Giant aneurysmal bone cyst of mandible: A case report and literature review
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Abstract
Aneurysmal bone cysts are infrequent in the craniofacial region. These cysts are non-neoplastic, expansile, and osteolytic in nature. Since these cysts are not lined by epithelium, they are considered as pseudocysts. Clinicopathological features and biological behavior of these cysts are enigmatic as they mimic various other pathogenetic lesions and two third of these cysts are associated with other bone pathology. Aim of the present paper was to report a case of aneurysmal bone cyst in 40-year-old female patient encountered in left mandibular posterior region which was surgically treated with hemimandibulectomy. The post-operative period was uneventful. Furthermore, emphasis was given to discuss its etiopathogenesis, clinicopathological variants, and other pathologies associated with this cyst.

Keywords
Aneurysmal bone cyst, denosumab, giant cells, mandible, pseudocyst

Introduction
Aneurysmal bone cyst (ABC) was initially recognized in 1893 by Van Arsdale and it was described as ossifying hematoma. ABC was initially identified and explained by Jaffe and Lichtenstein in metaphyseal region of long bones and vertebrae. Similarly, the first case of ABC in the jaw was recognized by Bernier and Bhaskar.¹ The World Health Organization defines ABC as “a benign tumor-like lesion with an expanding osteolytic lesion, consisting of blood filled spaces of varying size separate by connective tissue septa containing trabeculae or osteoid tissue and osteoclast giant cells.” The term “aneurysmal” defines the blown-out distention of the bone and “bony cyst” indicates thin shell of bone appearing as a blood filled cavity.²

Occurrence of ABC is more common in long bones accounting for 50% which is followed by vertebral column which is estimated to be 20%. Only 2% of cases have reported to occur in the craniofacial region, with predominance toward mandible. It inflicts young adults under the age of 20 years with slight female predilection.¹ ABC exists in two clinicopathological forms; Primary and secondary. Secondary lesions are arising from preexisting conditions such as fibrous dysplasia, giant cell granuloma, cementifying fibroma, and other unspecified lesions. Histopathologically, ABC can be classified into three variants; vascular, solid, and mixed type.³

The aim of the present case report was to discuss the clinicopathological aspects of ABC in 40 years female patient. Furthermore, attempts are made to explain the biological behavior and treatment protocol.

Case Report
A 40 years female reported with a chief complaint of swelling in the left lower back region of the jaw since 2 years. According to the patient’s history, swelling started with a size of small nodule of approximately 2 cm × 2 cm and has achieved the present size. Medical and family history appeared to be normal. Patient gave the history of trauma in domestic violence to the left mandibular area. Trauma was followed by post traumatic pain and swelling, which gradually subsided. On extra-oral
examination, facial asymmetry with increased volume on the left side of the face was noticed. Swelling appeared to be diffuse, extending anterior-posteriorly from body of the mandible which is 2 cm short of symphysis to the angle of the mandible including coronoid process, condylar process was spared. Superior-inferiorly it extends from the lower border of zygomatic arch to the base of the mandible. Skin over the swelling was normal and stretched [Figure 1]. On palpation, swelling was hard in consistency and skin over the swelling did not show increased temperature. There was no murmur or bruit noted on auscultation.

Intra-orally small nodular elevation was noted measuring about 2 cm × 2 cm, with the obliteration of the vestibule [Figure 2]. All molars on left side of the mandible were extracted because of the increased mobility due to trauma. On palpation, the swelling was nontender and firm without the rise of temperature.

Orthopanomograph showed a lytic and expansile lesion demonstrating "soap bubble" appearance with ill-defined lower border of the mandible. Buccal cortical plate was thinned out with complete destruction on the lingual side [Figure 3]. Based on the clinico-radiographic features, a provisional diagnosis of the aneurysmal bone cyst was noted and the differential diagnosis was listed which included ameloblastoma, giant cell granuloma, myxoma, desmoplastic fibroma, hemangioma. However, ABC was strongly considered based on the history of trauma, absence of bruits, thrill and pulse pressure. Furthermore, aspiration with needle yielded darkish red colored fluid which was in favor of vascular lesion. Incisional biopsy was recommended for the confirmation of the diagnosis. Biopsy specimen showed small blood filled cystic areas. Histopathological features revealed delicate connective tissue stroma consisting of predominant blood filled cavernous or sinusoidal spaces [Figure 4]. Abundant multinucleated giant cells were seen [Figure 5]. Stroma also consisted of numerous small endothelial lined blood vessels, inflammatory cells, and extravasated red blood cells. Based on all these features, a final diagnosis of the aneurysmal bone cyst was established.

