Case Report

A large aneurysmal bone cyst of mandible: A rare case report

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Abstract

An aneurysmal bone cyst (ABC) is a rare benign tumor-like lesion, described as an expanding osteolytic lesion consisting of blood-filled spaces of variable sizes separated by connective tissue septa. It is frequently accompanied by multiple cystic lesions due to aggressive hemodynamics with reactive bone formation and a genetic predisposition. This lesion has been classified as an atypical giant cell tumor or benign bone cyst. ABC has an incidence of 0.5% and comprises approximately around 1.5% of all non-odontogenic and non-epithelial cysts of the jaws. About 50% of the ABCs are reported in long bones and the vertebral column and only 2% have been reported to involve jaw bones. This case report gives an overview of a very large size ABC of the mandible in a 14-year-old male patient.

Keywords: Aneurysmal bone cyst, mandible, multilocular lesion, sinusoidal blood-filled spaces

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Submitted: 02-Nov-2022, **Revised:** 01-Feb-2023, **Accepted:** 16-Feb-2023, **Published:** 12-Sep-2023

INTRODUCTION

Aneurysmal bone cysts (ABC) are classified under non-neoplastic benign bone lesions, a rapidly growing, destructive benign bone tumor that was first described by Jaffe and Lichtenstein in 1942. Recent World Health Organization (WHO) classification of tumors of bone (2020) has considered ABC as a benign osteoclastic giant cell-rich neoplasm.[1] It is also classified as a pseudo cyst because of the lack of an epithelial lining. They are principally located in long bone metaphysis like femur and tibia (>50%) and spine (12–30%). The presence of these lesions in facial bones is infrequent, with a 2–12% incidence in the body. ABC accounts for 1–2% of all primary bone tumors of the craniofacial region. The lower jaw is more frequently affected than the maxilla with a proportion of 2:1 to 11:9. The body and the mandibular ramus are the frequently affected site that rarely involves condyle and coronoid process. The age of incidence of ABC is the first two decades of life with a slight sex predilection in females.[2]

The clinical signs and symptoms of these lesions are non-specific, hence may lead to difficulty in diagnosis. Occasionally, its presentation can be as a rapid expansile growing mass locally destructive that may be misdiagnosed as a highly aggressive or a malignant neoplasm. This rapid growth may be as a result of the erosion of one of the cortical plates. However, some authors consider atraumatic pathogenesis and local vascular alterations within a latent lesion as an explanation for this rapid progression of ABC. Although they are non-neoplastic lesions with possible local aggressiveness, differential diagnoses with other lesions

Access this article online

Quick Response Code:

Website: https://journals.lww.com/JPAT/

DOI: 10.4103/jomfp.jomfp_459_22

How to cite this article: Shamala S, Beeula A, Devi M, Hariganesh P. A large aneurysmal bone cyst of mandible: A rare case report. J Oral Maxillofac Pathol 2023;27:548-52.

like ameloblastoma, ossifying fibroma, Myxoma, giant cell granuloma, and sarcomas are considered.

CASE REPORT

A 14-year-old male patient reported to the outpatient department with a chief complaint of pain and swelling involving the lower left side of the face for the past 8 months [Figure 1]. The patient had no relevant medical history. Patient's past dental history reveals previous extraction of 37, curettage of the lesional area, and an incisional biopsy has also been performed in a different hospital, with a differential diagnosis of central giant cell granuloma, hyperparathyroidism, and ABC. Family history reveals that his parents and siblings were apparently normal and healthy. On clinical examination, extra orally, a diffuse swelling was seen in the left side of the face measuring about 10 × 12 cm in size extending from the ala-tragus line involving the body of the mandible, 2 cm beyond the lower border of the mandible crossing the midline. On palpation, the swelling was hard with mild tenderness, without any intraoral findings.

Orthopantomagram revealed a multilocular expansile radiolucent lesion of the left mandibular ramus and body of the mandible, with a sclerotic border and thinning out of cortices. Computed tomography mandible reported a large well-defined heterogeneous expansile osteolytic lesion involving the ramus, the angle, and the left half of the mandible with bony expansion. Significant cortical thinning and remodeling were seen in the left mandibular ramus and the lingual cortex. Cystic changes are also noted within the lesion with few internal septae resembling multi-loculation [Figures 2 and 3].

