



Republic of Iraq Ministry of higher education and scientific research

University of Al-Qadisiyah-College of Dentistry

2022-2023



---

---

## *Aneurysmal bone cyst*

---

---

A graduation project Submitted to the college of Dentistry in partial fulfillment of the requirement for the Degree of Bachelor in dental and oral surgery (B.D.S)

---

---

Prepared by:

**Hussein Saad, Hussein Alaa, Hassanen Hassan, Hussein Haleem**

Supervised by:

**Dr. Osama M. Ghazi**

---

---

**Signature:**

## شكر وتقدير

الحمد لله والشكر لله اولا واخرا...

لا يمكن للكلمات أن تعبر عن امتناني لأستاذي ورئيس لجنتي لما أبداه من صبر وتعليقات لا تقدر بثمن.

نقدم شكرنا وامتناننا إلى كل من اعاننا وساعدنا في الوصول الى هذه النقطة بفضلهم وجهودهم وبنصائحهم القيمة التي أبدوها لنا وبالأخص مشرف البحث الدكتور الفاضل أسامة محمد غازي

والى الهيئة التدريسية في كلية طب الاسنان عموما.

راجين من الله ان نكون قد أصبنا أكثر مما أخطأنا وان يستفاد مما بذل من مجهود،

املا ان نكون قد اعطينا الموضوع حقه

واسأل الله ان يعلمنا ما ينفعنا، وينفعنا بما يعلمنا.

ولنا من الله التوفيق.

## Contents

Abstract .....	4.
Introduction .....	4.
Epidemiology .....	4.
Etiopathology .....	5.
Clinical Presentation .....	6.
Diagnosis .....	6.
Treatment .....	8.
Prognosis .....	10.
Conclusion .....	11.
References .....	11.

## Abstract

Aneurysmal bone cysts (ABCs) are rare benign lesions of bone which are infrequent in craniofacial skeleton. ABCs are characterized by rapid growth pattern with resultant bony expansion and facial asymmetry.

## Introduction

Aneurysmal bone cysts (ABCs) are benign intraosseous lesions of head and neck region. ABCs are often considered as locally aggressive lesions with a potential for local recurrence and typically seen in the metaphysis of the long bones and vertebral column.<sup>1</sup>

The term aneurysmal bone cyst was first used by Jaffe and Lichtenstein in 1942 to describe two cases of erosive, expansile, blood-filled, cystic lesions in the vertebra of an 18-year-old boy and in the pubic symphysis in a 17-year-old boy.<sup>2</sup> ABCs are typically affect long tubular bones and spine.<sup>3</sup>

Approximately 3% of these lesions are seen in the head and neck region with the mandible being the commonest location.<sup>4</sup> The majority of the ABC cases are asymptomatic, but they may present with swelling and local pain.<sup>5</sup>

Imaging shows classic osteolytic lesions, whereas magnetic resonance imaging (MRI) reveals blood-filled lesions and fluid-fluid levels.<sup>6</sup> Histopathological study confirm the diagnosis.<sup>7</sup> Currently, the treatment of choice for ABCs is surgery such as complete excision, curettage and bone grafting.<sup>8</sup>

## Epidemiology

ABC is an uncommon non-neoplastic expansile osteolytic bone lesion of unknown cause .<sup>9</sup> The incidence of ABC is 0.14 per 100,000 population per year. ABC in the head and neck region are extremely uncommon clinical entity. There is slight female predominance of ABC in comparison to male.<sup>10,11</sup>

## Etiopathology

According to world health organization, the definition of aneurysmal bone cyst is benign cystic lesion of bone consisting of blood-filled spaces separated by connective tissue septa containing fibroblasts, osteoclast type giant cells, reactive and woven bone.<sup>12,13</sup>

The exact etiology of ABC is still elusive. This pathological condition is usually thought as reactive until reports of clonal cytogenetic abnormalities showing neoplastic properties. The lesion usually originates in isolation, although ABC may sometimes be associated with other bony pathology.<sup>14</sup>

