Giant Frontoethmoidal osteoma with orbital involvement- a rare case report with review of literature

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Giant Frontoethmoidal osteoma with orbital involvement—a rare case report with review of literature

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Abstract: Osteomas are benign tumours that involve paranasal sinuses more than other parts of body. Frontal sinus is most common while sphenoid is least one. They are usually asymptomatic and diagnosed incidentally on imaging for other reasons. Giant variety of frontoethmoid osteoma is very rare and only few cases are described in literature. Orbital involvement in giant frontoethmoid osteoma is even rarer. Due to threat to vision, this variety is an indication for surgical intervention followed by reconstruction of defect. As these tumours are very large, endoscopic approaches are less indicated and there is no clear consensus which open surgical approach is better. We are reporting such a rare case of giant frontoethmoid osteoma causing proptosis and restriction of eye movement that was treated with open surgical approach.

Key words: giant, osteoma, frontoethmoid

Introduction

Osteomas are rare benign bony mesenchymal tumours. They commonly involve craniofacial bony skeleton and jaw bones (12). They constitute about 1% of all bone tumours & 11% of all benign bone tumours (2). They are slow- growing, often asymptomatic & incidentally detected in 1% of skull X-Ray and 3% of the cranial Computed tomography (CT) Scans. These tumours involves frontal, ethmoid, maxillary & sphenoid sinus in decreasing order of frequency. Giant frontoethmoidal osteomas are very rare and patient usually present early as there is limited anatomical space (14,15,16). orbital and skull base involvement is very unusual in ethmoidal osteoma and can produce neurologic and ophthalmic manifestation hence surgical intervention is required in these patients(14,16). CT scan remains the gold standard to diagnose this pathology and surgery remains the treatment of choice for symptomatic frontoethmoidal osteomas. We are reporting such a rare case of frontoethmoidal osteoma that was treated surgically with special focus on various surgical options.
Case report

A 14 year male child referred from Otolaryngology department with chief complains of insidious onset, slowly progressive nasal blockage with difficulty in nasal breathing since last 4 years. Patients often had off and on mild headache for that he takes oral analgesic and his headache was relived after taking these medications. Gradually his mother notice broadening of his nasal bridge with deviation and bulging of his left eyes. CT scan of PNS showed a large frontothmoidal osteoma. Patient further evaluated in neurosurgery department. He was conscious and oriented. His vitals were within normal range. On neurologic examination, his higher mental status was normal. There were no cranial nerve deficits nor any deficit in sensory or motor system. His cerebellar examination was normal. Position of his right eye was normal but his left eyes was slightly bulging and deviated to left & there was slight restriction of left eye movement on superiomedial direction. There was no relationship of proptosis with bending forward. Visual acuity & fundus examination of both eyes was normal. On palpation, swelling was bony hard in consistency. A CT scan of the brain with PNS (Figure 1) revealed a hyperdense giant mass of approximately 7.2 cm of size occupying the left supraorbital region, frontothmoid region.

Surgical intervention was advised and possible complications and outcomes were explained to them in detail. After taking written and informed consent patient taken to operation theatre. Patient put on supine position with a bolster placed below the shoulder so that neck remains in extension. Bicoronal skin incision given and skin flap raised with pericranium lifted separately. Supraorbital bifrontal craniotomy done. As soon as dura separated from anterior cranial base, ivory white osteoma visualized. Frontothmoidal part of osteoma taken out with orbital part removed separately. Reconstruction of the anterior cranial base performed using pericranial patch. This was further reinforced by tissue glue. In post-
operative patient did well. There was no CSF leak from the nose and his proptosis was resolved with no diminution in vision. Post-operative CT scan (Figure 2) showed complete removal of osteoma. Histopathological examination confirmed the diagnosis of benign osteoma.

Discussion

Osteomas are rare benign bony tumours. They involve paranasal sinuses (13) most common, & found in the frontal sinus (71.8%) and less often in the ethmoid sinus (16.9%), maxillary sinus (6.3%) or sphenoid sinus (4.9%) (11). Osteomas of the other bony regions are also reported.

Osteomas affect less than 1% of population with slight male preponderance (17). 95% of the patients having osteoma remain asymptomatic (11). The average reported size of osteoma involving paranasal sinuses is less than 2 cm (12) but when the size exceeds more than 3cm they are termed as giant (4).

Most of osteomas are sporadic but few cases may have genetic predisposition like gardener syndrome (3). The pathogenesis of osteoma formation is not very clear and there are different theories (10,11,15). Traumatic theory of Gerber states that injuries suffered during puberty may cause the growth of osteoma from bone sequestra. Inflammatory theory which states that chronic sinusitis of paranasal sinuses can stimulate osteoblastic cell hence there is formation of osteoma. Most recent theory is embryologic theory which states that Osteoma arises from the remains of persistent embryologic cells located at the junction of the ethmoid and frontal sinuses. Traumatic and embryologic theories are most accepted one.

