Aneurysmal Bone Cyst of Mandible: A Case Report and Review of Literature

ABSTRACT

An aneurysmal bone cyst is a rare benign expanding osteolytic lesion of bone characterized by replacement with fibro-osseous tissue containing blood filled sinusoidal or cavernous spaces of variable size. Aneurysmal bone cyst are infrequent in craniofacial skeleton and among all cystic lesions that can be found in maxilla and mandible it is rare, most commonly found in long bones and vertebral column. The development of aneurysmal bone cyst is related to history of trauma and subperiosteal hematoma formation. In this article we present a case of 40-year-old female patient with an aneurysmal bone cyst involving right angle and ramus of mandible. Computed Tomography of mandible revealed a well-defined expansile lesion in the region of the angle and ramus of right hemi mandible with thinning of bony cortex and diagnosis was confirmed by histopathological examination. In our patient, en bloc (The resection of large bulky tumor virtually without dissection) resection allowed complete removal of the lesion and reconstruction plate was placed for rehabilitation. We have focused on differential diagnosis of lesions that are found at this location and histopathological examination remains the Gold Standard in confirmatory diagnosis of such lesions.

KEYWORDS aneurysmal bone cyst, ameloblastoma, osteolytic bone neoplasm, hemi mandiblectomy

INTRODUCTION

Aneurysmal bone cyst (ABC) is rare benign osteolytic bone neoplasm characterized by several sponge like blood or serum filled generally non-endothelialized spaces of various diameters that may contain osteoid tissue and osteoclast like giant cells.1,2

The origin of term “aneurysmal bone cyst” stems from two cases reported by Jaffe and Lichtenstein in 1942 in their article1. They noted two “peculiar cysts of large size” which they described as “aneurysmal bone cyst”. The word “aneurysma” emphasized the “blown-out” distended contours of affected bones and word “bone cyst” when the lesion enters through the thin shell of bone, it appears largely as a blood-filled cavity.

ABCs are infrequent in craniofacial skeleton and only 60–70 cases have been described in head and neck region. They are most commonly found in long bones 50% and vertebral column 20%. They are found in the metaphysis of long bones mainly in the tibia and the femur. Only about 2% of ABCs occurs in the jaws. The mandible is more commonly affected than maxilla and 50% of ABCs occur in the maxilla. About 90% of cases affect posterior mandible (body of mandible 40%, ramus 30%, angle 19%, symphysis 9%, Condyle 2%). There is a greater predilection in female gender (62%) and a greater predilection to occur in the first two decades of life (80% of patients are under 20 years of age). Although ABC is benign lesion it can be locally aggressive due to rapid growth and osteolytic destruction.

ABC can be classified into three types.

Conventional or vascular type (95%) presents as rapidly growing, expansive, destructive lesion causing cortical perforation and soft tissue invasion. The solid type (5%) presents as small asymptomatic lesion first seen as radiolucency on routine radiograph.

A third form or mixed variant shows features of both vascular and solid types. In this case report, we describe a case of ABC involving right angle and ramus of mandible with its clinical findings and radiological, histopathological investigations and treatment planned for the patient. Differential
diagnosis included Ameloblastoma, Cemento-ossifying fibroma, Fibrous dysplasia and Solitary bone cyst.

CASE REPORT

A 40-year-old female patient reported to the Department of Oral Medicine and Radiology with a complaint of swelling on right posterior region of the lower jaw, accompanied by dull and intermittent pain since 11–12 month (Fig. 1). Patient had reported to the Institute for same complaint 10 months back when a clinical and radiographic diagnosis of Ameloblastoma was made based on radiographic and CT findings (Figs. 2 and 3). The patient refused to do further advised investigations and did not report back. About two months ago, she reported exfoliation of mobile right posterior tooth. She visited a local Dentist for the same, who again referred her to the Institute for further management. The swelling in the same region had increased slowly to its present size with mild and continuous pain. The patient gave a history of extraction of lower right anterior teeth 3 months ago. Medical history, family history, habit history and social history were unremarkable.

