CASE REPORT

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

Srinivasa Prasad[1], Ashok M Raghaviah[2], Nitin Sharma[3], Einstein A[4], Saraswathi TR[5]
[1,4]Assoc. Professor, [2]Former Postgraduate Student, [3]Postgraduate Student, [5]Professor, [1,2,3]Department of Oral and Maxillofacial Surgery, [4,5]Department of Oral and Maxillofacial Pathology, Meenakshi Ammal Dental College, Chennai, India

Correspondence: *Dr. Srinivasa Prasad, Department of Oral and Maxillofacial Surgery, Meenakshi Ammal Dental College and Hospital, Alapakkam Main Road, Maduravoyal, India.*
E-mail: gsuja@rediffmail.com

ABSTRACT
Aneurysmal bone cyst (ABC) is an uncommon non-neoplastic lesion of the bones, usually affecting the long bones and spine. The rare jaw lesions are encountered in the body and ramus of the mandible. Commonly reported in the second and third decades of life, ABC's are characterized by a rapid growth pattern with resultant bony expansion and facial asymmetry. Surgical management usually consists of surgical curettage or resection. This paper describes a case of ABC in a 21-year-old female, that presented as an expansile bony mass in the parasymphyseal region. A brief review of existing literature on ABC is also made.
Key Words: Aneurysmal bone cyst, jaw cyst, jaw neoplasm, pseudocyst

INTRODUCTION

The World Health Organization defines aneurysmal bone cyst (ABC) as "a benign tumour-like lesion with an expanding osteolytic lesion consisting of blood-filled spaces of variable size separated by connective tissue septa containing trabeculae or osteoid tissue and osteoclast giant cells".[1] Although benign, ABC can be a rapidly growing and destructive bone lesion. The expansile nature of this lesion can cause pain, swelling, deformity, perforation or disruption of cortical plates, neurological symptoms (depending on its location) and pathologic fracture.

Jaffe and Lichtenstein were the first to recognize ABC as an intraosseous, osteolytic lesion chiefly affecting the metaphyseal region of long bones and vertebrae.[2] Bernier and Bhaskar described the first case of ABC in the jaws.[3] ABCs are most commonly found in the long bones and the vertebral column[4]. Only 1.9% of all ABCs occur in the jaws, representing 1.5% of all nonodontogenic cysts.[5] ABCs are found more frequently in the mandible than the maxilla (3:1) with preponderance for the body, ramus and angle of the mandible.[6] Involvement of other bones of the face such as zygoma and zygomatic arch has also been reported.[7-9] No sexual predilection has been established, but an age distribution toward young adulthood has been observed.[4] ABCs display variable aetiopathogenic, histological and radiographic characteristics.

CASE REPORT

A 21-year-old female patient reported to the outpatient department with the complaint of a hard, tender swelling in the right lower back teeth region since since months; with associated altered sensation in the right lower lip. She gave history of a small swelling that had started three months back and had since then increased in size with associated pain for the past one month. She gave history of facial trauma a year ago. The patient had history of irregular menstrual cycle and was under medication and also had a breast lump that regressed after taking medication. The patient had no significant family history.

Clinical examination was normal except for apparent facial disharmony with a firm, smooth, tender, diffuse swelling of 2.5 x 2 cms in the right mandibular parasymphyseal region. Intraorally, the expansion was evident in both the lingual and buccal vestibular regions with intact mucoperiosteal tissues [Figure 1]. There was no sinus opening or any discharge. Teeth in the involved areas were vital as checked by both thermal and electrical pulp testing.

Radiographic examination showed a unilocular radiolucency extending from the root of the right central incisor to the distal aspect of the root of right second premolar. Superiorly, the radiolucency was seen extending between the canine and premolars. Resorption of roots of the involved teeth was not observed. There was no discontinuity of the lower border [Figure 2]. Routine haematologic investigations were non-contributory. On aspiration, about 2 ml of blood was obtained.

