Bilateral aneurysmal bone cyst of maxilla: a case report and review of literature

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Abstract

Aneurysmal bone cyst (ABC) is an uncommon benign lesion originating in the craniofacial region and has seldomly been reported in Tanzania and other parts of the world. Between 2-12% of ABC are located in the head and neck region and 90% are found in the posterior mandible while less than 1% of the cysts are biopsed in the maxilla. In this study we report a case of ABC arising from both maxilla of a 19-years old African man as revealed by both roentgenographic and microscopic findings. The presenting symptoms of our patient were mainly painless swelling, difficult in chewing and closing the mouth. Extra-oral examination revealed difficult in closing the mouth, a large pinkish swelling protruding from the mouth and ulcerations on left and right side of the maxilla. The submandibular and submental lymph nodes were palpable bilaterally and were mobile. Intra-orally, the swelling involved the whole of left and right maxillary alveolar process. Only 48, 46, 45 were present on the mandible while the rest of the teeth were missing. Teeth marks were seen on the swelling. The swelling was bony hard on palpation and the gingivae had grown to reach the buccal sulcus and the palatal midline.The aetiology and pathogenesis of this interesting lesion remained unclear but several theories were reviewed and the differential diagnosis of the lesion was also discussed.

The treatment of choice of the case we performed was surgical excision and curettage of the cavity of the ABC, which is the gold standard, let alone many other treatment options. The treatment was directed towards complete removal of the lesion. Before entering, the lesion was adequately exposed to enable a rapid curettage to be done. Massive bleeding was encountered as soon as the lesion was entered and continued until complete curettage was accomplished and there was no further bleeding. The patient had a remarkable post-operative period and the outcome of the treatment was very good.

Key words: Aneurysmal, bone cyst, Maxilla, Craniofacial region, Curettage.

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Introduction

The aneurysmal bone cyst (ABC) is an infrequent bone lesion in the craniofacial region. ABC can usually be found in the metaphysic of long bones, mainly in the "tibia and femur,." but between 2-12% of ABC are located at the head and neck region [1,2]. Almost 90% of the cases in the head and neck region affect the body of mandible 40%, ramus 30%, angle19% but also symphysis 9% and condyles 2% [6,8,9,10]. An aneurysmal bone cyst of the maxilla is a non-neoplastic, uncommon, solitary bone lesion recognized by distinct radiographic and histopathological characteristics. It is a localized and quickly expandable bone lesion which can reach a considerable size. It is characterized by the replacement of bone by spongy fibro-osseoustissue and a locally destructive and multicystic lesion filled with blood. The median age at diagnosis is 13 years old and about 80% of the patients are under 20 years old[1,2,3] There is a slight greater incidence in women (62%) than in men (38%). Although ABC is a benign lesion, it can behave locally in an aggressive manner because of its rapid growth and its osteolytic capacity[2,3,4]. This lesion represents less than 1% of all the bone cysts biopsied [5]. There are many theories about its aetiology, mostly referring to alterations of the homeostatic-vascular equilibrium of the bone [5]. It is attributed to a circulatory disturbance leading to locally increased venous pressure but with an unclear aetiology.

What seems clear is that it is not a tumour, which is one of the most differential characteristics between the ABC and two other similar entities like Giant Cell

Tumour or the Ameloblastoma [5]. In the Macroscopic examination ABCs have sponge like appearance, consisting of blood-filled cavities separated by thin, fibrous septa.[1]. The main symptoms include dull pain and/or oedema. They usually have rapid growth. Depending on its location, other signs/symptoms can be found like headache, diplopia, loss of vision, proptosis, tooth mobility, hearing loss, etc.[1,2]. The Radiological features include: Cystic bone expansion. Honey-comb or soap bubble like inner structure. Sometimes a destruction of the bone cortex and periosteal reaction can be seen [1,2,4,7]. Histologically, 95% of ABC are osteolytic lesions, expansive with vascular lagoons of variable sizes, separated by connective tissue and includes bone trabecular, osteoid tissue and multinucleated giant cells. However, 5% of ABC are solid masses without a cystic compound of irregular density and contains multiple haemorrhagic foci with greater fibroblastic, osteoblastic and osteoclastic components. The large spaces in aneurysmal bone cysts are devoid of basement membranes and endothelial cells. The absence of endothelium may explain the abundance of haemorrhages in these lesions. However, immunocytochemical demonstration of endothelial antigen provides a valuable tool for the differential diagnosis between aneurysmal bone cysts and vascular tumours.

