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Case Report

# A Rare Case of Primary Intraosseous Calvarial Osteoblastic Meningioma of the Frontal Bone in a Young Child

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## Abstract

Meningioma is a common intracranial neoplasm, accounting for 15 - 20% of all brain tumours. The "Primary Extradural Meningioma (PEM)" is an uncommon manifestation and a rare separate subset which arises outside the intracranial compartment, accounting for just 1 - 2% of all meningiomas. Primary calvarial intraosseous meningiomas comprise two thirds of these extradural meningiomas. They are usually seen in the 4th or 5th decades of life and have a predilection for females.

With improved inter disciplinary training, diagnostics and team management, the scope and preview of Maxillofacial Surgery includes craniofacial procedures, done either alone by Maxillofacial Surgeons or as part of superspeciality teams. We present a rare case of a very large Primary osteoblastic intradiploic calvarial meningioma in a young 11 year old male child, with a clinical and radiographic appearance simulating an Osteoma or Fibrous dysplasia. The findings on radiographs, Computed Tomographic (CT) scans, Magnetic Resonance Imaging (MRI), the histological features and surgical treatment of this rare lesion are described.

***Keywords:*** Primary Intraosseous Meningiomas; Calvarial; Frontal Bone; Osteoblastic

## Introduction

Meningioma is the most common intracranial neoplasm, comprising about 15 - 20% of brain tumours [1]. Meningiomas arise from cells that make up the meninges, the membranes that surround the brain and spinal cord. Consequently, they occur intracranially or in the spinal canal. The cells of origin are usually the clusters of arachnoid cap cells forming the external surface of the arachnoid cap villi [1]. Most meningiomas are intradural lesions and are located in the subdural space. However, there exists a separate, rare subtype, accounting for 1 - 2% of all meningiomas, which are extradural in location [2].

These "ectopic" or "Primary Extradural Meningiomas" (PEMs), a term coined by Lang., *et al.* [2], have their origin separate from the dural covering or meninges of any part of the brain or spinal cord. They arise in locations other than the dura mater, such as, calvaria (52.5%), paranasal sinuses (11.5%), orbit (5.5%), neck (6%), chest (4.9%), skull base (3.8%) skin and salivary glands [3]. Extradural meningiomas are also reported to occur in the mandible, abdomen, upper limbs, shoulder, peritoneum and feet [3,4].

Extradural meningiomas that arise in the skull have been referred to as Calvarial, Intradiploic or Intraosseous. These Primary Intraosseous Meningiomas (PIMs) comprise approximately two thirds of all Primary Extradural Meningiomas (PEMs) [2,5]. These lesions affect the bone tissue of the skull and cause external lumps to form on the cranial bone. These tumours mostly affect adults in the fourth to fifth decades, especially females [6]. Osteoblastic or mixed osteoblastic-osteolytic lesions compose most of the intraosseous meningiomas, with purely lytic lesions the least common. The commonly reported craniofacial sites for Primary intraosseous meningiomas are calvarial, sphenoid and orbital bones with a predilection for the Frontoparietal regions, and to a lesser extent the maxilla and mandible [7]. Convexity areas and the skull base are the two major locations for intraosseous meningiomas.

Primary extradural meningiomas are classified by Lang and colleagues as [2]:

- Type I: Purely Extracalvarial
- Type II: Purely Calvarial (Intraosseous)
- Type III: Calvarial (Intraosseous) with Extracalvarial extension

According to the site of the tumour, Lang further subdivided Type II and III lesions as:

- (C): Convexity lesions
- (D): Skull base forms

Convexity intraosseous meningiomas usually grow as painless scalp masses, with a possible relationship to a cranial suture. Skull base meningiomas usually grow as painless masses, and may present with cranial nerve deficits [2].

A rare case of a Primary Osteoblastic Intraosseous/Calvarial meningioma in a young 11 year old male child is presented, causing a prominent dome shaped convexity/bossing of the frontal bone, falling into the category of Type II (C) according to Lang's classification.