Approximately 30% of ABCs are secondary to certain lesions such as giant cell tumor, osteblastoma, chondroblastoma, chondromyxoid fibroma, fibrous dysplasia or non-ossifying fibroma.<sup>15</sup>

The exact nature of histogenesis of ABCs are still unclear and it is classified into indeterminate tumor, intermediate malignancy and locally aggressive.<sup>16</sup> It was thought that intraosseous or subperiosteal bleeding due to abnormal venous circulation, activating the osteoclasts and inducing resorption of bone and local remodeling.<sup>17</sup> This theory is no longer approved for primary ABC, which rearrange USP6 on chromosome 17 but remains believable for secondary ABC which does not show translocation.<sup>16</sup>

The genetic aspect for causation of ABC may predominate, masking the other etiologies. Recently genetic and immunohistochemical studies showed that primary ABC is a tumor and not considered as a reactive tumor like lesion.<sup>18</sup> Moreover, the neoplastic nature of the ABC was evidenced by showing clonal chromosome band 17p13 translocations which place the USP6 oncogene under regulatory of the highly active CDH11 promoter.<sup>19</sup>

## Clinical presentation

ABCs may be found at any age group. However, ABCs are commonly seen in young patients in the first and second decades of life.<sup>20</sup> Approximately 80% of the ABCs are seen before the age of 20 years.<sup>21</sup> ABC primarily seen in young females.<sup>22</sup> ABC often present with pain and sometimes swelling or facial deformity in head and neck region. A short period of pain and swelling in affected bone is the characteristic features, although the clinical presentations are dependent upon the site affected. The main symptoms are edema and/or dull pain in head and neck and these lesions have rapid growth. Depending on the sites of the ABCs in head and neck region, other symptoms can be seen like headache, vision loss, proptosis, deafness.<sup>23</sup> The bony involvement may cause proptosis of the eye.<sup>24</sup> The proptosis of the eye is usually found in case of sinonasal involvement by ABC. Bony involvement particular long tubular bone cause fracture. Spinal involvement may result in pain, torticollis, stiff neck and rarely neurological symptoms.<sup>25</sup> The differential diagnosis of ABC includes vascular lesions, malignant lesions, eosinophilic granuloma, enchondroma or chondromyxoid fibroma, non-ossifying fibroma and fibrous dysplasia. ABCs can be seen in different grades in several other types of lesions such as unichameral cyst, ossifying fibroma, ameloblastoma, giant cell granuloma or even in fracture of the bone. ABC can affect any sites of head and neck region with long bones, spine and pelvis.<sup>26</sup>

## Diagnosis

The initial diagnosis of ABC can be done radiographically, with MRI being taken as the first choice for diagnosis. The computed tomography (CT) scan is helpful to delineate the bony lesions and extension of the ABC. The diagnosis of ABC should be firmly established and malignant lesions must be ruled out. This

implies not only help towards diagnosis of the ABC, but also any underlying bony pathologies inciting the origin of the ABC. The radiological features of ABCs include cystic bone expansion which shows honey-comb or soap bubble like inner structure. Sometimes radiological findings show destruction of the bone cortex and periosteal reaction.<sup>27</sup>

Histological features usually confirm the diagnosis. It is often unwise for a pathologist to render a diagnosis without a thorough inspection of the radiological films. If ABC is suspected radiologically, intra-operative frozen section is usually sought during excision/curettage. This is done to rule out the malignant lesions like telangiectatic osteosarcoma, usually the primary concern in radiological differential diagnosis. The gross pathology of ABC show sponge like, friable, hemorrhagic material which often gritty consisting blood-filled cavities separated by thin, fibrous septa. In case en-block resection of the tumor mass, cysts and cortical destruction can be appreciated. Majority of the ABCs in head and neck area measure between 1 to 10 cm in its maximum diameter.<sup>28</sup>

