Case Report

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Aneurysmal bone cyst of the mandible: a rare case report and literature review

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Introduction and importance: Aneurysmal bone cysts (ABCs) are benign, non-neoplastic cystic lesions composed of multiple blood-filled cavities, which are separated by connective tissue septa and they constitute ~6 and 1.5% of all cases of the skull and jaws lesions, respectively.

Case presentation: Herein, the case of an 80-year-old male with a histologically confirmed diagnosis of ABC involving the body of the left mandible was presented. The patient underwent partial mandiblectomy after confirmation of the diagnosis using orthopantomography, a computed tomography scan, and a tissue biopsy. The patient was free from pain after 1-year of follow-up, and the control orthopantomography showed no evidence of recurrence. This was followed by reconstruction of the healed part of the bone with titanium plates and a piece of bone from the lateral two-thirds of the left femur and thereafter he was cosmetically well.

Clinical discussion: Patients with ABCs present with expansile and radiolucent bone lesions, which may be associated with displacement and loss of teeth due to alveolar bone erosion. Complete excision of the lesions is mandatory for the prevention of recurrence and increased morbidity.

Conclusion: ABCs that involve the jaws are extremely rare and are more likely to pose a diagnostic challenge as they are more likely to be confused clinically with other expansile radiolucent bone lesions such as ameloblastoma, osteoblastoma, and giant cell tumor among many others. Also, those with extensive bone matrix formation may sometimes be confused histologically with other bone forming tumors including osteosarcoma. Recurrence is common but it can be avoided or minimized by complete resection of the lesion.

Keywords: aneurysmal bone cyst, bone lesion, mandible

Introduction

According to the WHO, an aneurysmal bone cyst (ABC) is defined as an expansile osteolytic lesion, which consists of blood-filled spaces separated by connective tissue septa that contain osteoid material and multinucleated giant cells[1]. Over 50% of the ABC cases occur in the metaphyseal part of long bones especially the femur and tibia and between 12 and 30% of such cases involve the spine[2]. ABC is extremely rare when compared with all types of jaw cystic lesions, and it constitutes ~1.5% of all nonodontogenic and nonepithelial cystic lesions of the jaws[3]. The average age of occurrence of ABCs is 13 years, and the majority of patients have an age below 20 years[4], with a slight female preponderance[2,3].

ABC is a rapidly growing and destructive bone lesion, which is characterized by the replacement of the normal bone matrix with fibro-osseous tissue containing blood-filled sinusoidal or cavernous spaces[5]. The exact cause of ABC remains unknown; however, chromosomal translocation t(16;17)(q22;p13) has been considered to be an etiologic factor in the development of ABCs[6]. ABCs can also arise de-novo when they are primary or as secondary lesions when they are in co-existence with other lesions, for example ossifying fibroma[7,8]. Currently, ubiquitin specific protease 6 (USP6) or Tres2 has been found to be upregulated in a subpopulation of patients with ABCs indicating that probably is a true neoplasm[9].

HIGHLIGHTS

- Aneurysmal bone cysts (ABCs) present as unilocular lesions radiologically and are associated with the loosening and displacement of teeth.
- The ABCs of the jaws are more likely to pose diagnostic challenges clinically due to their expansile and radiolucent appearance observed also in other common bone lesions.
- Meticulous preoperative investigations are mandatory for discriminating ABCs from their great mimickers malignancies such as osteosarcoma.
- Complete excision of ABCs prevents or reduces significantly the chances of recurrence.

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In this study, the case of an 80-year-old-male with a destructive ABC involving the body of the mandible and a literature review were described.

Case report

We present the case of an 80-year-old male with a 1-year history of a slightly painful swelling on the left side of his face. His past medical history indicated that he was diagnosed with type 2 diabetes mellitus for the past 6 years and he was on regular oral antidiabetics. Nevertheless, his family history was unremarkable. The patient reported no history of trauma to his left jaw prior to developing the lesion described. Drug history, family history, including any relevant genetic information, and psychosocial was uneventful. On physical examination, the patient had facial asymmetry due to the swelling on the left mandible. Extraorally, the overlying skin was intact and the swelling was slightly firm on palpation. Intraorally, the gingival lining mucosa around the swelling was normal, the cortical plate was bulging both bucally and lingually. He had no element of trismus and could open his mouth without difficulty. Neither the area innervated by the left mandibular nerve nor the left facial nerve had numbness.

