

ANEURYSMAL BONE CYST OF MAXILLA- A RARE CASE REPORT AND REVIEW

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Abstract

An Aneurysmal bone cyst (ABC) is a rare benign lesion seen as a locally destructive, rapidly expansile, mostly affecting long bones and vertebrae. The association of ABC with Cemento-ossifying fibroma (COF) is extremely rare with only few cases reported so far in maxilla. We are herewith reporting one such case of hybrid lesion in a 15-year old boy, who came with a solitary swelling of right maxilla and showing partial obliteration of buccal vestibular sulcus and paraesthesia over right cheek region. Coronal Computed tomography (CT) of Para nasal sinus (PNS) view revealed large expansile lesion with thinning of cortex in right maxillary sinus involving hard palate and right alveolar process. Histological features were confirmative of the diagnosis of Aneurysmal Bone Cyst of Maxilla secondary to Cemento-ossifying fibroma. Treatment consisted of the enucleation of the lesion with surgical curettage through an intraoral approach under general anaesthesia.

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Introduction

The World Health Organisation defines an Aneurysmal bone cyst (ABC) as " a benign tumour like lesion with an expanding osteolytic nature, consisting of blood filled spaces of variable sizes separated by connective tissue septa containing trabeculae or osteoid tissue and osteoclastic giant cells.^{1, 2} Bernier and Bhaskar were the first to report ABC in an oral site. In 1950, Jaffé and Lichtenstein used the term

"aneurysmal bone cyst" for the first time in literature.^{3, 4, 5, 6}

The pathogenesis of ABC is still in doubt. Most of these cysts are considered as congenital primary lesions that may coexist with other osseous pathologies. Jaffé and Lichtenstein have suggested that development of ABC may be due to hemodynamic alterations or an arteriovenous malformation leading to resorption, connective tissue replacement and osteoid formation or subperiosteal intraosseous hematoma.^{7, 8} Other different theory considers that ABC's are secondary lesions related to degeneration of a pre-existing bone lesion such as central giant cell granuloma, fibrous dysplasia, ossifying or cementifying fibromas histiosarcoma, hemangioendothelioma, fractures and trauma.^{9, 10}

The Radiological features of ABC are not pathognomonic. The lesion may appear as a cystic, multilocular, soap bubble or honey comb or moth eaten radiolucency causing expansion or destruction of the bony cortices. Histologically, the ABC consists of fibrous connective tissue

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stroma with numerous blood filled caverns or sinusoids, which may undergo thrombosis. Hemosiderin is present in variable amounts and osteoid and reactive bone formation is variable.¹⁰

Cemento-ossifying fibroma is a well demarcated fibro-osseous lesion composed of fibrocellular tissue and mineralized material of varying appearances, including mineralizing spherules named cementicles. The lesion is well encapsulated and typically may be surgically shelled out from the surrounding bone with ease. It can occur in both the jaws, but more frequently occurs in mandible, arising from the periodontal ligament affecting tooth bearing areas of the jaws. The early tumour often manifests as a well-circumscribed radiolucent lesion, and it is almost impossible to be distinguished from fibrous dysplasia. With the maturing of the tumour, the more intensive forming of spherules resembling cementum are present, and radiological image shows calcifications within the osteolytic lesion. Old lesions can be purely radiopaque.¹¹

Herein we report a rare case of Hybrid lesion comprising of ABC of the maxilla with extensive local involvement and bony destruction, arising from cemento ossifying fibroma that was treated by enucleation with surgical curettage.

Case Report

A 15-year old boy presented with a solitary diffuse swelling of the right cheek region for seven months secondary to trauma. Mild, continuous dull pain was associated with swelling, subsided on using analgesics in two months, but there was an increase in size of swelling to present size in seven months. On extra oral examination, there was about 5 x 6.5 cm in size swelling over right cheek region, asymptomatic with bowing of right zygoma and obliteration of right nasolabial fold. [Figure-1] Swelling was hard and associated with paraesthesia over right cheek region, with no difficulty in breathing and eye sight problems.

