

Case report of unilateral axial proptosis due to Aneurysmal bone cyst.

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Abstract:

Aneurysmal bone cyst is rare condition presenting with proptosis. A 2 years old child presented with a gradual, progressive painless protrusion of left eyeball since 15 days & bleeding from left nasal cavity. On examination the proptosis was non pulsatile and non tender. Patient was uncooperative for visual acuity, anterior segment was normal, and a mass found in the left nasal cavity. Ultrasound of left eye showed a retro orbital mass of size 16x14 mm. MRI brain with orbit showed a cystic lesion around left superior & middle ethmoidal sinus 25x26x35mm, with post contrast thick peripheral enhancement and mild compression of intracranial optic nerve. ENT surgeon performed Functional endoscopic sinus surgery (FESS) which revealed a left side nasal cavity mass with diseased tissue from maxillary ostium, periorbital tissue, prolapsed dura and from supra orbital region left side sphenoid ostium, that was removed, and sent for histological examination. Anterior and posterior ethmoidectomy was done. The proptosis due to aneurysmal bone cyst of ethmoid bone was confirmed by histopathology.

Keywords: Aneurysmal bone cyst, Axial proptosis, Functional endoscopic sinus surgery (FESS).

Introduction:

Aneurysmal bone cyst (ABC) is a benign tumour like, expansive osteolytic lesion consisting of blood-filled spaces and channels divided by connective tissue septa that contain osteoid tissue and osteoclastic giant cells. Aneurysmal bone cyst occurs more frequently in females than in males and there is 10-15 percent recurrence even with treatment⁽¹⁾. The most common locations of aneurysmal bone cyst are long bones (50%), followed by flat bones, vertebral column (20%). Only 2% are found in the head & neck area with mandible and maxilla being the most frequent sites being involved. Involvement of ethmoidal sinus, is extremely rare.

ABC was first described by Jaffe and Lichtenstein in 1942.⁽²⁻³⁾ Their cause is not entirely clear but presumed to be due to alteration in local haemodynamics related to venous obstruction or arteriovenous fistula with bone resorption or due to haemorrhage secondary to trauma.⁽⁴⁾ Secondary origin of the lesion from a pre-existing lesion such as

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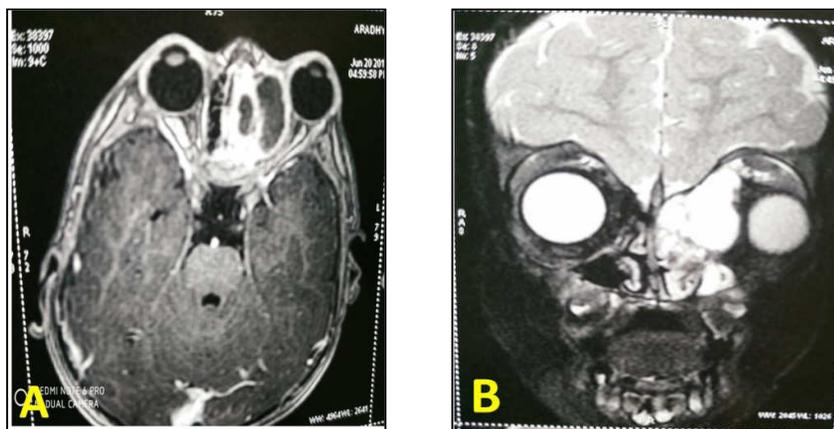
giant cell tumour, fibrous dysplasia, osteblastoma, osteosarcoma, chondroblastoma, chondromyxoid fibroma, unicameral bone cyst has been well documented. The lesions often produce symptoms due to compression of adjacent structures. ABC of ethmoid bone usually presents with epistaxis, headache, and nasal obstruction when involves the paranasal sinuses and nasal cavity. Severity of symptoms depends on the location and the rate of growth of the lesion.

Due to the rarity of ABCs in paranasal sinuses and the emerging role of functional endoscopic sinus surgery (FESS) over the last decade. We hereby report an extensive lesion of ethmoidal aneurysmal bone cyst treated with FESS.

Case Report:

Parents of a 2 years old female child residing in Sihor district of Madhya Pradesh, India presented with complains of protrusion of her left eyeball since 15 days & bleeding from her left nasal cavity since 5 days. On examination patient was uncooperative for visual acuity. Slit lamp examination showed normal anterior segment in both eyes. Ultrasound examination of left eye showed a retro orbital mass of size 16x14 mm. MRI study of brain was performed 1.5T HDXT using T1 and T2 W SE, FLAIR & GRE sequences in axial, sagittal & coronal planes. Post contrast T1 W image was acquired in all three planes. A Cystic lesion was seen in superior ethmoid sinus with dehiscence in the left lamina papyracea and extraconal extension leading to marked proptosis and infero-lateral displacement of eyeball. Lesion was measuring 25(SI) X 26(RL) X 35(AP) mm leading to marked compression of left medial rectus and optic nerve. The T2WI reveal fluid level with post gadolinium scan revealed heterogeneous thick peripheral enhancement. Features were suggestive of aneurysmal bone cyst of ethmoid bone. Mild restriction noted in DWI. The eye ball, its layer and lens appeared normal. Features were suggestive of an aneurysmal bone cyst of ethmoid bone. (Image 1 A & B).

