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Complete endoscopic resection of an ossifying fibroma involving the skull base & orbit with superolateral extension upto the lateral orbital wall.

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Abstract

A young girl presented to us with gradual progressive loss of vision, headache & unilateral proptosis. A CT scan revealed a radio dense mass occupying the right orbital apex & expanding laterally into the roof of the orbit & supraorbital region. The lesion was completely resected endoscopically.

Key words

Ossifying-fibroma, endoscopic, orbit, skull base, ethmoid

Introduction

Ossifying fibroma is a benign encapsulated fibro-osseustumor made up of fibrous tissue & mature bone wherein normal bone is replaced with fibrocellularstromawith focal mineralization of the bony trabeculae. Thepathological classification of ossifying fibromas is based on the pattern of mineralization into 3 types namely – juvenile psammomatous type, juvenile trabecular type & cement-ossifying type. Although the juvenile types have a lot of similarities they can be distinguished from each other based on many features, the most striking of them being their location.The psammomatous type is more common in the paranasal sinuses & orbit whereas the latter being more common in the maxilla. The cement-ossifying type is more common in the mandible. Diagnosis usually cannot be made solely based on clinical or radiological findings but is a combination of these 2 with pathological findings as well.

Case report

A 29 year old woman presented to us with history of headaches for about a year; of the dull aching type & was of moderate in severity, more during the early morning & did not subside with sleep. Patient also presented with protrusion of the right eye with complete loss of vision in the same eye & restricted ocular mobility in the convergent & upward gaze.

On examination, we found a pink smooth capsulated mass in the region of the Right ethmoids&spheno-ethmoidal recess extending upto the anterior end of the middle turbinate. Coronal CT scans revealed a tumor at the orbital apex with well-defined, expansile mixed soft tissue & bone density lesion. The lesion had the classical finding of thick bony walls surrounding a fibrous centre. Hence, a presumptive diagnosis of a craniofacial fibro-osseus lesion was made. Patient was also subjected to an enhanced T1 fat saturated MRI which helped us identify all the surrounding soft tissues at risk during surgery. The lesion was centred on the orbital apex area pushing the orbital wall laterally & inferiorly, extending above the roof of the orbit &intracranially but not penetrating the dura. The lateral extent was upto the orbital component of the greater wing of sphenoid & the lesion was crossing the mid orbital plane beyond the optic nerve.

It was decided that since the patient did not have vision for a very long time, the tumor could be delivered with the endonasal endoscopic approach since no further damage could be

done. This would also prevent an external scar in a young female. Under general anesthesia, the middle turbinate was partially resected on the right side for the purpose of access. A Haddad flap was elevated on the left side so as to cover the possible skull base defect. Posterior septectomy was then performed to get good access & bimanual manoeuvrability. Mucosa around the tumor was then removed with a 40 Rad microdebrider& the tumor was isolated from all sides. A Mucocele was encountered since the mass was obstructing mucous outflow. The tumor was found to be adherent to the periorbital, medial rectus muscle & the optic nerve & had to be removed off these structures with careful dissection. The tumor was then peeled off &separated from the middle fossa dura with a suction periosteal elevator & delivered. The tumor was yellow – white in color & gritty in some places but crumbly & cheesy in some areas.

With a 70 degree endoscope & a 60 rad microdebrider, the region above and lateral to the globe was cleared of the tumor until the lateral orbital wall was cleared.The endoscope was placed in the right nasal cavity and the 60 rads microdebrider was used from the left nasal cavity through the septectomy. This was possible due to removal of bone in the roof of the orbit allowing adequate visualization of the superolateral orbital angle ensuring complete tumor removal. Finally the tumor was completely removed from the dural attachment, the periorbital & the superolateral extension. The exposed dura was then covered with a Haddad flap for early & primary healing.

Histopathological examination revealed an encapsulated tumor with a matrix of randomly distributed mature lamellar bone & fibrous stromal mixture going in favor of ossifying fibroma. Center of the mass had immature bone whereas the peripheral area had mature bone. The patient has been followed up for a year now with CT scans every 6 months with no evidence of recurrence so far.

Discussion:

Ossifying fibromas are a rare entity within the orbit & paranasal sinuses & are generally found involving the mandible. They belong to the group of benign fibro-osseus lesions comprising Fibrous dysplasia, Cemento-ossifying fibroma & juvenile active ossifying fibroma. The paranasal sinus location is associated with a much aggressive behaviour than their mandibular counterparts [1].

Clinically, fibrous dysplasia has a self-limiting growth.with skeletal maturity. These tumors can be identified on a CT scan by their clear demarcation from the surrounding tissue.

There are many modalities of management of such a case; options being the open technique by the Fronto-temporo-orbito-zygomatic (FTOZ) approach or by the closed endonasal endoscopic approach to the orbital apex. In the earlier days, reaching the supero-lateral wall of the orbit would have been difficult but today with the help of curved microdebrider blades & angled endoscopes; it is possible to expand the horizons of endoscopic surgery. However, surgery must be performed by a surgeon well versed with the anatomy of this region & should have had previous experience with endoscopic skull base surgery. Case selection is an important part as well. This case was treated with the endoscopic approach despite the lateral extent of the tumor upto the lateral orbital wall which goes against the principles of Endoscopic skull base surgery but since the disease was long standing & the patient did not have vision for a long duration, any hope of residual vision after tumor removal would have been over-optimistic. Hence, the open approach did not have any distinct advantage over the endoscopic approach in such a case; rather it would leave the patient with a scar or deformity. However, the optic nerve was not damaged during the entire procedure of the surgery despite no hope for vision restoration.