Considering the recent history of trauma, clinical presentation, radiographic features and positive blood aspiration, a provisional diagnosis of aneurysmal bone cyst was made and excisional

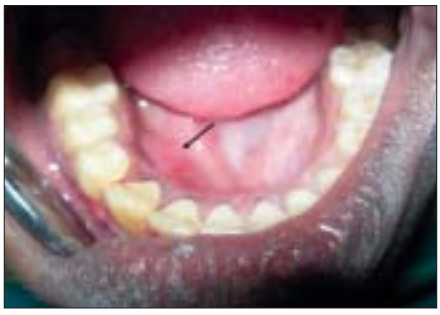

Figure 1: Preoperative intraoral photograph of the lesion

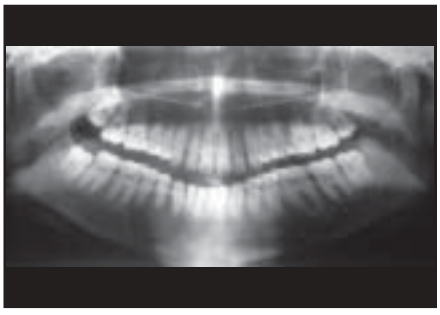

Figure 2: OPG showing unilocular radiolucency in the right anterior body of the mandible

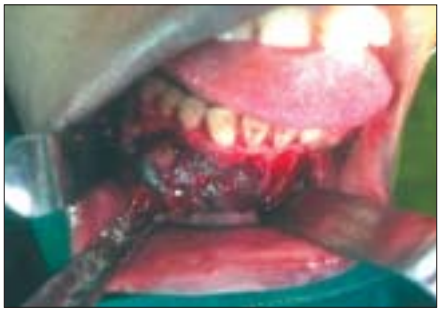

Figure 3: Intraoperative lesion before curettage

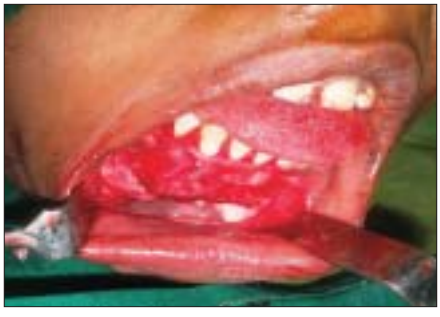

Figure 4: Intraoperative photograph after curettage

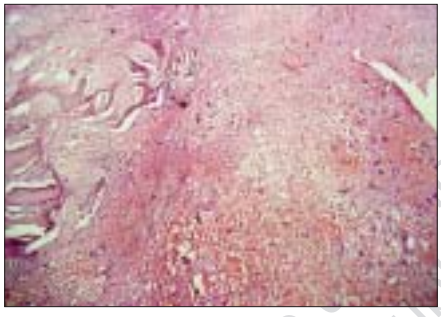

Figure 5: Fibrous connective tissue with numerous sinusoidal blood-filled spaces, bony trabeculae and multinucleated giant cells (H and E, 4x)

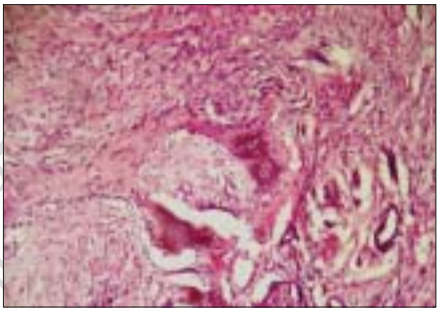

Figure 6: Immature bony trabeculae (H and E, 10x)

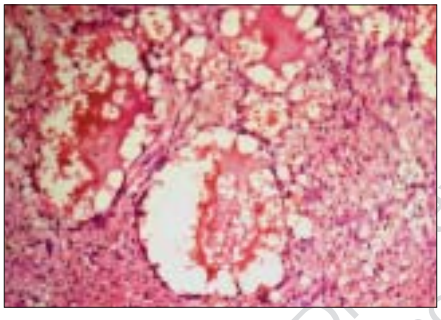

Figure 7: Sinusoidal spaces filled with RBCs (H and E, 10x)

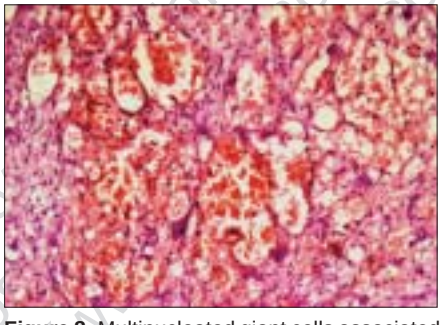

Figure 8: Multinucleated giant cells associated with the sinusoidal spaces (H and E, 10x)

