Aneurysmal Bone Cyst of the Ethmoid: Apropos of a Case and Review of the Medical Literature

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This is a case report of aneurysmal bone cyst of the ethmoid. It is a benign process but with high capacity of local damage. Because of its resemblance with other expansive pathologies of the nasal cavity, it needs to be taken into account to ensure proper diagnosis and treatment.

Key words: Aneurysmal. Bone. Cyst. Ethmoid.

INTRODUCTION

The aneurysmal bone cyst (ABC) is an expansive, non-neoplastic bone lesion, consisting of blood-filled sinusoidal or cavernous spaces. The most common location of the ABC is the metaphyses of long bones and in 30% of cases they are secondary to other bone processes. They typically present in individuals under the age of 30 years, with a mean age of 16 years on debut.

CLINICAL CASE

We present the case of a 13-year old patient, with no personal history of interest, with a feeling of swelling of the right eye and retroocular pain lasting 15 days. In the last 24 hours she had also perceived decreased ipsilateral visual acuity. In the last months she had not presented hyposmia, nose bleeds, nasal respiratory insufficiency, or epiphora.

The endonasal physical examination reveals a neoformation covered with healthy mucosa. A slight proptosis of the right eye is observed with eye motility that is mildly painful, but normal and unaccompanied by diplopia; upon exploration a smooth, localized mass, occupying the upper half of both nasal cavities is noted in the upper two thirds of both nasal cavities, predominantly the right one, causing expansion and remodelling of both medial walls of the sockets without producing bone destruction and with no intracranial spread.

In the magnetic resonance (Figures 3 and 4), intralesional septa and high intensity fluid-fluid levels can be observed.

Figure 1.
Computerized axial tomography. Remodelling of the medial wall of the right socket by the mass is visible without total destruction.
in both the T1- and T2-weighted sequences, a hyperintensity that persists in the fat-suppressed sequences.

We carried out endoscopic nasosinusal surgery (ENSS), evacuating approximately 10 mL of bloody material, and created a pouch in the cyst by placing a silastic lamina from one nasal cavity to the other, surrounding the posterosuperior part of the nasal septum. The pathology study reveals cystic walls with proliferation of spindle cells and multinucleated giant cells with images of mitosis without atypia, compatible with ABC. The patient remains asymptomatic and without any signs of relapse 10 months later.

DISCUSSION

ABC is a benign bone lesion, the aetiology of which is not entirely clear. On the one hand, it has been postulated that it is a local haemodynamic disturbance causing an increase in venous pressure and bone resorption, which is replaced by connective and osteoid tissue. On the other hand, areas similar to ABC have been found in other lesions (fibrous dysplasia, giant cell tumours, etc.) and it has been suggested that the lesion is secondary to a microcyst, due to cell oedema in the primary lesion. In a review of 66 benign bone neoplasms, Biesecker et al. found that one third had an associated ABC. Two different types of ABC are considered: a typical variant (95% of the cases) and a solid variant (5%) that are indistinguishable on clinical and radiological presentation and differ one from another solely because the solid form does not present cavernous and sinusoidal spaces, but does present a greater number of mitotic figures and definitive diagnosis of ABC depends on the pathology study of the specimen. In one work carried out at the Mayo Clinic with 238 cases of ABC, the authors found that the most common location is the metaphyses of the long bones, followed by flat bones (above all, the pelvis) and the spinal column. Less than 5% of these lesions are located on the skull and face, and the most common location in this area is the mandible and maxillary bones. They conclude that it is made up of a fibrous matrix crossed by cavernous and sinusoidal spaces, some of which may be covered with endothelial cells, and comprising an irregular distribution of spindle cells and multinucleated giant cells. Osteoid tissue was found in practically all the specimens analyzed in this study.

Fifty per cent of the cases presented formation of new reactive bone on the borders of the lesion. They describe the presence of mitotic figures in close to 90% of the samples, without finding atypias in a single case. ABC is a more or less rapid growth with local bone destruction, leading to the clinical manifestation of pain and swelling. Only 9 cases of ABC have been found to involve the ethmoid bone in the entire body of medical literature, 4 of which were in females and 5 in males. The mean age at debut was 11.6 years (11 months-20 years), and the mean time since onset was 19 months (2 weeks-8 years).

The most commonly detected presenting symptoms (89%) in this review were related to the socket: diplopia, blurry vision, proptosis, loss of visual acuity, nasolacrimal
obstruction, among others, and there was only one case that presented with headache. This clinical manifestation was accompanied by nasal obstruction in 4 patients, facial tumefaction at the level of the inner corner of the eye in 3, nose bleeds in 2, rhinorrea in 1, and anosmia in 1.

The radiological studies revealed bulging and lateral displacement of one or both laminae papyraceae of the ethmoid bone in all cases, with the resulting eye involvement. Moreover, in 3 of them (33%), the maxillary sinus is involved, intracranial extension is present in another 3, and growth toward the nasopharynx in 1. The computerized tomographic images were characteristic, revealing expansive growth with bulging and thinning of the adjacent cortical bone. In turn, the magnetic resonance is highly suggestive, with a well-defined image of fluid-fluid levels separated by septa.

The treatment applied consisted of external ethmoidectomy (associated with maxillectomy or frontoethmoidectomy) in 4 patients, bifrontal craniotomy (one associated with revision surgery to excise the nasopharynx extension) in the 3 cases with intracranial extension, and of the remaining 2 cases, ENSS was performed in one and a subfrontal approach was used in the other. None of the authors report relapses or post-operative complications.

There appears to be a consensus in the sense that the treatment of ABC in any location is eminently surgical, but in light of its progressive growth, capacity for local destruction, and spread to soft tissues, many groups have considered the possibility of a more aggressive initial therapy. A 19% recurrence rate is reported in cases that were treated with debridement, whereas not a single patient of those whose surgery included broad safety margins presented recurrence.

Several studies have been published on aneurysmal bone cysts treated by means of percutaneous CT-guided sclerotherapy with very good results, albeit its use is not advisable in lesions located very close to the central nervous system, as these can entail serious complications, and, in cases that cannot be approached percutaneously, direct intrallesional sclerotherapy may be carried out. Radiotherapy is not recommended, as several cases of evolution into sarcoma have been reported following this treatment.

CONCLUSIONS

ABC is a benign process that is common in other locations of the body, but exceptional when it appears in the nasal cavities. Nevertheless, it should be taken into account when making a differential diagnosis. It can only be confirmed by means of the pathology study of the surgical specimen. In English-language medical literature, various surgical treatments using an external approach have been described. In our case, we performed an endonasal endoscopic surgical technique and the patient was disease-free 10 months after the surgery.

REFERENCES