## Case Report

An 11 year old male child was brought by his parents for a large swelling over the right side of his forehead. History revealed that it had been noticed approximately six months ago and had progressively increased in size ever since, to reach its present dimensions. There was no history of any previous trauma to the region, or of

any associated pain or discharge from the swelling. The child had two recorded epileptic attacks at the ages of 1month and 8 years respectively. He was not on any anticonvulsant therapy. On examination, there was a large, localised, dome shaped swelling measuring approximately 7 x 6 x 3 cm3 in the right anterior aspect of the frontal bone, extending from the coronal suture region to the right supraorbital rim and bridge of the nose (Figure 1A-1D). The surface of the swelling appeared smooth and the overlying skin was pinchable and demonstrated no specific alterations in its consistency, colour, texture or thickness, and the child denied any changes in sensation. On palpation, the swelling was bony hard in consistency, non-tender, with diffuse borders which blended imperceptibly with the surrounding normal appearing bone of the forehead. All neurological examinations were unremarkable.

Radiographs (Postero-Anterior and Lateral views) revealed a diffuse radio-opaque thickening and hyperostosis of the outer cortical table of the frontal bone, on the right side, extending from the coronal suture region down to the supra-orbital rim and bridge of the nose (Figure 1E, 1F). The enlargement was dome shaped and the radio-opaque mass exhibited a "Ground glass" appearance with features of a "sun-ray"/"sunburst" appearance towards the periphery (Figure 1F). The diploeic space in this region appeared hazy and enlarged. The findings were consistent with a Right sided frontal osteoma or Fibrous dysplasia.

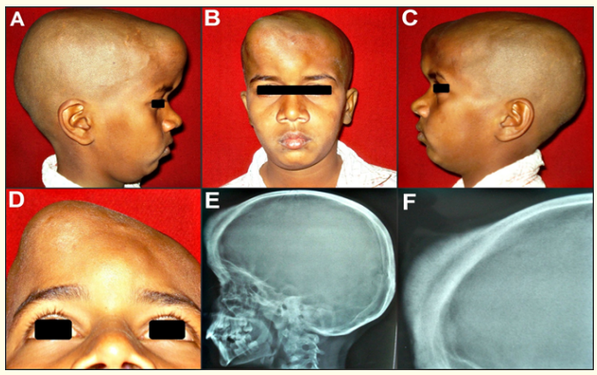


**Figure 1:** (A, B, C, D) A 11 year old male patient with a large dome-shaped, bony hard swelling in the right anterior aspect of the frontal bone, extending from the coronal suture region to the right supraorbital rim and bridge of the nose. (E) Lateral view skull radiograph, revealing a diffuse radio-opaque thickening and hyperostosis of the outer cortical table of the frontal bone, with an enlarged and hazy diploeic space. (F) A typical "Ground glass" radio-opacity with features of a "sun-ray"/"sunburst" appearance towards the periphery.

A provisional diagnosis of Osteoma of the frontal bone was made on the basis of clinical and radiographic features, while the differential diagnosis included Osteoblastoma, Chondrosarcoma, Intraosseous meningioma and Fibrous dysplasia.

Plain, non-contrast cranial/cerebral axial CT scans, using 5 mm serial contiguous sections revealed a bony hyperdense outgrowth arising from the right frontal bone with no obvious intracranial extension (Figure 2A-2C). There was a diploeic expansion of the calvarium in this region with evidence of any parenchymal lesion seen in the brain and the basal ganglia, thalami, cerebellum, brain stem, ventricular system, basal cistern and cortical sulcal spaces all appeared normal. The focally thickened lesion expanding the right frontal region of the calvarium enhanced densely after contrast administration. Pre- and Post- contrasted cranial/cerebral Magnetic Resonance Imaging showed a homogeneous, hypodense expansion of the right frontal bone, with no evidence of intracranial extension. However, a dural reaction was evident adjacent to the intradiploeic lesion (Figure 2D-2F). This was more suggestive of the rare Primary Intraosseous meningioma, rather than an Osteoma or fibrous dysplasia of the frontal bone. MR imaging provided a better anatomic delineation in the evaluation of the soft tissue component, in terms of the dural reaction without dural extension of the lesion.

