**A UNUSUAL CASE OF ANEURYSMAL BONE CYST IN MAXILLA: A CASE REPORT**

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**ABSTRACT**

Aneurysmal bone cysts of the maxilla are very uncommon, and the effective treatment is surgical excision. We present our experience of an aneurismal bone cyst found in a young girl in maxillary region. Evaluating the aneurismal bone cyst and using an external approach for removing the cyst adds upto the experience of removing these maxillary aneurismal bone cysts, with no recurrence and better cosmetic results.

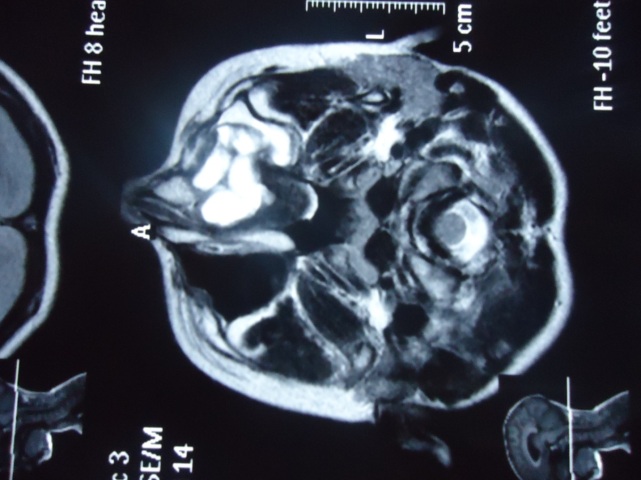
**Keywords:** Aneurysmal Bone Cyst

**INTRODUCTION**

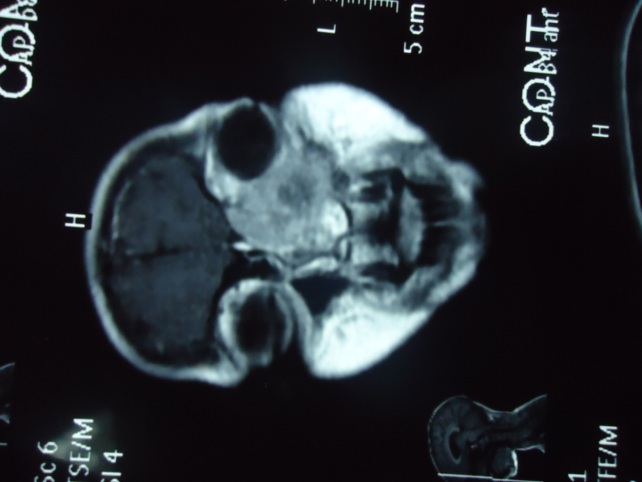
The incidence of Aneurysmal bone cyst in head and neck region is less then 4% and a cyst is maxillary region is further very rare less then 2%(3). So there may be a delay in diagnosing such a cyst. Etiologically aneurysmal bone cyst has been widely regarded as a reactive process of uncertain cause since its initial description by Jaffe and Lichtenstein in 1942. Aneurysmal bone cyst, abbreviated as ABC, is an [osteolytic](https://en.wikipedia.org/wiki/Osteolytic) [bone](https://en.wikipedia.org/wiki/Bone) [neoplasm](https://en.wikipedia.org/wiki/Neoplasm) characterized by several sponge-like blood or serum filled, generally non-[endothelialized](https://en.wikipedia.org/wiki/Endothelium) spaces of various diameters[[1]](https://en.wikipedia.org/wiki/Aneurysmal_bone_cyst).The term is a misnomer, as the lesion is neither an [aneurysm](https://en.wikipedia.org/wiki/Aneurysm) nor a [cyst](https://en.wikipedia.org/wiki/Cyst). The only symptoms of aneurysmal bone cyst would include gradually increasing swelling, with pressure symptoms on the adjacent structures and very rare pain. The skin temperature around the bone may increased. A bony swelling may be evident,and would keep on growing in size. There might be restriction of movements in adjacent joints. Aneurysmal bone cyst are common pathology seen in the long bones but in the head and neck region the incidence is very less, usually less