

A Classic Appearance of an Uncommon Lesion: A Unique Presentation and Review of an Aneurysmal Bone Cyst

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Received: August 09, 2020

Published: November 18, 2020

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Abstract

Aneurysmal bone cysts (ABCs) are rare in the head and neck region, and ABCs of the nasal cavity are even less common. Ascertaining a diagnosis may be difficult owing to their variable clinical presentation. Herein, we document the case of a 17-year-old male who presented with symptoms of nasal obstruction and proptosis of his eye. Imaging revealed a “classic” intra-cystic fluid-fluid level which, while not completely unique to ABCs, lends itself to diagnosing these lesions. The common presentation, diagnostic methodology, and treatment of aneurysmal bone cysts are also discussed.

Keywords: Aneurysmal Bone Cyst; Nasal Cavity; Radiographic Imaging; Pediatrics

Introduction

Aneurysmal bone cysts (ABCs) are benign neoplasms most commonly seen in long bones of the pediatric population [1]. Rarely, they involve the craniofacial skeleton and even less commonly are found within the nasal cavity and paranasal sinuses [1,2]. Surgery is the mainstay of treatment for definitive diagnosis, and to rule out synchronous malignancy. We report an ABC centered in the right nasal cavity of a 17-year-old male and present a review of the clinical and imaging characteristics of this lesion.

Case Presentation

A 17-year-old male with no significant past medical history presented with 3 months of worsening, right-sided nasal obstruction and right sided proptosis. He denied vision changes, focal numbness, epistaxis, or other symptoms. Physical exam was significant for right sided proptosis and anterior rhinoscopy revealed a leftward deviation of his septum. A large, right-sided nasal mass was visualized anteriorly and appeared to be separate from the middle and inferior turbinates.

A magnetic resonance imaging (MRI) brain with and without intravenous (IV) contrast, with a skull base protocol, as well as a computed tomography angiogram (CTA) of the patient's head

and sinuses were obtained. A 7.1 x 4.1 x 4.9 cm enhancing, complex mass with numerous fluid levels centered in the right nasal cavity was visualized (Figure 1a and 1b). The mass extended into the right maxillary and ethmoid sinuses and thinning of the right lamina papyracea and displacement of the right orbital contents was seen. Posteriorly, the mass extended into the sphenoid sinuses bilaterally. No abnormal intracranial enhancement was observed. On CTA the right internal maxillary artery was noted to extend to the mass (Figure 2).

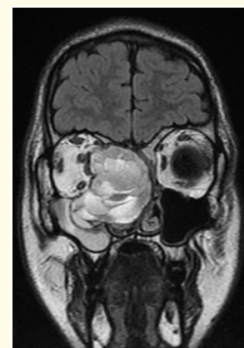


Figure 1: MRI brain with IV contrast T2 FLAIR at the level of the nasal cavity; a: Coronal; b: Axial.

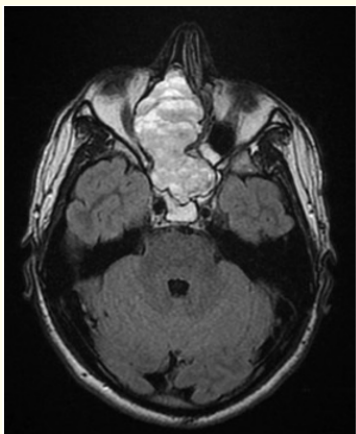


Figure 2: CTA head at the level of the maxillary sinus. White arrow showing internal maxillary artery coursing toward the mass.

Prior to surgery, interventional radiology performed a diagnostic angiogram with possible embolization of a feeding vessel, however no target vessel was identified. The patient then underwent endoscopic removal of the nasal mass with image guidance assistance during which a mass containing both bony and soft tissue features with cystic components was encountered (Figure 3). It extended into the right maxillary sinus, ethmoid air cells, along the skull base, and posteriorly into the sphenoid sinuses where there was osteoneogenic bone along the posterior wall. The mass was successfully excised endoscopically after which the patient was discharged home and recovered well. Final pathology was consistent with an aneurysmal bone cyst.



