Aneurysmal Bone Cyst Of The Anterior Ethmoid: Management Of This Rare Entity

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INTRODUCTION

Aneurysmal bone cysts (ABCs) are rare, benign, non-neoplastic, osteolytic and expansile lesions of the bone [1]. Approximately 2% of ABCs are encountered in the head and neck region with the jaws being the most frequently involved sites of the facial skeleton [2]. Aneurysmal bone cysts originating in the ethmoid sinuses are rare with only 22 reported cases in the English literature [3], [4] and [5]. They are more common in individuals under the age of 30 years and females have been reported to have an increased incidence [4]. The average age at diagnosis for ABC occurring in the ethmoids is 11.6 years [3]. There is no known racial predilection. The aetiology and pathogenesis of ABC is not known, however it is thought that a preceding trauma or vascular disturbance causes a reactive process that results in the ABC [5]. As a consequence of the increased venous pressure haemorrhage occurs which in turn causes osteolysis, promoting further haemorrhage and cyst amplification [5]. Pre-existing lesions such as giant cell tumor, chondroblastoma, chondromyxoid fibroma, nonossifying fibroma, osteoblastoma, fibrosarcoma, fibrous histiocytoma, osteosarcoma, and fibrous dysplasia have been reported in a third of cases [6]. We present the case of an 8 year old male with an ABC of the right ethmoid sinus managed in our paediatric tertiary referral centre. We also review the patient characteristics, investigation and management of other reported cases of ethmoidal ABC in the English literature, and suggest a management strategy for these cases.

CASE REPORT

An 8 year old boy presented to the department with a three month history of right epiphora and a right medial canthal mass. He complained of right nasal obstruction. Examination revealed a proptosed and lateralised right eye with a decreased range of movement. A firm medial canthal mass could be palpated and a right nasal mass was seen on flexible nasoendoscopy. Ophthalmological opinion revealed a visual acuity of 0.34 in the right eye and 0.02 in the left eye. He had 3mm of right proptosis with lateral displacement of the right globe. Furthermore there was some limitation of extraocular muscle motility on the right eye with a high intraocular pressure. His cycloplegic refraction revealed a large astigmatic refractive error on the right side.

The CT of the head revealed an expansile mass which appeared to be centred on the anterior aspect of the ethmoid air cells on the right, with a thin surrounding shell of bone (figure 1a coronal, 1b axial).
The enveloping rim of bone was incomplete in places. The mass was expanding into the anterior aspect of the maxillary antrum and also the nasal cavity, displacing the right globe laterally. Magnetic Resonance Imaging demonstrated a largely cystic mass with high signal intensity on T1 images suggesting haemorrhage (figure 2a, 2b). The lesion showed haemorrhagic components of different ages and multiple fluid levels. In the periphery of the lesion there was evidence of some solid tissue and T2-weighted scans also suggested fluid levels. Septal enhancement was noted between these cystic areas. The appearances were suggestive of changes consistent with the diagnosis of an ABC and possibly a primary aneurysmal bone cyst although the site was rather unusual. The adjacent bone was noted to be normal.

On rigid nasal endoscopy under general anaesthesia, a hard right nasal mass was noted with normal overlying mucosa (figure 3). On biopsy, under endoscopic guidance the
anterior and inferior aspects of the tumour were removed after aspiration of 10 millilitre of what appeared to be old blood (figure 4).

**Figure 5**
Figure 3. Right nasal tumour

The tumour was cleared from the right anterior ethmoid to the lamina papyracea, and cleared posteriorly from the posterior maxillary sinus wall, to the postnasal space. Due to the extent of disease and nature of the ABC it was not possible to fully remove all the disease from the skull base, due to the high risk of a cerebrospinal fluid leak and residual cyst wall was left on the skull base. The lateral cyst wall was excised via an external approach using a small Lynch Howarth incision. The hard bone-like wall of the ABC was carefully dissected off the medial wall of orbit; sharp dissection and curettage was required. Residual cyst was left laterally, as tumour was indistinguishable from the lamina papyracea, and due to the increased risk of damage to orbital structures. Postoperatively the patient made a good recovery, and on follow-up with repeat MRI imaging at 12 months there is no sign of recurrence.