Intraoral Examination showed diffuse solitary swelling occupying right maxilla associated with obliteration of right buccal vestibular region extending from upper right lateral incisor to upper right maxillary second molar regions. Swelling was hard and asymptomatic with no difficulty in mouth opening. No evidence of associated teeth tenderness and no disturbance in occlusion and no mobility.

Spacing was observed between upper right incisors and canine teeth regions. Swelling extended palatally occupying right side of hard palate up to the midline [Figure-2]. Considering the above, provisional diagnosis was given as fibrous dysplasia involving right maxilla.



Figure -1.



Figure -1, 2. Pre-operative photographs showing swelling over right cheek region and hard palate up to the midline.

Pulp vitality test for associated teeth was performed which showed that upper right second premolar and first molar teeth were nonvital and rest all the teeth were vital. Digital Orthopantomograph [OPG] revealed large radiolucency extending from upper right central incisor to second molar teeth region and occupying right maxillary sinus and right nasal fossa without break in infraorbital rim. [Figure-3] Fine needle aspiration cytology [FNAC] showed numerous red blood cells [RBCs] suggestive of hemorrhagic fluid. The blood chemistry and hematologic profiles were within normal limits. Coronal CT of P.N.S view revealed large rounded expansile lesion of bone with thinning of cortex without the destruction of internal wall of right maxillary sinus involving hard palate and

right alveolar process with no evidence of calcifications. [Figure- 4]



Figure-3. Digital Orthopantomograph [OPG] showing well defined large radiolucency with displacement of teeth 14, 15, scalloping borders occupying right maxilla



Figure-4. Coronal Computed Tomography [CT] of PNS view showing large radiolucency with thinning of cortex in right maxilla.

The clinical presentation and the radiographic appearance of this lesion could have been associated with, fibrous dysplasia, central ossifying fibroma, Traumatic bone cyst, ameloblastoma, Aneurysmal bone cyst, myxoma or central giant cell granuloma. Incisional biopsy of the lesion at upper right premolar and first molar buccal vestibular region was performed to confirm the nature and diagnosis of the lesion. Histological features showed sinusoidal spaces filled with red blood cells and lined by fibro cellular connective tissue, surrounded by many mature bony trabeculae enclosing fibro cellular and vascular marrow tissue. Focal thick areas of the connective tissue capsule showed collections of multi nucleated giant cells and plump ovoid cells. Based upon these features, diagnosis was

suggestive of Aneurysmal bone cyst involving right hard palate and maxillary sinus. [Figure-5]

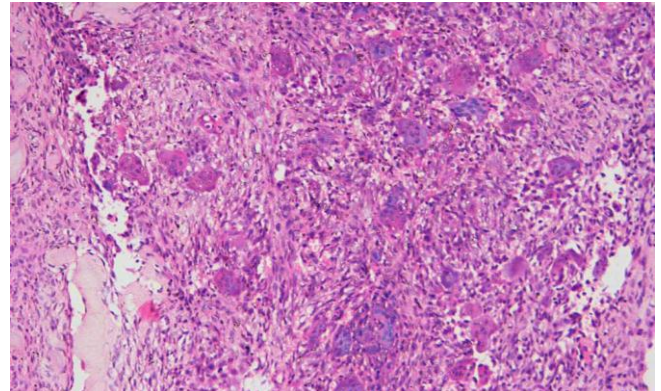


Figure- 5. Microphotograph showing vascular spaces separated by septa containing multinucleated giant cells and fibroblasts (H&E 250×).