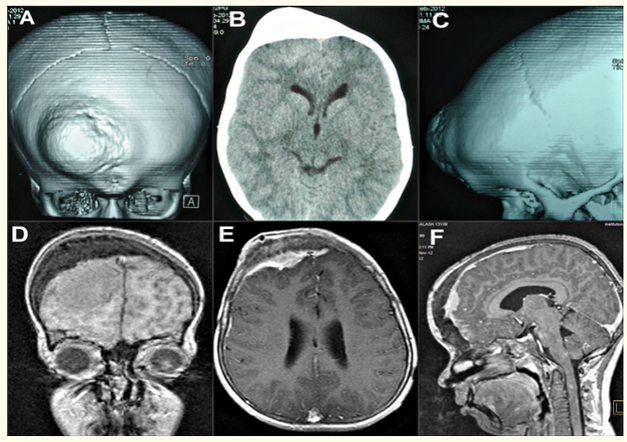


**Figure 2:** (A, B, C) Plain, non-contrast enhanced cranial/cerebral CT scans, using 5mm serial contiguous sections revealed a bony hyperdense outgrowth arising from the right frontal bone with no obvious intracranial extension. (D, E, F) Pre- and Post- contrasted cranial/ cerebral Magnetic Resonance Imaging showed a homogeneous, hypodense expansion of the right frontal bone, with no evidence of intracranial extension. However, a dural reaction is evident adjacent to the intradiploeic lesion.

Owing to the large size of the calvarial hyperostotic mass, producing considerable contour deformity and owing to ambiguity as to the precise diagnosis of the lesion, the patient was planned for marginal resection and debulking of the bony mass with surgical recontouring of the frontal bone under General anaesthesia. As there were no neurological symptoms associated with possible existence of an intracranial component, such as headache, seizures and focal neurological signs, such as limb weakness, dysphagia

etc, and absence of any evidence of an intracranial extension as observed on CT or MRI, a total surgical excision and craniectomy were decided against. It was planned to approach the bony mass by elevation of the scalp flap over a bicoronal incision.

After the routine investigations, the patient was taken up for surgery under General Anaesthesia via Oro-endotracheal intubation with intermittent positive pressure ventilation (IPPV). He was scrubbed and draped with aseptic isolation of the surgical site. A bicoronal incision line was marked (Figure 3A), Adrenalin 1:200000 infiltrated and haemostatic sutures were placed on either side of the proposed incision line (Figure 3B). Incision was made through the skin, subcutaneous tissue and galea, down to the periosteum and the frontal bone was exposed. The bony lesion was clearly distinguishable from the adjacent normal bone, by its pinkish-white colour and spongy and porous texture (Figure 3C, 3D). It was softer than the normal cortical bone of the calvarium, and was quite vascular somewhat resembling cancellous bone. Osteotomes were used to resect the lesional bone in layers (Figure 3E, 3F), till the dome shaped bulge in the region was eliminated. Recontouring and finishing was completed using vulcanite trimmers under copious saline irrigation (Figure 3G, 3H). Haemostasis was achieved and closure was completed after the placement of a vacuum assisted closed suction drain (Figure 3I).

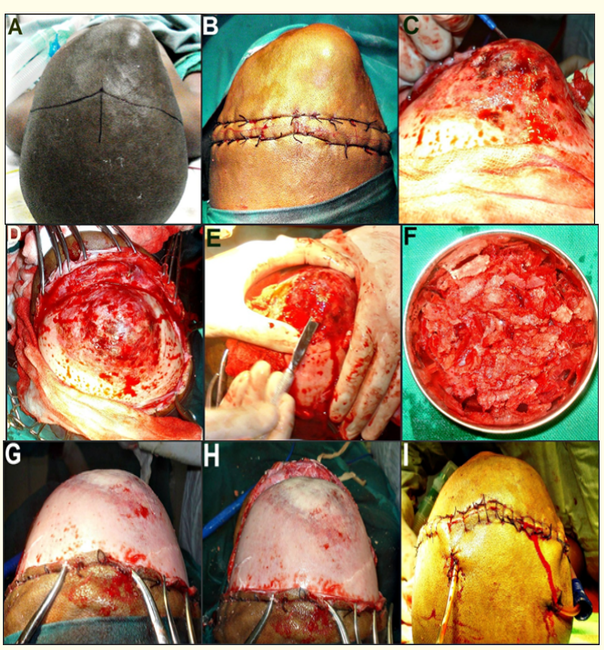


**Figure 3:** (A, B) Bicoronal incision line marked and haemostatic sutures placed on either side of this line. (C, D) Bony lesion clearly distinguishable from the adjacent normal bone, by its pinkish colour, spongy and porous texture indicative of high vascularity. (E, F) Osteotomes used to resect the relatively soft lesional bone in layers. (G, H) Recontouring and finishing completed with elimination of the dome-shaped bulge. (I) Closure completed after placement of a vacuum assisted closed suction drain.

