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Prikazi bolesnika/
Case reports

EXTRA-ORAL APPROACH TO THE TREATMENT OF GIANT ANEURYSMAL BONE CYST/ *Case report*

EKSTRAORALNI PRISTUP U LEČENJU DŽINOVSKE ANEURIZMATIČNE KOŠTANE CISTE/ *Prikaz slučaja*

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Key words
Aneurysmal bone cyst; jaw cyst; jaw neoplasm; pseudocyst; surgical treatment

Ključne reči
Aneurizmatična cista kosti, cista vilica, tumori vilica, pseudociste, hirurško lečenje

Abstract

Aneurysmal bone cyst is a benign osteolytic lesion. Although it was first described by Jaffe and Liechtenstein in 1942 its generation mechanism, natural history or ideal treatment are not yet fully understood. Here we describe the case of a 17 years old male patient with a large tumor in the chin of several years of evolution, which had progressively protruded into the mouth floor displacing the tongue and impairing swallowing. According to its clinical and radiological characteristics we considered this tumor to be in the active phase of the Capanna classification. The size of the tumor indicated that extra-oral approach was safest. After total extirpation of the lesion, we proceeded to bone reconstruction with a ground corticocancellous bone autologous graft from the right iliac bone, using a titanium micro-mesh that closely fitted the defect holding the graft in place.

INTRODUCTION

The World Health Organization defined Aneurysmal Bone Cyst (ABC) as a benign osteolytic lesion characterized by blood-filled cystic spaces of variable size, separated by connective tissue septa, with a fibroblastic stroma containing giant multinuclear cells, osteoid and bony tissue. (1)

This entity was first described by Jaffe and Lichtenstein in 1942. (2)

However, the first published case of mandibular location of ABC was reported by Bernier and Bhaskar in 1958.(3) After all this time, the mechanisms of generation, natural history and optimal treatment of this entity are not yet fully understood. It has been proposed that these lesions could correspond to a vascular degenerative process occurred during the development of some benign bony lesions. Although appealing such a proposal is however not supported by general pathological findings, with a few exceptions. In general, the outcome studies of tissue samples pose difficulties for differential diagnosis against giant-cell tumor, chondroblastoma, hemangioma, osteoblastoma, non-ossifying fibroma, fibrous dysplasia, chondromyxoid fibroma and other benign and malignant tumors.

Here we present a case of a large-sized ABC located on the mandibular symphysis that required extraoral approach for complete removal and subsequent careful reconstruction.

CASE REPORT

A 17 years old male patient presented with a large-sized tumor in the chin of several years of evolution, which had recently protruded into the mouth floor, pushing the tongue and impairing swallowing. His dentist had performed endodontics on the anterior inferior sector. As he encountered the vital teeth, he referred the patient to our hospital for further examination.

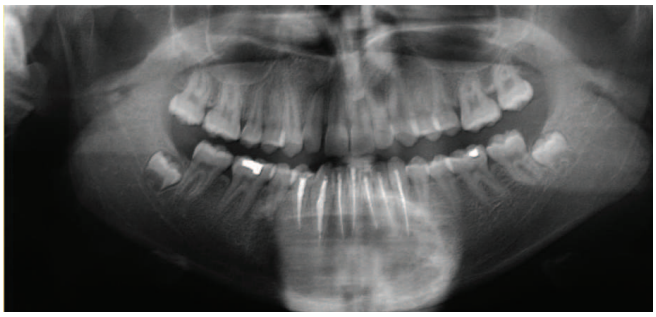

Figure 1.- Orthopantomography showing the image of a lytic lesion in the mandibular symphysis, extending from left premolar to right canine.