Laboratory investigations showed a normal white blood count with its differentials. Also, platelet count, and hemoglobin level were normal. Orthopantomography (OPG) of the skull showed a radiolucent and unilocular lesion involving the body of the left mandible extending from tooth 35 to 33 with anterior displacement of tooth number 34 (Fig. 1). A three-dimensional computed tomography (CT) scan showed the presence of a unilocular and radiolucent lesion occupying the body portion of the left mandible and it was extending to involve teeth 33 and 35 with evidence of anterior displacement of the tooth 34 (image not included).

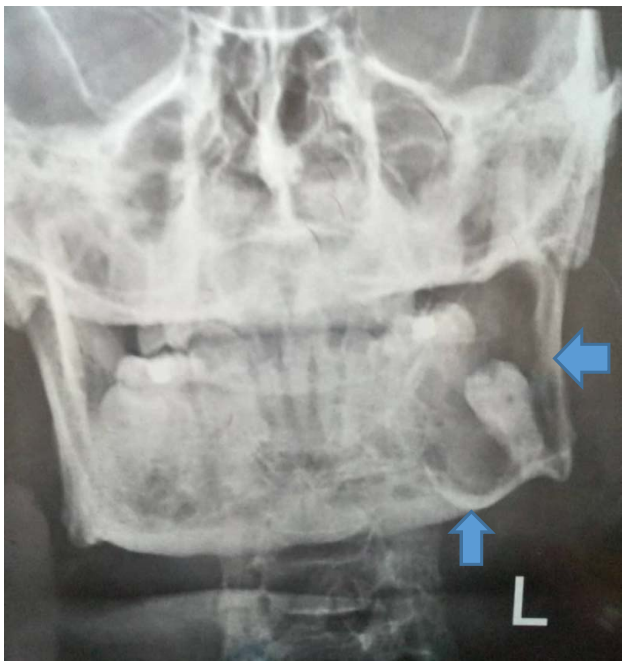

Figure 1. Anteroposterior view of the orthopantomography showing a radiolucent lesion together with thinning of the cortical plates, expansion, and anterior displacement of the tooth number 34 (arrows).

Considering the clinical manifestations as well as the OPG and CT scan findings, the clinical diagnosis was considered to be ameloblastoma. The surgical excision of the lesion was performed by a maxillofacial surgeon and a medical registrar. Under general anesthesia, a mucoperiosteal flap was raised followed by a partial mandiblectomy. The affected part of the mandible was successfully removed and hemostasis was achieved. Intraoperatively, the cortical plates were thinned and expanded both buccally and lingually; however, there was no resorption of the root of tooth 34. The patient was prescribed with prophylactic antibiotics and analgesics and on the 10th day postoperatively, he was discharged home.

Grossly, the specimen was received in formalin consisted of a single bony tissue measuring about 6.5 cm greatest diameter with central unilocular cystic lesion. The cut surface revealed a unilocular cyst containing blood stained fluids and a single molar tooth (Fig. 2). Microscopically, the lesion showed numerous blood-filled spaces of variable sizes devoid of endothelial lining, also noted focal areas of fibrosis, osteoid formation, and multiple

Figure 2. Gross appearance of the excised mass together with a tooth. The mass consists of irregularly thin wall areas with bone spicules.