Considering the severity and extent of the lesion, hemimandibulectomy was performed with the preservation of condyle for further reconstructive procedures. Post-operative period was uneventful and wounds healed normally.

Figure 1: Preoperative extra-oral view revealed swelling on left side of the face

Figure 2: Preoperative intra-oral view revealed nodular elevation with the obliteration of the vestibule

Figure 3: Preoperative orthopanomograph showed a lytic and expansile lesion demonstrating "soap bubble" appearance with ill-defined lower border of mandible including coronoid process on left side. Condylar process was spared

Figure 4: Photomicrograph revealed numerous blood filled cavernous spaces devoid of endothelial cells within the immature connective tissue (H/E, ×10)
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Challenged the concept of reactive nature of ABC and demonstrated through cytogenetic studies that primary ABCs exhibit chromosomal translocation t (16;17) (q22;p13). Based on these findings, it can be assessed as gene translocation leading to primary ABCs and pre-existing lesion transform into ABCs. Present study is considered under secondary type as there is a history of trauma before the initiation of the lesion.

Peak incidence of occurrence has found out to be in the second decade of life with a slight predilection for females. Considering the craniofacial region, mandible is most commonly affected. ABC produces firm swelling which is rarely associated with pain. Enlargement of swelling appears to be rapid with the mobility of associated tooth, resorption of root, and malocclusion. In contrast to the above findings, present case evolved slowly and there is no evidence of malocclusion. In most of the occasion, enlargement is so severe that only thin cortical plates or periosteum remains which causes "egg-shell" cracking. Clinical diagnosis of ABC is difficult as it share similar findings with lesions such as traumatic bone cyst, giant cell tumor, hyperparathyroidism, fibrous dysplasia, desmoplastic fibroma, fibrous histiocytoma, hemangioma, osteogenic sarcoma, etc. Aspiration of blood from the lesion is evident of some vascular lesion or ABCs. The absence of bruits, thrills and pulse pressure aid in clinically differentiating ABCs from other vascular lesions.

ABC evolves through four radiographic stages; initial, active growth, stabilization, and healing. Initial phase is characterized by well-defined areas of osteolysis with a minute elevation of periosteum. Initial phase is followed by "active growth" phase which shows rapid destruction of bone with the development of characteristic "blown-out" radiographic pattern. The 3rd phase is the period of stabilization characterized by the maturation of bony shell which has a characteristic "soap-bubble" appearance. Final stage is the healing phase characterized by progressive mineralization and ossification resulting in the transformation into dense bony mass. Present case showed third phase of radiographic appearance which was characterized by "soap-bubble" appearance.

One study has reported both fine needle aspiration cytology and incisional biopsy are incapable of achieving the final diagnosis. This study also documented macroscopic examination of the entire surgical specimen is essential for achieving the accurate diagnosis. It is also mandatory to examine the whole tissue to exclude the relationship with other pre-existing lesion. Aspiration of a dark red or brownish hemorrhagic fluid favors the diagnosis of ABC. Microscopically, the lesion show numerous blood filled cavernous or sinusoidal spaces within the loose connective tissue. These sinusoidal areas are devoid of endothelial cells. Predominant populations of multinucleated giant cells are seen. Stroma consists of small endothelial lined blood vessels, inflammatory cells, and fibroblasts. Connective tissue showed evidence of trabeculae of reactive and woven bone. Histopathological features of the present case are consistent with above-mentioned features.

Literature review revealed best treatment options include curettage, enucleation, sclerotherapy, diagnostic and therapeutic embolization, block resection, reconstruction, and systemic calcitomin therapy. All the treatment protocols depend on the site, size, and extent of the lesion. Denosumab is been recently implemented in the treatment of ABC, which reduces...
the formation of osteoclastic giant cells by selectively binding to the Rankl receptor.\(^9\) About 26-56% of ABC tends to reoccur within the 1\(^{st}\) year of surgical treatment. Reoccurrence is mainly attributed to the inadequate removal of the tumor due to under exposure of the surgical field or difficulty in accessing the extension of the tumor. In case of esthetic deformity, high risk of fracture and loss of mandibular continuity few authors advice immediate reconstruction of the defect with the use of autogenous graft.\(^4\) In the present case, due to severity of the lesion and total destruction of the left body of the mandible, hemimandibulectomy was carried out. Patient is under regular follow up for a year after surgical treatment. There is no evidence of reoccurrence.

**Conclusion**

ABC is considered as a pseudocyst which is categorized under the reactive lesion. However, its clinicopathological features, etiopathogenesis, and biological behavior remain enigmatic. ABC mimics various other lesions clinicopathologically which mandates accurate diagnostic protocol for suitable treatment. Further, research is required to assess the pathogenesis and biological behavior which is essential for the treatment and prognosis.

**References**
