Case Report

Aneurysmal bone cyst: Rarity in mandible and its ambiguity with Central giant cell granuloma

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ABSTRACT

The aneurysmal bone cyst is an uncommon lesion which has been found in most bones of the skeleton, although the majority occur in the long bones and in the spine. It was first described as a distinct clinical entity by Jaffe and Lichenstein in 1942 to describe the characteristic "blow-out" of the bone seen in the radiographs of the lesion. In the past, the lesion has been classified as an atypical giant cell tumor or benign bone cyst. We report a case of an aneurysmal bone cyst in an 18-year-old patient who reported with the chief complaint of swelling on the right side of the face since 4 months. It was non-tender, non-fluctuant, and hard in consistency. Radiographic examination revealed a large, expansile, multilocular lesion suggestive of benign odontogenic tumor. Complete enucleation was carried out and the final histopathologic diagnosis of aneurysmal bone cyst was given.

Key words: Aneurysmal bone cyst, central giant cell granuloma, giant cell, enucleation, giant cell

INTRODUCTION

The aneurysmal bone cyst (ABC) is a benign intraosseous lesion characterized by blood-filled cavernous spaces of varying sizes associated with a fibroblastic tissue containing multinucleated giant cells and osteoid. This disease has been known as a pathologic entity since 1942,[1] and several reports in the medical and dental literature have clarified the pathology and confirmed the benign nature of this lesion, although the etiology remains obscure.[2]

Most of the lesions are associated with the mandible. They are located in the molar regions of the mandible and maxilla. A number of mandibular cases extend posteriorly to involve the angle and ascending ramus. The histopathology of the central giant cell granuloma of the jaws and the ABC is almost identical; the only difference is the blood-filled cavernous spaces seen in the ABC.[2]

The primary ABC is the entity which arises as itself from the initial stages of development. The secondary ABC is a secondary manifestation developing in a pre-existing lesion altered by hemorrhage, cystic degeneration, or some other pathological process.[3]

CASE REPORT

An 18-year-old female patient reported to the Department of Oral Medicine with the chief complaint of swelling on the right side of the face since 4 months. The onset of the swelling was gradual. It had rapidly increased to the present size within 2 months. There was no history of trauma, though the patient gave a history of pressing hard over the swelling with her right palm before 2 months.

Extraoral examination revealed diffuse swelling on the right side of the mandible, extending anteroposteriorly from the midline to the mandibular angle and superoinferiorly from approximately 3 cm above the inferior border of the mandible till 2 cm below it along the length of the body of the mandible, of approximately

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6 × 5 cm in size. There was obvious facial asymmetry. Overlying skin was normal and there were no signs of inflammation, bleeding, or pus discharge. Expansion and bowing of inferior border of mandible was seen [Figure 1]. On palpation, there was local rise in the temperature as compared to the opposite side. It was hard in consistency, non-fluctuant, non-reducible, non-pulsatile, non-compressible, and non-tender. There was no egg-shell crackling or paresthesia noted. One right submandibular lymph node was enlarged, palpable, soft, mobile, and tender.

On intraoral examination, a diffuse swelling in the mandibular right buccal vestibule, extending anteroposteriorly from the midline to mandibular second molar region and superoinferiorly from the gingival margin to the depth of the buccal vestibule, of approximately 6 × 3 cm in size was noted. There was obliteration of the buccal vestibule and expansion of the buccal cortical plate. Lingual tipping of right mandibular first premolar, second premolar, first molar, and second molar was observed. There were no carious, discolored, or fractured teeth. The third molar was not seen clinically [Figure 2]. On palpation, it was hard in consistency. Expansion of the buccal cortex and expansion of lingual cortex in the region of 46 and 47 was noted. Mandibular right and left central and lateral incisors were Grade I mobile. Dull, cracked-pot sound on percussion of the teeth of the entire right quadrant was noted.

With consideration of all the above findings, a provisional diagnosis of benign odontogenic tumor was given.

All of the mandibular teeth gave normal response on the electric pulp vitality test.

