

Case Report

Giant fronto-ethmoidal osteoma with orbital and intracranial extension: a case report

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ABSTRACT

Osteomas of the paranasal sinuses are often small, asymptomatic and incidentally diagnosed on radiological imaging. Giant osteomas, although rare, are clinically significant due to their propensity to cause orbital and intracranial complications. Complete surgical resection is usually possible with good outcome and low recurrence rate. We report a case of 62 years female who presented with proptosis and blurring of vision. Radiological work up revealed giant fronto-ethmoidal osteoma measuring 7.9×5.1×4.8 cm extending into the orbit and along the anterior skull base. Open surgical excision of the osteoma was done via left fronto-orbital approach. This is one of the largest fronto-ethmoidal osteomas reported in the literature till date.

Keywords: Giant, Fronto-ethmoidal, Osteoma, Intracranial extension

INTRODUCTION

Osteomas are benign osteogenic tumors formed as a result of proliferation of cancellous or cortical bone. Paranasal sinuses, especially the frontal and ethmoidal sinuses, are the most commonly affected sites in the craniofacial region. The average size of an osteoma ranges from 0.2 to 3 cm. Those with a size >3 cms or weighing >110 gm are termed as 'giant' osteomas. Such giant osteomas are relatively rare, but clinically significant as they may lead to morbid complications secondary to intra-orbital and intracranial extension.¹

CASE REPORT

A 62 years female presented with painless, progressive proptosis of the left eye since 1 year. She also developed double vision associated with limitation of the left eye movements since last 4 months. Examination revealed visual acuity of 6/60 with relative afferent pupillary defect in left eye and normal ishihara colour plate testing. Protusion of left eye along with outward and downward

deviation of left eye was evident. (Figure 1A) Hertel exophthalmometry revealed 6 mm proptosis on left side. Anterior segment and fundus examination were normal. Systemic clinical examination did not suggest any inflammatory or infectious process.

A computed tomography (CT) scan of the orbit revealed a well-defined, large, multilobulated, hyperdense mass measuring 7.9×5.1×4.8 cms, occupying the left supraorbital region, left frontal and ethmoid sinuses. (Figure 1B, D, E) Magnetic resonance imaging (MRI) of the orbit showed an exophytic mass arising from the outer wall of the sinuses involving the extraconal compartment of the left orbit and extending posteriorly in the subfrontal region compressing the brain, terminating in the left parasellar region (Figure 1C). The radiological features were suggestive of osteoma. This mass was excised through a left fronto orbital approach. A supraorbital frontal craniotomy was done and osteoma was exposed at fronto-ethmoidal sinuses and supraorbital region. Supraorbital rim was involved by the mass. The mass was removed en bloc by a single stage surgery.

Defect in the dura was repaired and frontal sinus was exteriorized. Pericranial flap is used to cover the floor of the anterior cranial fossa. Reconstruction of the supraorbital rim and frontal bone was done with titanium mesh (Figure 2A, 2C). The patient had an uneventful postoperative recovery. Histopathologic report confirmed a diagnosis of osteoma with osteoblastic and osteoclastic activity.

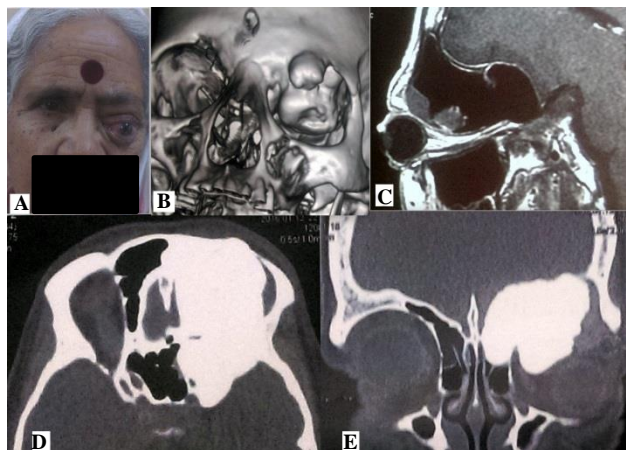


Figure 1: (A) Patient photo demonstrating the degree of proptosis with downward and outward displacement of the left eyeball; (B) 3D CT showing involvement of supraorbital rim and intraorbital extension of the osteoma; (C) MRI brain showing extension of the osteoma along the anterior skull base with compression of the frontal lobe; (D, E) CT orbit (axial and coronal) showing well defined, multilobulated, giant osseous lesion arising from frontoethmoidal sinuses and extending into the orbit, pushing the eyeball inferolaterally.

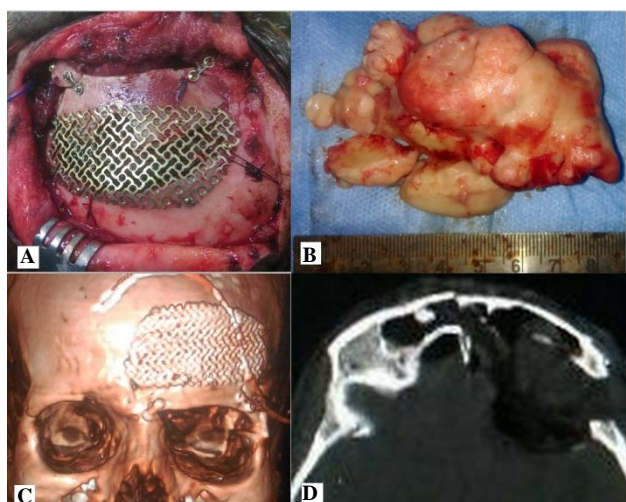


Figure 2: Reconstruction of supraorbital rim and frontal bone with titanium mesh (A) Intra op photo; (B) Clinical photograph of the excised giant osteoma; (C) Post op 3D CT; (D) Postoperative CT showing complete excision of Giant osteoma.