Clinical examination revealed swelling on the lower right side of the face resulting in facial asymmetry. Extra orally a single well defined, oval swelling was present extending antero-posteriorly approximately 7 cm from midline of mandible and supero-inferiorly extending 5 cm from the right outer canthus of the eye to 2 cm below right inferior border of mandible (Fig. 4). The skin over swelling was normal. The swelling was approximately 4 cm × 4 cm in its maximum dimensions. On palpation swelling was oval in shape, well defined with smooth surface, bony hard consistency and tenderness was present, temperature was normal. It was non-compressible, non-fluctuant and non-pulsatile in nature. Temporo-mandibular joint and mandibular movements were normal.

Intra-oral examination showed missing 48 and 43. A diffuse swelling in relation to right first and second molar with vestibular obliteration and the expansion of buccal and lingual cortical plates with intact mucosal tissue was evident. There was no sinus opening or any discharge present. Clinical diagnosis of Ameloblastoma was given and a differential diagnosis of ossifying fibroma or Cemento-ossifying fibroma was given.

In radiographic investigations orthopantomogram and CT mandible were performed. Conventional radiographs and CT mandible showed a transition from a unilocular radiolucent lesion at first visit to a predominantly multilocular lesion at the second visit.
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OPG (Fig. 5) showed a single, round, well-defined multilocular lesion showing expansion of inferior, posterior border of mandible. Axial CT scan (Fig. 6) revealed a well-defined non-enhancing expansile lesion at angle & ramus of right hemi-mandible measuring 3.2 × 3.3 cm in maximal axial dimension. Coronal CT Scan (Fig. 7) showed the lesion to be measuring 3.7 cm in height, with expansion and thinning of both the buccal and lingual bony cortices with irregular breaks at many places. 3D reconstructed CT images (Fig. 8) show relatively well defined, round to oval, mass at the right body and the angle on the mandible region with surface showing the discrete irregular appearance and extending up to the ramus region. Radiographic impression was most likely suggestive of an aggressive lesion like Ameloblastoma.

So considering clinical presentation and radiographic features, the Diagnosis of Ameloblastoma affecting the R hemi-mandible involving the body, angle and ramus was made. Differential diagnosis considered were Odontogenic myxoma, central hemangioma, aneurysmal bone cyst, Odontogenic keratocyst and Traumatic bone cyst. Routine hematological investigations were within normal limits.

Surgical treatment was planned. It consisted of en bloc resection of right mandible from distal of 45 till the base of right coronoid process and condyle 46 was extracted during the surgery (Fig. 9a). Primary reconstruction of the defect was carried out with titanium reconstruction plate. This can be appreciated in the post-surgical OPG (Fig. 10).

The excised specimen was sent for histopathological examination. Before making sections, gross examination of the resected mandibular segment with the tumor mass was done. The specimen shows the presence of well circumscribed, bony hard tumor mass measuring 3 × 3 × 1 cm. External surface shows bluish discoloration. Also, it shows the presence of single tooth (47) along the alveolar margin. The superior alveolar bone appears uninvolved and the inferior alveolar bone shows tumor mass (Fig. 9a). On cut section, it shows hemorrhagic and necrotic areas (Fig. 9b). Histopathological sections from the excised specimen under 10 × (Fig. 11a) and 40 × (Fig. 11b) magnification revealed many dilated blood vessels containing blood and at places showing bone and osteoid tissue. There was no evidence of malignancy. The histologic picture was consistent with ABC. Based on histopathological findings, the final diagnosis of aneurysmal bone cyst of the right body and angle of the mandible was made. The patient was in regular follow up for next 1 year and there was no evidence of any residual lesion and recurrence.

