Aneurysmal Bone Cyst of the Ethmoid Sinus in a Fifteen-Month Old Girl: A Rare Entity

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Abstract

Introduction: Aneurysmal bone cyst is a vascular bone lesion, occurring most commonly in long bones. Only 2% occurs in the head and neck and it is very rare in the ethmoid sinus.

Case Report: A fifteen-month-old girl presented with proptosis in the right eye. MRI revealed an expansile lesion having fluid levels with a multiloculated appearance in the right ethmoid sinus extending to the right orbit. Biopsy revealed that the lesion was an aneurysmal bone cyst. The patient underwent a complete surgical removal of the mass through a frontal craniotomy and endoscopically through the nose.

Conclusion: ABC is one of the rare differential diagnosis of nasal hemorrhagic mass in a pediatric population. A characteristic appearance on MRI helps to diagnose a relatively common lesion in an extremely rare location. Complete surgical removal is the gold standard with the lowest risk of recurrence.

Keywords: Aneurysmal Bone Cyst; Ethmoid; Orbit; CT MRI; Fluid-Fluid Levels

Introduction

Aneurysmal Bone cysts (ABC) are vascular tumors consisting of blood-filled sinusoidal spaces and they rarely occur in the head and neck region. Involvement of ethmoid sinus is very rare [1,2]. ABC can be a primary or secondary tumor to certain pathological bone lesions. Radiologic findings include a well-defined, expansile, multiloculated bony lesion with sharp, smooth rounded margins bordered by continuous or interrupted thin shell of bone, and fluid-levels spaces on CT scans and MRI [3-5]. The aggressiveness of the disease warrants early diagnosis and proper management to prevent recurrence.

In this article, a fifteen month-old girl presented with proptosis due to a right nasal mass. The patient was diagnosed with this ABC of the ethmoid sinuses.

Case Report

A fifteen-month-old girl, previously healthy, presented for right progressive exophthalmos since one month associated to epistaxis. On physical examination, a right proptosis was present with a moderately congested conjunctiva (Figure 1). The fundoscopic exam was normal. Fibroscopy was performed and a large mass was found filling the right nasal cavity.

Figure 1: Proptosis of the right eye.

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MRI revealed the presence of a multiloculated expansile lytic lesion of the right anterior ethmoid air cells measuring 3.5 x 2.5 x 2.7 cm in its three axis dimensions. It shows intermediate signal intensity on T1W images, heterogeneous high signal intensity on T2W images with multiple fluid-fluid levels and heterogeneous important enhancement on injected sequences. This lesion exerts a mass effect on the ipsilateral orbit with destruction of the lamina papyracea, displacing the medial rectus muscle and causing a grade II exophthalmos. It invades the cribriform plate on the same side leading to a small intracranial extension in the anterior cranial fossa (Figure 2).

**Figure 2:** a) Axial T1W image shows heterogeneous intermediate signal intensity lesion in the right anterior ethmoid air cells with proptosis. b) Axial T2W image shows multiple fluid-fluid levels in the lesion with destruction of the lamina papyracea and displacement of the medial rectus muscle. C and d) sagittal and coronal T2 W showing intracranial extension.

CT was also performed and showed an expansile bone lesion with thinned ethmoid sinus and orbit walls (Figure 3).

**Figure 3:** CT axial and coronal cuts showing extension of the tumor through the lamina papyracea into the right orbit and through the cribriform plate into the anterior cranial fossa.

Endoscopic biopsy was done (Figure 4). The immunohistochemical study revealed that the lesion is vimentin and SMA: positive, desmin, CD34 and CD99: negative. These findings along with reactive bone formation within the tumor are suggestive of ABC (Figure 5).

**Figure 4:** Mass filling the right nasal cavity seen with a rigid endoscope.
Surgical excision was then planned. A bicoronal incision was made, then the scalp flap was rotated anteriorly and frontal craniotomy performed (Figure 6). The superior orbital rim and part of the orbital roof were removed. The optic canal was then unroofed. A defect in the anterior cranial fossa floor was detected through which the tumor was seen to extend intracranially. The superior bulk of the tumor was removed through this open approach using a microscope reaching the basisphenoid. The remaining of the tumor was removed endoscopically through the nose with a 30-degree endoscope. The right periorbita was incised and the tumor was dissected off the medial rectus muscle. The defect in the anterior cranial fossa floor was covered using the temporalis muscle fascia.