then <4%. Commonly affected sites are [metaphyses](https://en.wikipedia.org/wiki/Metaphyses) of [vertebra](https://en.wikipedia.org/wiki/Vertebra), [flat bones](https://en.wikipedia.org/wiki/Flat_bones), [femur](https://en.wikipedia.org/wiki/Femur) and [tibia](https://en.wikipedia.org/wiki/Tibia)[[3]](https://en.wikipedia.org/wiki/Aneurysmal_bone_cyst). Approximate percentages by anatomical sites would vary from: skull and mandible (4%), spine (16%),clavicle and ribs (5%),upper extremity (21%),pelvis and sacrum (12%),femur and lower leg (24%). Aneurysmal bone cysts arise as a consequence of increase venous pressure and resultant dilation and rupture of local vascular network. Primary cause has been regarded as arterio venous fistula within the bone. Histologically, they are classified in two variants. The classic (or standard) form (95%) Has blood filled clefts among bony trabeculae. The solid form (5%) shows [fibroblastic](https://en.wikipedia.org/wiki/Fibroblast) proliferation, osteoid production and degenerated calcifying fibromyxoid elements. It is common in age group of 10–30 years. it is second most common tumor of spine and commonest benign tumor of pelvis in [pediatric](https://en.wikipedia.org/wiki/Pediatric) population. Incidence is slightly more in males than females (1.3:1).Diagnosis is based on a radiograph well-defined, expansile, lytic lesion is observed, expansion of cortex gives the balloon-like appearance. Fluid filled cavities appearing on MRI larger lesions may appear septated. Differential diagnosis include unicameral bone cyst, Giant cell tumor, Telangiectatic osteosarcoma ,Secondary aneurysmal bone cyst. Treatment is always surgical removal. [Radiation is contraindicated](https://en.wikipedia.org/wiki/Curettage) [curettage](https://en.wikipedia.org/wiki/Curettage) is performed on some patients, and is sufficient for inactive lesions. The recurrence rate with curettage is significant in active lesions, and marginal resection has been advised. Recurrence rate of solid form of tumour is lower than classic form, it is 25% in classic form.