Post-operative recovery of the patient was smooth and uneventful. The patient was placed on a protocol of antibiotics, pain relief and other supportive medications and was discharged on the seventh post – op day.

Histopathological examination of the excised specimen showed fragments of mature bone, with the bone marrow spaces replaced by typical relatively uniform-appearing meningothelial cells forming whorls and lobules (Figure 4A-4D). The features were consistent with an intraosseous meningioma of the meningothelial subtype (WHO Grade I). A high vascular density was observed, with numerous vascular channels and tumour cell whorls interspersed between and replacing the bony trabeculae (Figure 4C, 4D). Typical meningothelial and syncytial cells surrounded the numerous vascular channels (Figure 4E, 4F).

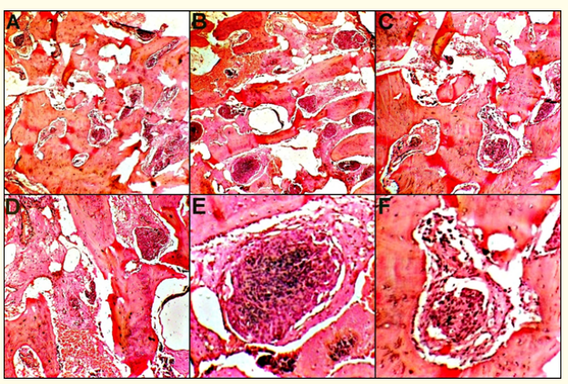


**Figure 4:** (A, B) Intraosseous meningioma of the meningothelial subtype, exhibiting high vascular density (HandE, x40) (C, D) Numerous vascular channels seen adjacent to trabeculae of mature lamellar bone (HandE, x60). (E, F) Mature bone with the marrow spaces replaced by typical uniform appearing menigothelial cells with a synctial growth pattern forming lobules and whorls (HandE, x80).

The child was followed up every two months, and there was no evidence of recurrence at the end of one and a half years (Figure 5), and is still under regular radiographic and neurological follow up.

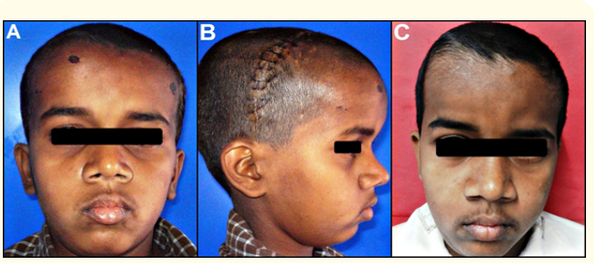


**Figure 5:** (A, B) Post-operative appearance of the patient after 10 days. (C) Post-operative appearance after one and a half years, with no evidence of recurrence.

## Discussion

The average reported age of patients presenting with Primary intraosseous meningioma is 50.5 years, with a slight female predominance of 1.65:1 [8]. Our case was rare, being found in a young 11 year old male child.

Various theories have been proposed to explain the origin of the "Primary intraosseous/calvarial meningiomas". Misplacement and entrapment of ectopic meningothelial cells/meningocytes or

arachnoid cap cells into cranial sutures during moulding of the head at birth, or during sutural closure after birth is one hypothesis. Another suggests that dura or arachnoid becomes encased by calvarial bone following a cranial fracture secondary to trauma, leading to intraosseous meningioma formation. Yet another theory suggests that as meninges are mesenchymal in origin, the Primary extradural meningiomas at other locations such as skin, paranasal sinuses, orbits and salivary glands may develop from pluripotent mesenchymal cell precursors, either pinched off during closure of the neural tube or as an inappropriate response to an unidentified stimulus [9].

The term "Primary extradural meningioma" highlights the origin of these tumours as being separate from the dural coverings of brain or spinal cord, differentiating them from "Primary intradural meningiomas" with or without extracranial extension. Although some authors have emphasized that this group of primary extradural meningiomas should have no connection to the dura mater or any other intracranial structure [10]. Other reports have included tumours with intracranial growth to the group [9].