Figure 3: Endoscopic view of the right nasal cavity with partially resected aneurysmal bone cyst highlighting the bony septations of the tumor.

On post-operative visits, the patient initially had mild crusting and mucosal edema on rigid nasal endoscopy. These findings ultimately resolved. Clinical symptoms including nasal obstruction also resolved completely. He is currently asymptomatic and with no signs or symptoms of persistent disease or recurrence.

Discussion

Aneurysmal bone cysts represent an uncommon benign pathology of the head and neck (H&N) region, with estimates of only 2 - 6% of cases involving the craniofacial skeleton [1,3]. Of the documented H&N occurrences, the majority of cases are limited to the mandible and maxilla, with only scattered case reports of sinonasal cavity involvement [1,2]. Teenagers and young adults are most commonly affected, with a male predominance [4].

ABCs are believed to arise either de novo (primary) in approximately 70% of cases, or secondary to a local trauma or other bone lesions [3,4]. The etiology and pathophysiology are debated. Theories include local hemorrhage leading to cyst formation, or genetic rearrangements and translocations [4]. There are also reports of ABCs with co-existing lesions, including osteomaligancies or benign bone lesions [5]. Clinical presentation depends on lesion subsite, but generally in the head and neck, symptoms are due to local mass effect, including nasal obstruction, proptosis, peri-orbital cellulitis, visual disturbances, and epistaxis [1,3-6]. Hnenny, *et al.* described a case involving the skull base of a pregnant woman that predictably included neurologic manifestations, such as anosmia, ataxia, and hearing loss [7].

Diagnosing ABCs may be difficult owing to its variable clinical presentation. Both MRI and CT imaging is useful and necessary for surgical planning. Radiographically, ABCs are osteolytic lesions on CT and T2 hyperintense on MRI with a classic “intra-cystic fluid-fluid” level [3]. Smaller cysts may arise from larger ones and have been described as “soap bubble” in appearance [8]. Nonetheless, this manifestation is not unique to ABCs and can be seen in telangiectatic osteosarcomas and giant cells tumors among other lesions [5]. Tissue sampling with an incisional biopsy is the gold-standard for obtaining a diagnosis, as less invasive techniques have demonstrated to be inconclusive [9,10].

Macroscopically, an enlarged bony cavity containing fluid (commonly blood) is seen, which accounts for its descriptive name [7]. Hemorrhage and fibrous septations may be present, but their overall gross appearance is non-specific [1]. These septations, com-

prised of spindle cells and cystic spaces lacking endothelium, are basic microscopic features of ABCs [1].

The goals of management of ABCs rest on disease eradication with preservation of maximal healthy tissue. Surgery remains the definitive treatment choice, and most advocate for complete surgical resection if possible, as there is risk of underlying malignancy [3]. In addition, numerous other treatment options, including radiation therapy and immunotherapy, have been reported [5]. For example, Denosumab, a monoclonal antibody binds to the cytokine receptor activator of nuclear factor-kappa B ligand (RANKL), has shown efficacy in case reports and small case series [11]. These alternative treatments can be particularly helpful in tumors where complete resection is especially high-risk or morbid; the skull base, for instance.

Recurrence is common with rates in the literature ranging from 10 - 60% [12]. Risk factors for recurrence are age less than 10 years old, method of excision (curettage), lesions near growth plates, and certain histologic features. In the Orthopedic literature, there is suggestion that surgeons are more cautious in resecting lesions adjacent to growth plates, which may account for higher recurrence rates [12]. Given the risk of recurrence and the nature of paranasal sinus surgery, close follow-up, possibly with surveillance imaging, is warranted.

Conclusion

We report a case of a 17-year-old male who presented with nasal obstruction and proptosis and was found to have an aneurysmal bone cyst of the nasal cavity. An MRI brain demonstrated an enhancing, complex mass with intra-cystic fluid-fluid levels. This is consistent with documented case reports of ABCs and emphasizes imaging's utility in helping provide insight into this benign neoplasm's diagnosis. Although rare, inclusion of ABCs in the differential of an intranasal mass should be considered.

Funding Support

None.

Conflict of Interest

The authors declare that there is no conflict of interest.

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