Aneurysmal bone cyst of the ethmoid on fibrous dysplasia: A usual association within a rare location

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Abstract

Aneurysmal bone cyst is a non-neoplastic lesion consisting of blood filled sinusoidal spaces that expand from the affected bone. The most common locations of aneurysmal bone cysts are the metaphysis 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 involved, involvement of the ethmoidal bone is extremely rare. Their occurrence on a pre-existing bone lesion is described. Imaging features may assist in diagnosis by demonstrating blood-fluid levels, which are a characteristic finding in these lesions. We present computed tomography and magnetic resonance imaging findings of aneurysmal bone cyst of the ethmoid sinus on fibrous dysplasia in a 12-year-old female.

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Introduction

Aneurysmal bone cysts are extremely rare in the head and neck region and even rarer in sinuses. It is a benign multicystic mass that is locally destructive and rapidly expandable. Hemorrhagic fluid and septated appearance are the characteristic features of aneurysmal bone cysts [1]. We present the case of an aneurysmal bone cyst on fibrous dysplasia with the particularity of being localized on ethmoidal cells.

Case report

A 12-year-old male presented with a slow growing swelling on both sides of the nose over a period of 2 years. There was a history of epistaxis solved spontaneously during this period. The patient also gave a history of anosmia. Physical examination of the lesion showed a nontender swelling on the paranasal region responsible for hypertelorism and proptosis of both eyes.

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Computed tomography (CT) of the paranasal sinuses confirmed an expansile boney mass centered over the ethmoid sinuses with individualization within the mass of a ground glass appearance of the vomer bone (Fig. 1). Magnetic resonance imaging (MRI) revealed the presence of a large cystic expansile mass causing the expansion of the ethmoid sinuses. The septated mass caused displacement of the orbits laterally and anteriorly, and revealed the presence of multiple fluid levels within (Fig. 2). The mass also showed enhancement of its internal septation following contrast administration (Fig. 3). The tumor extended into the anterior cranial fossa, the right nasal fossa, and filled both choanas.

Discussion

Aneurysmal bone cysts typically involve the long bones of the extremities, membranous bones of the thorax and pelvis, or vertebrae. The midline of the skull base is not the site of predilection for aneurysmal bone cysts and involvement of ethmoid sinus is extremely rare. There have been occasional reports of involvement of sphenoid and maxillary sinus [2].

Their cause is not entirely clear but presumed to be due to alteration in local hemodynamics related to venous obstruction or arteriovenous fistula with bone resorption or due to hemorrhage secondary to trauma. Secondary origin of the aneurysmal bone cyst from a pre-existing lesion such as giant cell tumor, fibrous dysplasia, osteoblastoma, osteosarcoma, chondroblastoma, chondromyxoid fibroma, and unicameral
bone cyst has been well documented. In our case, it is secondary to a fibrous dysplasia.

The lesions often produce symptoms due to compression of adjacent structures or as a result of pathologic fracture, rather than by themselves [3].

On CT, aneurysmal bone cysts are typically lytic, expansile and surrounded by a thin shell of bone. Within the cyst, fluid and blood levels are present. The CT seems to be very efficient in detecting ground glass matrix lesions of the fibrous dysplasia. MRI allows a better analysis of the endocranial extension. It shows a lobulated lesion, surrounded by a thin border and crossed by internal septations that appear in low signal on all sequences and enhance after gadolinium administration. These septa delimit cysts that appear in high signal on T2-weighted images and low signal on T1-weighted images. The larger ones have fluid levels related to the deposition of different products of degradation of hemoglobin [2].

Although the aneurysmal bone cyst is not a malignant tumor and has no potential for metastasis, its evolutionary potential, highly destructive character in some cases, and its inaccessibility in this particular case make the treatment very difficult [4].

The standard of treatment remains curettage and grafting to fill the bone void, but the adjuvant treatment methods to reduce recurrence are numerous. Most commonly, orthopedic oncologists use curettage, a high-speed burr, and local adjuvants to remove the tumor prior to reconstruction. Aneurysmal bone cysts in anatomic locations where surgery would cause significant morbidity are most often treated with embolization or radiotherapy and with increasing frequency, medical management with denosumab [5].

Conclusion

Aneurysmal bone cysts of the ethmoidal bone are very rare. CT and MRI allow the identification of blood lakes with fluid levels approving in most cases the diagnosis. They can also reveal other signs related to a pre-existing bone lesion. In addition, MRI seems to be very efficient in establishing an accurate extension assessment especially in this location to guide surgical possible procedures.

Declaration of Competing Interest

The authors declare that they do not have a conflict of interest.

REFERENCES


