Unilateral Proptosis in Young Man: A Rare Manifestation of Intraosseous Meningioma

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Abstract
Benign primary intraosseous meningioma presented as unilateral proptosis is a rare case in our settings. The reported occurrence is 1%-2% of all meningiomas. This type of tumour has numerous nomenclature that represents its location outside the dural component, such as ectopic, extracranial, calvarial and extraneuraxial. Although it is a benign entity, the tumour can cause significant disfigurement, especially in the context of young individuals. We report a rare case of intraosseous meningioma of frontal and greater wing of sphenoid bone presented as proptosis in a young patient.

Keywords: Progressive Proptosis; Young Patient; Intraosseous Meningioma

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Introduction
Primary intraosseous meningiomas are relatively uncommon skull bone lesions that contribute to less than 2% of all meningiomas [1]. It is a term used to describe a subset of extradural meningiomas that arise in bones in approximately two thirds of all extradural meningiomas [1]. The large number of intraosseous meningiomas arises in the frontoparietal and orbital bones [3]. The involvement of two bones simultaneously with unusual radiography findings makes the diagnosis of this condition rather challenging. In this case, CT and MRI inevitably compliment the clinical findings in making a diagnosis. We report a rare case of intraosseous meningioma of the skull base, involving the greater wing of sphenoid and the inferior part of the frontal bone in a young patient.

Case Report
A 19-year-old Malay male was presented to our department with a progressive, painless proptosis of the left eye for a 5 year duration. The proptosis started at the age of 14, and had gradually increased until it caught his mother’s concern. Questioning during the first consultation revealed association with eye redness, but vision was good. No symptoms of hyperthyroid were reported. There was no significant history of trauma, history to suggest infectious causes such as that of fever, contact with patient with known Tuberculosis, and no history of loss of appetite or weight loss.

On examination, vision in the left eye was 6/18, which improved to 6/9, while the vision of right eye is was normal. There were 6 mm non-axial proptosis in left eye with diplopia on downward gaze, and restriction to up gaze (Figure 1). The conjunctiva is mildly injected at the temporal quadrant with a slight chemosis at the superotemporal quadrant. The optic nerve functions were normal and optic discs appearance were normal. Examination of right eye was unremarkable.
Figure 1: Extraocular motility. Nine cardinal positions of gaze. The patient complained of diplopia on downward gaze only and shows a restriction to up gaze in the top middle photograph. He also had overreaction of left eye in down gaze position in the bottom middle photograph.

Laboratory tests revealed normal results for blood count, erythrocyte sedimentation rates, thyroid function, venereal disease research laboratory, antinuclear antibody, Mantoux, and renal and liver functions. A skull radiograph showed erosion of the greater wing of the sphenoid bone (Figure 2). A Computed Tomography study revealed an expansile mixed lytic and sclerotic lesion with a well corticated margin of his left sphenoid and inferior part of the frontal bone (Figure 3, 4). Magnetic resonance imaging demonstrated a well-delineated lesion with post-contrast enhancement of the lesion (Figure 5, 6).

Figure 2: Plain skull radiograph shows erosion of left greater wing of sphenoid bone.

Figure 3: CT scan showing expansile lytic and sclerotic bone lesion at the lower part of left greater wing of sphenoid.
Figure 4: CT scan showing expansile lytic and sclerotic bone mass at the inferior part of left frontal bone.

Figure 5: MRI post Gadolinium T1 showing hyperintense bone mass at the left frontal bone.

Figure 6: MRI post Gadolinium T1 showing hyperintense bone mass at the left frontal bone.

The provisional diagnosis is intraosseous meningioma; differential diagnosis of fibrous dysplasia and diploeic metastasis were considered. The patient was co-managed with the maxillofacial and neurosurgical team. This patient was managed conservatively as he was not keen for any surgical intervention and under frequent monitoring, for the progression of the condition. During a 3 year follow-up, there was no progression of the proptosis. Vision in both eyes remained good, and optic nerve functions were normal.

Discussion

Primary intraosseous meningioma, or, a simpler term, ectopic meningioma, is a rare disease that contributes to less than 2% of all meningioma [1]. Several nomenclatures describe this unusual disease entity, including extradural, extracranial, or extraneural or secondary meningioma [2].

