

Aneurysmal Bone Cyst Plus In an 8 Year Old Female, A Case Report.

¹Swapnil Moghe, ²Nitin Saini, ³Ajay Pillai, ⁴Anjali Moghe, ⁵Kavita Pillai

¹(Assistant Professor, Department of Oral & Maxillofacial Surgery, Peoples Dental Academy, Bhopal, India)

²(Assistant Professor, Department of Oral & Maxillofacial Pathology, D.A.V (C) Dental college, Yamunanagar, India)

³(Associate Professor, Department of Oral & Maxillofacial Surgery, Peoples Dental Academy, Bhopal, India)

⁴(Lecturer, Department of Oral Medicine & Radiology, Peoples Dental Academy, Bhopal, India)

⁵(Dental Surgeon, Bhopal, India)

Summary: Aneurysmal bone cyst (ABC) is rare benign lesions of bone which are infrequent in craniofacial skeleton. ABC's are characterized by rapid growth pattern with resultant bony expansion and facial asymmetry. We describe a case of ABC in a 08 year old Female patient affecting the right side of the anterior maxilla with expansion and thinning of the buccal & palatal cortical plates. Treatment consisted of surgical curettage of the lesion. A one year follow-up showed restoration of facial symmetry and complete healing of the involved site.

I. Background

Aneurysmal bone cyst (ABC) has been recognized since 1893 when it was described as an ossifying hematoma by Van Arsdale¹. Jaffe and Lichtenstein were the first to recognize ABC as an intraosseous, osteolytic lesion, chiefly affecting the metaphyseal region of long bones and vertebrae. Bernier and Bhaskar described the first case of ABC in the jaws in 1958^{2,3}. ABC is a benign cystic lesion of bone, composed of blood-filled spaces separated by connective tissue septa containing fibroblasts, osteoclast-type giant cells and reactive woven bone⁴. Fifty percent of ABCs arise in the long bones and 20% in the vertebral column. It accounts for 1.5% of the non odontogenic, nonepithelial cysts of the mandible^{1,5}. It is found more frequently in the mandible than the maxilla (3:1) with preponderance for the body, ramus and angle of the mandible. It affects young persons under 20 years of age with no gender predilection^{5,6}.

ABC can be classified into three types. Conventional or vascular type (95%) manifests as a rapidly growing, expansive, destructive lesion causing cortical perforation and soft tissue invasion. The solid type (5%) may present as a small asymptomatic lesion first noticed as radiolucency on a routine radiograph or as a small swelling^{7,8}. A third form or mixed variant demonstrates features of both the vascular and solid types. It may be a transitory phase of the lesion because sudden activation or rapid enlargement of stable lesions has been reported⁸.

II. Case Presentation

A 08 -year-old female patient reported to the Department of Oral and Maxillofacial Surgery with a complaint of an asymptomatic swelling in the right upper front teeth region since 2 months, which had gradually increased to the present size. Her medical and family history was unremarkable and there was a history of trauma. On extraoral examination, facial asymmetry was apparent with a diffuse swelling involving the right nasomaxillary region, measuring approximately 4 × 5 cm as seen in figure 1. The swelling was firm and nontender. Intraoral examination revealed a diffuse swelling in relation to right front teeth with vestibular obliteration as seen in figure 2 & 3. Egg shell crackling was present with partial extrusion of 11. Bluish swelling on anterior hard palate on right side was seen. On aspiration, blood-tinged fluid was obtained and electrical pulp testing showed that the involved teeth were non vital.

Orthopantomogram, occlusal radiograph of maxilla & intraoral periapical radiograph shows well defined unilocular radiolucent area involving 11,12,53,54 & erupting 14. Displacement of involved teeth is seen. Pathological root resorption is seen i.r.t 53 as seen in figure 4.

The surgical exposure of the lesion is shown in figure 5. The cystic content was excised in toto, along with extraction of offending tooth. Soft tissue specimen brownish in color, measuring about 2.8 x 2.6 x 1.4 cms, was firm in consistency. Cut surface shows micro- cavities in centre filled with brownish fluid. Tissue is grossly encapsulated as seen in figure 6 & 7.

H & E stained section shows large blood filled cystic spaces of varying sizes, surrounded by connective tissue comprising of osteogenic & spindle shaped fibrogenic flattened mesenchymal cells. Cellular osteoid &

many multinucleated osteoclast like giant cells are seen in a predominantly cellular stroma. Focal areas showing myxoid changes & erythrocytes spilled in the connective tissue as seen in figure 8.

III. Investigations

1. Orthopantomogram
2. IOPA
3. Occlusal
4. Complete blood count (CBC)
5. Incisional biopsy done.

IV. Differential Diagnosis

Differential diagnosis included eosinophilic granuloma, giant cell tumor, nonossifying fibroma, unicameral bone cyst, fibrous dysplasia, chondroblastoma, chondrosarcoma, chondromyxoid fibroma, Ewing's tumor, and, in older patients, metastatic carcinoma or myeloma. The lesions are often eccentric and irregular in structure and sometimes show calcification in the central areas. As a rule, the cortex is thin, but there is rarely a cortical defect or a soft tissue mass.

V. Treatment

Treatment of ABC is usually directed toward complete removal of the lesion. This may prove difficult at times since the lesions are often multilocular and may be divided by multiple bony septae³. The treatment modalities are percutaneous sclerotherapy, diagnostic and therapeutic embolization, curettage, block resection and reconstruction, radiotherapy and systemic calcitonin therapy⁵. Self-healing cases have also been reported on long-term follow-up¹⁷. Several authors recommend immediate reconstruction of the defect with autogenous grafts in cases of esthetic deformity, high risk of fractures and loss of mandibular continuity^{1,6,15}. Simple curettage is associated with high recurrence rates varying from 21 to 50%. But Motamedi *etal*³ have reported that initial resection is not necessary and have not noted any recurrences following surgical curettage of mandibular lesions. The present case was treated by curettage and regularly monitored. There was no evidence of any residual lesion after 1 year of follow-up.