The following radiographic investigations were carried out:
Mandibular right lateral occlusal topographic view [Figure 3] showed well-defined, corticated multilocular radiolucency extending from the left mandibular first premolar to the right second molar region. Expansion and thinning of buccal and lingual cortices was noted. Lingual tipping of the mandibular right first premolar, second premolar, first molar, and second molar was observed.

Oral pantomograph [Figure 4] showed well-defined, corticated multilocular radiolucency extending from the left mandibular first premolar to the condylar process of the mandible, sparing the condylar head. There was thinning and expansion of the inferior border of the mandible. No lateral or apical root resorption, tipping, or displacement of the teeth was noted. Superior scalloping between the roots of the anterior teeth was noted.

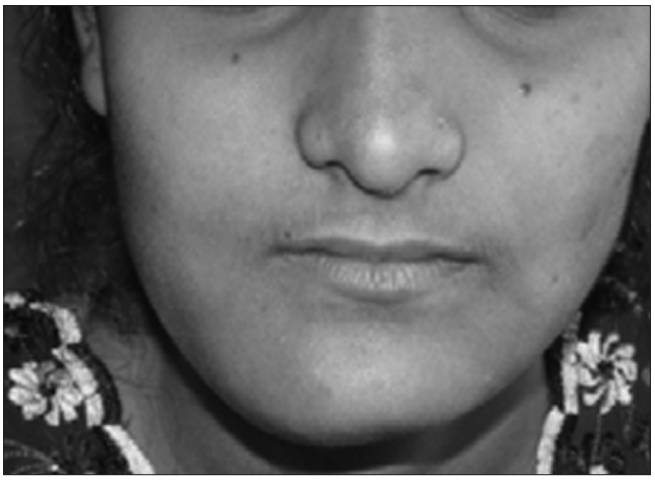

Figure 1: Extraoral swelling on the right side of the face

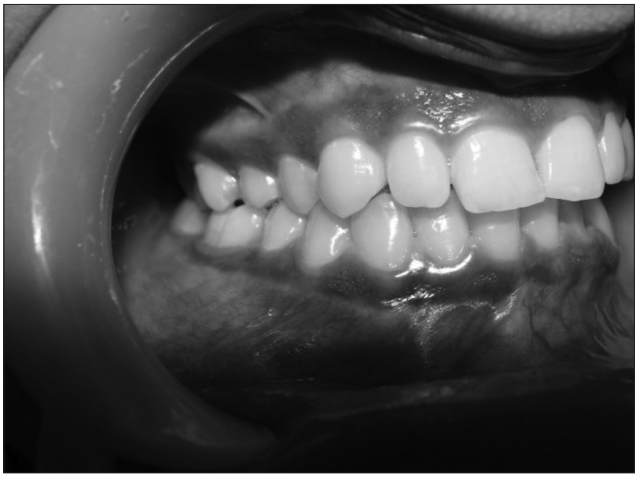

Figure 2: Intraoral swelling in the right buccal vestibule

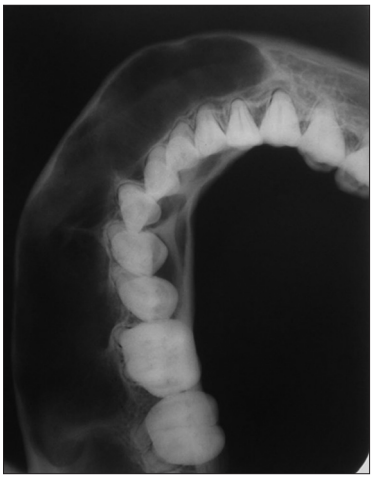

Figure 3: Mandibular right lateral occlusal topographic view showing well-defined, corticated multilocular radiolucency extending from the left mandibular first premolar to the right second molar region

Axial section [Figure 5] showed a well-defined, expansile, cystic, multilocular hypodense lesion, 5.5 × 2.2 cm in size. There was break in the continuity of the lingual cortex in the region of ascending ramus. It

showed CT value of 15–30 HU, likely to be thick cystic fluid with no significant enhancement on contrast study. Suggested pathology was benign odontogenic cyst or tumor.

The most probable clinicoradiographic diagnosis given was of central giant cell granuloma.

