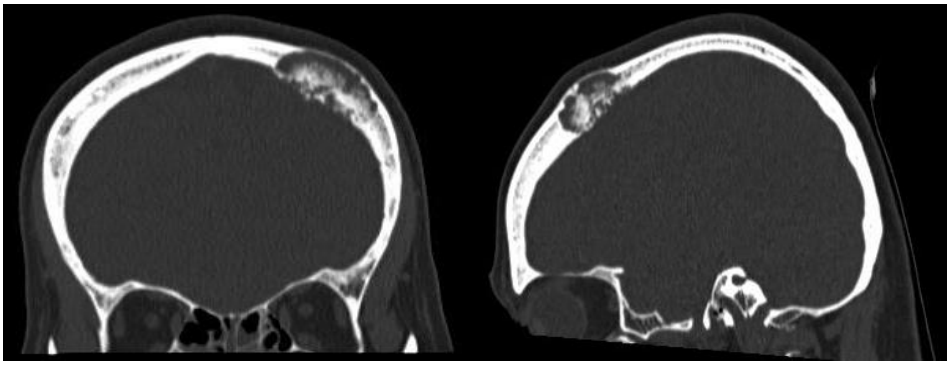


Fig. 1: CT images showing hyperostotic and osteolytic areas involving the left frontal bone, without brain parenchymal involvement.

After detailed counseling and consent, patient underwent left Frontal Craniotomy and wide excision of the bony lesion under general anesthesia. There was hyperostosis of the frontal bone with hemorrhagic areas within the lesion, along with erosion of the inner and outer table of frontal bone. (Fig: 2) There was no involvement of Dura mater, by way of any discoloration or extension of the bony tumor into the Dura. Post-operative period was uneventful, and she is being planned for an elective cranial defect closure (Cranioplasty) at a later date.

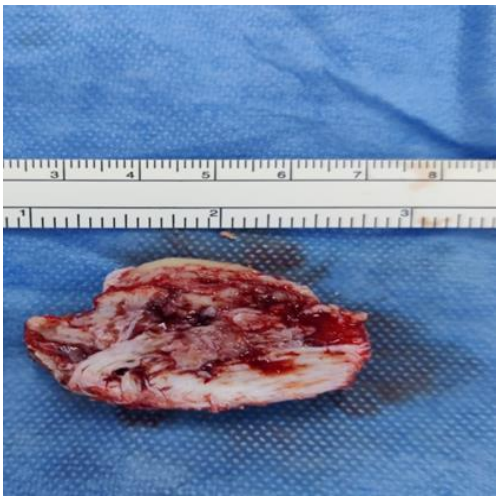


Fig. 2: Surgical specimen showing frontal bone tumor.

The entire cranial bone involving the tumor was subjected to pathological examination. The gross specimen was about 4.5x3 cm with evidence of abnormal reddish gray tumor which was eroding the inner table. Histological studies showed moderate cellular neoplasm with meningothelial cells exhibiting dual architectural pattern. There was well formed concentric whorls mixed with spindle shaped cells and Psammoma bodies. Nuclear pleomorphism and mitotic figures were seen (<4/10 High power fields). The tumor permeates and entraps the bony trabeculae(Fig: 3). The final histopathological diagnosis was Meningioma-Transitional type (WHO grade 1). In view of the final diagnosis, the patient merits regular follow-up.