VI. Discussion

The term "aneurysmatic" refers to the "blow-out" effect or expansion of the affected bone that appears in these types of lesions⁹. The ABC of the jaw is a pseudocyst lacking epithelial lining^{6,9}. It comprises 5% of all the lesions of the cranial and maxillofacial bones³ and is most common in those regions of the skeleton where there is both a relatively high venous and marrow content. This explains the rarity of ABC in the skull bones, in which there is low venous pressure⁵.

The etiology of ABC is controversial. According to Steiner and Kantor, the ABC can develop as either a primary or secondary lesion associated with other bone diseases. Levy *et al.* had proposed that a history of trauma and subperiosteal hematoma formation is an essential factor in the development of ABC. Struthers and Shear have also concluded that ABC can occur as a secondary phenomenon in a pre-existing lesion and that central giant cell granuloma appears to be the most common of these lesions¹. Tillman *et al.* have reported 95 cases with no history of trauma⁸. In the present case also, there was no history of trauma. Jaffe and Lichtenstein refer to alterations in local hemodynamics causing increased venous pressures and engorgement of the vascular bed in the transformed bone, leading to resorption, connective tissue replacement and osteoid formation^{1,8}. Hernandez *et al.* classified ABC as primary and secondary. 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 degeneration of pre-existing lesions such as a cyst, tumor or fibrous lesion¹. The two lesions could exist independently. Hence, ABC is considered as nonneoplastic, fibrodysplastic, noncystic bone entity^{1,10}. In the present case as no history of trauma was reported, the etiology could be either due to alterations in local hemodynamics or degeneration of any pre-existing lesion at the involved site. Panoutsakopoulos *et al.* had described three cases of ABC with chromosomal anomalies, involving band 16q22¹¹. Familial incidence of ABC has also been reported in literature^{12,13,14}.

ABCs are most commonly found in long bones and vertebral column; 1.9% are reported to occur in jaws. An unusual location for ABC, i.e., mandibular condyle and coronoid process has also been reported^{8,9,15}. ABC is extremely variable in clinical presentation, ranging from a small, indolent, asymptomatic lesion to rapidly growing, expansile, destructive lesion causing pain, swelling, deformity, neurologic symptoms, pathologic fracture and perforation of the cortex³.

The radiological features of ABC in the jaws are quite conflicting; the bone is expanded, appears cystic resembling a honeycomb or soap bubble and is eccentrically ballooned. There may be destruction or perforation of the cortex and a periosteal reaction may be evident⁶. It may appear radiolucent, radiopaque or mixed. In our case, a unilocular radiolucency causing expansion of the cortical plates and thinning of the palatal border of the maxilla with root resorption of the involved teeth was present. The diagnosis based on radiographic appearance is impossible because there are other lesions having similar radiographic appearance, such as ameloblastoma, myxoma, central giant cell granuloma, odontogenic cysts or central hemangiomas of the bone¹⁶.

Histologically, ABC consists of many sinusoidal blood-filled spaces set in a fibrous stroma, with multinucleated giant cells and osteoid. Hemosiderin is present in variable amounts and there is evidence of osteoid and bone formation. This description is characteristic of the "classic or vascular" form⁹. The histologic features in our case were consistent with the above-mentioned features. Solid form is the other histological type, which is a noncystic variant with solid gray-white tissue, hemorrhagic foci and abundant fibroblastic and fibrohistiocytic elements with osteoclast-like giant cells, osteoblastic differentiation areas with osteoid and calcifying fibromyxoid tissue. The mixed form demonstrates elements of both vascular and solid types⁹.

There was no evidence of any residual lesion after 1 year of follow-up.

VII. Conclusion

As the radiological features of ABC are varied, resembling many lesions, histopathologic analysis is a must for the diagnosis.

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Learning Points/Take Home Messages

1. The radiological features of ABC in the jaws are quite conflicting; the bone is expanded, appears cystic resembling a honeycomb or soap bubble and is eccentrically ballooned. Hence ABC should be correlated histopathologically.
2. The use of histological markers is useful in reaching to a diagnosis of ABC.

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Figure 1: Diffuse swelling involving right side of face



Figure 2: Vestibular obliteration on buccal aspect.



Figure 3: Vestibular obliteration on palatal aspect.



Figure 4: OPG Shows pathological root resorption with 53.



Figure 5: Surgical exposure of the lesion.



Figure 6: Soft tissue specimen brownish in color, measuring about 2.8 x 2.6 x 1.4 cms.



Figure 7: Cut surface shows micro- cavities in centre filled with brownish fluid.

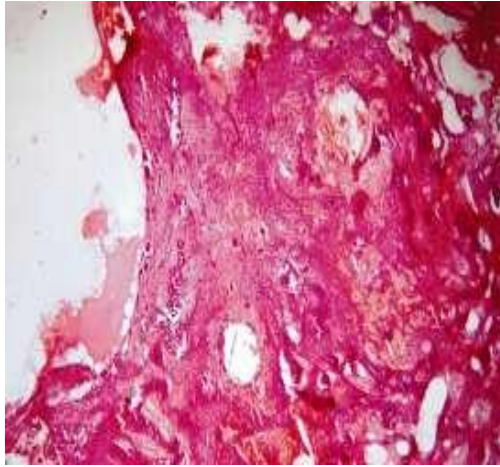


Figure 8: Focal areas showing myxoid changes & erythrocytes spilled in the connective tissue.