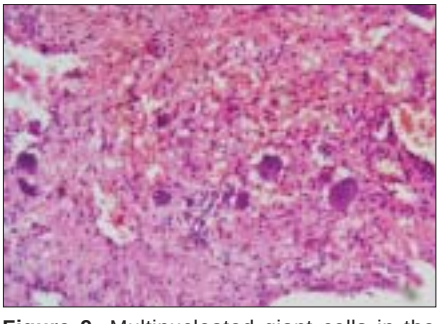

Figure 9: Multinucleated giant cells in the fibrous tissue (H and E, 10x)

biopsy of the lesion was planned under general anesthesia.

An intraoral crevicular incision was extended up to first molar with a vertical release incision in the midline. The buccal cortex was exposed which was egg-shell thin, but not perforated. The lingual cortex was also expanded. A bluish-brown, soft, vascular connective tissue was exposed on peeling the outer cortical plate [Figure 3]. The tissue was totally curetted out, until there was no bleeding, indicating complete removal [Figure 4]. The incision was closed with a 3-0 catgut. The patient's postoperative course was uneventful and the wound healed normally.

The excised specimen was sent for histopathological examination. Histological sections from the specimen revealed multiple blood-filled sinusoidal spaces surrounded by fibrous tissue, with the presence of cluster of giant cells. Mature and immature bony trabeculae and hemorrhage and hemosiderin deposits were observed [Figures 5-9]. The histologic picture was consistent with aneurysmal bone cyst. The patient is on regular follow-up for the last one year and is asymptomatic.

DISCUSSION

The true etiology of ABC is unknown. Most believe that ABC are the result of a vascular malformation within the bone. The cause of the malformation is however a topic of controversy. Three theories concerning the cause and pathogenesis of ABC have been proposed; possible traumatic origin resulting in subperiosteal or intra-medullary hemorrhage with misguided reparative processes;[3] altered hemodynamic state leading to a dilated congested vascular bed, causing resorption and erosion of bone with expansion of the lesion; and a secondary phenomenon that occurs in a primary osseous lesion. In our

case, trauma could have contributed to the development of the lesion. Panoutsakopoulos *et al.*[10] described three cases of ABC with chromosomal anomalies; band 16q22 being involved in all three patients. Similar findings were reported by Herens *et al.*[11] Familial incidence of ABC has also been reported in the literature.[12-14]

The radiographic features are not pathognomonic and there is no consensus in the literature in this regard. The "blown-out" appearance in radiographs gives the appearance of a "bone cyst". The lesion may appear unicystic or as a unilocular, multilocular (soap-bubble or honeycomb) or moth-eaten radiolucency, causing expansion, perforation or destruction of the bony cortices. There may also be an associated periosteal reaction with reactive new bone formation, resulting in a peripheral sclerotic border in some cases.[8,9,15-17] Our case presented as a unilocular radiolucency; however, resorption of roots of the involved teeth was not observed in spite of the close proximity of the radiolucency to the roots.

Histologically, ABC consists of a fibrous connective tissue stroma with numerous blood filled caverns of sinusoids, multinucleated giant cells and osteoid. Hemosiderin is present in variable amounts and osteoid and bone formation are variable.[17] Since a normal epithelial lining is lacking, the lesion is also referred to as pseudocyst. The histologic appearance in our case was consistent with the abovementioned features.

The clinical course of the ABC is inconsistent, ranging from a self-limited lesion to an aggressive, rapidly destructive lesion mimicking malignancy.[6,15,18,19] 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;[20] no treatment,[19-23] simple curettage,[24] cryotherapy,[20,26,27] excision, block resection and microvascular bone grafting,[24] therapeutic embolization and open packing.[25]

Recurrence of jaw lesion, although uncommon, has been attributed to inadequate access and thus incomplete removal of the lesion,[5,8,17,18,21] 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.