A hemi-mandibulectomy was performed and the excised specimen was submitted for histopathological reporting [Figure 4].

The specimen was grossed and the cystic lesion was approached by opening the overlying soft tissue, which revealed a cavity filled with blood clots and soft tissue and multiple bits of the specimen were processed, sectioned, and stained for histopathology reporting.

Histopathologically, the H&E stained soft tissue sections of the postoperative specimen revealed delicate fibrovascular connective tissue areas, which appear myxomatous in some areas with stellate fibroblast and fibrovascular in a few areas. The section also revealed, large sinusoidal spaces filled with Red blood cells (RBC) and numerous capillaries of varying sizes with

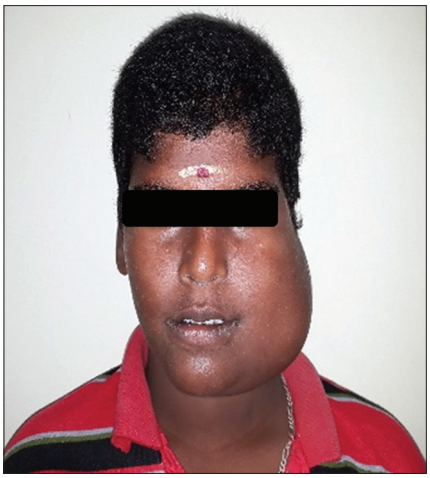

Figure 1: Unilateral diffuse swelling involving the left side of the face

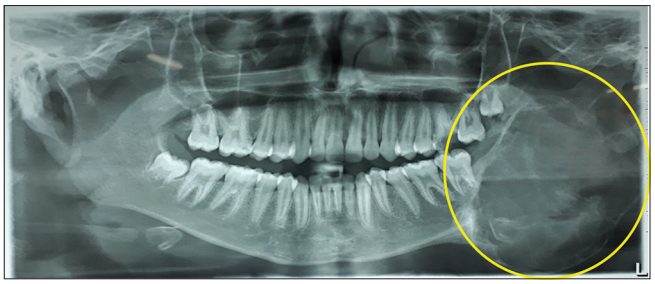

Figure 2: OPG reveals radiolucency, eccentric ballooning, and loss of cortical bone with few septations

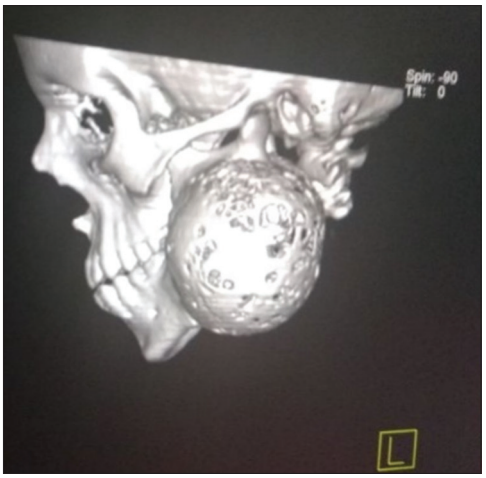

Figure 3: CT mandible—large well-defined heterogeneous expansile osteolytic lesion

peripheral hyalinization. Periphery of the sections revealed numerous bony trabeculae with osteoblastic rimming and osteocytes within lacunae resembling reactive bone [Figure 5]. One area of the section reveals numerous multinucleated giant cells within a fibro-cellular background. Most of the giant cells seem to be associated with bony trabeculae with the presence of reversal lines [Figure 6]. The entire area of connective