Relatively few cases of aneurysmal bone cyst have been cytogenetically characterized, yet abnormalities of the short arm of chromosome 17 appear to be recurrent. These cytogenetic and molecular findings expand our knowledge of chromosomal alterations in aneurysmal bone cyst, further localize the critically involved 17p breakpoint, and provide an alternative approach by using Fluoresce In Situ Hybridization (FISH) technique for detecting 17p abnormalities in non dividing cells of aneurysmal bone cysts.[15]. The latter could potentially be utilized as an adjunct in diagnostically challenging cases.

The clinical course of the ABC is inconsistent, ranging from a self-limiting lesion to an aggressive, rapidly destructive lesion mimicking malignancy.[18,19,20] Treatment of ABC is usually directed towards complete removal of the lesion. Before entering the lesion, it should be adequately exposed to enable a rapid curettage to be done. Curettage may prove difficult at times since the lesions are often multilocular and may be divided by multiple bony septae. Massive bleeding may be encountered as soon as the lesion is entered and will continue until complete curettage is accomplished. Continued bleeding after completion of the procedure indicates incomplete curettage. In such cases, irrigation of the cavity enables identification of the source of bleeding and prompt curettage. The role of such factors explains the multitude of treatment modalities available for ABC; [21] no treatment, [20,21,22,] simple curettage,[23] cryotherapy, [21] excision, block resection and microvascular bone grafting, [24] therapeutic embolization and open packing.

Case report

A 19 years old young man presented to our hospital (Muhimbili National Hospital-MNH) in November 2006 with a painless, slow growing swelling involving the left and right upper jaws for the past 16 years. The swelling was noticed during childhood at 3 years. The review of system for the patient was unremarkable except for difficult in chewing. Extra-oral local examination revealed difficult in closing the mouth, with a large pinkish swelling protruding from the mouth and the left and right side of upper jaw had focal ulcerations. However, there were three scattered teeth which were seen on the swelling. The submandibular and submental lymph nodes were palpable bilaterally, all measuring about 2 cm in greatest diameter and were mobile. An intra-oral examination revealed a swelling involving the whole of the left and right maxillary alveolar process. Only 48,47, 46, 45 were present on the mandible while the rest of the teeth were missing because of extraction as narrated by the patient although he could not tell the reason behind the multiple tooth extractions. Teeth marks were also seen on the swelling. The swelling was bony hard on palpation. The gingivae had grown to reach the buccal sulcus and the palatal midline (figure 1). An incisional biopsy of the swelling was done. The tissue was formalin fixed and paraffin embedded and processed routinely by Haematoxylin and Eosin (H&E) method. Microscopically it showed areas of variable histological appearance as depicted in the photomicrographs; (figure A) – showed young fibroblasts in connective tissue stroma separated by dilated blood vessels from bone trabeculae with focal areas of haemorrhage while (figure B) revealed blood-filled vascular spaces, mostly devoid of basement membrane and endothelial cells around bone trabeculae. Other areas (figure C) revealed expansile osteolytic lesion, blood-filled spaces and connective tissue septa containing bone trabecula and osteoid. In one area of the lesion (figure D) there was extensive recent haemorrhage around the bone trabecula.

The patient was then planned for an operation whereby the extirpation of the cyst was performed. Using an intra-oral approach mucoperiosteal flap was raised; a muco-buccal and muco-labial incision that extended from the right to the left maxillary tuberosities was made. Thereafter, the lesion was adequately exposed to enable a rapid thorough curettage. We experienced some difficulties during curettage because the lesion was multilocular divided by multiple bone septa in addition to massive bleeding when it was opened. Haemostasis was achieved by using bone wax, diathermy, compression and remodeling. Remodeling of the expanded alveolar process and maxillary tuberosities was done in order to re-contour the alveolar process for the future prosthesis. After remodeling the muco-periosteal flap on the palatal side was striped off, reduced in size and then stitched back in position. The patient had a remarkable post operative period with good recovery (figure2). The patient was then discharged from our hospital two weeks later. He was to report back 6 months after discharge. When the patient reported back the general condition during this time was very good and local examination both extra-orally and intra-orally revealed well covered alveolar ridges by mucous membrane with no undercuts. At this time evaluation of the patient was made and full dentures were made for upper and lower jaws in order to improve his cosmetic appearance and function satisfaction (figure 3).

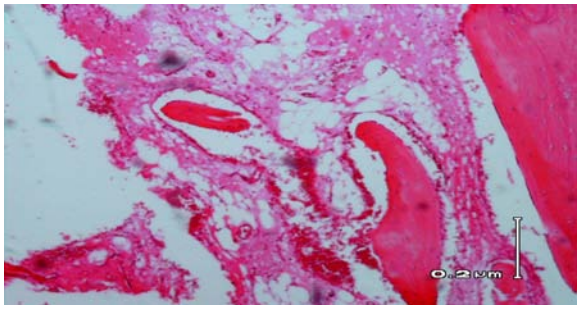

Figure A: Aneurysmal bone cyst (x 40). Young fibroblasts in connective tissue stroma separated by dilated blood vessels from bone trabeculae. Areas of focal haemorrhage are seen.