Filling the defect with bone grafts is not required as bone repair is rather rapid.[19,22,23] Use of radiation has not been advocated as it may fail to arrest the lesion and can result in sarcomatous changes.[28-30]

ACKNOWLEDGMENTS

The authors thank Dr. R.S. Neelakandan, Professor and Head, Department of Oral and Maxillofacial Surgery, Meenakshi Ammal Dental College, Chennai for his critical review and comments on the manuscript.

REFERENCES

1. Kramer IR, Pindburg JJ, Shear M (1992): *In*: Histological typing of odontogenic tumours, 2nd edn. Berlin: Springer-Verlag; p. 32.
2. Jaffe HL, Lichtenstein L (1942): Solitary unicameral bone cyst with emphasis on the roentgen picture, the pathologic appearance and pathogenesis, Arch Surg, 44:1004-25.
3. Bernier JL, Bhaskar SN (1958): Aneurysmal bone cysts of the mandible oral Surg Oral Med Oral Pathol, 11:1018-28.
4. Giddings NA, Kennedy TL, Knipe KL, Levine HL, Smith JD (1989): Aneurysmal bone cyst of the mandible, Arch Otolaryngol Head Neck Surg, 115:865-70.
5. Eldeeb M, Sedano HO, Waite DE (1980): Aneurysmal bone cyst of the jaws. Report of a case associated with fibrous dysplasia and review of the literature, Int J Oral Surg, 9:301-11.
6. Trent C, Byl FM (1993): Aneurysmal bone cyst of the mandible, Ann Otol Rhinol Laryngol, 102:917-24.
7. Eveson, JW, Moos KF, MacDonald (1978): Aneurysmal bone cyst of the zygomatic arch, Br J Oral Surg, 15:259-64.
8. Struthers PM (1984): Shear Aneurysmal bone cyst of the jaws, Clinicopathological features, Int J Oral Surg, 13:85.
9. Struthers PM (1984): Shear: Aneurysmal bone cyst of the jaws. Pathogenesis, Int J Oral Surg, 13:92.
10. Panoutsakopoulos G, Pandis N, Kyriazoglou I, Gustafson P, Mertens F, Mandahl N (1999): Recurrent t(16;17)(q22;p13) in aneurysmal bone cysts, Genes Chromosom Cancer, 26:265-6.
11. Herens C, Thiry A, Dresse MF, Born J, Flagothier C, Vanstraelen G, *et al.* (2001): Translocation (16;17)(q22;p13) is a recurrent anomaly of aneurismal bone cysts, Cancre Gene Cytogene, 127:83-4.
12. Vicenzi G (1981): Familial incidence in two cases of aneurysmal bone cyst, Ital J Orthop Traumatol, 7:251-3.
13. Power RA, Robbins PD, Wood DJ (1996): Aneurysmal bone cyst in monozygotic twins: A case report, J Bone Joint Surg BR, 78:323-4.
14. Leithner A, Windhager R, Kainberger F, Lang S (1998): A case of aneurysmal bone cyst in father and son. Eur J Radiol, 29:28-30.
15. Kalantar Motamedi MH, Khodayar A (1993): Aneurysmal bone cyst mimicking a malignancy, J Oral Maxillofac Surg, 51:691.
16. Laskin DM (1985): Oral and maxillofacial surgery, Vol 2. CV Mosby, St Louis, p. 477-8.
17. Shafer WG, Hine MK, Levy BM (1983): A textbook of oral pathology, Saunders, Philadelphia; p. 149-52.
18. Hardee PS, Whear NM, Morgan PR (1992): Aneurysmal bone cyst of the maxilla-an association with tooth resorption, J Cranio Maxillofac Surg, 20:266-9.
19. Hernandez GA, Castro A, Castro G, Amador E (1993): Aneurysmal bone cyst versus hemangioma of the mandible.