The final histopathology report confirmed the diagnosis of ABC.

Few days post operatively, the exophthalmos resolved and the patient was discharged home (Figure 7).

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MRI was repeated six months post-operatively with no evidence of recurrence.

**Discussion**

The first description of aneurysmal bone cyst (ABC) dates back to 1942 when Jaffe and Lichtenstein published two cases of "peculiar blood-containing cysts of large size" [3]. ABC occurs most commonly in the femur, tibia, humerus, spine, and pelvis [6]. ABCs are classified into primary and secondary, with the former being characterized by the lack of history of trauma or pre-existing lesion and is associated with fibrous dysplasia, giant cell tumors, chondroblastoma, chondromyxoid fibroma, non-ossifying fibroma, fibrous histiocytoma, osteoblastoma, and osteosarcoma among others [7].

Of all locations ABCs affect the head and neck region in less than 2% of cases with the mandible being the most commonly affected structure [8]. In 1982, Kimmelman, et al. reported the first case of ABC of the sphenoid sinus in a child [9]. In 1990, Hady, et al. reported the first case of ABC involving the maxillary sinus [10].

To our knowledge sixteen cases of primary ABC of the ethmoidal sinus (excluding our case) have been reported in the literature so far and exceptionally at an age as young as our case subject [8,11,12]. ABC of the ethmoid sinus had no gender predilection in the cases published in the literature before 2006 and a 5:1 female to male ratio after 2006. The average age at presentation is 28.2 years (5 months-90 years), the two extremes of this interval were reported by Segall, et al. [11] and Janjua, et al. [13]. Most patients present with symptoms attributable to the ocular involvement such as diplopia, blurry vision, proptosis, decreased visual acuity and nasolacrimal obstruction. In a review by Ruiz de la Cuesta F, et al. [12], these symptoms were present in 89% of patients with ABC of the ethmoidal sinus, only one patient presented with isolated headache. Associated nasal symptoms including obstruction, epistaxis, rhinorrhea and anosmia were present in eight patients. They reported tumefaction at the level of the inner corner of the eye in 3 patients [12]. Of the previously cited symptoms, our case subject had obvious proptosis and epistaxis.

Radiologic studies have a major role in diagnosing ABC of the ethmoidal sinus with a characteristic appearance on MRI described as fluid-fluid levels separated by septa. CT scan is essential to determine the relationship of the ABC to the lamina papyracea, maxillary sinus and skull base, and the mass is often described as well-defined, expansile, multiloculated bony lesion with sharp, smooth rounded margins bordered by continuous or interrupted thin shell of bone [3-5].

Regarding the management of this entity, curettage has a recurrence rate of 19% when ABC at all locations are considered [14]. Sclerotherapy, although showing promising results [15], is dangerous when transcutaneous approach requires maneuvering near vital neurologic and vascular structures. Radiotherapy is a tempting option considering the difficulty of surgical excision, however this modality might lead to sarcomatous transformation of the ABC [16].

Because it is a locally destructive lesion and in close proximity to important structures, complete surgical resection seems to be the only reasonable option with the lowest risk of recurrence [17]. Surgical strategies include approaching the corresponding sinus with an external incision, bifrontal craniotomy and endoscopic sinus surgery. Our patient was surgically treated using a combination of both bifrontal craniotomy and endoscopy with no peri-operative and post-operative complications. There was no evidence of recurrence 6 months after surgery.

**Conclusion**

Aneurysmal Bone Cyst (ABC) is a benign multicystic mass that is locally destructive and rapidly expandable. It is extremely rare in the ethmoid sinuses. It should be considered as a differential diagnosis of a hemorrhagic nasal mass in children. It can be diagnosed by specific radiologic findings containing a well-defined, expansile, multiloculated bony lesion with sharp, smooth rounded margins bordered by continuous or interrupted thin shell of bone, and fluid-filled spaces on CT scans and MRI. Complete surgical removal is the best therapeutic approach whenever possible, offering the lowest recurrence rate.
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Disclosure

No conflicts of interest to disclose.

Bibliography


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