Primary intraosseous meningioma of the calvarium presenting as a solitary osteoblastic lesion. Case report

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ABSTRACT
Background: Intraosseous meningiomas are extradural meningiomas, which are rare lesions arising in the skull. They are likely to originate from intraosseous entrapment of arachnoid cells. They are often asymptomatic, but may cause symptoms depending on the location and size. Surgical resection is usually used to treat patients. Radiation therapy and chemotherapy may be needed as an adjuvant therapy in tumors, which cannot be completely resected.

Clinical case - We report a case of a 42–year old male patient presenting with painless, gradually progressing swelling over the right frontoparietal region for 4 years. CT scan of the brain with contrast revealed an expansive osteoblastic lesion with lobulated outlines involving right frontoparietal bone, with obliteration of diploic space, and with associated enhancing diffuse extra-cranial soft tissue component, also with a thin extra-axial soft tissue component suggestive of an osteoblastic intraosseous meningioma. Surgical resection was performed. Biopsy was confirmatory.

Conclusion - Intraosseous meningiomas are osteoblastic or osteolytic. Osteoblastic lesions mimic fibrous dysplasia. They are extradural meningiomas, which are rare lesions arising in the skull.

Keywords: Meningioma, intraosseous, extradural, osteoblastic, calvarial.

INTRODUCTION
Meningiomas can be intradural or extradural. Usually, meningiomas are the primary intradural lesions and are located in the subdural space. Extradural meningiomas, arising in locations other than the dura mater, constitute 1-2 % of all meningiomas [1]. Primary intraosseous meningioma (PIOM) is a subset of extradural meningiomas that arise from bone with no neural attachment [2]. Majority of intraosseous meningiomas arise from cranial bones with few cases arising from mandibles. Extradural meningiomas are also named as ectopic, secondary, extra-calvarial, cutaneous, primary extraneuraxial meningiomas. Extradural meningiomas arising in the skull are named as calvarial, intradiploic and intraosseous [3]. Another nomenclature used is primary extradural meningiomas, which differentiates this tumor from those originating in the dura with extracranial extension, and from extracranial meningiomas, that are distinct metastases from primary intradural meningiomas.

CASE REPORT
A 42-year old male patient presented with painless gradually progressive swelling over the right frontoparietal region for the last 4 years. Computed tomography (CT) of the brain was performed without (Figure 1) and with contrast (Figures 2), showed an expansive osteoblastic/sclerotic lesion with lobulated outlines involving right frontoparietal bone, with obliteration of diploic space, and with associated enhancing diffuse extra-cranial soft tissue component, also with a thin extra-axial soft tissue component suggestive of an osteoblastic intraosseous meningioma. Surgical resection was performed. Biopsy was confirmatory.

Conclusion - Intraosseous meningiomas are osteoblastic or osteolytic. Osteoblastic lesions mimic fibrous dysplasia. They are extradural meningiomas, which are rare lesions arising in the skull.

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A diagnosis of Type III intraosseous meningioma was made. CT brain angiography showed mild hypertrophy of the right superficial temporal artery, which appeared more tortuous as compared to its counterpart. It was stretched and draped over the bony mass, however it was not supplying the mass. No arterial supply to the mass from any intracranial arteries was noted. The anterior third of the superior sagittal sinus was in close contact with the mass. Histopathology showed a tumor with cells in a nested and whorled pattern. Syncytial cell pattern was present. Focal sheets of cells showed an infiltration into surrounding soft tissue. Underlying bone was involved by the tumor. Tumor cells were positive for vimentin and focally positive for EMA. Cytokeratin was negative. MIB labeling index was 2-3%. Diagnosis was meningothelial meningioma (Grade I).

Image 1 - Plain axial brain CT showing expansive osteoblastic lesion in the right frontoparietal region with slightly hyperdense intracranial and extracranial soft tissue components.

Image 2 - CECT Brain axial (a-c), coronal (d) showing an expansile osteoblastic lesion in the right frontoparietal region with an enhancing intracranial and extracranial soft tissue component.
Image 3 – axial CT brain bone window (a-d) showing an expansile osteoblastic lesion in the right frontoparietal bone with obliteration of diploic space.

Image 4 – Coronal (a), Sagittal (b) CT of the brain bone windows showing an expansile osteoblastic lesion in the right frontoparietal bone with obliteration of diploic space.
DISCUSSION

Meningioma constitutes about 20% of primary intracranial tumors. Winkler in 1904, first described a meningioma in extradural locations [4]. Different hypothesis exists regarding the intraosseous and primary extradural meningioma. They usually arise from arachnoid cap cells or ectopic meningiocytes, trapped in the cranial sutures during moulding of the head at birth [5]. Calvarial meningiomas are prone to develop malignant changes (11%) compared with intracranial meningiomas (2%) [1].