**CASE REPORT**

A young girl ten years old girl presented in outpatient department of ENT. She was resident of district Okara. Patient was in usual state of health one year back when she developed Left side facial prominence over the left maxilla. It gradually started to increase in size. After about 2 months she also developed left sided nasal obstruction. In about six months time the swelling on the left maxillary region increased and it was accompanied with left eye ball protusion aswell. The left maxillary swelling was gradual in onset and painless. There were no associated complaints of eye except for the protusion of the eyeball. Radiological investigation including computed tomographic scan was carried showing well-defined, expansile lytic lesions giving the classic balloon-like appearance. Magnetic resonance imaging studies showed fluid filled cavities with septated Figure 1, Figure 2.

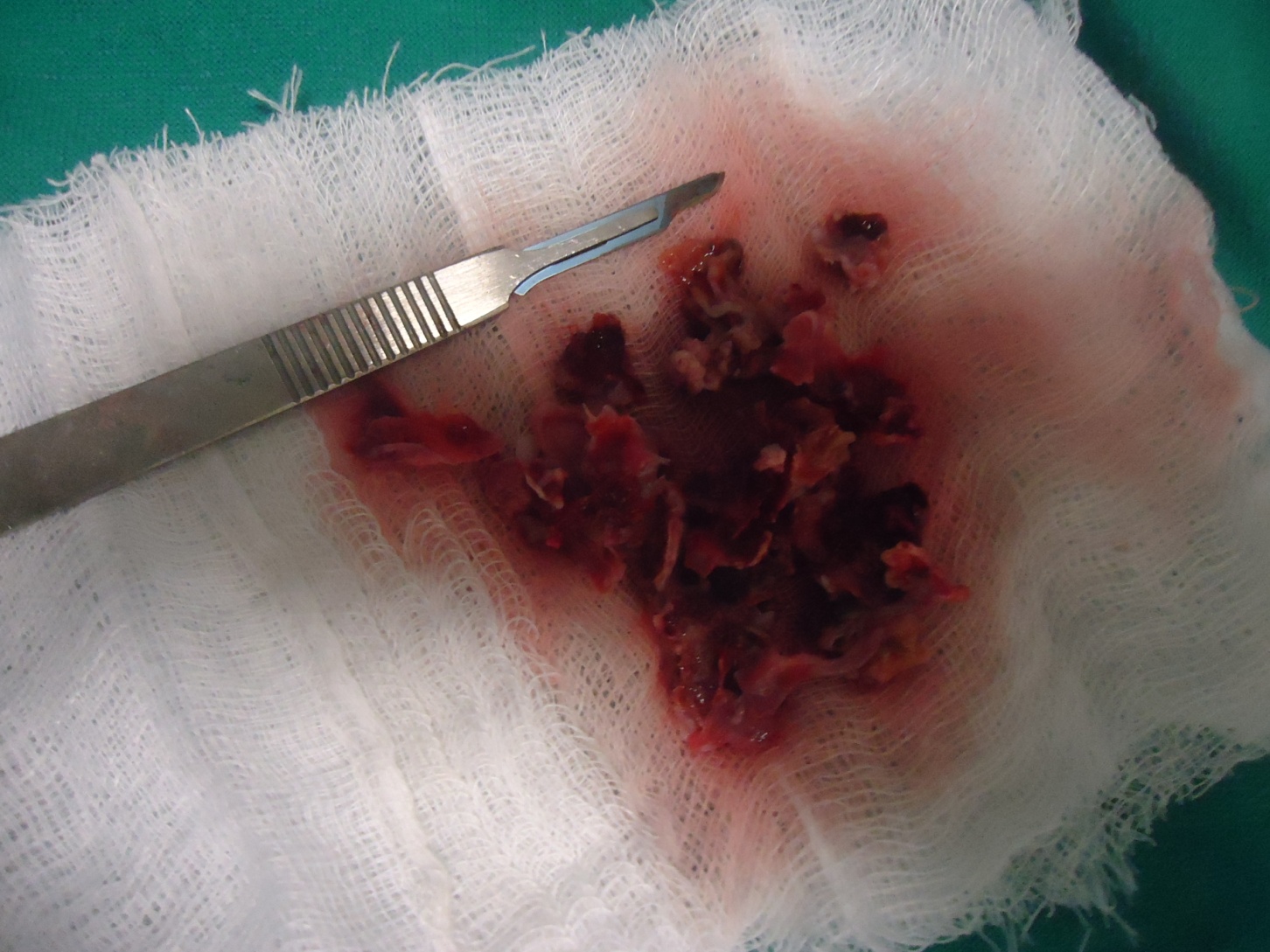


**Figure 1:** MRI Coronal View



**Figure 2:** MRI Axial View

Once the diagnosis was established the treatment was planned for marginal surgical excision using lateral rhinotomy incison . The cyst was excised intoto and the recovery of the patient was unremarkable. The histopahthology of the specimen ( Figure 3) confirmed after diagnosis, and the patient was followed up for quarterly for next two years with no recurrence.