The microscopic picture of ABC shows a stroma consisting of fibroblasts, multinucleate giant cells and bone, as well as cystic spaces usually filled with blood and consisting of increased number of giant cells lining the cavity (Figure 1). Histologically, the cavities lack endothelial cover and full of blood. These are contoured with fibrous septa, enclosing fibroblasts, inflammatory lymphohistiocytic elements, siderophages and osteoclastic giant cells.<sup>29</sup>

In initial phase, mitosis may be strong but not abnormal. There are no elastic or smooth muscle fibers. There is reactive osteogenesis inside the immature septa, consisting a thin network of woven or lacy osteoid or a greater number of mature trabeculae. There is strongly calcified basophil fibrochondroid matrix in more than

one-third of cases. The number of cystic components in ABC can vary. ABC can be divided into types such as classic (95%) and solid (5%).<sup>30</sup> In the head and neck region, the solid type of ABC is usually called as giant cell reparative granuloma, central giant cell granuloma or central ossifying fibroma. The common coexisting lesions with ABC include chondroblastoma and giant cell tumor.<sup>30</sup>

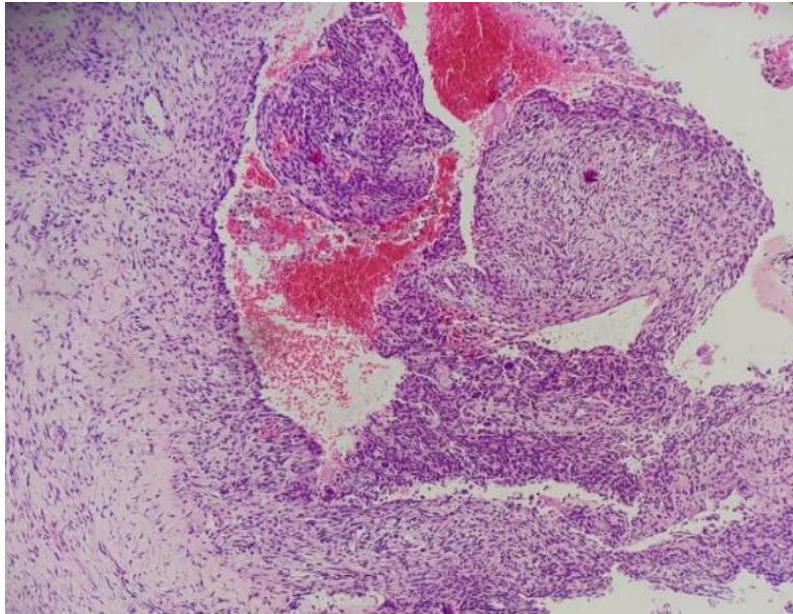


Figure 1: Microphotograph of histopathological picture of the aneurysmal bone cyst.

## Treatment:

### Surgical

Surgical excision is the treatment of choice in ABC.<sup>31</sup> Wide resection of the tumor prevent local recurrence but wide resection may require reconstruction which increase the cost of treatment.<sup>32</sup>

Currently, Enbloc resection is not an option in majority of patients with ABC because of the resulting disability/required reconstructive surgery in this benign disease. So, intralesional curettage with or without bone grafting is still commonly used treatment, but pose a risk of local recurrence of approximately 20%.<sup>32</sup>



Marginal resections may be helpful in very expansive form of ABC. Curettage and aspiration of cyst without filling gives good results in comparison to intra-cyst resection. The chance of risk can be minimized by association with curettage to cryotherapy or argon plasma coagulation (with high chance of fracture). In case of fracture or fragilization, osteosynthesis may be helpful if diagnosis of ABC is certain. Complications for surgical excision of ABC in head and neck regions are uncommon. In some cases, facial paresis may be found following surgical excision of ABC. Facial paresis following surgery can be managed with conservative treatment.<sup>33</sup>

## Medical

Local intra-cystic injection of demineralized bone powder, bone marrow, calcitonin, bone substitute and doxycycline are related to contradictory results and sometimes need a large number of procedures.<sup>34</sup>