Image 1 MRI study of Brain & orbit showing cystic lesion.



Nasal endoscopy revealed a cystic tumour-like mass obstructing the left nasal cavity, The ENT surgeon performed a Functional endoscopic sinus surgery (FESS). Adhesions of the mass to left middle turbinates were noted. Left nasal septal cartilage was partially eroded by the mass. Diseased tissue from maxillary ostium, periorbital tissue and diseased tissue from

supra orbital region of left side sphenoid ostium was identified and removed. Left sided uncinectomy with anterior ethmoidectomy was done, prolapsed dura was also excised. Histopathology of resected specimen confirmed aneurysmal bone cyst of ethmoid bone. Diagnosis of Aneurysmal bone cyst was done & proptosis relieved by FESS. (Image 2 & 3).

Image 2 Preoperative image showing the Proptosis



Image 3 Postoperative image after FESS.



ENT surgeon & neurosurgeon propose to remove the rest of the orbital mass in subsequent follow up.

Discussion

ABCs are benign cystic lesions that destroy and expand the affected bone. Radiologic appearance similar to vascular aneurysms lead to the name 'ABC'. Primary and secondary lesions can be differentiated by the fact that primary ABCs have no history of trauma or other associated tumors, while one third of the lesions occur secondary to other tumors such as giant cell tumor or chondroblastoma. Due to the uneventful medical history of our young patient the lesion can be defined as 'primary' ABC.

Symptoms are mostly described with headache, ptosis, rhinorrhoea, strabismus, exophthalmos, swelling, vision loss and nasal obstruction. Severity of symptoms depends on the location and the rate of growth of the lesion. The patient in our report suffered from proptosis and nasal bleeding.

In the review by Hrishikesh KA et al. they report that lesions affecting the skull base are more likely to present with neurological deficits including anosmia, ataxia, otalgia, facial numbness and hearing loss. However, there were no any neurological deficits in our patient.⁽⁵⁾

Fluid levels can be seen in other lesions such as telangiectatic osteosarcoma, giant cell tumour and chondroblastoma. The fluid levels are probably due settling of degraded blood products within the cysts.⁽⁶⁾ In our study the presence of fluid–fluid levels, cyst within a cyst appearance and lack of restricted diffusion within the sinonasal contents was seen

Diagnosis is often delayed due to the uncommon location of the lesion.⁽⁷⁾ Further, treatment of an ABC in the paranasal sinuses is challenging due to its proximity to crucial anatomical structures. In our cases by doing a FESS, proptosis was relieved.

Angiography is used when there is concern about the vascular nature of the lesion on cross-sectional images and to demonstrate arterio-venous shunt when present. The typical histopathological findings of ABC reveals fibrous proliferation, new bone formation, multinucleated giant cells, blood-filled cyst and spindle shaped endothelial cells which were

present in our case.⁽⁸⁾

Treatment of choice is complete surgical excision which may range from conservative surgery like endoscopic surgery, curettage and enucleation to paralateral rhinotomy and bifrontal craniotomy for ethmoid ABC. Pre-surgery Selective arterial embolization may also be beneficial. forms are medical management such as interferon alfa-2a and radiation therapies are reserved for unresectable lesions. An underlying lesion should be sought for in cases refractory to treatment.^(5,8)

In spite of surgical resection, complete clearance is sometimes impossible at the skull base. In such cases, further surgical procedures to debulk the lesion may be needed. For refractory cases in adults, limited success with radiotherapy has been reported in cranial ABCs, albeit with an accepted risk of sarcomatous degeneration.⁽⁹⁾ However, as there is a paucity of information for ethmoidal lesions particularly shell of bone, having multiple fluid levels within in a patient helped to confidently diagnose ABC. In children, there exists no clear consent for radiotherapy.

In conclusion, presence of a well-defined, expansile, multiloculated bony lesion with sharp, smooth rounded margins bordered by continuous or interrupted thin shell of bone, having multiple fluid levels within in a patient helped to confidently diagnose ABC.

The confirmatory histo-pathological diagnosis remains mandatory to exclude malignant lesions. The high recurrence rate of the lesion necessitates a regular annual follow-up after the surgical resection.

Minimal invasive FESS may be used for tumour debulking as symptomatic treatment in radically non-resectable lesions.

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