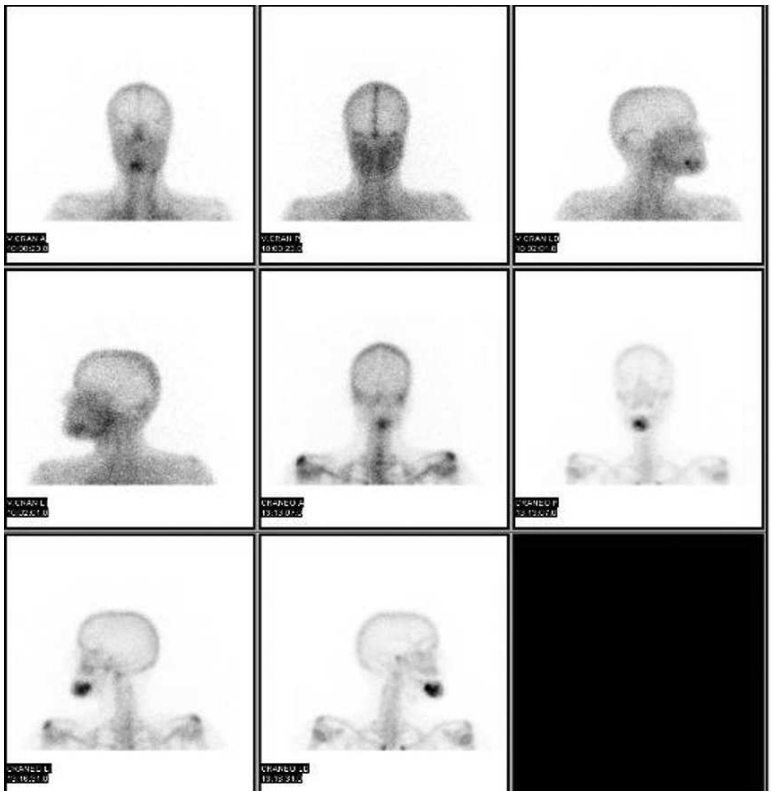

Figure 2.- The gammagraphy shows a noticeable increase in the uptake of the radiolabeled ligand in the chin area, both in the early and the late phases of the study.

An orthopantomography revealed a lytic lesion on the mandibular symphysis, extending from 43 to 34 (Fig. 1).

Selective bone gammagraphy, performed 5 minutes and 2 hours after intravenous administration of Tc99m-HMDP showed high uptake of the radiolabeled ligand in the chin region with higher involvement of the left hemimandible, observed both in the early and the late phases of the study (Fig. 2). Nuclear magnetic resonance revealed a large mass taking up the whole mouth floor, though it failed to provide any further data to help diagnose (Fig. 3). Patient examination was completed with computerized axial tomography, which revealed an expansive lytic lesion on the anterior region of the mandibular arch, which included septa. The cortical was thinned and appeared to be absent at some points, 38 x 35 x 33 mm (anterior-posterior, transverse and craniocaudal diameters, respectively). Heterogeneous highlighting was observed after intravenous contrast administration, which contacted with the roots of the lower incisors and canines and with both teeth canals, apparently invading the left one. No soft tissue mass was identified outside the lesion. Submandibular adenopathy of non-radiologically-significant size and morphology were observed (Figs. 4 and 5). All of these findings were suggestive of a solid non-aggressive lesion, which was suspected of fibrous dysplasia, on the basis of the patient's age, location of the lesion and radiological image.

Figure 6 shows the bone distension resulting from tumor growth; while Figure 7 shows the corticotomy performed to access it. Figure 8 shows that the tumor was completely removed while the mouth floor was left untouched. After total extirpation of the lesion, we proceeded to bone reconstruction with ground corticocancellous bone autologous graft from the right iliac bone, using a titanium micro-mesh that closely fits to the defect holding the graft in place (Fig. 9).The microscopic description of the tumor included proliferation, mainly fibroblastic benign, where small differentiation nests of trabecular

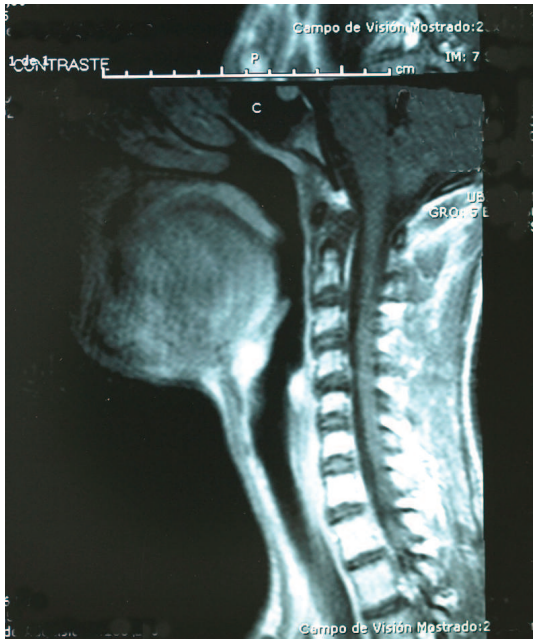

Figure 3.- The nuclear magnetic resonance revealed a large mass taking up the whole mouth floor.