Injection of absolute alcohol also shows good result with low rate of complications. It may need repeat injection, if cyst shows incomplete result or cure.<sup>35</sup>

Intralesional sclerotherapy with alcohol is an effective mode of treatment. Injection of methyl prednisolone acetate should be avoided, as it may exacerbate the lesion.<sup>24</sup> Radiotherapy is also an effective one, but the chance of malignant transformation limits its use in ABC. However, radiotherapy is sometimes preferred in recurrent spinal lesions inaccessible to other treatment modalities.<sup>37</sup>

Interferon alpha-2a therapy is helpful for treatment of vascular tumors in the head and neck region such as ABC and giant cell tumors. It can also be used for controlling unresectable lesions and act as effective adjunct to the surgical excision, when administered before surgery for reducing the tumor size. Chemotherapy is a less common option for ABCs.<sup>38</sup>

Artery embolization is an efficient adjuvant treatment for ABCs.<sup>39</sup> It commonly calcify within 2 to 4 months after embolization.<sup>40</sup> This therapy is usually helpful for ABCs in head and neck regions those are difficult to resect because of their locations like deep seated sites. The embolization techniques are often done before surgical resection. The adverse effects of the arterial embolization include ischemia to other vessels, nerves and organs with blood supply from same artery that is embolized.<sup>39</sup>

## Prognosis

There is approximately 20% of the patients with ABC show recurrence.<sup>41</sup> The recurrence rate can be reduced by complete surgical excision or surgical excision with cryosurgery.<sup>42</sup>

One report shows that selective arterial embolization provides favorable results. As 90% of recurrence after surgery occur within 2 years, close observation with serial radiograph is required for at least two years.<sup>43,44</sup>

The treatment options of ABC have evolved over the years. Resection of the lesion is not an option in majority of cases leaving intralesional procedures like curettage as standard of care.<sup>45</sup> Because of the recurrence rate more than 50%, several adjuvant treatments have been employed. The adjuvant treatments include polymethyl methacrylate (PMMA)/ bone cement, phenol, argon, ethanol and cryotherapy. Less invasive techniques are aggressive biopsy, selective arterial embolization, sclerotherapy.<sup>46,47</sup>

## Conclusion

Aneurysmal bone cyst is expansile, non-neoplastic bony lesion, consisting of sponge like cavities of different sizes which contain either blood or serum. ABC notoriously affects the vertebral column and long bones in the body. Rarely, ABCs seen the head and neck region. ABCs of the head and neck area in pediatric patients pose a diagnostic challenge to otolaryngologists or pediatricians because of their rarity and non-specific symptomatic presentations. As ABCs in the head and neck region often present as a rapidly growing, expansive and destructive lesion, it must be crucial for clinicians or pediatricians for this clinical entity in pediatric patients to diagnose accurately and treat correctly with appropriate method.

## References

1. 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):1-6.
3. 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):1-6.
4. Calliauw L, Roels H, Caemaert J. Aneurysmal bone cysts in the cranial vault and base of skull. *Surgical neurol*. 1985;23(2):193-8.
5. Muratori F, Mondanelli NI, Rizzo AR, Beltrami G, Giannotti S, Capanna R et al. Aneurysmal Bone Cyst: A Review of Management. *Surgical technol int*. 2019;35:325-35.
6. Blanchard M, Abergel A, Williams MT, Ayache D. Aneurysmal bone cyst within fibrous dysplasia causing labyrinthine fistula. *Otol Neurotol*. 2011;32(2):11.
7. Swain SK, Samal S, Mohanty JN, Choudhury J. Nasopharyngeal carcinoma among the pediatric patients in a non-endemic region: our experience at a tertiary

care teaching hospital in Eastern India. Egyptian Pediatr Association Gazette. 2020;68(1):1- 6

9. Ramirez AR, Stanton RP. Aneurysmal bone cyst in 29 children. J Pediatr Orthop B. 2002;22:533-9.

10. Nielsen GP, Fletcher JA, Oliveira AM. Aneurysmal bone cyst. In: Fletcher BJA, Hogendoorn PCW, Mertens F, editors. WHO classification of tumours of soft tissue and bone. Lyon: IARC. 2013;348-9.