Murchison ET al[2] have reported a 22% complication rate with the endoscopic endonasal approach to the orbital apex namely CSF leaks,

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reduced visual acuity, diplopia etc.
The most common complication of such an approach is a CSF leak as mentioned by Brodish BN et al [3]& this justifies the use of the Headed flap to cover the skull base defect created.

There have been some articles published previously such as that by Post &Kountakis[4]&Choi YC et al[5] where in large sinonasal OFs' have been removed through the endoscopic approach adding to the growing evidence that endoscopic approach is a good alternative to the open approach with the advantage of minimally invasive surgery & better outcomes in terms of cosmetics, lesser hospital stay, reduced chances of infection & reduced morbidity of associated with a craniotomy.

Conclusion

Endoscopic resection of these tumors is a definite advantage over the open approach provided the surgery is performed by a surgeon with a good hold over the skull base anatomy. Case selection also plays an important part in the success of the surgery. Cases should be chosen such that the approach leaves the patient with least morbidity lest the purpose of this approach will be defeated. Follow up of these cases at regular intervals is very important so as to detect recurrences at the earliest when the tumor is small which makes it easy to resect.

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Images

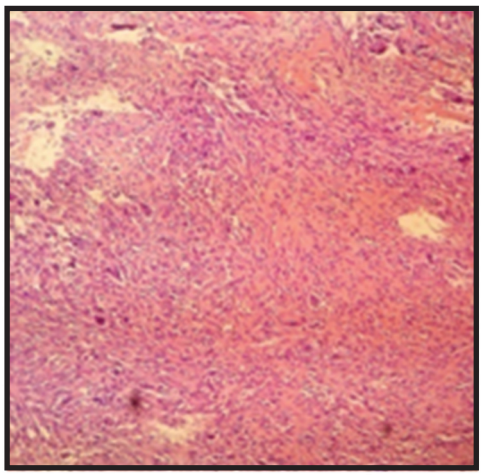
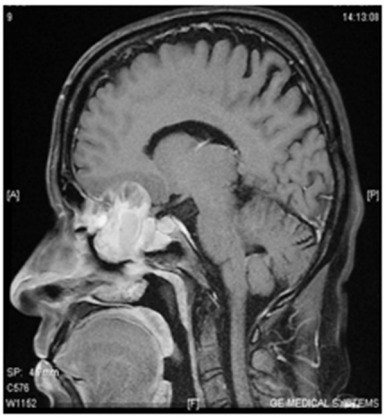


Image showing randomly distributed lamellar bone with fibrous stroma suggestive of ossifying fibroma.



A Sagittal T1W MRI showing well-defined, expansile mixed soft tissue & bone density lesion extending from the orbital apex posteriorly

almost upto the frontal sinus anteriorly. Observe that the center of the lesion is more fibrous than the periphery.

A Coronal T1W MRI showing lesion pushing the orbit inferolaterally& in close proximity to the medial rectus & superior rectus muscles. There is also intracranial extension & laterally the tumor extends upto the superolateral orbital wall.

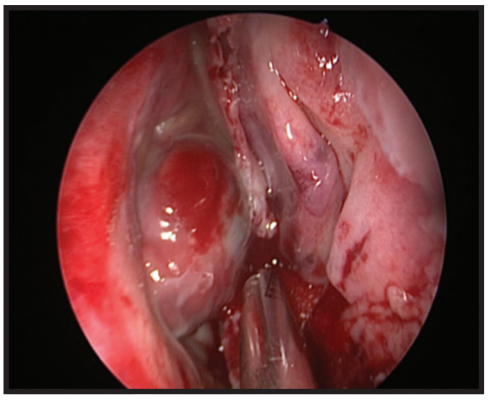
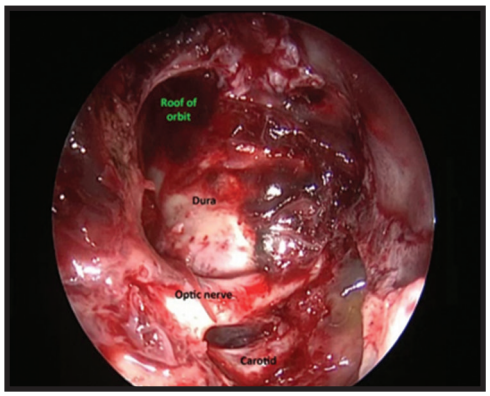


Image showing a reddish pink smooth encapsulated margin the right ethmoids&spheno-ethmoidal recessextending anteriorly upto the anterior end of the middle turbinate.



Labelled image showing the orbital apex with complete removal of the tumor from the optic nerve &dura. Superolateral clearance is seen in the tunnel above the orbit.

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