bony structures with a lattice-like arrangement could be observed. The fibroblastic proliferation showed no cellular atypia, while collagen fibers and small focal osteoid pieces were observed. In general, it tended to limit hematic cavernous cavities. Mitotic figures were rare. Scattered osteoclast-like multinuclear giant cells were found. Differentiation of fibromyxoid material with a chondroid "aura" was not observed. The focuses, where the lattice-like pattern of small bony trabeculae could be observed, showed small vessels, low inter-trabecular cell density and no cellular atypia (Figs. 10 and 11). On the basis of these findings, primary aneurysmal bone cyst was diagnosed. Follow-up controls were conducted 6 and 12 months after intervention. The patient was asymptomatic and the bone defect had regenerated.

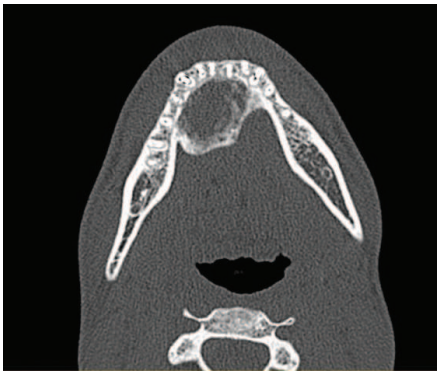

Figure 4.- The lesion contacted with the roots of the lower incisors and canines.

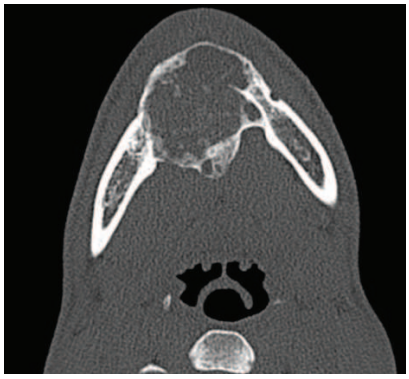

Figure 5.- Expansive lytic lesion in the anterior region of the mandibular arch.

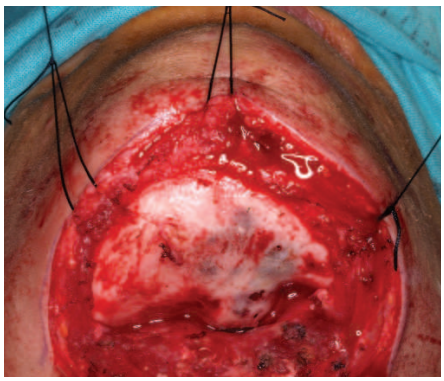

Figure 6.- Tumor growth into the moth floor.

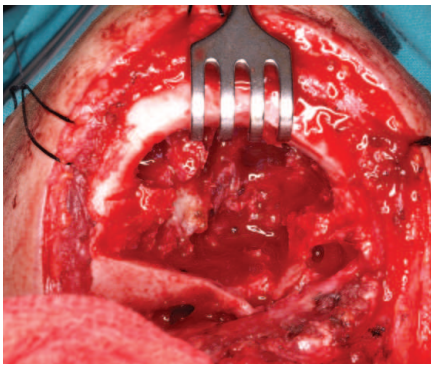

Figure 7.- Corticotomy to access the cyst.

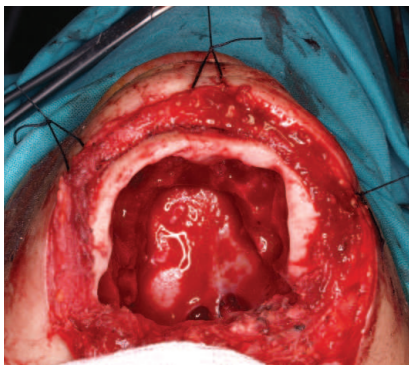

Figure 8.- Complete extirpation of the tumor

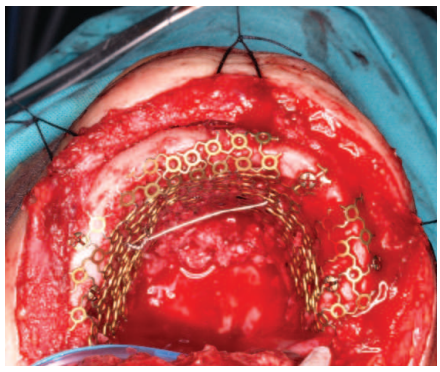

Figure 9.- Reconstruction with ground corticocancellous bone autologous graft and titanium micromesh.