DISCUSSION

Osteomas are infrequent tumors representing only about 1% of all bone tumors. They are benign, slow growing and most commonly affect the fronto-ethmoidal sinuses (95% of cases). Osteomas of the sphenoid and maxillary sinus have been reported as well. The prevalence in the general population is only about 0.43-1% with males being more commonly affected than females.² Although the exact etiology remains elusive, local trauma and infection (sinusitis) are thought to play an important role in stimulating osteoblast proliferation and tumor formation. An embryological origin is also postulated, given the fact that osteomas more commonly arise at the junction of frontal and ethmoidal sinuses, a place where membranous and cartilaginous tissues meet during the developmental stage. Multiple osteomas also occur as a part of Gardner's syndrome, an autosomal dominant hereditary disorder characterized by intestinal polyposis, osteomas and soft tissue tumors. Hence, in all patients presenting with osteomas, a colonoscopy is a mandatory investigation to look for presence of intestinal polyps.³

Majority of these tumors are small in size and remain asymptomatic, being incidentally diagnosed on X-rays and CT scans. The giant ones, however, can produce a variety of symptoms depending on their size, location and direction of tumor growth. A large frontal sinus osteoma may cause facial deformities and may lead to obstruction of sinus drainage causing headache (most common), sinusitis, frontal mucocoele and the 'vacuum sinus' syndrome. Given their proximity to the orbit, giant ethmoidal osteomas commonly lead to orbital invasion causing various ocular symptoms such as proptosis, diplopia, hypopsia, epiphora, ptosis and rarely, amaurosis and orbital emphysema. Intracranial extension is more common with giant frontal osteomas, resulting in meningitis, cerebral abscesses, intracranial mucocoele, pneumocephalus and cerebral edema.¹⁻⁴

A fronto-ethmoidal osteoma can be accurately diagnosed on a CT scan, where it typically appears as a thick, sclerotic, homogenous and well defined mass.¹ A contrast enhanced CT brain with orbits and paranasal sinuses helps in determining the exact tumor size, location and extension to adjacent structures. MRI is useful in cases with ocular/intracranial symptoms to further delineate the extent of soft tissue involvement.⁴ Histologically, osteomas consist of mature bone similar to a normal bone, but with a decreased bone marrow. They are classified into three types- the ivory (dense, mature lamellar bone and little fibrous stroma), mature (mature lamellar bone tissue with abundant fibrous stroma) and mixed variety.³ The ivory type tends to occur over the outer table of the skull while other types tend to involve the inner table or rarely, the diploic space. Important differential diagnoses of the fronto-ethmoidal osteoma include osteosarcoma, osteoblastoma and fibrous dysplasia. The typical radiological and pathological

features of these tumors should be looked for to help differentiate them from a benign osteoma.³

A conservative treatment approach, by observation and periodic follow up with imaging, is recommended in most of the asymptomatic cases, particularly in elderly patients. Surgery is indicated in patients who are symptomatic, show significant growth of osteomas on follow up scans or those with giant tumors extending beyond the sinus borders. Orbital and sphenoid osteomas, however small they may be, should be considered for early surgical intervention due to their potential to cause compression of the visual pathway.^{2,3} The optimal treatment consists of en bloc resection or curettage of the tumor either by endoscopic or open surgical procedure. The selection of surgical approach depends upon the tumor size, tumor location, extension of the tumor and surgeon's preference. Complete resection of giant fronto-ethmoidal osteomas via endoscopy alone is difficult and the classical approach is still considered to be an external one viz. fronto-orbital approach, transfacial approach with lateral rhinotomy and bicoronal bifrontal osteoplastic technique. In a review by Cheng et al, only 3 out of 45 cases of giant fronto-ethmoidal osteomas could be successfully treated by an endoscopic approach alone. Seven cases required a combined endoscopic and open approach, while the rest 35 cases were successfully treated via an open approach alone.³ Reconstructive procedures, which are often required after resection of giant osteomas, are difficult to achieve with endoscopy alone. Hence, we resorted to an open approach in our case, given the size and extensive nature of the tumor as well as the need to reconstruct the superior orbital rim. Complete surgical excision can be curative. Recurrence rate up to 10% are reported in the literature with incomplete excision.

CONCLUSION

Giant fronto-ethmoidal osteomas with orbital and intracranial extension are very rare lesions. Presenting with a plethora of symptoms, CT scan is accurate in establishing the diagnosis. Being benign in nature, en bloc surgical resection of these tumors is usually curative. Although endoscopic approach with/without open resection is preferred for small to medium sized tumors, an open surgical procedure remains the gold standard treatment for giant fronto-ethmoidal osteomas.

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