The typical radiological appearance of Primary Intraosseous Meningioma is hyperostotic (60%), though 20 - 35% may appear lytic and these carry a worse prognosis and a malignant potential [2,3,9]. A mixed hyperostotic and osteolytic picture has been reported in around 6% of cases. If there is a lesional "ground-glass" appearance, the diagnosis may point to Fibrous dysplasia or Osteoma, as was seen in the case presented. On CT imaging, PIMs appear as hyperdense regions of cranial expansion. There may be lesional enhancement on contrast administration, as was in our case. The typical PIM MRI appearance is that of a T1 hypodense lesion, hyperintense on T2-weighted images. There is usually homogeneous enhancement after intravenous contrast administration. Although homogeneous enhancement is typical, often the intraosseous component may not enhance, only the adjacent dura [11] this was so in our case too.

The review of Lang., *et al.* [2] of primary extradural meningiomas states that calvarial meningiomas that erode the inner table of the skull with dural reaction or invasion, cannot be excluded from being classified as "primary extradural" lesions.

So, our case which showed a distinct dural reaction by the calvarial/intraosseous lesion is rightly classified as a primary extradural meningioma, with dural reaction, nevertheless with no intracranial extension. Radiographs, CT scans and MRI findings in our patient support this designation. The possible origin in this patient may have been the entrapment of meningothelial cells in a prior healed skull fracture or embryonic rests of the arachnoid cap cells within the coronal suture of the developing calvaria.

Due to the paucity of documented cases and published research on Primary Extradural meningiomas, definitive management guidelines do not exist [12]. The generally recommended management is marginal surgical excision (with curative intent) [11]. In symptomatic cases, such as those with a history of recurrent seizures, total tumour removal with a wide surgical resection followed by cranial reconstruction by cranioplasty is the preferred choice of treatment [9,11]. In situations where complete excision of PIMs is not possible and only subtotal resection has been carried out, because of the lesion being very large, as was in our case, or in complex skull base tumours, PEM debulking to decompress neural structures and surgical recontouring for aesthetic outcomes is advised [11]. Our patient presented with just signs and symptoms of a painless, slowly expanding and enlarging bony mass, producing a gross visible contour deformity. There were no neurological symptoms associated with an intracranial/dural component, such as headache, seizures, cranial nerve deficits, anosmia, changes in mentation, urinary incontinence etc or focal neurological signs, such as limb weakness, dysphagia, somnolence, apathy or disinhibited behaviour etc. Hence he was taken up for a subtotal excision of the bony mass with surgical recontouring of the frontal bone deformity.

The alternative option involving total excision of the involved bone and cranioplasty at first presentation was avoided, as it carried a significant risk of bleeding, infection and seizures and was considered unnecessary in the absence of dural extension by the lesion.

It has been recommended that residual lesions be followed up radiologically for evidence of any recurrence. Recommended adjunctive treatments are similar to those applied to intracranial meningiomas; radiotherapy is generally recommended annually for five years, thereafter every 2 years up to 10 years post-surgically, as recurrence if at all usually occurs within 10 years [2,11]. Other adjuvant treatment modalities have also been recommended, for example, gamma knife surgery, chemotherapy and bisphosphonate therapy [6]. The overall recurrence rate and prognosis for intraosseous extracranial tumours is unknown due to both the small number of cases and degree of follow up.

Our patient is presently under a rigorous monthly radiological follow up, and after a year now, has shown no evidence of recurrence.

## Conclusion

A variety of lesions may be considered in the differential diagnosis of osteoblastic/hyperostotic, radio-opaque lesions of the calvaria, such as osteoma, fibrous dysplasia, osteosarcoma, Paget's

disease etc. For osteolytic types, one should consider hemangioma, chondroma, chondrosarcoma, multiple myeloma, metastatic cancer, eosinophilic granuloma, plasmacytoma etc. Signs, symptoms and radiological appearances may be of little help in distinguishing one diagnosis from the other. Given this uncertainty, it is nevertheless important to consider Primary intraosseous calvarial meningioma in the differential diagnosis of an osteoblastic/hyperostotic radio-opaque calvarial lesion, especially if it is near a suture line, such as the coronal suture or the pterion, as was in the case presented, or if associated with a prior skull injury. From the case presented, it is clear that it was possible to obtain adequate tissue sample to make a definitive diagnosis as well as simultaneously achieve an excellent cosmetic result by a subtotal excision and remodelling the contour of the involved area of the skull. This would be of help to clinicians not in only verifying their diagnosis, but also to get very valuable cues in the management of such rare conditions.

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**Volume 2 Issue1 January 2018**