Lang and colleagues have classified primary extradural meningiomas as type- I Purely extracalvarial tumors, Type II – Purely calvarial tumors and Type III – calvarial tumors with extracalvarial extension. Each category is further divided into two subsets, depending on anatomical location: C – convexity, and B – Skull base. Hence, intraosseous meningioma is Type II and III primary extradural meningioma [2].

Common locations of intraosseous meningiomas are frontoparietal and periorbital regions [6]. They are usually firm and painless with intact overlying skin, and are usually detected incidentally. Neurological symptoms and signs are usually absent. But it may present with headache, vague sensations in the head, seizures, vomiting, dizziness and tinnitus, which may precede months to years prior to the diagnosis. Though, skull base intraosseous meningioma is painless and slow growing, may present with cranial nerve deficiency like ophthalmoplegia or visual field defects, proptosis, deformity, atrophy of the optic nerve. Lesions involving paranasal sinuses or nasal cavity may present with nasal obstruction or epistaxis.

Intraosseous meningiomas can be osteoblastic or osteolytic. The majority is osteoblastic. X-ray of the skull often shows hyperostosis, irregular calcific foci and atypical vascular markings. CT scan with bone window shows focally thickened hyperdense lesion causing an expansion of calvarium. It is usually hyperdense on plain CT with CT value +65-+85 HU and shows dense enhancement on contrast study as intradural meningiomas. Due to bone expansion and ground glass appearance of intraosseous meningiomas, it mimics fibrous dysplasia. Entities, which mimic osteoblastic intraosseous meningiomas, are osteoma, fibrous dysplasia, Paget's...
disease, meningioma en plaque, osteosarcoma. Fibrous dysplasia usually stops its growth after puberty, while intraosseous meningioma appears after puberty and continues to grow slowly. Fibrous dysplasia lacks tumors blush on angiography, which is seen in meningiomas. Serum Alkaline phosphatase levels are raised in Paget's disease [3, 6].

Osteolytic intraosseous meningiomas are rare and are seen as lytic lesions on skull X-ray. On CT scan, they show expansion, thinning and interruption of the inner and outer cortex. They are hyperdense with respect to the brain in plain CT, and show homogeneous enhancement in contrast view. Differential diagnosis for osteolytic meningiomas are hemangiomas, dermoid, epidermoid, Brown tumor, Giant cell tumor, Aneurysmal bone cyst, eosinophilic granuloma, plasmacytoma and metastases [3, 6].

MRI findings in both osteolytic and osteoblastic subtypes of intraosseous meningiomas are as seen in intradural meningiomas i.e., hypointense on T1WI, hyperintense on T2WI with marked homogeneous enhancement on contrast study. Though dural tail is not observed, gadolinium enhancement of the underlying dura may be seen due to dural irritation or tumor invasion [6].

On histopathology, the most common lesion is meningotheliomatous meningioma. Other histological types are psammomatous, transitional, microcystic, choroidal, fibroblastic, atypical and malignant [6].

The diagnostic criteria of an intraosseous meningioma are lack of involvement of the brain, arachnoid and dura; location should be epidural and intraosseous; and presence of histopathological features of a meningioma [7, 8].

Ectopic meningiomas can arise from the calvarium, perineural sheath of cranial nerves, paranasal sinuses, orbit, salivary glands. Primary intraosseous meningioma is a subset of extradural meningioma, arising in the bone and constituent 2/3rd of all extradural meningiomas. These are usually observed at or near suture lines, usually coronal or pterion sutures, or at previous fracture sites. The age ranges from 7 months to 82 years with male to female ratio 1:1.1[7]. They are usually treated with a wide surgical excision. Adjuvant therapy like radiation therapy, chemotherapy, Gamma knife surgery and biphosphate therapy is considered in unresectable tumors causing neurological deficient or demonstrating malignant changes. Wide surgical resection is followed by the placement of high density polyethylene for cranial reconstruction; it is the treatment of choice [3, 7].

**CONCLUSION**

Intraosseous meningiomas are extradural meningiomas, which are rare lesions arising in the skull. Majority of them are osteoblastic, mimicking fibrous dysplasia. Osteolytic lesions are rare. Though usually asymptomatic, neurological symptoms may occur depending on the location and size. It should be considered as differential diagnosis in radiography of the skull with osteoblastic or osteolytic lesions. Tumor resection and cranioplasty is curative. Adjuvant therapy like radiotherapy and chemotherapy is needed in tumors, which cannot be completely resected.
REFERENCES


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