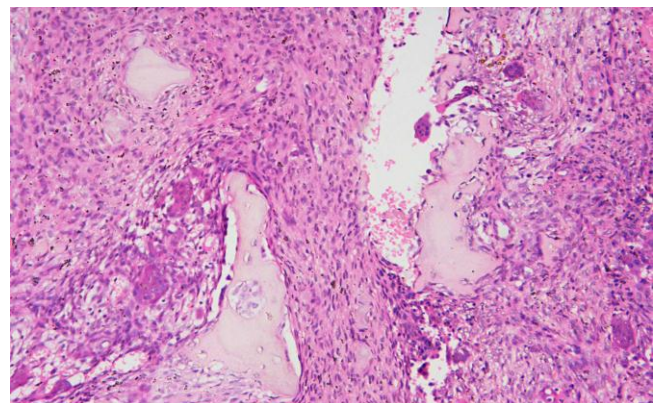


Figure-6. Photomicrograph showing bony trabeculae and layers of cementum-like calcifications embedded in thin fibrous tissue (H&E stained, 4×)

Surgical curettage with enucleation was performed under General anaesthesia with incision at right maxillary buccal vestibular region and right maxillary third molar was removed intraoperatively. Cyst enucleation and chemical cauterization was done and the cavity was packed with white head varnish gauze. The excised lesion was sent for histopathological examination that revealed presence of highly cellular connective tissue with many spindle cells with vesicular nuclei intermingled with clusters of multinucleated giant cells and many trabeculae of osteoid and woven bone and few acellular cementum admixed with sinusoidal spaces [Figure- 6] containing RBCs with increased vascularity. The diagnosis was confirmed as

Aneurysmal bone cyst secondary to cemento-ossifying fibroma.

Patient was under regular follow up for cavity dressings postoperatively. Postoperative radiographs with repeat digital OPG and PNS views [Figure-7, 8] were taken 1 year later and no recurrence was noticed for 3 years.

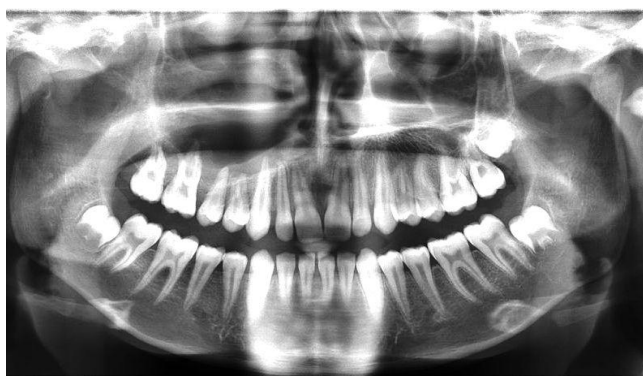


Figure -7. Post-operative Digital Orthopantomograph



Figure -8. Post-operative P.N.S Radiograph

Discussion

Aneurysmal bone cyst is an uncommon, benign, expansile pseudo cyst, usually appears in the second decade of life. ABC occurs very rarely in the jaws, about 160 cases have been reported and two thirds were located in mandible (the body of the mandible 40%, the ramus 30% and the angle 19%) and one third in the maxilla. However, only 2% of the cases are found in the head and neck, and only 22 cases of ABC

located in the maxilla have been reported. The average age of occurrence is 14.3 years with no gender predilection. ABC occurs in regions of the skeleton where there is both a relatively high venous pressure and high marrow content. It can attain greater dimensions and can cause symptoms depending upon the severity and location of lesion. Provisional diagnosis of ABC is difficult because of its similarity to other lesions. The radiographic appearance is not diagnostic.¹¹ Definitive diagnosis of ABC can be made only after histopathological examination.

Peculiar features seen in the present case of ABC were involvement of maxilla with rapid expansion secondary to trauma. Radiological features showed well-defined radiolucency involving expansion of maxilla and alveolus, with internal septae and resorption of involved teeth and thinning of cortex of maxillary sinus with no discontinuity in internal wall of maxillary sinus and infra-orbital rim and no calcifications were noticed.