DISCUSSION

Although extensively used the term ABC is actually not suitable to name this entity. It is not an aneurysm, since there is no endothelial lining of the vascular spaces, and it is not a cyst, since it lacks epithelial lining. Therefore, this term is currently being replaced by central benign giant cell tumor or central giant cell lesion. [4] The etiology of ABC is still unknown. It is not known whether these lesions appear de novo or result from a vascular accident occurring in a pre-existing lesion. Most authors believe that they result form a vascular malformation in bone. Three different theories have been proposed on the origin and pathogenesis of ABC: a) traumatic origin, which would involve subperiostic or intramedullary hemorrhage with altered repair processes; b) hemodinamic alterations, which promote dilatation and congestion of the vascular bed, thus causing bone resorption and erosion, plus enlargement of the lesion; and c) lesion secondary to an earlier primary bone lesion. [5] ABC usually appears on long bones and vertebrae; it seldom occurs in the maxillary bone. Only 92 cases of maxillary ABC have been published up to date, in 75 articles in English. [6] Like in our reported case, this entity usually occurs in people younger than 20 years, with an average age of 16.5 years. [6]

The clinical presentation of ABC may be widely variable. Most cases evolve indolent and asymptomatic (54.7%), although pain (37.2%) or fast destructive growth may occur; 28% of cases have a background of traumatism in the area where the lesion appears. [6] Further, less frequent symptoms are: limited mouth opening, alterations in sensitivity or local paresthesia, nasal obstruction, diplopia, epiphora, epistaxis, strabismus, exophthalmos, teeth-related symptoms such as tooth mobility or displacement. Very infrequently it can be associated with pathological fracture, with only one case described in the reviewed literature. [7] The here reported case involved progressive painless growth without further accompanying symptoms.

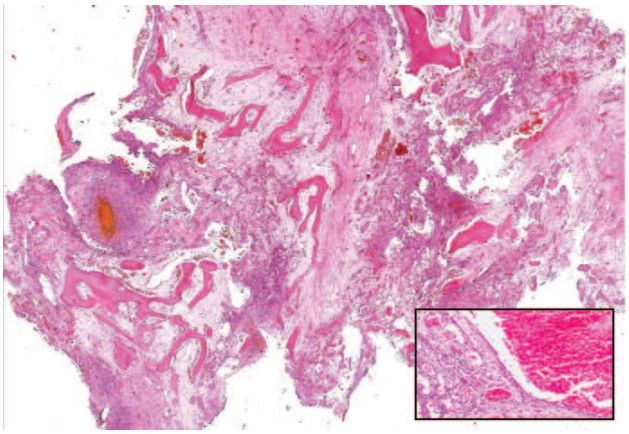

Figure 10.- Cancellous bone destroyed and dried out by fibroblastic proliferation with multinuclear giant cells and pieces of small bony trabeculae in a lattice-like arrangement.

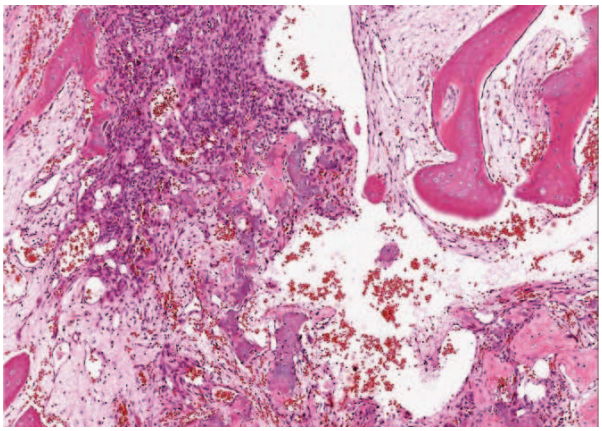

Figure 11.- Mitotic figures are very rare; cellular atypia is not observed.

