Case Report
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# Multidisciplinary management of a rare aneurysmal bone cyst of the temporal bone: A case report

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## Abstract

Aneurysmal bone cyst (ABC) is a benign tumor-like lesion that is described as "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. Although they can occur in any bone, they are most common in the femur, tibia, and vertebrae. Aneurysmal bone cysts of the skull are unusual lesions but can occur in the different parts of the skull bone. We describe here a 15 years old female from the rural part of Ethiopia who presented with a right-side temporal aneurysmal bone cyst that grew for four years.

**Keywords:** Bone neoplasm, Aneurysmal bone cyst, Temporal bone

## Background

Aneurysmal bone cysts (ABC), according to the World Health Organization histological typing of bone tumors, are defined as benign lesions with blood-filled spaces and connective tissue septa containing osteoid tissue and osteoclast giant cells (1). Even though, ABC are neither an aneurysm nor a cyst, rapid growth and expansion of the cancellous and cortical bones resulting in “ballooned out distension” of the periosteum creates the wrong perception of malignant growth in these benign lesions. ABC are commonly seen in long bones or vertebrae of patients younger than 30 years of age (1–5), with slight female predominance (Male: Female: 1.04-1.18) (4, 6). Aneurysmal bone cysts in the skull bone are unusual lesions accounting for 3-6% of ABC but have been described in the literature as occurring in the frontal, temporal, parietal, occipital, and base of the skull bone. (3,4,7,8).

In a systematic review of ABC of craniofacial origin by Rehman et al (9), the male-to-female ratio in long bones was reversed with a slightly more predominance in male patients for ABC of the cranial bone (Male: Female (1.17:1). These ABC are commonly extradural but can present as intradural lesions extending to the subdural space and these lesions have dire prognosis (7). Skull aneurysmal bone cysts are benign, but expansive and locally aggressive lesions of the bone that can present as isolated rapidly expanding tender bony masses with intact scalp skin or be associated with generalized headaches. Basal ABC on the contrary can present with raised intracranial pressure, proptosis, or associated cranial nerve palsies (7, 10).

Diagnostic tests for ABCs include plain radiographs which show an expansive cystic lesion with a honeycomb or a soap-bubble appearance and computed tomography scan which delineates the bone architecture of the tumor, a thin cortex of newly formed bone surrounding the multiloculated lesion originating in the diploe and expanding both intracranially and extracranially which can later be confirmed with histopathology examination (3,5,7,8). Histological examination shows blood-filled spaces surrounded by compressed fibrous tissue rather than endothelium. Multinucleated giant cells of the osteoclast type throughout the fibrous stroma are also common (4,5). Although there are reports of spontaneous regression in the literature (4), complete surgical excision of the mass is the treatment of choice because of the high recurrence rate of up to 59% when simple curettage was done (4,5,8). The ABC are vascular tumors and are liable to bleed during surgery (3). Treatment by surgical excision ranges from enucleation and curettage to a conservative resection. Total removal is associated with a cure and an excellent long-term outcome (4). Recurrence in the skull is rare unlike ABC in other sites (11) and recurrences can be treated with radiotherapy (12). Here, we describe a 15 years old female who presented with a right-side temporal aneurysmal bone cyst that progressively grew for four years.

## Case description

A 15-year-old female adolescent presented to the neurosurgery clinic with a four years history of progressively growing painful mass over her right temporal area associated with decreased hearing in the right ear, tinnitus, and intermittent headache. There was no history of any trauma, ear discharge, vertigo, nausea, vomiting, or seizures. On physical examination, there is a 6cm X 6cm hard mass on the right temporal region that is tender to palpation and fixed to the underlying bone but with no fixity to the overlying skin. All other neurologic examinations were normal except Weber's test lateralized to the right ear and Rinne’s test that showed (Air conduction) AC>BC (Bone conduction) suggesting a conductive hearing loss. She also had House Brackmann Grade II facial palsy on the right side. A non-contrast CT scan of the head (Figure 1-3) revealed 8.5x7.3x8.7cm extra-axial expansile multiloculated osteolytic mass with an erosion of the petrous, tympanic and mastoid parts of the temporal bone which appeared thinned out and scalloped with a complete blockade of the external auditory meatus.

