ANEURYSMAL BONE CYST OF THE FRONTAL BONE: A CASE REPORT
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INTRODUCTION: Aneurysmal bone cyst is an uncommon in skull bone. The reported incidence in the skull is 1% of all aneurysmal bone cysts. Most of the cases are associated secondary phenomenon like pre-existing fibrous dysplasia. We present the imaging features and pathology of a primary ABC of the frontal bone, with no underlying bony abnormality or pre-existing trauma.

CASE REPORT: A 16-year-old boy presented with insidious onset gradually progressive, painless swelling in the right side of the forehead over 6 months. Physical examination revealed an ill-defined, firm to hard consistency, non-tender mass (measures 3x4cm), fixed to frontal bone with free surface situated just superior and posterior to the right orbital rim with downward displacement of right eye. The skin over the bone was normal. Exophthalmos was present on the right side. The upward gaze of the right eye was restricted, but visual acuity was normal. There was no focal neurological deficit.

CT scan head showed an expansile intradiploic lesion in right bone. It is heterogeneous mass with multiple septi. Multiple fluid levels are seen within the lesion. The mass extended medially into the supero-medial part of bony orbit and sinonasal cavity on right side, superiorly into anterior cranial fossa compressing frontal lobe and posteriorly up to sphenoid bone.

MR imaging was performed 1.5T seimence Machine. It showed an intradiploic lesion in the frontal bone with multiple fluid–fluid levels and T2 hypo intense internal septations. The lesion showed extra axial mass effect on ipsilateral frontal lobe. Inferiorly the lesion extended into supero-medial extraconal orbital compartment causing bulging of medial wall with extension into and filling of right sinonasal cavity. The lesion extends posteriorly into the sphenoid bone. The lesion is hypo intense on T1WI with a few hyper intense areas suggestive of bleed. On contrast admiration the lesion showed multiple enhancing septi with non-enhancing loculi.

Catheter angiography was done which showed dens tumour blush with feeders from supratrochlear and supraorbital branch of the right internal maxillary artery and right ophthalmic artery and the feeders were embolized by PVA particles to decrease the bleeding risk during surgery.

The patient underwent right frontal craniotomy with excision of the tumour followed by right lateral nasal rhinotomy and excision of the intranasal and right maxillary part of the tumour. The cranial defect was reconstructed with moulded titanium mesh. Postoperatively the patient recovered well with his proptosis reverting back with acceptable cosmetic outcome for his forehead.

Histo pathological examination showed blood filled space surrounded by fibroblasts and dead bony spicules and presence of osteoclastic giant cells.
DISCUSSION: Aneurysmal bony cyst (ABC) of cranial bone is very rare consisted 1% of all aneurysm bone cyst. [1] Other uncommon locations are temporal, [2] parietal and occipital bones, as well as in the facial bones. Among the facial bones, ABC most commonly involves the mandible. [3,4] They usually present at second or third decade of life without any sex predilection. [5] They usually present as firm to hard scalp mass with free skin. Occasionally, it may involve intracranial space, [6] the sinonasal cavities and orbit makes it very difficult to resect.

Primary ABC occurs because of haemorrhage in the bone as a result of increased venous pressure. The haemorrhage is thought to lead to osteolysis. The osteolysis, in turn, causes further haemorrhage, leading to further growth of the tumour. The venous pressure in the calvarium is low; ABC is uncommon in the facial skeleton. On the other hand, ABCs are common in long bones, where the venous pressure is high and the marrow content is greater. [7]

The ABC may be primary or secondary. Trauma is an important etiological factor for secondary ABC. The other condition coexist with ABC are unicameral bone cyst, non-ossifying fibroma, giant cell tumour, chondroblastoma, fibrous dysplasia, osteofibrous dysplasia, fibrous histiocytoma, osteoblastoma and cartilaginous hematoma of chest wall of infants. [5,6] In our case there is no evidence of any associated condition nor had history of trauma.

CT scan is superior to plain radiology for localizing the bony involvement. MR imaging gives better soft tissue characterization of the tumour. It also defines the extension into the cranium, Sino nasal cavity and orbit. Multiple small fluid levels are important characteristics of aneurysmal bone cyst on CT and MRI, which represents sedimentation of red blood cells within blood filled cavities. [8, 9] Other features noted are multiple enhancing fibrous septa, blooming on GRE images. In our case multiple cystic cavies with fluid-fluid levels and enhancing septa were seen.

The treatment is total excision as it has recurrence. These lesions are highly vascular making impose problem during surgery. So pre surgical embolization of arterial feeders can be done to decrease the bleeding risk. [10, 11] In our case preoperative embolization could not be performed as blood supply from ophthalmic artery. So sclerotherapy was performed with sodium tetradecyl sulphate. Following sclera therapy near complete removal of the lesion was possible with minimal blood loss.

Pathologically, these cysts contain multiple blood filled cavities separated by multiple thin fibrous septa lined by multinucleated giant cells. These cystic spaces do not have an endothelial lining. Other additional features are blue bone, woven bone, degenerated calcifying fibromyxoid stroma reported by Rosai. [12] The risk of recurrence is increased with an increase of mitotic figures. All the features except nuclear atypia or mitotic figure were seen in our case. So the chance of recurrence is low, but need to be followed up.

CONCLUSION: Intradiploic aneurysmal bony cyst with intracranial, orbital and sinonasal extension is very rare. CT scan is better to characterize the bony part of tumour. MR imaging is imaging of choice to define the lesion along with its extension. Preoperative embolization or sclera therapy help in decrease the bleeding risk during surgery.
CASE REPORT

REFERENCES:

Fig. 1: MR T2 coronal image (A) showed an intradiploic lesion in the frontal bone with multiple areas of fluid-fluid level and intervening hypo intense septi. (B) Coronal T1 weighted MR image shows subacute blood products in the superior aspect of the lesion. (C) On the post-contrast T1 weighted sagittal image, there is intense enhancement of the peripheral solid component of the tumour. The central cystic components do not enhance.
Fig. 2: Cerebral angiogram showed dense vascular blush with feeders from right ECA branches (A) and right ophthalmic branches (B). The extracranial feeders were embolized with PVA particles (C).
Fig. 3: Right frontal craniotomy with excision of the tumour.

Fig. 4: H & E 4x: histopathology section showing blood filled space surrounded by fibroblasts and dead bony spicules. Inset (H & E, 20x) show presence of osteoclastic giant cells.

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