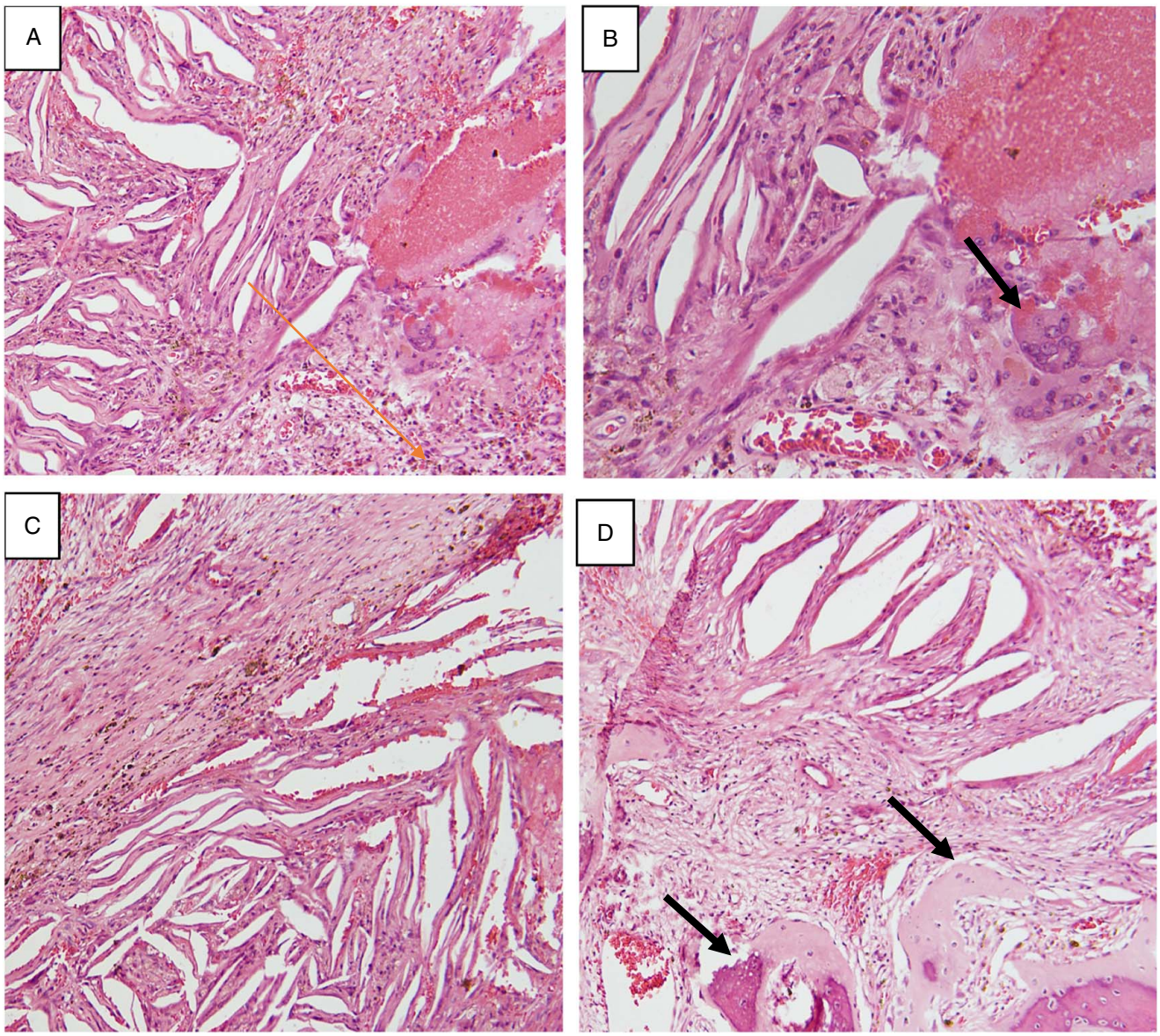

Figure 3. (A) Formation of cystic spaces filled with blood, (B) Proliferating multinucleated giant cells (arrow) (haematoxylin and eosins, ×200), (C) Focal area of fibrosis with cystic spaces without lining endothelial cells, and (D) Laying down of the bone matrix by the osteoblastic cells (arrows) (haematoxylin and eosins, ×200).

osteoclastic-like giant cells [Fig. 3 A, B, and C]. The final histopathological diagnosis of ABC was concluded. The patient was free from pain after 1-year of follow-up and the control OPG showed no evidence of recurrence. This was followed by reconstruction of the healed part of the bone with titanium plates and a piece of bone from the lateral two-thirds of the left femur and he was cosmetically well. This work has been reported in line with Surgical CAse REport (SCARE) criteria[10].