The clinicoradiographic differential diagnosis considered was of odontogenic keratocyst, ABC, ameloblastoma, and traumatic bone cyst.

Aspiration and incisional biopsy was carried out from the right mandibular premolar–molar region. Cytological entities seen were mainly red blood corpuscles. Small bits of tissue composed of chiefly the connective tissue component showing a fibrocellular stroma and a few inflammatory cells were seen. Young fibroblasts were numerous in the stroma. Only in focal areas, thin, 2–3-cell-thick epithelial lining was evident. It was absent in all the other areas. The cavity mainly contained blood.

The diagnosis of dental cyst was given. The liver function tests, renal function tests, serum calcium, serum phosphorous, and serum alkaline phosphatase levels were normal. The liver function tests were advised in our patient in order to assess serum bilirubin, serum albumin and globulin, and prothrombin time. The above-mentioned tests were thought to be useful as the future treatment plan for the patient was surgical curettage. These tests give the idea regarding the integrity of the clotting mechanism and general metabolism of the body.

Thorough curettage of the lesion was done at once and a dressing of betadine gauze roll was placed in the bony cavity. The dressing was changed every 2 days with progressive smaller sizes of the gauze roll. This was carried out for a period of 8 weeks. The developing third molar tooth was extracted. Arch bars were placed for maxillary and mandibular arches. Intermaxillary fixation with elastics was done in the posterior region to support the redundant inferior borer of the mandible. The epithelial lining and the gross pathology specimen were sent for histopathologic examination. The patient was followed up every 15 days after the treatment to evaluate healing and bone formation.

The H and E stained sections demonstrated the following details:
Lesional tissue in several fragments showed large, irregular sinusoidal spaces engorged with blood and devoid of any endothelial lining [Figure 6]. These spaces did not show thrombosis. Varying amounts of hemosiderin was also present.

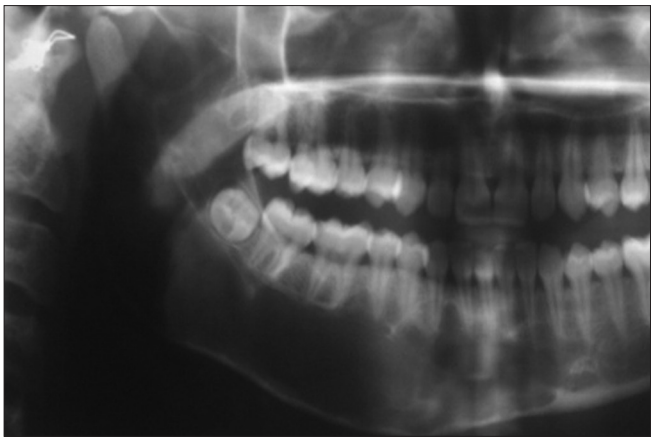

Figure 4: Cropped OPG showing well-defined, corticated multilocular radiolucency extending to the condylar process of the mandible sparing the condylar head

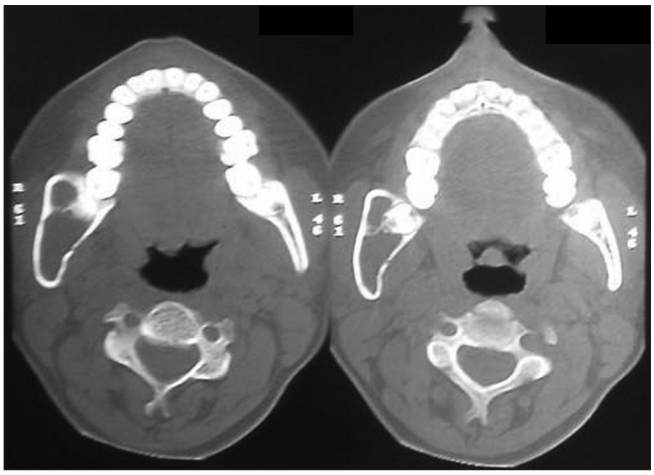

Figure 5: Axial computed tomography section shows a well-defined, expansile, cystic, multilocular hypodense lesion