Lang and colleagues [3] classified this tumour into three types to reduce confusion due to variation of the term used in the literature. Type I is a purely extracalvarial tumor, type II is a purely calvarial tumor and type III is a calvarial tumour with extracalvarial extension. Each category is further divided into convexity (C) or skull base (B) subtypes based on their anatomical location [3]. Based on this classification, this patient fit into skull base subtypes, but not into any other type described. Agrawal et al. [4] found that the tumour was difficult to classify, especially since it depends on the radiographic appearance and location involved. Skull base intraosseous meningioma is a slow growing tumour, and the location causes different presentations compared to its convexity counterparts [5]. In this case, the involvement of 2 bones simultaneously is unusual, and we do not know whether the tumour spread from the greater wing of the sphenoid bone, or vice versa. Simultaneous involvement of 2 bones is also possible in this case, in which to date, there is no reported case of simultaneous involvement of the frontal bone and the greater wing of sphenoid.
Different theories exist regarding the origin of intraosseous meningiomas. One hypothesis is that this tumor arises as a result of aberrant differentiation of multipotent mesenchymal stem cells. Misplacement of meningocytes or arachnoid cap cells after differentiation may explain the origin of some intraosseous meningioma [6]. Alternatively, intraosseous meningioma may originate from arachnoid cap cells trapped in the cranial sutures, possibly during the molding of the cranium at birth [7]. Other theories postulated that arachnoid cap cells becoming trapped within skull fractures or sutures are a result of trauma [8]. Thus, no single theory could explain the exact origin of the tumour.

Intraosseous meningiomas affects men and women equally, with a slight female preponderance [3]. These tumours predominantly affect patients at a later age, with a median patient age at diagnosis in the fifth decade [3]. Presentation of this type of tumour in a young age such as in our case is an uncommon occurrence. The most common locations for intraosseous menangiomas are the frontal, parietal and orbital regions [3]. Other unusual sites reported in the literature are the paranasal sinuses, the nasal cavities, oral cavities, parapharyngeal space, and along the perineural sheath of the cranial nerves [3]. Very rare occurrences in the finger have also been reported [3].

Conventional radiographs are usually of limited value in making the diagnosis of intraosseous meningioma, since it is a non-specific, and the superimposed bony structure may mask the findings. The majority of intraosseous meningioma is of the osteoblastic subtype. In this subtype, skull radiograph shows hyperostosis, uneven foci of calcification and atypical vascular marking in 30-60% of cases [9]. In our case, the plain radiograph exhibits erosion of the greater wing of the sphenoid, which represents rare osteolytic subtypes. The radiographic appearance, however, depends largely on the site of the tumour. Tokgoz et al. [10] reported a well-defined solitary lytic lesion of the tumour located at the calvarial bone. Unusual findings necessitates further imaging of this kind of tumour. In other reports of sphenoid bone involvement, they rely solely on CT scan and MRI findings.

The most common CT scan findings in intraosseous meningioma are hyperostosis, which is described as a thickened and sclerotic bone [11]. The lesion appeared hyperdense on a nonenhanced CT scan, and enhances homogenously post contrast. Other less common findings are osteolytic and mixed pictures of osteolytic and sclerotic bone lesions [4]. The purely osteolytic mass usually involves the scalp, known as the calvarial meningioma [4]. Mixed type is the rarest type, as seen in our case, and has been reported in 6% of cases [11]. Close to these findings is fibrous dysplasia, but both entities can be distinguished based on the patient's age. Intraosseous meningiomas typically appears at a later age and grow gradually. In contrast to fibrous dysplasia, the tumour usually appears at younger age and stops growing after puberty [11]. From radiograph findings, fibrous dysplasia does not enhance post contrast [11].

The distinctive MRI findings of the intraosseous meningioma demonstrate homogeneously dense contrast enhancement of the tumor [11]. This finding is important, as it helps to distinguish this lesion from meningioma en plaque, and other forms of osteoblastic skull lesions, such as fibrous dysplasia, osteoma and Paget disease [11]. We also rely on clinical findings, as this patient systemically does not show any bony lesion or bony changes elsewhere, as can be found in Paget disease. After 3 years follow-ups, he did not show any rapid progression or deterioration of the disease with the benign entity of the tumour.

The treatment of choice is a wide surgical excision of an intraosseous meningioma, with cranial reconstruction [11]. The surgery is indicated in this type of tumour if it shows signs of compression on the optic nerve, debilitating proptosis and significant disfigurement. In this case, the patient should have ideally undergone surgical excision due to the presence of 6-mm proptosis, causing significant disfigurement. However, the patient was not keen for any surgical intervention. Furthermore, upon follow-up of the patient, he did not show any imminent signs of compression on the optic nerve, and the proptosis was not worsening. Depending of the site of the lesion, the total resection of the tumour may not be possible in skull base lesions. Preoperative assessment with a three-dimensional CT may allow fitting of a custom-made implantation at the site of defect during the removal of the tumour [12]. Apart from the surgical resection, monitoring of the progression, and recurrence, are equally important. Although most of the tumour is benign, malignant changes were reported in a small number of cases.

In conclusion, benign primary intraosseous meningioma presented as unilateral proptosis in a young patient is a rare presentation. CT and MRI findings are both inevitable in making a diagnosis in this type of tumour. No definite consensus has been discussed on the optimum treatment but surgical excision with bone reconstruction is the mainstay of treatment.
References