Discussion

Regarding the head and neck region, ABCs occur more commonly in the maxillofacial bones compared to skull bones due to high venous pressure as well as high marrow content in the former, and for that reason; ABC is rarely encountered in low venous pressure skull bones[5]. The mandible is affected three times more than the maxilla, and the lesions frequently affect the molar and ramus regions of the mandible compared to other parts of the jaw[2].

Patients with ABCs commonly present clinically with a local swelling, which is usually tender[11]. The swelling tends to pulsate if the lesion breaks through the inner table of the cranium[5]. Mild to moderate pain has also been reported in some patients. Patients with ABCs are at a high risk of developing pathological fractures due to the aggressive and osteolytic nature of the lesion[2]. The most severe and feared complication of ABCs is spinal cord compression, which may lead to neurological deficits[12]. Affected children especially those with long bones involvement, may present with limb deformity and decreased height as a result of impaired growth of the metaphysis of the affected bone[13].

Histologically, ABCs are divided into three variants, which include solid, vascular, and mixed type. The vascular type is the most common type and it accounts for 95% of all the cases of ABC similar to the case described in this report. This variant manifests as a rapidly growing, expansile, and destructive bone lesion causing cortical perforation and soft tissue invasion[14]. The less common solid variant consists of focal hemorrhagic and areas of fibrosis and it constitutes 5% of all the cases of ABC, and usually patients present with small and radiolucent lesion usually asymptomatic[15]. The mixed variant demonstrates features of both vascular and solid types[16,17]. This variant is considered to be a transitory phase of the lesion since sudden and rapid enlargement of stable lesions has been reported[2–4].

Concerning treatment options for ABCs, it has been shown that, the extent of the lesion is usually used in deciding whether the patient should undergo either surgical resection or curettage[2]. The first treatment approach was described in the work of Aiba *et al.*[18] on ABCs, which included curettage and reconstructing the defect with bone graft, which remains the mainstay of modern treatment. However, this approach was later found to be associated with high rates of recurrence[13]. Self-healing after long-term follow-up of patients with ABCs has also been reported[19]. Surgical resection is also another option for treating patients with ABCs[1]; however, despite local control through surgical resection, this approach has been associated with postoperative pain, limb length discrepancies, muscle weakness, and decreased ranges of motion[20]. The management approach for our patient was very different from most of the cases reported in the literature because the lesion was mimicking ameloblastoma and therefore, he was managed by partial mandiblectomy.

Recurrence rates range from 20 to 30% in different groups and it occurs most frequently within the first year after surgery[3]. This is usually associated with insufficient excision or curettage of the lesion, especially in soft tissue invasive cases. Studies recommend immediate reconstruction of the defect with autogenous grafts in cases of esthetic deformity as well as those with a high risk of fractures and loss of mandibular continuity[1,10,14]. Motamedi *et al.* reported that initial resection is not necessary and did not observe any recurrences following surgical curettage of mandibular lesions[21]. In the present case, there was no evidence of any residual lesion after 12 months of follow-up, and this could be attributed to the radical nature of the surgical treatment done.

Conclusion

ABCs of the jaws are extremely rare as compared to those of long bones and clinically, they tend to mimic other odontogenic tumors particularly ameloblastoma and keratocystic odontogenic lesions as it was in the present case. Additionally, ABCs have a possibility of undergoing malignant transformation to osteosarcoma with or without a prior history of radiation. Therefore, thorough and meticulous preoperative investigation including MRI and biopsy is mandatory to establish the diagnosis and prevent unnecessary radical surgical approaches such as partial mandiblectomy, which may be associated with morbidity and not be cost-effective.

Ethical approval

Ethics approval is not required for case reports. The ethical approval was exempted by the Institution Review Board (IRB) of the Makerere College of Health Sciences (MakCHS).

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

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Author contribution

J.J.Y. and E.D.M.: acquisition, organizing, and writing the first draft of the manuscript; Z.S.A. and E.O.: preparation of the images. All authors reviewed the manuscript critically.

Conflicts of interest disclosure

The authors declare that they have no conflicts of interest.

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Guarantor

James J. Yahaya agreed to be the guarantor and accepted to have full responsibility for the work.