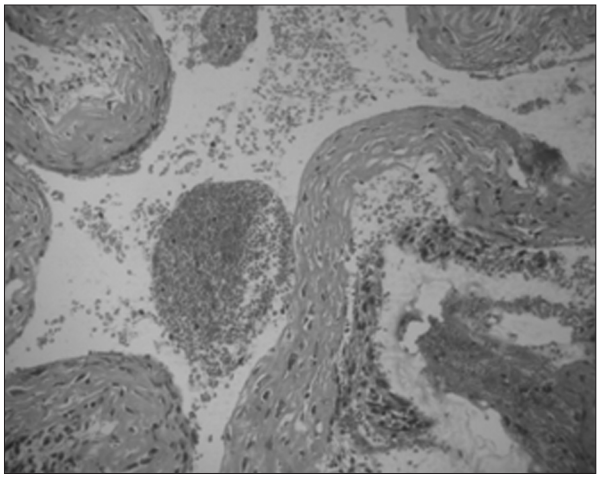

Figure 6: Evidence of large, irregular sinusoidal spaces engorged with blood, devoid of epithelial lining

Other features noted were osteoid formation and giant cells at places. There was no evidence of malignancy. The final diagnosis given was primary ABC.

DISCUSSION

ABC is a rare non-epithelialized pseudocyst of the jaws. It is rarely found in the jaws and comprises 1% of all nonepithelial and non-odontogenic cysts of the jaws. It was first recognized as a distinct clinical and pathological entity by Lichtenstein *et al.* in 1942.[1] Bernier and Bhaskar in 1958 were the first to describe the presence of this lesion in the jaws.[3] Despite its recognition in the jaws since 1958, it is still an uncommon finding in the facial bones. According to Jaffe, the word aneurysmal was used in relation to the blow-out distension of part of the affected bone area.[1,3] The initial part is formation of the microcysts.[4] This entity in some respects is similar to simple bone cyst.[5]

Aneurysmal cysts of the jaws produce firm swellings which have been described as painful in fewer than half of the reported cases. The swelling and malocclusion frequently become progressively worse and the rate of enlargement is often described as relatively rapid. Occasionally, there is a history of recent displacement of teeth which remain vital.[6,7] When the lesion perforates the cortex and is covered by periosteum or only a thin shell of bone, it may exhibit springiness or egg-shell crackling, but is non-pulsatile. Bruits are not heard. There may be some difficulty in opening the mouth if there is impingement of the lesion on the capsule of the temporomandibular joint. During surgery and upon removal of the bone shell, dark venous blood wells up and bleeding may be difficult to control.

The etiology and pathogenesis of this lesion is still debatable. According to the patient's histories, trauma does not seem to play a significant etiologic role. Lichtenstein[1,3] in 1950 postulated that ABC is a manifestation of altered hemodynamics. There is a presence of circulatory disturbance leading to congestion. Pressure secondary to vascular dilatation and engorgement may result in bone resorption, with deposition of fibrous connective tissue, osteoid, and new bone.