Report of a long-term follow-up of a self-limiting case oral Surg Oral Med Oral Pathol, 76:790-.
20. Marcove RC, Sheth DS, Takemoto S (1995): The treatment of aneurysmal bone cyst, Clin Orthop, 311:157-63.
21. Motamedi MH, Yazdi E (1994): Aneurysmal bone cysts of the jaws: Analysis of 11 cases. J Oral Maxillofac Surg, 52:471-5.
22. Kalantar Motamedi MH (1998): Aneurysmal bone cysts of the jaws: Clinicopathological features, radiographic evaluation and treatment analysis of 17 cases, J Cranio Maxillofac Surg, 26:56-62.
23. McQueen MM, Chalmers J, Smith GD (1985): Spontaneous healing of aneurysmal bone cysts. A report of two cases, J Bone Joint Surg Br, 67(2):310-2.
24. Schreuder HW, Veth RP, Pruszczynski M (1997): Aneurysmal bone cysts treated by curettage, cryotherapy and bone grafting, J Bone Joint Surg Br, 79(1):20-5.
25. Motamedi MH (2002): Destructive aneurysmal bone cyst of the mandibular condyle: Report of a case and review of the literature, J Oral Maxillofac Surg, 60:1357-61.
26. Malawer MM, Marks MR, McChesney D (1988): The effect of cryosurgery and polymethylmethacrylate in dogs with experimental bone defects comparable to tumor defects, Clin Orthop Relat Res, 226:299-310.
27. Marcove RC (1982): A 17-year review of cryosurgery in the treatment of bone tumors, Clin Orthop Relat Res, 163:231-4.
28. Cohan W (1984): Sarcoma arising in irradiated bone, Cancer, 1:3.
29. Cole L (1965): Radiation carcinogenesis: The sequence of events, Science, 150:1782.
30. Eisenbud L, Attie J, Garlick J (1987): Aneurysmal bone cyst of the mandible oral Surg Oral Med Oral Pathol, 64:202.

Source of Support: Nil, **Conflict of Interest:** None declared.

Living Legends: Dr. T. R. Saraswathi

Dr. T. R. Saraswathi, born on 20th October, 1942 in Chennai, did her BDS and MDS from Tamil Nadu Government Dental College, Madras University. Then she worked as a Research Fellow and Assistant Reader at the Dental wing, Madras Medical College. She underwent a epidemiological and oral cytology training programme in Tata Institute of Fundamental Research, Mumbai. In this programme, she actively participated in epidemiological study of oral cancer and precancerous lesions among the rural population of various parts of India. Then she participated in the Oral Cancer Control Project organized by Government of Tamil Nadu in collaboration with WHO at Kancheepuram, Tamil Nadu. During this period, she was awarded the WHO fellowship to pursue higher studies related to oral precancerous lesions at London University, UK, leading to MSc in Oral Pathology. She was the first woman dental surgeon from Tamil Nadu to be awarded such a prestigious fellowship.

Dr. Saraswathi worked as Reader and Professor at various places which include Arignar Anna Memorial Cancer Institute and Hospital, Kancheepuram, Raja Muthiah Dental College, Chidambaram, Kilpauk Medical College, Tamil Nadu Government Dental College, Ragas Dental College and presently she is working as a Professor in Meenakshi Ammal Dental College, Chennai.

Dr. Saraswathi has conducted and organized various research projects. One of the prominent studies among them being "the oral cancer control programme – A pilot project organized at Arignar Anna Memorial Cancer Institute, Kancheepuram". This was an epidemiological Research involving the urban population of Kancheepuram town for the early detection, prevention and control of oral cancer along with cervical cancer. The project was conducted on collaboration with World Health Organization. This work was appreciated by eminent oral pathologists like Professor JJ Pindborg and Dr. Jacob Zacharia.

In Saudi Arabia, she organized and established the oral diagnosis clinic and oral pathology department. Other than the professional service, she also took part in teaching the dental surgeons during their training period, and also worked as a co – investigator for the project "relationship of dental caries and nutritional status in Saudi school children, Abha, K.S.A".

In collaboration with National Medical Research Council, Ministry of Health, Singapore, she worked as a co-investigator in a randomized controlled trial to study the effect of topical steroids with cyclosporine in the management of oral lichen planus.

She has delivered lectures at various national and international gatherings and is a recipient of Dr. Chitambalam award. She has contributed to more than 40 publications. She is also one of the past presidents of Indian Association of Oral and Maxillofacial Pathologists. As an appreciation of her service to the field of Oral Pathology and to IAOMP, she was awarded the 'Lifetime contribution to IAOMP' award in 2006.