**Figure 3:** Surgical Specimen

**DISCUSSION**

Aneurysmal bone cyst, abbreviated as ABC, is an [osteolytic](https://en.wikipedia.org/wiki/Osteolytic) [bone](https://en.wikipedia.org/wiki/Bone) [neoplasm](https://en.wikipedia.org/wiki/Neoplasm) characterized by several sponge-like blood or serum filled, generally non-[endothelialized](https://en.wikipedia.org/wiki/Endothelium" \o "Endothelium) spaces of various diameters.[[1]](https://en.wikipedia.org/wiki/Aneurysmal_bone_cyst#cite_note-sd-1)The term is a misnomer, as the lesion is neither an [aneurysm](https://en.wikipedia.org/wiki/Aneurysm) nor a [cyst](https://en.wikipedia.org/wiki/Cyst)[[1]](https://en.wikipedia.org/wiki/Aneurysmal_bone_cyst#cite_note-sd-1). The major symptoms of aneurasymal bone cyst include pain and gradual increase in the size of sweeling. The skin temperature around the bone may increase, a bony swelling may be evident, and movement may be restricted in adjacent joints.[[2]](https://en.wikipedia.org/wiki/Aneurysmal_bone_cyst#cite_note-TMJ-2). Commonly affected sites are [metaphyses](https://en.wikipedia.org/wiki/Metaphyses) of [vertebra](https://en.wikipedia.org/wiki/Vertebra), [flat bones](https://en.wikipedia.org/wiki/Flat_bones), [femur](https://en.wikipedia.org/wiki/Femur) and [tibia](https://en.wikipedia.org/wiki/Tibia).[[3]](https://en.wikipedia.org/wiki/Aneurysmal_bone_cyst#cite_note-rosai-3) Aneurysmal bone cyst has been widely regarded a reactive process of uncertain cause since its initial description by Jaffe and Lichtenstein in 1942. Many hypotheses have been proposed to explain the cause and pathogenesis of aneurysmal bone cyst, and until very recently the most commonly accepted idea was that aneurysmal bone cyst was the consequence of an increased venous pressure and resultant dilation and rupture of the local vascular network. However, studies by Panoutsakopoulus *et al.* and Oliveira *et al.* uncovered the clonal neoplastic nature of aneurysmal bone cyst. Primary cause has been regarded arteriovenous fistula within bone.[[4]](https://en.wikipedia.org/wiki/Aneurysmal_bone_cyst#cite_note-ped-4). Histologically, they are classified in two variants. The classic (or standard) form (95%) has blood filled clefts among bony trabeculae. [Osteoid](https://en.wikipedia.org/wiki/Osteoid) tissue is found in stromal matrix. The solid form (5%) shows [fibroblastic](https://en.wikipedia.org/wiki/Fibroblast) proliferation, osteoid production and degenerated calcifying fibromyxoid elements. They can also be associated with a *TRE17*/[*USP6*](https://en.wikipedia.org/wiki/USP6) translocation.[[5]](https://en.wikipedia.org/wiki/Aneurysmal_bone_cyst#cite_note-pmid20418905-5)Aneurysmal bone cysts may be intraosseous, staying inside of the bone marrow. Or they may be extraosseous, developing on the surface of the bone, and extending into the marrow. A radiograph will reveal a soap bubble appearance. On a radiograph, well-defined, expansile, lytic lesion is observed. Expansion of cortex gives the lesion a balloon-like appearance. Larger lesions may appear septated[[6]](https://en.wikipedia.org/wiki/Aneurysmal_bone_cyst" \l "cite_note-who-6). Following conditions add to the differential diagnosis of aneurasymal bone cyst:[[7]](https://en.wikipedia.org/wiki/Aneurysmal_bone_cyst" \l "cite_note-ddsp-7)Unicameral bone cyst, Giant cell tumor, Telangiectatic osteosarcoma, Secondary aneurysmal bone cyst. The treatment of ABC can include [Curettage](https://en.wikipedia.org/wiki/Curettage) is performed on some patients[[8]](https://en.wikipedia.org/wiki/Aneurysmal_bone_cyst" \l "cite_note-pmid16170183-8).We have preferred the external approach in our patients due to the size and location of the cysts in the maxillary regions. Follow-up for 2 months period revealed that the patient was treated without any complications. The only disadvantage was the incisional scar.

**CONCLUSION**

Aneurysmal bone cysts are very rare, so most otolaryngologists surgeons will probably see few cases in their lifetime. In order to a guideline for surgeons, we can say that these cysts can be easily dissected from surrounding tissues and excised totally. Therefore, no excessive surgery is required, so, in our opinion, external approach and complete excision of the cyst with no recurrence is important.

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