The outcomes of X-ray and TAC studies are not pathognomonic and there is no consensus in the literature in this regard. This could be partially due to the existence of variants in terms of development: there is a relatively solid, defined and less destructive growth pattern; and there is the so-called vascular growth pattern with less defined margins and generally more destructive. X-ray studies may reveal unicystic, multilocular or moth-eaten appearance. In all cases, it causes expansion, perforation or destruction of the bone cortex, as shown in the images of this article. Additionally, a periosteal reaction with bone formation can be observed, which results in a peripheral sclerotic rim. The clinical diagnosis is rather difficult due to the similitude between this entity and other lesions such as: ameloblastoma, giant-cell tumor, brown tumor of hyperparathyroidism, myxoma, traumatic bone cyst or keratocyst. Blood aspiration from the lesion should lead the clinician to suspect of a vascular lesion or an ABC. The absence of noise, vibration or pulse may help in the clinical differentiation of a vascular lesion from an ABC. This is essentially a diagnosis of exclusion. A definitive diagnosis can only be reached after incisional biopsy. Only after a vascular lesion has been ruled out, biopsy can be pursued. (8)

On the basis of the above commented clinical and radiologic characteristics, Capanna et al. (9) classified ABCs in: a) cysts in an inactive phase, with well-defined sclerotic margins; b) cysts in an active phase, with some alterations in the periosteum but still with defined margins; and c) aggressive cysts with signs of osteolysis and undefined margins. Although these authors were concerned with long bones, we consider that this classification might be applied to ABC with a maxillary location and that it could even have a certain prognostic value, useful in the choice of treatment and the management of possible recurrence. The case described here could be considered to be in the active phase of this classification.

In 95% of cases, the typical histopathologic findings correspond to the vascular form, which involves an osteolytic expansive lesion with vascular lakes of variable size, separated by connective tissue including bony trabeculae, osteoid tissue and multinuclear giant cells. Our reported case corresponded to this histopathologic variant. In the remaining 5% of cases the solid variant, consisting of a solid mass of variable density and without a cystic element, should be considered.

The treatment for ABC depends on different factors: the patient's age, the location and size of the lesion, and possible occurrence of concomitant diseases. Performing a biopsy previous to the treatment is crucial both to confirm the diagnosis and its clinical form. The Capannas's classification. (9) might help establishing a suitable treatment plan. Inactive lesions may be treated with curettage. Selective arterial embolization or injection of sclerosing agents may considerably help in the treatment. Inactive ABCs shall be carefully monitored after the initial treatment. Since these patients are usually children, careful curettage should be the initial treatment of election. Resection should be considered if enlargement of the lesion, pain or destruction of neighboring structures were observed. (6)

The rate of recurrence is about 20-30%. Recurrence is most frequently observed during the first year of treatment and usually attributed to an inadequate approach that results in incomplete extirpation, especially in cases where extension into the soft tissues has occurred. (5) Many authors have recommended primary bone reconstruction with autologous grafts whenever the resulting defects makes it necessary, e.g. due to potential negative cosmetic outcome or risk of fracture or mandibular discontinuity. (10, 11, 12,13)

In our patient, the size of the tumor suggested that an extra-oral approach was safest, preventing the risk of vascular or nerve lesions, as well as of lesions to the Wharton ducts.

Apstrakt

Aneurizmatična koštana cista je osteolitička benigna lezija. Iako je prvi put opisana još 1942 od Jaffe-a i Lihtenštajna, mehanizam nastanka, priroda bolesti i idealna terapija još nisu u potpunosti shvaćeni. Opisan je slučaj 17 godina starog muškarca s velikim tumorom u bradi od nekoliko godina evolucije, koji je progresivno prodirao pod jezik u usta i ometao gutanje. Prema kliničkim i radiološkim karakteristikama smatrali smo da je tumor u aktivnoj fazi po Capanna klasifikaciji. Veličina tumora pokazala je da je ekstra-oralni pristup bio najsigurniji. Nakon potpune ekstirpacije lezije, uradili smo rekonstrukciju koštanog defekta presadom kosti (mešavine korteksa i spongioze), kao autolognog grafta, uzetog sa desne ilijačne kosti, koristeći titanijumsku mikro-mrežicu kao ležište koja se sasvim popunjava koštanim graftom (presadom), ispunjavajući na taj način defekt i držeći presađenu kost na mestu postoperativnog defekta.

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