Data availability statement

Not applicable.

Provenance and peer review

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References

[1] Urs AB, Augustine J, Chawla H. Aneurysmal bone cyst of the jaws: clinicopathological study. J Maxillofac Oral Surg 2014;13:458–63.
[2] Sharma PK, Kundu ZS, Lamba A, *et al.* Diaphyseal aneurysmal bone cysts (ABCs) of long bones in extremities: analysis of surgical management and comparison with metaphyseal lesions. J Clin Orthop Trauma 2021;18:74–9.
[3] Manor E, Kachko L, Puterman MB, *et al.* Cystic lesions of the jaws – a clinicopathological study of 322 cases and review of the literature. Int J Med Sci 2012;9:21–6.

[4] Kasatwar A, Patil S, Durairay D, *et al*. Recurrence of aneurysmal bone cyst of the mandible and its management: A case report and overview. J Datta Meghe Inst Med Sci Univ 2021;16:199–201.
[5] Ugga L, *et al*. Spectrum of lytic lesions of the skull: a pictorial essay. Insights Imaging 2018;9:845–56.
[6] Ahmad T, Naous R. Subperiosteal aneurysmal bone cyst with florid ossification: a rare subtype. Case Rep Pathol 2020;2020:1–5.
[7] Yeom HG, Yoon JH. Concomitant cemento-osseous dysplasia and aneurysmal bone cyst of the mandible: a rare case report with literature review. BMC Oral Health 2020;20:1–6.
[8] Salehinejad J, Shaban B, Mehri T, *et al*. Synchronization of calcifying odontogenic cyst and aneurysmal bone cyst: a case report. J Dent Mater Tech 2017;6:83–8.
[9] Li L, *et al*. Ubiquitin-specific protease USP6 regulates the stability of the c-jun protein. Mol Cell Biol 2018;38:1–11.
[10] Agha RA, *et al*. The SCARE 2020 guideline: updating consensus Surgical CAse REport (SCARE) guidelines. Int J Surg 2020;84:226–30.
[11] Dawod MS, Alisi MS, Rabab'a H, *et al*. Surgical management of aneurysmal bone cyst of the pubis: a case report and review of literature. Int Med Case Rep J 2022;15:287–92.
[12] Novais EN, Rose PS, Yaszemski MJ, *et al*. Aneurysmal bone cyst of the cervical spine in children. J Bone Jt Surg 2011;93:1534–43.
[13] Park HY, *et al*. Current management of aneurysmal bone cysts. Curr Rev Musculoskelet Med 2016;9:435–44.
[14] Al-Maghrabi H, Verne S, Al-Maghrabi B, *et al*. A typical presentation of giant mandibular aneurysmal bone cyst with cemento-ossifying fibroma mimicking sarcoma. Case Rep Otolaryngol 2019;2019:1–5.
[15] Singh A, Majeed A, Mallick S, *et al*. Solid variant of aneurysmal bone cyst masquerading as malignancy. J Clin Diagnostic Res 2017;11:ED35–6.
[16] Devi P, Thimmarasa VB, Mehrotra V, *et al*. Aneurysmal bone cyst of the mandible: a case report and review of literature. J Oral Maxillofac Pathol 2011;15:105–8.
[17] Pelo S, Gasparini G, Boniello R, *et al*. Aneurysmal bone cyst located in the mandibular condyle. Head Face Med 2009;5:1–6.
[18] Aiba H, *et al*. Treatment of aneurysmal bone cysts using endoscopic curettage. BMC Musculoskelet Disord 2018;19:1–7.
[19] Acan AE, Basci O, Havitcioglu H. Aneurysmal bone cyst healing response with intramedullary lengthening nail. Acta Orthop Traumatol Turc 2018;52:232–5.
[20] Flont P, Kolacinska-Flont M, Niedzielski K. A comparison of cyst wall curettage and en bloc excision in the treatment of aneurysmal bone cysts. World J Surg Oncol 2013;11:1–8.
[21] Deventer N, Schulze M, Gosheger G, *et al*. Primary aneurysmal bone cyst and its recent treatment options: a comparative review of 74 cases. Cancers (Basel) 2021;13:1–12.