**Figure 6**
Figure 4. Right nasal tumour following biopsy

The specimen consisted of pieces of chronically inflamed nasal mucosa with underlying thin plates of woven trabecular bone rimmed by osteoblasts. The tissue showed extensive fascicular spindle cell proliferation. The tissue was markedly more cellular around the bony trabeculae which showed evidence of remodelling. A few osteoclasts were present around the bone and the intertrabecular spaces showed oedematous fibrous tissue. There were sparse lymphocytes and eosinophils in the tissue together with groups of haemosiderin laden macrophages. There was focal fresh haemorrhage. The appearances were consistent with an aneurysmal bone cyst (figure 5a, 5b).

Following biopsy confirmation of the ABC an excision of
Figure 7
Figure 5a. Photomicrograph demonstrating a blood-filled cyst, devoid of an endothelial lining and surrounded by reactive fibroblasts (haematoxylin-eosin, original magnification x10)

Figure 8
Figure 5b. The cyst walls are composed of dense fibrous tissue containing scattered collections of multinucleated giant cells (osteoclasts), osteoid and reactive new bone (haematoxylin-eosin, original magnification x20)

Further histology confirmed the diagnosis of the ABC. On microscopy it was shown that the majority of the lesion consisted of dense fibrous tissue, some areas of which contained trabecular bone. Haemorrhagic lakes surrounded by multinucleated giant cells and rims of reactive fibroblasts were seen. A second histopathological opinion was sought which reaffirmed the diagnosis of an ethmoidal ABC. There was no evidence of another lesion in the submitted tissue to suggest that this represented a secondary aneurysmal cyst. Furthermore there was no evidence of malignancy.

DISCUSSION
In the Ethmoids ABC commonly present with epiphora, proptosis, orbital mass, orbital deformity, nasal obstruction, headache and rarely epistaxis [3], [4], [5], [7] and [8]. The average time from onset of symptoms to diagnosis is approximately 19 months, ranging from 2 weeks to 8 years in earlier reports.

Imaging plays a key role in the diagnosis of ABC with findings pathognomonic for the disease entity. Preoperative radiographic studies are also essential to differentiate ABC from other expansile neoplastic lesions [8]. Classically the CT scan reveals a cystic lesion with bone destruction and new bone formation. MRI imaging shows septa and a fluid-fluid level with evidence of haemorrhage in the cystic lesion [8]. Fluid-fluid levels have been reported to be present in 87.5% of late-stage ABC [8]. This finding is important in differentiating ABC from lesions such as giant cell tumours [8]. If there is any doubt regarding the diagnosis biopsy or angiography is recommended [9]. Features of ABC on angiography elsewhere in the body are a characteristic pathological circulation with a patchy contrast distribution that is persistent in the venous phase, however findings may be absent in cranial lesions indicating that the mass is avascular [10].

Macroscopically, pathological examination typically shows an ABC to be composed of individual cysts filled with unclotted liquid blood and blood-tinged serous fluid [10]. Microscopically blood-filled cavernous spaces are seen, with a paucity of endothelial cells [10]. Septa composed of spindle-celled fibrous tissue, separate the ABC, which contains multinucleated giant cells and possibly osteoid tissue [10].

The preferred mode of treatment is total surgical extirpation of the ABC [9]. Simple curettage is associated with a high recurrence rate of up to 50% [9]. A lower recurrence rate has been achieved with the use of a high-speed bur [6] and [9]. Recurrences tend to occur within 2 years of surgery and during this period patients should remain under close observation [9] and [11].