Figure 4: Surgically excised hemi-mandibulectomy specimen with lesion

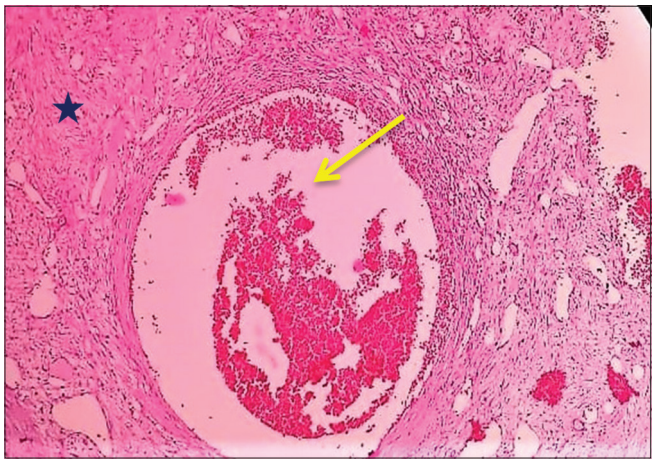

Figure 5: (10×) H&E stained section with blood-filled sinusoidal space (yellow arrow) within CT stroma

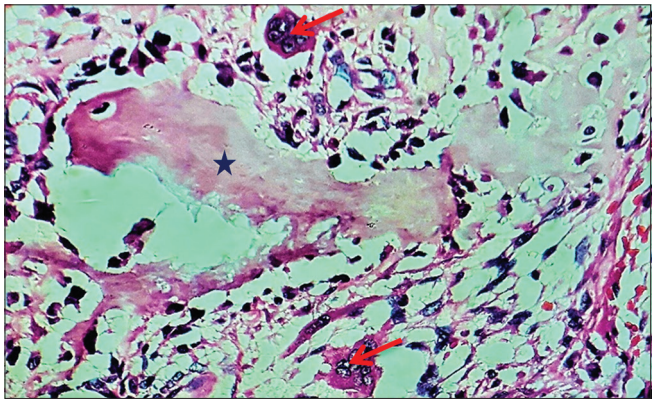

Figure 6: (10×) H&E stained section with osteoid and multinucleated giant cells (red arrow)

tissue was diffusely sprinkled with lymphocytes. Some of the capillaries seem to be thrombosed and large pools of RBCs are seen with hemosiderin pigment. On clinicopathological correlation, the lesion was histopathologically reported as ABC.

DISCUSSION

The term "aneurysmatic" refers to the "blow-out" effect or expansion of the affected bone that appears typical in ABC. Jaffe and Lichtenstein first described ABC in 1942 as a non-neoplastic blood-filled bone lesion with the presence of giant cells and bony trabeculae.[3] The WHO defines ABC as a benign intra-osseous lesion, characterized by blood-filled spaces of varying size associated with a fibroblastic stroma containing multinucleated giant cells, osteoid, and woven bone.[4,5] ABC develops mostly in maxillofacial bones depending on high venous pressure and high marrow content. It comprises 5% of all the lesions of the cranial and maxillofacial bones among which only 2% of the lesions appear in the jaws. The mandible is commonly affected when compared to the maxilla and mostly molar and ramus regions of the mandible are the frequent sites. Recent studies have suggested the pathophysiology of ABC to resemble giant cell tumors of the bone.[6,7]

The etiology of ABC is controversial. Levy *et al*.[8] have reported that the development of ABC is associated with a history of trauma and subperiosteal hematoma formation, while Tillman *et al*.[9] have reported 95 cases with no history of trauma. Jaffe and Lichenstein stated that the development of ABC can be due to increased venous pressure and repletion of the vascular bed in the transformed bone caused by the alteration of local hemodynamics related to resorption, connective tissue replacement, and osteoid formation. Hernandez *et al*.[10] classified ABC as primary and secondary. The primary could be congenital or acquired and could originate from pre-existing AV malformations. The congenital type is seen in children and young adults with no history of trauma, whereas the acquired type is found in adults with a history of trauma. The secondary type is postulated to be associated with the degeneration of pre-existing lesions such as a cyst, tumor, or fibro osseous lesion. Struthers and Shear assumed that ABC was secondary to a pre-existing lesion and that central giant cell granuloma was the most common of these lesions. Swing *et al*.[11] suggested that ABC was derived from another type of benign lesion like a giant cell tumor in which some changes have made possible the communication between the stroma and medullary vessels. When this communication is maintained, an ABC is established, whereas if the connection is interrupted, a giant cell granuloma occurs.[5,12,13]

The characteristic clinical features of ABC are slow growing, asymptomatic, expansile, inconspicuous, or as a sudden, rapidly expanding lesion with resultant bony

destruction and facial asymmetry mimicking malignancy or a false aneurysm.[12,14]

Few classification systems have been proposed according to the nature, morphological appearance and activity of ABC. In 1969, Dabska and Buraczewski classified the progression of ABC into four phases such as 1. initial phase, 2. growth phase, 3. stabilization phase, and 4. healing phase. In 1985, Capanna *et al.* [Table 1][15] classified the lesion in long bones, into five subgroups.