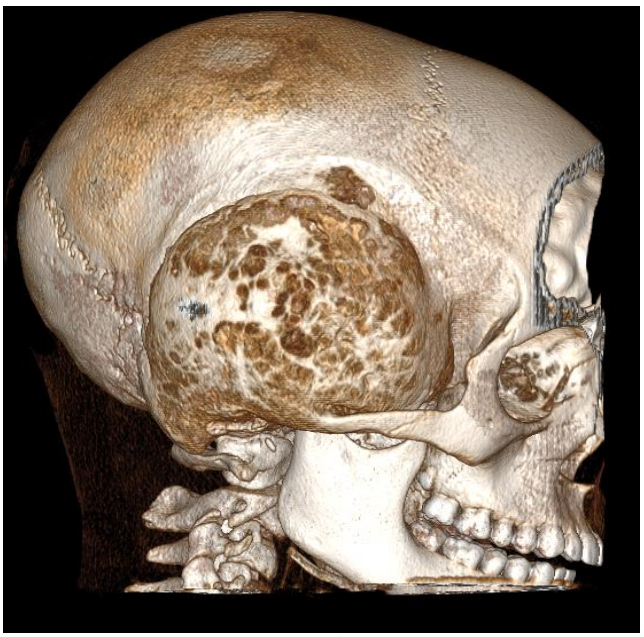


**Figure 1** - a 3D reconstructed image of the right temporal ABC

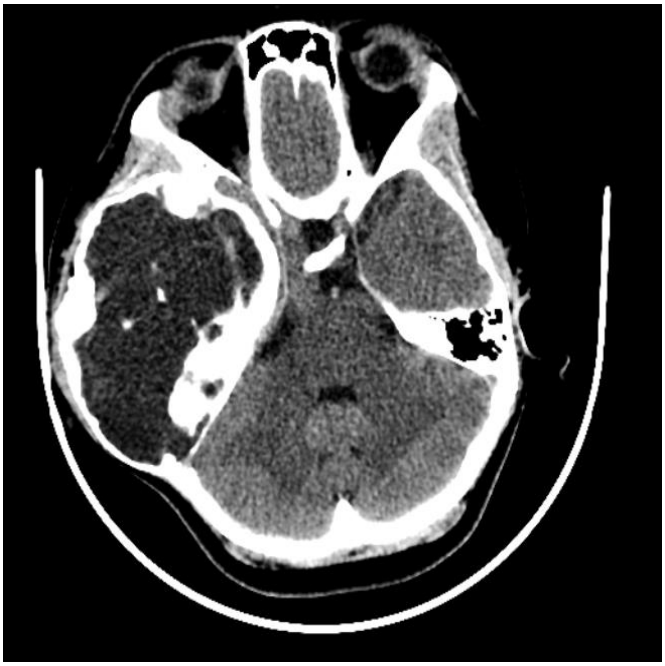


**Figure 2-** Brain window of CT scan of right temporal ABC

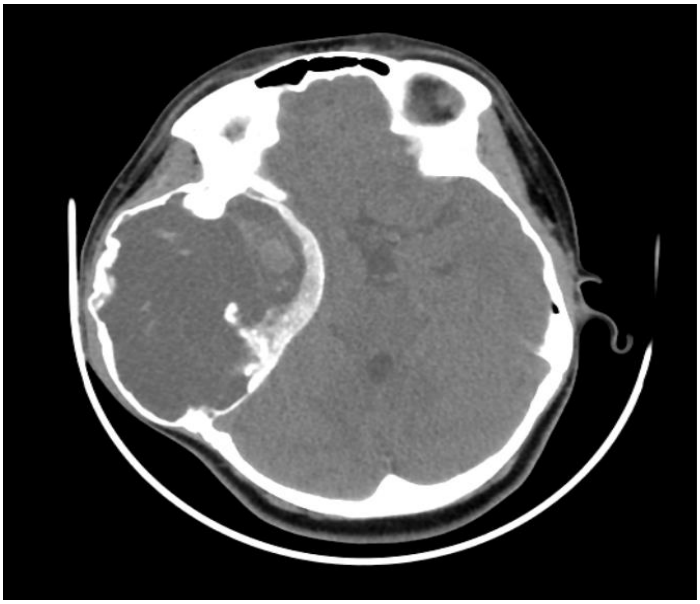


**Figure 3-** A bone window of CT scan of Right temporal ABC

The tumor was approached with a temporo-frontal incision (orbit-zygomatic approach) and gross total excision of the cyst was done. Skull bone appeared thinned out and fragile with greenish and hemorrhagic multi-cystic lesions that eroded the middle fossa floor. Postoperatively, there was a significant improvement in the temporal swelling but similar findings of hearing tests and no worsening of the facial palsy. After 10 days of a smooth postoperative course, the 8.5cm x 7.5 cm bone defect (Figures 4 and 5) was reconstructed using autologous split rib grafts from the right sixth and seventh ribs (Figure 6) by the plastic and reconstructive surgical team. The patient had a smooth post-operative course and has not returned to the hospital for further care 2 years post-surgery. Good health and no recurrence were reported on phone follow up calls conducted at three, six and twelve months.