13. Rosenberg AE, Nielsen GP, Fletcher JA. Aneurysmal bone cyst. In: Fletcher CDM, Unni KK, Mertens F, eds. World Health Organization Classification of Tumours. Pathology and Genetics of Tumours of Soft Tissue and Bone. Lyon: IARC Press. 2002;338-9.

14. Swain SK, Debta P, Samal S, Mohanty JN, Debta FM, Dani A. Endoscopic Treatment of Sinonasal Ossifying Fibroma: A Case Report. Indian J Public Health. 2019;10(09):1697-1700.

15. Swain SK, Mohanty JN. Chondroma of the nose. Libyan J Med Sci. 2020;4(2):87-9.

16. Shetty N, Hegde P, Singh H, Gulia A. Aneurysmal Bone Cyst-Review. J Bone Soft Tissue Tumors Vol. 2020;6(1):17-20.

17. Hermann AL, Polivka M, Loit MP, Guichard JP, Bousson V. Aneurysmal bone cyst of the frontal bone-A radiologic-pathologic correlation. J radiol case rep. 2018;12(7):16-24.

18. Leithner A, Machacek F, Haas OA, Lang S, Ritschl P, Radl R et al. Aneurysmal bone cyst: a hereditary disease? J Pediatr Orthop B. 2004;13(3):214-7.

20. Swain SK, Samal S, Sahu MC. Chondrosarcoma at the sinonasal region. BLDE University J Health Sci. 2019;4(1):30-3.

21. Mankin HJ, Hornicek FJ, Ortiz-Cruz E, Villafuerte J, Gebhardt MC. Aneurysmal bone cyst: a review of 150 patients. J Clin Oncol. 2005;23(27):6756-62.

22. Hamilton HB, Voorhies RM. Tumours of the skull. In: Wilkins RH, Rengachary SS, eds. Neurosurgery. New York: McGraw-Hill. 1996;2:1503-28.

23. Swain SK, Sahu MC. An unusual giant isolated mucosal malignant melanoma of nasal cavity-A case report. *Egyptian J Ear, Nose, Throat Allied Sci.* 2017;18(2):151-3.
25. Boriani S, De Iure F, Campanacci L, Gasbarrini A, Bandiera S, Biagini R et al. Aneurysmal bone cyst of the mobile spine: report on 41 cases. *Spine.* 2001; 26(1):27-35.
26. Gutierrez LB, Link TM, Horvai AE, Joseph GB, O'Donnell RJ, Motamedi D. Secondary aneurysmal bone cysts and associated primary lesions: imaging features of 49 cases. *Clin imaging.* 2020;62:23-32.
27. Swain SK, Sahu MC. An extensive fibrous dysplasia of anterior skull base area of a 12-year-old boy-A case report. *Pediatrics Polska.* 2016;91(6):636-9.
28. Swain SK, Mohanty S, Singh N, Samal R. An unusually Giant Hematoma threatening to the laryngeal airway. *Int J Otorhinolaryngol Clin.* 2014;6(3):92-4.
29. Vergel De Dios AM, Bond JR, Shives TC, McLeod RA, Unni KK. Aneurysmal bone cyst. A clinicopathologic study of 238 cases. *Cancer.* 1992;69(12):2921-31.
30. Kransdorf MJ, Sweet DE. Aneurysmal bone cyst: concept, controversy, clinical presentation and imaging. *Am J Clin Path.* 1995;164:573-80.
31. Varshney MK, Rastogi S, Khan SA, Trikha V. Is sclerotherapy better than intralesional excision for treating aneurysmal bone cysts? *Clin Orthop Related Res.* 2010;468(6):1649-59.
32. Swain SK, Samal S, Das S, Padhy R. A Large Intraoral Sublingual Schwannoma in a Pediatric Patient: A Case Report. *Iranian J Otorhinolaryngol.* 2021;33(5):333-7.
33. Shiels WE, Mayerson JL. Percutaneous doxycycline treatment of aneurysmal bone cysts with low recurrence rate: a preliminary report. *Clin Orthop Related Res.* 2013;471(8):2675-83.
34. Swain SK, Sahu MC, Baisakh MR. Extranodal nasofacial natural Killer/T-cell lymphoma: Our experiences at a tertiary care hospital of Eastern India. *Apollo Med.* 2018;15(2):88-93.

