Aneurysmal bone cyst of the ethmoid sinus: A case report

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Summary

Background: Aneurysmal bone cyst is an expansile bone lesion, non-neoplastic in nature, occurring most commonly in long bones. It is uncommon in facial bones and exceptionally rare in ethmoid bone. Ten cases of aneurysmal bone cysts of ethmoid bone have been reported so far.

Case Report: A young adolescent presented with decreased vision and pain in the right eye. MRI revealed an expansile lesion having conspicuous fluid levels with a multiloculated appearance in the right ethmoid bone extending to the right orbit. CT was done to characterize better bone details. Both biopsy of the lesion and histopathology of resected specimen confirmed aneurysmal bone cyst.

Conclusions: A characteristic appearance on MRI and CT examinations helped to confidently diagnose a relatively common lesion in an exceedingly rare location.

Key words: aneurysmal bone cyst • ethmoid • orbit • CT • MRI • fluid-fluid levels • blood

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Background

Aneurysmal bone cyst (ABC) is a non-neoplastic lesion consisting of blood filled sinusoidal spaces which expand from the affected bone. The most common location of ABCs are the metaphyses of long bones, followed by flat bones. Only 2% of all are found in the head and neck area, with mandible and maxilla being the most frequent sites being involved, involvement of ethmoid sinus is extremely rare [1–4]. Only ten cases of ABC with involvement of the ethmoid bone have been found in the literature; about half of them with orbital extension [1,2].

Case Report

A thirteen year old male presented with decreased vision in the right eye associated with pain, redness and discharge. On examination right proptosis was present, conjunctiva was mildly congested, ocular movements were painful but no limitation of movement was seen. Visual acuity was found to be 6/24 in the right eye and 6/6 in the left eye.

MRI revealed presence of a lesion in the right ethmoid sinus showing multiple, conspicuous fluid levels with septations and a multiloculated appearance (Figure 1). The lesion extended into the right orbit causing displacement of the ocular globe antero-laterally, while the medial rectus and superior oblique muscles along with the optic nerve were displaced laterally (Figure 1A–C). Furthermore, a small focal intracranial extension of the lesion was seen into the anterior cranial fossa. The lesion also showed spreading out into the right nasal cavity and into the infundibulum of right maxillary sinus (Figure 1D). A bulge of the nasal septum to the left was also noticed. The contrast enhancement of thickened mucosa was seen within the left ethmoid cells, right maxillary sinus and slightly less in other paranasal sinuses (Figure 1D). The lesion measured 52×26×38 mm in the anteroposterior, mediolateral and superoinferior plains, respectively. In addition to the fluid levels, the lesion showed hypointense to hyperintense signal in T2W images and predominantly isointense signal with variably hyperintense areas in T1W images, suggesting blood products within the cysts (Figure 1A,B). The contrast enhancement of the septations and solid component was also seen.

CT was done following the MRI to characterize bone details. The images showed expansile bone lesion thinning
Subsequently biopsy was performed and histopathology concluded ABC. During surgery, approximately three fourth of the mass was resected. Parts of the mass lesion close to ocular globe, optic nerve, in the sphenoid sinus and near cribriform plate were not touched. Histopathological examination of the resected specimen confirmed the lesion to be an ABC, the same as what was diagnosed in the presurgical biopsy and in the imaging. No pre-existing lesion was evident from the surgical specimen. On the day of discharge visual acuity of the right eye and ocular movements were normal.

Discussion

ABCs, first described by Jaffe and Lichtenstein in 1942 are benign, expansile, lytic lesions of bone; pathologically consisting of thin walled blood filled cavities, lacking normal endothelium and vascular lamina. 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 [1,2,5–7]. Secondary origin of the lesion from a pre-existing lesion such as giant cell tumour, fibrous dysplasia, osteoblastoma, osteosarcoma, chondroblastoma, chondromyxoid fibroma, unicameral bone cyst has been well documented [1,2]. The lesions often produce symptoms due to compression of adjacent structures or as a result of pathological fracture, rather than by themselves [5].

A constellation of the following features that is of patient’s age less than 20 years, the lesion being expansile bordered by thin low signal rim, having a lobulated or multiloculated appearance with fluid levels on fluid levels on MRI or CT is not pathognomonic but highly suggestive of ABC [5,4]. In addition, each cyst is often surrounded by thin well defined low signal rim on both, T1W and T2W images [5]. On CT, ABCs are typically lytic, expansile and are surrounded by thin shell of bone, which may be partially invisible or eroded. Fluid levels are usually seen within the cyst. The lesions usually show sharp, smooth, rounded soft tissue margin indicating containment by periosseous membrane [4,6]. The visualization of fluid levels can be improved after a period of
patient immobilization before CT examination and viewing the images using narrow window settings [6].

Fluid levels also can be seen in other lesions such as telangiectatic osteosarcoma, giant cell tumour and chondroblastoma. The thin, well defined margins of aneurysmal bone cyst help to distinguish it from other lesions. The fluid levels are probably due settling of degraded blood products within the cysts [5,6,8].

ABCs typically involve the long tubular bones of the extremities, membranous bones of thorax and pelvis or vertebrae [4,6,7,9]. ABCs are less common in head and neck area but predominant sites are paranasal sinuses followed by mandible [9]. Lorne et al stated that diagnosis must be firmly established and malignancy ruled out, this includes diagnosing primary underlying lesion [9]. The most common precursor lesion was found to be giant cell tumour, followed by osteoblastoma, angioma and chondroblastoma [9].

Radiological differential diagnosis of ABC of head neck would include giant cell tumour, giant cell reparative granuloma, hemorhagic cyst, telangiectatic osteosarcoma, metastasis, plasmocytoma and also fibrous dysplasia [4,10–12]. Giant cell tumours are usually seen in older age group. Giant cell reparative granulomas usually have previous history of trauma, both of these present with hypointense signal in T2W and T1W images due to hemosiderin and/or fibrous components [4,13]. Fibrous dysplasia of the skull commonly involves the ethmoid sinuses and is expansile, but it usually has a characteristic ground glass appearance on CT. The cortical, margins unlike in aneurysmal bone cyst, are intact with minimal thinning and usually no fluid levels are seen. Plasmocytoma usually has diffuse marked contrast enhancement [4,11]. Telangiectatic osteosarcoma may have radiological features akin to the aneurysmal bone cyst but is more aggressive in appearance and the age group is older [10]. On MRI, metastatic lesions show as replacement of normal marrow signal on T1 weighted images, they usually have hyper-intense signal on T2 weighted images and show enhancement of bone and soft tissue component with gadolinium [12].

It may be difficult to differentiate the lesion histologically from giant cell tumour, fibrous dysplasia, ossifying haemato ma and cavernous haemangioma of bone due to presence of multiple giant cells in all the above [4].

A neoplastic basis in primary ABC was evidenced by demonstration of clonal chromosome band 17p13 translocations that place the USP6 oncogene under the regulatory influence of highlyactive CDH11 promoter [14].

Complete surgical excision is the treatment of choice, it may range from conservative surgery like curettage, enucleation and endoscopic surgery to more aggressive ones like paralateral rhinotomy and bifrontal cranietomy for ethmoid ABC. Selective arterial embolization in large lesions may help to improve results of surgery. Other 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 [1,4,9].

Conclusions

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 young patient helped to confidently diagnose ABC, although in an exceedingly rare location.

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