Only very few cases of ABC associated with COF were reported. Ellis and Walters reported two cases of ABC arising from cementifying fibroma.³ Unlike their cases, the present case had history of trauma with rapid growth of swelling in short duration. Biesecker's proposed that ABC is actually a vascular complication of another primary bony pathologic lesion, similar to the present case.⁶

Another case of ABC arising in an area where a cementifying fibroma was excised earlier was reported by Robinson.¹² Rapid growth is usually associated with younger age group when associated with secondary ABC. Most of the cases reported involved the left side of maxilla unlike the present case. Reported cases of Maxillary ABCs listed in Table-1,^{7, 13, 14, 15, 16, 17, 18, 19} and all are diagnosed based on histopathological findings.

Most of the reported cases of ABC in maxilla were either primary or secondary to Trabecular or Psammomatoid variants of juvenile ossifying fibroma unlike the present case. An interesting observation in the present case was that two histological patterns were present within the same lesion, with very few cases reported. ABC has a high recurrence rate, which varies from 26% to 56% in most cases, within the first year after treatment. Therefore, regular follow-up of the patient is recommended, with periodic imaging guidance and histopathological

examination until complete osseous repair and remodelling of the affected area have taken place.

Year	Author(s)	Age (yrs)	Sex	Primary or Secondary
1959	Bhaskar et. al.	22	F	P
1960	Wang	8	F	P
1962	Vianna and Horizonte	18	M	P
1964	Yarington et. al.	48	F	S (Giant cell Granuloma)
1968	Gruskin and Dahlm	20	M	P
	Nosaka and Nakazawa	8	M	P
	Tillman et. al.	NS	NS	NS
1969	Byrd et. al.	17	F	P
1970	Lejeune & Bordonon	17	F	P
1972	Ellis & Walters (2 cases)	17	M	S (cementifying fibroma)
	Romanuk and Becker	10	M	P
	Komorn	26	F	P
1975	Powell & Glaser	16	F	P
	Roa	12	M	P
1977	Ruiter et. al.	NS	NS	NS
1978	Cornyn et. al.	9	M	NS
	Reyneke	18	M	P
1979	Boyd	27	M	S (Ossifying fibroma)
1980	Blayney et. al. ^[10]	14	F	S (cementifying fibroma)
	Goldman & Sission ^[11]	NS	NS	S (fibrous dysplasia)
1981	Salmo et. al. (2 cases)	55	F	P
1985	Robinson	13	M	S (cementifying fibroma)
1986	Zachariades et. al.	35	F	P
	A.W Blayney et. al.	14	F	S (cementifying fibroma)
1990	Takimoto et. al.	15	F	S (Fibrous dysplasia)
	Hady et. al.	13	F	P
1992	Hardeee et. al.	29	M	P

Abbreviations: P- primary S-Secondary M-Male F- Female NS- not specified

Table-1: Recorded maxillary Aneurysmal bone cysts of maxilla

1993	Matt	12	F	P
	Medeiros et. al.	19	F	P
	Cohen	13	M	P
1994	Cheyne et. al.	14	F	P
1996	Bataineh	25	M	P
2003	Justin Weir et. al. ^[12]	10	F	NS
2004	Zhang X et. al. (4 Cases) ^[13]	NS	NS	NS
	Sánchez AP et. al. ^[14]	14	F	P
2005	Pablo Guzman G	15	F	P
2007	Saheeb BD et. al. ^[15]	5	NS	S (cementifying fibroma)
2009	Kavita Mardi et. al.	18	F	P
	Tang IP, Shashinder et. al.	17	M	P
2010	Sheth MB et. al.	8	F	P
2011	Barcellous Silva et. al.	9	F	S (Tr JOF)
	Sankaranarayanan et. al.	6	F	S (Tr JOF)
	Lee HM, Cho KS et. al. ^[16]	NS	NS	P

Abbreviations: P- primary S-Secondary M-Male F- Female NS- not specified

Table-1: Recorded maxillary Aneurysmal bone cysts of maxilla [continued]

Conclusion

ABC of maxilla represents an enigmatic pseudo cyst with variable clinical and radiological presentations, therefore posing a diagnostic dilemma. Even though occurrence of ABC is rare in maxilla, it should be included in differential diagnosis of traumatic swellings as rapid growth of this lesion may lead to complications like loss of eyesight and hearing. Early diagnosis and effective treatment with long term follow-up are essential considering their aggressive behaviour and high recurrence rate.