35. Lambot-Juhan K, Pannier S, Grévent D, Péjin Z, Breton S, Berteloot L et al. Primary aneurysmal bone cysts in children: percutaneous sclerotherapy with absolute alcohol and proposal of a vascular classification. *Pediatric radiol.* 2012;42(5):599-605.
36. Adamsbaum C, Mascard E, Guinebretiere JM, Kalifa G, Dubousset J. Intralesional Ethibloc injections in primary aneurysmal bone cysts: an efficient and safe treatment. *Skeletal radiol.* 2003;32(10):559-66.
37. Feigenberg SJ, Marcus Jr RB, Zlotecki RA, Scarborough MT, Berrey BH, Enneking WF. Megavoltage radiotherapy for aneurysmal bone cysts. *Int J Radiation Oncol Biol Physics.* 2001;49(5):1243-7.
38. Sun ZJ, Zhao YF, Yang RL, Zwahlen RA. Aneurysmal bone cysts of the jaws: analysis of 17 cases. *J Oral Maxillofac Surg.* 2010;68 (9):2122-8.
39. Mendenhall WM, Zlotecki RA, Gibbs CP, Reith JD, Scarborough MT, Mendenhall NP. Aneurysmal bone cyst. *Am J Clin Oncol.* 2006; 29:311-5.
40. Saez N, Sharma GK, Barnes CH, Lu Y, Hsu FP, Huoh KC et al. Solid variant of aneurysmal bone cyst of the temporal bone. *J Pediatr Orthop.* 1997;17(4):440-3
41. Song W, Suurmeijer AJ, Bollen SM, Cleton-Jansen AM, Bovée JV, Kroon HM. Soft tissue aneurysmal bone cyst: six new cases with imaging details, molecular pathology, and review of the literature. *Skeletal radiol.* 2019;48(7):1059-67.
42. Swain SK, Sahu MC. An unusually giant myxoma of the maxilla in a child—A case report. *Pediatrics Polska.* 2016;91(5):476-9.
43. De Cristofaro R, Biagini R, Boriani S, Ricci S, Ruggieri P, Rossi G, Fabbri N, Roversi R. Selective arterial embolization in the treatment of aneurysmal bone cyst and angioma of bone. *Skelet Radiol.* 1992;21:523-7.
44. Vergel De Dios AM, Bond JR, Shives TC, McLeod RA, Unni KK. Aneurysmal bone cyst. A clinicopathologic study of 238 cases. *Cancer.* 1992;69(12):2921-31.
45. Park HY, Yang SK, Sheppard WL, Hegde V, Zoller SD, Nelson SD et al. Current management of aneurysmal bone cysts. *Curr reviews musculoskeletal med.* 2016;9(4):435-44.

46. Batische F, Schmitt A, Vendevre T, Herbreteau D, Bonnard C. Aneurysmal bone cyst: a 19-case series managed by percutaneous sclerotherapy. *Orthopaedics and Traumatol Surg Res.* 2016;102(2):213-6.

47. Palmerini E, Ruggieri P, Angelini A, Boriani S, Campanacci D, Milano GM et al. Denosumab in patients with aneurysmal bone cysts: a case series with preliminary results. *Tumori.* 2018;104(5):344- 51.