DISCUSSION

The ABC as a distinct pathogenic entity was first described in literature by Jaffe and Lichtenstein3,9. Jaffe postulated...
that aneurysmal bone cyst may be secondary phenomenon due to hemorrhagic “blown-out” in a preexisting lesion which may be destroyed in process. Lichtenstein also suggested a vascular origin but postulated that the lesion was the result of “local circulatory disturbance”. It could be thrombosis of a sizable vein or perhaps an arterio-venous communication. The term “aneurysmatic” refers to the “blown-out” or expansion of the affected bone that appears in this type of lesions.
The majority, approximately 80% of patients presenting with ABC are less than 20 years old. More than half of all such lesions occur in long bones and 12–30% cases occur in the spine. The pelvis accounts for about half of all flat bone lesions. They are rare in the jaw and mandible is affected twice as frequently as maxilla. ABC accounts for 1.5% of non-odontogenic, non-epithelial cysts of mandible. Familial incidence of ABC have also been reported in literature. Investigators have proposed two forms of ABC: “primary aneurysmal bone cyst” in which no pre-existing lesion is identified and those in which the lesions is secondary to identifiable precursor. The most common of these are giant cell tumor that accounts 19–39% and other common precursor lesions include fibrous dysplasia, non-ossifying fibroma, solitary bone cyst, eosinophilic granuloma and osteosarcoma.

**Histologically, it can be of three variants**

1) Conventional or Vascular type (95%): Osteolytic lesion with blood filled cavities and sinusoidal spaces, separated by fibrous connective tissue septae with osteoid trabecular. Hemosiderin and giant cells can be found.

2) The solid type (5%): This form is non-cystic variant with osteoclast like giant cells. Osteoblastic differentiation areas with osteoid and calcifying fibromyxoid tissue seen.

3) A mixed variant shows feature of both vascular and solid type. It may be a transitory phase of the lesion because sudden activation or rapid enlargement of stable lesion is reported.

The natural history of aneurysmal bone cyst evolves through four radiologic stages; Initial phase, Active phase, Stabilization phase and Healing phase. In the initial phase the lesion is characterized by a well-defined area of osteolysis with a discrete peristeme. This is followed by growth phase in which lesion grows rapidly with progressive destruction of bone and development of "blown-out" radiologic appearance. The growth phase is succeeded by a period of stabilization in which characteristic "soap-bubble appearance" develops as a result of maturation of bony shell. Final healing results in progressive calcification and ossification with the lesion transformed into dense bony mass.

Clinically, there is firm, non-tender to slightly tender swelling which enlarges progressively expanding and perforating bony cortex and displacing teeth. A history of rapid growth is elicited as a result of erosion of cortical plate. The overlying mucosa remains normal.

The radiological features of ABC in jaws are variable. Lesions are radiolucent in most cases but can be radiopaque or mixed. The multilocular effect gives this cyst characteristic "honeycomb" and "soap-bubble" like appearance. Destruction or perforation of cortex along with subperiosteal reaction display "sun-ray" or "Moth-eaten appearance." In our case, the ABC represented a well-defined, unilocular, expansile lesion, with thinning of bony cortex and irregular breaks at many places.

Diagnosis based on only radiographic examination may be misleading as lesions such as Ameloblastoma, myxoma, central giant cell granuloma, central hemangiomma of bone has the similar radiographic appearance. Histopathological examination remains gold standard in diagnosis of ABC.

Treatment of ABC is directed towards surgical excision and curettage of cavity, but it is difficult many times since the lesions are multicellular and divided by septae. Recently embolotherapy has been used to limit blood loss at surgery. When surgery is not possible the other treatment modalities considered are sclerotherapy, en bloc resection and reconstruction, systemic calcitonin therapy. Immediate reconstruction of defect with autogenous grafts is recommended when suspected high risk of fracture and loss of mandibular continuity is there. The radiotherapy is not recommended because of possibility of radiation induced tumors.

The recurrence rate varies from 20 to 50% with simple curettage and most frequent within the first year after surgery. Insufficient curetage or excision of the lesion, especially in soft tissue invasive cases leads to recurrence. The present case was treated by en bloc resection followed by reconstruction with titanium reconstruction plate. There was no evidence of recurrence after 1 year of follow up.

**CONCLUSION**

The clinical presentation and radiologic appearance of ABC is extremely variable resembling many other lesions, and histopathological analysis is must for the diagnosis.

**CONFLICT OF INTERESTS**

The authors declare that they have no conflicts of interest.

**FINANCIAL DISCLOSURE**

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**REFERENCES**