References

- Motamedi MH, Yazdi E. Aneurysmal bone cyst of the jaws: Analysis of 11 cases J Oral Maxillofac. Surg. 1994; 52:471-5.
- Sheth MB, Sujan SG, Poonacha KS. Maxillary Aneurysmal bone cyst: Report of a rare case. J Ind. Soc. Pedo. Prevent. Dentistry 2010;28:4: 307-310.
- Bernier JL. & Bhaskar SN: Aneurysmal bone cysts of the mandible. Oral. Surg. 1958;11:1018-1028.
- Jaffe HL. & Lichtenstein L. Solitary Unicameral cyst, with emphasis on roentgenographic picture, pathologic appearance, and pathogenesis. Arch. Surg. 44:1004-25.
- Zachariades et al. Aneurysmal bone cyst of the jaws. Review of Literature and report of 2 cases Int J. Oral Maxillofac. Surg. 1986;15:534-540.
- Ellis & Walters. Aneurysmal bone cyst of the maxilla. J Oral Surgery 1972; 34: 1:26-32
- Sanchez AP. et.al. Aneurysmal bone cyst of maxilla. J Craniofac. Surg. 2004;15:1029-33.
- Tang IP, Shashinder S, Loganathan A, Anura M, Zakarya S, Mun KS. Aneurysmal bone cyst of the maxilla. Singapore Med J 2009; 50:9: e326-28
- Silva CAB et.al. Trabecular Juvenile Ossifying Fibroma with Aneurysmal Bone Cyst: A Rare Presentation. J Paedia. Dentistry 2011;33:5:388-91.
- Guzmán P, Araya ABJ, Roa J, Brevis L, Torres P. Aneurysmal bone cyst of the maxilla. Report of one case Rev Méd Chile 2005; 133: 1355-1360
- Sopta J, Mijucic V, Dražić R, Tulić G, Tepavčević Z. Cemento-ossifying fibroma of jaws- correlation of clinical and Pathological findings. Cilm Oral Invest 2011; 15:201-207
- Sankaranarayanan et.al. Hybrid lesion of Maxilla. J Oral Maxillofac Pathol. 2011; 15:3: 299-302.
- Bataineh AB, Sarajevo. Aneurysmal Bone Cysts of the Maxilla: A Clinicopathologic Review J Oral Maxillofac. Surg. 1997: 55:1212-1216.
- Blayney, Tayeb. The 'hybrid' fibro-osseous lesion. J Laryngology & Otology 1986;100:291-302.
- Goldman & Sisson. Fibrous dysplasia of maxilla with its relation to Aneurysmal bone cyst. Proc Inst Med Chic. 1980: 33:1:29-31
- Weir J, Harris N, Sergi C. Pathology quiz case: aneurysmal bone cyst (ABC) of the maxilla. Arch Otolaryngol Head Neck Surg. 2003; 129:12:1345.
- Zhang X, Huang J, Zhan S, Fu X. Exploration of diagnosis and treatment of maxillary aneurysmal bone cyst (report of 4 cases and review of literature. Lin Chuang Er Bi Yan Hou Ke Za Zhi. 2004;48:9:553-554
- Saheeb BD, Ojo MA, Obuekwe ON. Aneurysmal bone cyst: a primary or secondary lesion? Niger J Clin Pract. 2007 ;10:3:243-6.
- Lee HM, Cho KS, Choi KU, Roh HJ. Aggressive aneurysmal bone cyst of the maxilla confused with telangiectatic osteosarcoma. Auris Nasus Larynx. 2012 ;39:3:337-40.