Another proposal was formulated by Bernier and Bhaskar[3] who suggested that though ABC has nothing in common with giant cell turnout of bone, it resembles the central giant cell reparative granuloma of the jaws. The only difference between the two is the presence of blood-containing spaces. They further suggested that both lesions represent overzealous attempts of the connective tissue to replace a hematoma (possibly arising from trauma) in the bone marrow. If the hematoma maintains a circulatory connection with the damaged vessel, an ABC results. If it is obliterated, then the lesion is a giant cell reparative granuloma. A third hypothesis was given by Biesecker *et al.*[1] where they proposed that a primary bone lesion initiates an osseous, arteriovenous malformation, and thereby creates, via its hemodynamic forces, a secondary reactive lesion of the bone. This leads to the production of an abnormal vascular component, an arteriovenous fistula, by the precursor lesion of the bone. The expansile hemodynamics of the fistula would erode and resorb adjacent bone, producing a labyrinth of vascular channels bounded by an expanding periosteal shell of bone. The osseous reparative processes would produce reactive giant cells and active stromal cells lining vascular passages surrounded by extensive fibroblastic proliferation and bone formation. Struthers and Shear[4] also partly supported the hypothesis that the ABC is secondary to a primary bone lesion and the hemodynamic theory where they believed that the ABC was a result of a secondary change in a central giant cell granuloma. They observed that central giant cell granuloma appears to have a propensity to form microcysts because of its loose, fibrillar connective tissue stroma. Localized areas of relative ischemia in the stroma lead to intercellular edema and formation of microcysts. These microcysts tend to enlarge because of further collapse of the stroma and coalesce with each other. Enlargement also occurs as a result of hemodynamic or osmotic forces. Rupture of thin-walled vessels causing hemorrhage into the microcysts occurs because of loss of stromal support around the vessels. With the expansion of these spaces, pressure resorption of the surrounding medullary bone occurs leading ultimately to endosteal resorption of the cortical plates. Once the cortical plates are reduced to only a thin shell of bone, blow-out of the lesion may occur, with its thin bony shell lifted. The concept that the ABC is a secondary phenomenon arising in a pre-existing bone lesion has gained considerable support, and there is undoubtedly good evidence to sustain it according to Hillerup and Hjorting-Hansen.[5] As early as 1940, Ewing described what appears to be an ABC and suggested that it was a benign giant cell tumor modified by communication with large blood vessels. Jaffe in 1950 proposed that the cyst may result from modification of some other lesion of bone, most of which may be destroyed by hemorrhage. Clough and Price in 1968 suggested that the cyst may be either a primary occurrence or a secondary phenomenon in both benign and malignant lesions of bone.

It produces a radiolucent area which causes an ovoid or fusiform expansion of the bone and may balloon the cortex.[8] It is usually unilocular, but others have been described as having faintly discernible septation or trabeculations, and some as being multilocular or honeycomb-like. The teeth may be displaced and root resorption has been described.[9] Radiological differentiation from other expansile jaw lesions is difficult. The ABC has been reported to evolve through

four radiologic stages: initial, active growth, stabilization, and healing.[10] In the initial phase, the lesion is characterized by a well-defined area of osteolysis with discrete elevation of the periosteum. This is followed by a growth phase in which the lesion grows rapidly with progressive destruction of bone and development of the characteristic "blown-out" radiologic appearance. The growth phase is succeeded by a period of stabilization, in which the characteristic "soap bubble appearance" develops as a result of maturation of the bony shell. Final healing results in progressive calcification and ossification, with the lesion transforming into a dense bony mass. Goaz and White[11] claim that the margins of the lesion depend on its stage of development and are generally somewhat irregular and less distinct than those of odontogenic cysts but more distinct than those of central malignancies. Regezi and Sciubba[12] also state the margins are slightly irregular. In contrast, Laskin *et al*.[13] report that the borders are usually well defined, in some cases, with an onion-skin effect at the cortex. Some investigators[14,15] claim that diagnosis of ABC from plain radiographs is difficult since the lesions have a thin cortical shell that can only be clearly shown by CT. CT is stated to image the borders and extension of the lesion into soft tissues better.[16-18]

The treatment of the ABC is determined by the nature of any associated lesion. There is no uniform treatment and management of ABC due to its varied nature. The usual treatment of choice is curettage as it is a benign lesion.[19] The failure to remove the lesion completely has been associated with a recurrence, although there has been a report of a case whereby the lesion regressed spontaneously. Some authors have also recommended supplementing curettage with cryotherapy. The defect can be filled up with bone chips prior to cryosurgery. Segmental resections are performed with immediate bone grafting if the lesions have been found to be extensive and cause functional and cosmetic deformities.[20] Radiation is not recommended as sarcomatous change has been reported in these lesions after irradiation.[21] A high recurrence rate of 53–66% has been reported for ABC in the jaws. Therefore, a close follow-up of the cases is recommended.

Depending on the size, site, and extent of the lesion, the treatment options range from curettage, enucleation, percutaneous sclerotherapy, diagnostic and therapeutic embolization, block resection and reconstruction, and systemic calcitonin therapy. The recurrence rate is fairly high, ranging from 19 to 50% after curettage and approximately 11% after resection.[22]

Considering the age of the patient and morbidity associated with the invasive surgical procedure, enucleation and curettage of the lesion was opted as the treatment of choice and a regular follow-up was maintained.

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