To date 22 cases, including the report above can be found in the English literature. The management strategies in these cases have been documented as fully as possible in table 1. Articles where only the abstract was available in English were included in order to obtain demographic data and also any information regarding the management of these cases was extracted where available.
On reviewing all the cases of ABC of the ethmoids in the English literature, it is clear that disease in this location occurs more commonly in children (16/22 cases). There appear to be more case reports from the U.S.A (6/22 cases); although due to the limited number of cases this may not reflect a geographical location with an increased incidence, nor are there racial or ethnic group predilections. The tumours appear to be equally distributed amongst females and males. The presenting symptoms are consistent with those reported earlier. An antecedent history of recent trauma was only documented in 3 cases. Imaging is crucial to the diagnosis of this condition. Computed tomography is crucial to delineate the bony anatomy, and to aid the decision regarding the best surgical approach. MRI is fundamental to the diagnosis with the key finding of fluid-fluid levels. In order to confirm the diagnosis prior to the definitive surgical procedure biopsies of the lesion are essential; and as these are found mainly in children, the lesion can be examined at the same time under general anaesthesia. Opthalmological opinion was sought and reported in the majority of cases; a preoperative and postoperative ophthalmology review and eye test is mandatory. Due to the rarity of ABC of the ethmoid the management of each patient should be discussed in a multidisciplinary setting. The surgical approach ideally should be minimally invasive and endoscopic to aid cosmesis; however the ultimate treatment goal is disease clearance. The surgical procedure should be tailored to each individual case. In the literature so far, solely endoscopic, combined endoscopic, paralateral rhinotomy and bifrontal craniotomy approaches have been used; thus alluding to the differences in the extent of the disease and also perhaps surgical expertise. Postoperatively vigilant follow-up is required in the first two years as this appears to be when recurrences are most likely.

**CONCLUSION**

Aneurysmal bone cysts are composed of blood filled anastomosing cavernous spaces separated by cyst like walls. The pathogenesis of these lesions is unclear. Radiological investigations are essential in the diagnosis of ABC. These lesions although non malignant may be rapidly expansile causing progressive destruction. Treatment of these lesions in the ethmoids by surgical resection and long term follow up is necessary to check for any recurrence.

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References

1. Cakirer S, Cakirer D, Kabukcuoglu F: Aneurysmal bone
cyst of the orbit: a case of rare location and review of the
2. Fyrmpas G, Constantinitis J, Televantou D,
Konstantinitis I, Daniilidis J: Primary aneurysmal bone
cyst of the maxillary sinus in a child: case report and review of
the literature. Eur Arch Otorhinolaryngol; 2006; 263:
695-698.
V, Jain R: Aneurysmal bone cyst of the ethmoid sinus:
Uncommon manifestation of a rare case. European Journal
of Radiology Extra; 2009; 70: e49-e51.
4. Ozdamar Y, Acaroglu G, Kazanci B, Saka C, Kazanci B:
Aneurysmal bone cyst of the ethmoid presenting with
proptosis and epiphora. Orbit; 2010; 29:149-151.
5. Hino N, Ohtsuka K, Hashimoto M, Sakata M:
Radiographic features of an aneurysmal bone cyst of the
6. Marcel W, Mandra M, Malinowska I, Pajak J,
Wlaszczuk P, Klczewska E: Aneurysmal bone
7. Gan YC, Hockley AD: Aneurysmal bone
cysts of the cranial in children. Report of three cases and
discussion of the literature. J Neurosurg; 2007; 106:
401-406.
8. Durig JS, Klainguti G: Treatment of a complication after
surgical removal of an aneurysmal bone
cyst of the ethmoid and frontal bone. Klin Monbl Augenheilkd;
9. Mouchon Y, Pharaboz C, Garcia JF, Rigaud A,
Bassoulet J, Cosnard G: Aneurysmal bone
cyst of the ethmoid. Apropos of a case. J Radiol; 1988; 69:
103-107.
10. Kubicz S, Sobieszczen-Sadowska I: A case of
aneurysmal bone cyst of the ethmoid and frontal bone in an
11. Guilemany JM, Alobid I, Blanch JL, Ballestero F, Alos
L, Mullol J: Orbitoethmoid aneurysmal bone cyst. Case
report and literature review. Rhinology; 2004; 42: 164-166.