Symptomatology of osteomas depends upon the location. Overall most common symptoms is headache (10). Sometimes osteoma may involve anterior cranial base and patients may have cerebrospinal fluid leak, meningitis, pneumatocele (5) and brain abscess (8). As the orbit is very close to ethmoid sinus, giant osteoma of frontoethmoidal region may involve the orbit and can produce proptosis, diplopia and other ocular symptoms (16). Very rare complications like amaurosis (18) and orbital emphysema is also reported in frontoethmoidal osteoma.

There are three different types of osteoma viz. Ivory, mature and mixed (12). This is very crude classification. Each osteoma has all these kind of cells but proportion of each cell varies. More mature cells migrate from the centre and align themselves to periphery and this is the reason that there is no recurrence after leaving some peripheral part of the osteoma (17).

CT scan of the paranasal sinuses is the investigating modality of choice. It can exactly delineate which part of the craniofacial skeletal is involved but it lacks the sensitivity and specificity of MRI for soft tissue detail like optic nerve involvement. Radionuclide scan can be performed that shows more radionuclide uptake in active growing lesions.

Osteoblastoma, osteoid osteoma (19), fibrous dysplasia, ossifying fibroma and chondroma (9) are major differential diagnosis of osteoma. There is no need for active management for osteoma except sphenoid osteoma (11) that threaten orbital...
apex or optic canal and a giant frontoethmoidal osteoma with involvement of orbit.

There is no clear consensus among surgeons that which surgical treatment is better for frontoethmoidal osteoma. Open and endoscopic surgical approaches can be used to treat these pathologies. The choice of surgical approach is based upon tumor location, dimension, extension and experience of surgeon. There is rapid expansion in endoscopic technology and now a day’s endoscopic approaches are more commonly performed in small and medium size osteomas (13). Less morbidity, less blood loss, no operative scar, close and direct visualization of tumor and less hospital stay are usual benefits of endoscopic approach (13). Long learning curve, huge instrument cost, difficult intraoperative control of bleeding, inability of bony reconstruction and non-familiarity with endoscopic anatomy of paranasal sinuses are usual drawbacks of endoscopic approach. Open surgical approaches remains treatment of choice for giant frontoethmoidal osteoma. The osteoplastic flap technique, anterior surgical exposure (craniofacial, transcoronal, and transcutaneous paranasal approaches), external fronto-ethmoidectomy, and lateral rhinotomy have all been described in the literature as possible techniques in the resection of giant osteomas that extends beyond the ethmoid sinus (6,7,14,21). Recurrent sinusitis, cranial nerve injury, ptosis, ophthalmic complication, meningitis, postoperative CSF leak and bleeding are the most common complications after open surgical procedures (1,20). In our case it was huge frontoethmoidal osteoma with involvement of orbit & requires post-operative reconstruction that renders endoscopic approach less favorable.

The goal of surgery is complete excision of lesion. During surgery of frontoethmoidal osteoma with orbital involvement, nearby vital structures may get damaged like cribiform plate, optic nerve, eye globe, anterior and posterior ethmoidal arteries. Good aesthetic outcome is also important as far as possible (16). Tumours should be removed enbloc as far as possible and if surgeon is not able to do so than central hard part of osteoma should be drilled out and peripheral thinned rim of tissue should be taken out separately (4). Same technique should apply to orbital osteomas. In our case we first approach anterior skull base intracranially- extradurally & removed frontoethmoidal part of osteoma enbloc. Orbital part removed separately. Postoperative defect after removal of giant frontoethmoidal osteoma should repair as far as possible. Small defect can be repair using pericranial patch or galeal-pericranial patch. Large defect should be repair using autologous calvarial bone graft, as this graft provides natural cranial contour. Synthetic implants can also be used for correction of postoperative defect. We offered reconstruction of bony defect but due to financial constrains, patient and his parents refused for the same. Post-operative recurrence of osteoma is very unusual but few case reports are described of rapid recurrence (2). CSF leaks, meningitis, post-operative bleeding, and injury to vital structures like optic nerve and lacrimal apparatus injury are the common postoperative complications.
Conclusion

Giant frontoethmoidal osteomas with orbital invasion are very rare lesions. Most of the existing literature on this pathology is in the form of case reports or small case series hence no definitive approach is superior. In today's era both surgical and endoscopic approaches can be used. In case of giant osteomas due to invasion of nearby structures like orbit, endoscopic approaches are less useful. Our case is very unique and adds a rich knowledge to the existing literature and reaffirms the open surgical procedure as preferred modality of treatment in case of giant frontoethmoidal osteoma.

References