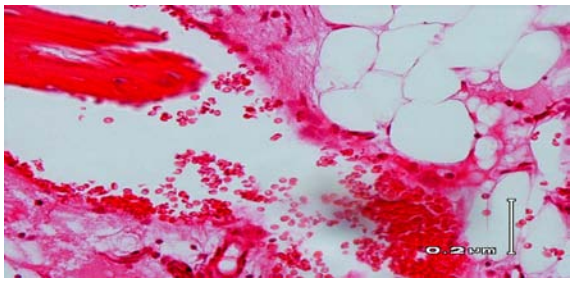

Figure B: Aneurysmal bone cyst (x 40). Blood-filled vascular space, mostly devoid of basement membrane and endothelial cells around a bone trabecula.

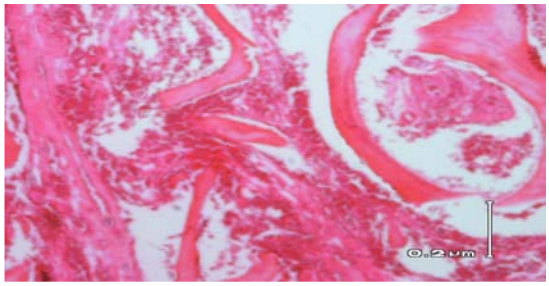

Figure C: Aneurysmal bone cyst (x40). Expansile osteolytic lesion, blood-filled spaces. Connective tissue septa containing trabeculae bone and osteoid separate the blood-filled spaces.

Figure D: Aneurysmal bone cyst (x 40). with extensive areas of recent haemorrhage around bone trabecula.

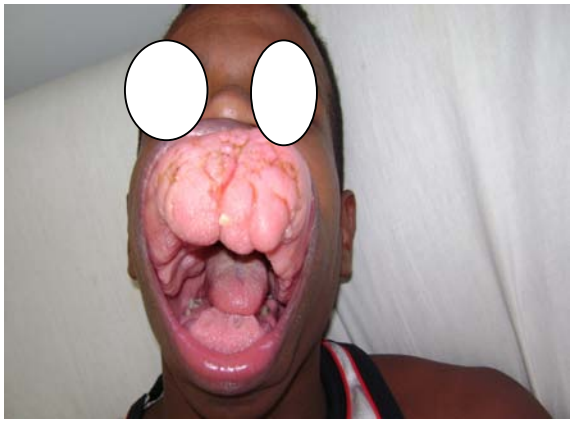

Figure 1:Aneurysmal bone cyst (ABC) is seen arising from the maxilla showing a sponge -like appearances. Also teeth marks are seen on the swelling.

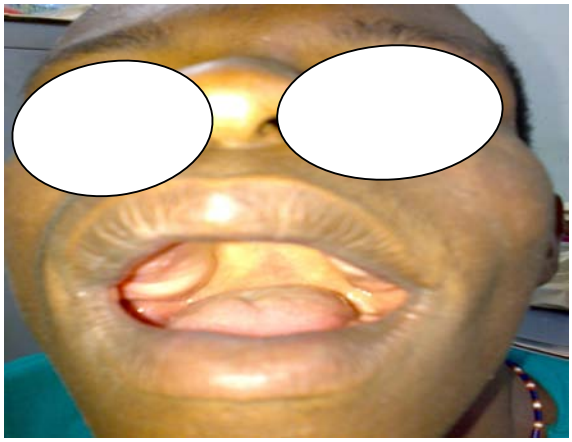

Figure 2: Post operative appearance of the maxillary alveolar process six months later

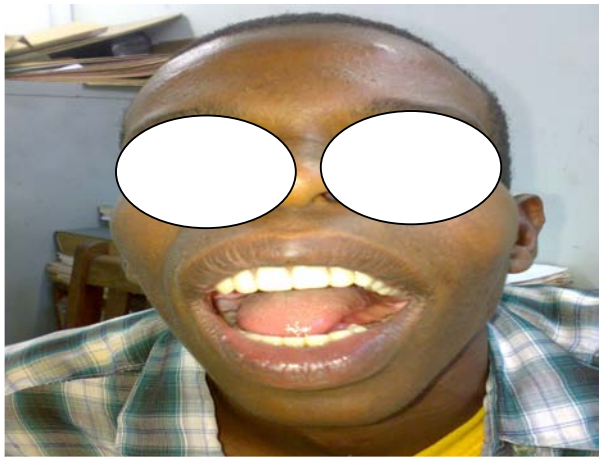

Figure 3: Partial dentures is seen in the oral cavity following surgical removal of ABC