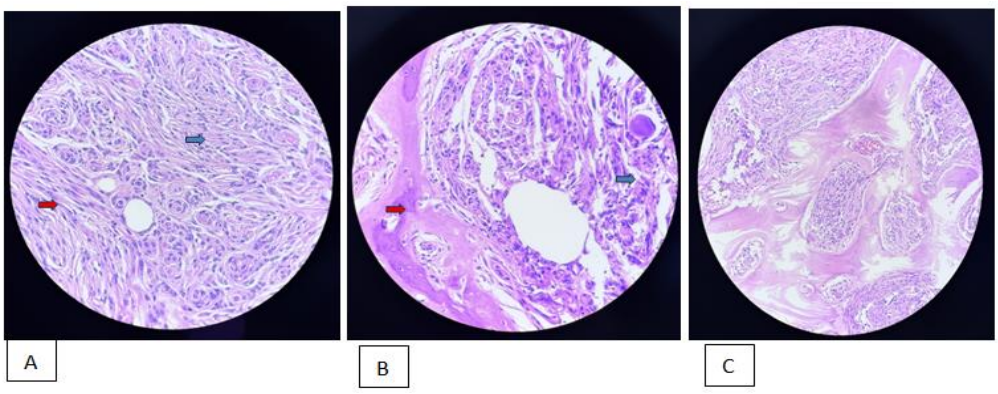


Fig: 3.

Fig: 3(A): High power view (H&Ex400) reveals well-formed concentric whorls of epithelioid meningothelial cells (red arrow) and admixed spindle cells. Tumor cells possess round to oval nuclei, inconspicuous nucleoli with minimal nuclear pleomorphism and infrequent mitotic figures (blue arrow)
Fig:3(B): High Power View (H&Ex400) shows tumor permeating and entrapping bony trabeculae (red arrow) and Psammoma body (blue arrow)
Fig:3(C): (H&E stain, original magnificationx200) shows tumor infiltrating between bony trabeculae.

DISCUSSION

Intra-osseous Meningioma is a rare subtype of Meningioma, and is classified as primary extradural Meningioma. It represents 1-2% of all Meningioma, and arises outside the Dura mater, originating within the calvarial bones and extending either intra or extra-cranially.[1,2]

The pathogenesis of Intra-osseous Meningioma is not well understood, however the following hypotheses have been proposed.

1. Entrapment of arachnoid cap cells within the cranial sutures during embryogenesis.
2. Traumatic displacement of meningothelial cells.
3. Differentiation from pluripotent mesenchymal cells.[1,3]

Based on classification proposed by Lang FF et al[1], Primary extradural Meningioma is classified into three types.
Purely Extra-calvarial (Type I)
Purely Calvarial or Intra-osseous (Type II)
Calvarial with Extra-calvarial extension (Type III)[1]

Our report is that of Type II Intra-osseous meningioma.
Clinically, these tumors present as a slowly growing, painless scalp swelling. Usually, there are no features of raised intracranial tension, or focal neurological signs, unless there is an intracranial extension. Commonly affected part is the frontal or parietal bone. Patients seek medical attention as the concern is related to cosmetic deformity.

Imaging studies, specifically Computed Tomography scan is particularly indicated to assess the extent of bony involvement. Imaging findings reveal hyperostotic (osteoblastic) lesion frequently and less commonly osteolytic variants which may mimic more aggressive pathologies like Metastasis or Multiple myeloma.[2,3] The present report imaging shows both osteolytic and osteoblastic areas within the frontal bone with no intracranial extension.

The treatment modality for such lesions is complete wide excision of the tumor with clear margins, followed by cranioplasty for reconstruction.

Histopathologically, intra-osseous Meningioma shows typical fibroblastic or meningothelial patterns. Immunohistochemistry studies typically show positivity for Epithelial membrane antigen and Vimentin. The majority of the lesions are benign (WHO Grade 1), although atypical and malignant variants have been reported especially with osteolytic lesions.[3,6]

In the present case, the histopathology showed typical meningothelial pattern with Psammoma bodies, along with few areas of mitotic activity and nuclear pleomorphism. This correlates with the radiological findings of both osteoblastic and osteolytic areas in the tumor. Transitional Meningioma denotes an aggressive variant, and hence long term follow up is recommended. In case of recurrence or subtotal excision, adjuvant radiotherapy is recommended.[2,4]

CONCLUSION

The overall prognosis of Intra-osseous meningioma is good following complete excision. However, lesions with osteolytic features or atypical histopathology may recur and hence long term follow up is recommended. This case report highlights the pure Intra-osseous Type II variant of Primary extradural meningioma.

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