In 2020, the WHO classification of tumors of bone proposed three main histological components, namely, cellular, fibrillar, and osteoid components [Table 2].

These mechanisms are not exclusive to ABC and are also reported in other tumors, such as giant cell tumor and chondroblastoma, explaining the lytic component of the lesions.[1]

The radiographic features are not pathognomonic. The lesion may appear as unilocular, multilocular, soap bubble or honeycomb, or moth-eaten radiolucent patterns causing expansion, destruction of bone, perforation of the cortices, or an associated periosteal reaction with the reactive new bone formation with a peripheral sclerotic border.[3,4] Diagnosis based on only radiographic examination may be misleading due to the radiographic appearance being similar to those of other lesions such as ameloblastoma, myxoma, central giant cell granuloma, odontogenic cysts, or central hemangiomas of the bone.[16]

Table 1: Capanna's classification of lesions in long bones

Type	Description
Type I	Centrally located lesions without a distinct outline or slightly expanded outline. Mostly reported to involve the short bones of the hand and feet.
Type II	Involves the entire bone segment and is expansile with cortical thinning.
Type III	Most common type is eccentric metaphyseal lesions, which typically involve only one cortex.
Type IV	Least common type of ABC. It is sub-periosteal and grows away from the bone.
Type V	Lesions located periosteally that expand peripherally and finally penetrates the underlying cortex.

Table 2: Histological component of the lesion in the long bones (WHO)

Cellular component	Fibrillar component	Osteoid component
Cellular components including multinucleated giant cells resembling osteoclasts expressing high levels of receptor activator of nuclear kappa B (RANK) and neoplastic stromal mononuclear and myo/fibroblastic cells that express high levels of RANK ligand (RANKL).	Fibrillar component that made up of a collagenous extracellular matrix.	Osteoid component including organic bone matrix secreted by osteoblasts. Mitoses are also seen. RANKL is a tumor necrosis factor stimulating osteoclasts by binding to RANK.

Histopathologically, the "classic or vascular" form of ABC consists of many sinusoidal blood-filled spaces set in a fibrous stroma, with multinucleated giant cells, osteoid, and bone formation. Hemosiderin is also present in variable amounts.[2,17]

As it is histologically similar to the browns tumor, it is recommended to study the levels of calcium, phosphate, and enzyme alkaline phosphatase.[4]

Treatment of ABC is usually directed toward the complete removal of the lesion. The various treatment modalities are per-cutaneous sclera therapy, diagnostic and therapeutic embolization, curettage, block resection and reconstruction, radiotherapy, and systemic calcitonin therapy. Self-healing cases have also been reported on long-term follow-up.[18,19]

Panoutsakopoulos *et al.*[20] described three cases of ABC with chromosomal anomalies, involving band 16q22. It is recently identified that recurrent chromosomal translocations involving the USP6 (ubiquitin-specific protease) gene confirmed ABC and there was USP6 rearrangement present in approximately 65–70% of ABC cases, with CDH11-USP6 fusion in 30%.[1,14]

Recurrence rates range from 20 to 30% in different groups and it occurs most frequently within the first year after surgery, mainly attributed to incomplete removal of the lesion.[5,8,12]

CONCLUSION

ABC is a benign but aggressive lesion that affects predominantly children and young adults. ABC involves any bone of the skeleton frequently long bones and involvement of the jaw bone also should be considered. Though radiographic features of ABC seem to be characteristic, biopsy is mandatory to exclude other bone neoplasms prior to the treatment plan. This rare case report has been presented for its incidence in a child involving the lower jaw with an unusual size.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship
Nil.

Conflicts of interest
There are no conflicts of interest.

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