Discussion

Bilateral aneurysmal bone cysts of the maxilla in craniofacial region have seldom been reported[1,2]. In our case ABC arose in both maxilla in a 19 years man. The

median age at diagnosis of ABC in some studies has been found to be 13 years old and about 80% of the patients are under 20 years old [2,3]. This age distribution is in agreement with our case. However, previous studies have also shown a slight greater incidence in women than in men [2,3], although the single case we report was a male patient. Between 2-12% of ABC are located in the head and neck region [1,2] and almost 90% of the cases affect the posterior mandible. The case being reported arose from the maxilla bilaterally. The aneurysmal bone cyst is an uncommon lesion of the jaws and cases involving the maxilla have been reported infrequently [14].

An aneurysmal bone cyst of the maxilla is a non-neoplastic, uncommon, solitary bone lesion recognized by distinct radiographic and histopathological characteristics. It is described as a localized and quickly expanding benign tumor, which can reach a considerable size. It is characterized by the replacement of bone by spongy fibro-osseous tissue and a locally destructive and multicystic lesion filled with blood. In the reported case, the tumour had a slow growing rate during childhood (at 3 years) and had remained so till 16 years. When the patient was admitted at the age of 19 years, the tumour was rather rapid and had reached a considerable size and the bone was replaced by expandable spongy fibro-osseous looking tissue, locally destructive, multicystic and filled with blood. There are many theories about its aetiology, mostly referring to alterations of the homeostatic-vascular equilibrium of the bone [3]. What seems clear is that it is not a tumour [4], which is one of the most differential characteristics between the ABC and two other similar entities like the Giant cell tumour or the Ameloblastoma [5]. The main symptoms include dull pain and/or oedema. They usually have rapid growth and depending on its location, other signs/symptoms can be found like headache, diplopia, loss of vision, proptosis, tooth mobility, hearing loss, etc. In this case the ABC was located in the maxilla and the patient presented mainly with a slow growing painless swelling with difficult in chewing and closing the mouth. In addition, our case had multiple losses of teeth on the mandible from 44 to 38 although the reason for the missing teeth could not immediately be established from the patient. However, is a known fact that the presence of intraoral tumour could lead to poor oral hygiene as a result of difficulty in brushing due to the swelling, pain and bleeding. The patient could have the ended up with development of periodontal disease, loosening and mobility of dentition with subsequent tooth extractions. This fact might have been the reason in our case being reported which had multiple loss of teeth on the mandible.

One of the most controversial issues of ABC refers to whether its origin is a primary or secondary lesion. Some authors [3], consider it as a secondary to trauma followed by the development of subperiosteal haematoma; while others [4], suggested that ABC is derived from another benign lesion like giant cell tumour in which some changes have made possible the communication between stroma and medullaris vessels. When this communication is maintained, aneurysmal bone cyst should be established and if the communication is interrupted, a giant cell granuloma is constituted. Lichenstein [2], added that may be some alterations of local circulation may enhance the venous pressure and therefore result in a subsequent dilatation and resorption of the bone adjacent to the vessels. In summary there is no consensus about the pathogenesis of the ABCs.

The ABC in our study was of vascular variety. It is known that this vascular variety behaves more aggressively including local invasion of the adjacent tissue in contrast with the solid type. This behaviour of ABC was also evidenced in our case whereby initially had a slow growth rate but later become rapid with local invasion of the adjacent tissue.

Concerning the treatment, although many options have been performed, the gold standard, which we also used in our case, is still surgical excision and curettage of the cavity [16,17]. The use of radiotherapy is not recommended because of radioinduced tumours.

Conclusion

The clinical course of the ABC is inconsistent, ranging from a self-limiting lesion to an aggressive, rapidly destructive lesion mimicking malignancy. Treatment of ABC is usually directed towards complete removal of the lesion. Before entering the lesion, it should be adequately exposed to enable a rapid curettage to be done. Curettage may prove difficult at times since the lesions are often multilocular and may be divided by multiple bony septae. Massive bleeding may be encountered as soon as the lesion is entered and will continue until complete curettage is accomplished. Continued bleeding after completion of the procedure indicates incomplete curettage. In such cases, irrigation of the cavity enables identification of the source of bleeding and prompt curettage. The role of such factors explains the multitude of treatment modalities available for ABC; no treatment, simple curettage, cryotherapy, excision, block resection and microvascular bone grafting, therapeutic embolization and open packing. Recurrence of jaw lesion, although uncommon, has been attributed to inadequate access and thus incomplete removal of the lesion, for which block resection is advocated. Our lesion was not a recurrence and there was no evidence of any residual lesion. This factor led us to opting for curettage as the treatment of choice.

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