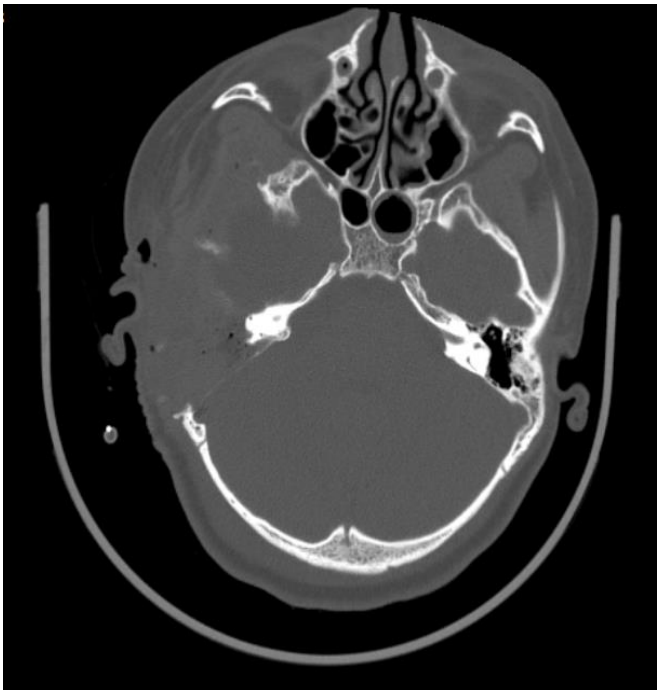


**Figure 4-** Postoperative bone defect after excision of Right temporal ABC

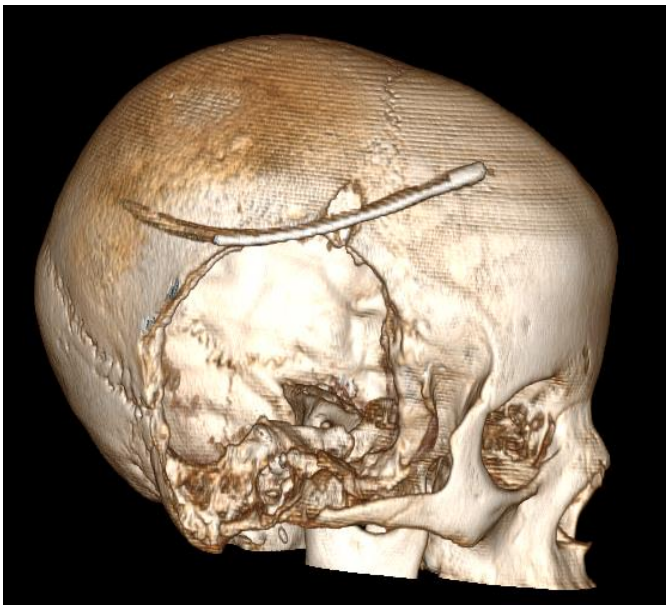
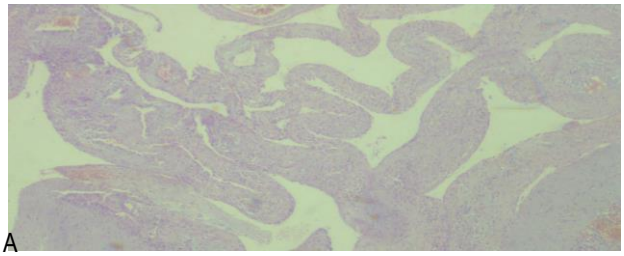


**Figure 5** - 3D reconstruction of CT scan depicting cranial defect postoperatively

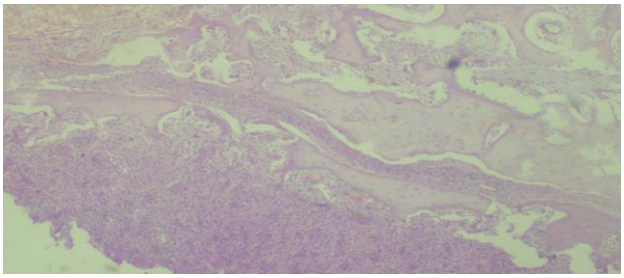
## Discussion

Aneurysmal bone cyst was first described in 1942 by Jaffe and Lichtenstein as a rare and benign disease that shows rapid growth, with osteolysis and cystic lesions that appear as an expansive arterial aneurysm (13). ABC are commonly seen in the long bones and vertebrae and rarely occur in the cranial bones (3,10). The pathogenesis of ABCs has not yet been clearly described but it has been hypothesized to occur primarily as a congenital or acquired vessel wall anomaly of intraosseous blood vessels that occurs as a result of a chromosomal translocation t(16;17) (q22;p13)(5) or as a secondary lesion occurring on pre-existing trauma site, chondroblastoma, giant cell tumor or fibrous dysplasia(3,5). The clinical presentation of craniofacial ABC will depend on the cranial bone from which it arises and can range from tender or non-tender masses, and worsening headaches, to decreased hearing and variable levels of visual disturbances. (9) The site of the mass on the temporal bone and the erosion of petrous and mastoid parts of the temporal bone and physical blockade of the external ear canal in our patient explain the presenting symptoms of tender mass, hearing loss, and facial palsy that will gradually improve with surgical removal of the mass.

Histologic examination confirmed the diagnosis revealing irregular cystic spaces separated by cellular septae containing fibroblast and blood vessels (Figure 6A) and a solid region of cyst abutting normal bone architecture (Figure 6B).



A



B

**Figure 6.** Histologic examination showing A- Irregular cystic spaces separated by cellular septae containing fibroblast and blood vessels. B Solid region of cyst abutting normal bone architecture

Surgical excision of the tumor is the preferred treatment for ABC (4,5,7,8) which was also done in our patient that resulted in a skull defect measuring 8.5x7.5cm that was later reconstructed by autologous rib graft (Figure 7, 8) to recreate a protective physical barrier, a natural convex contour of the calvarium which was a good autologous calvarial reconstruction option (14) in the absence of alternative alloplastic materials in a resource-limited setup like ours.

**Figure 7-** Intraoperative picture of split rib graft used to reconstruct the cranial defect

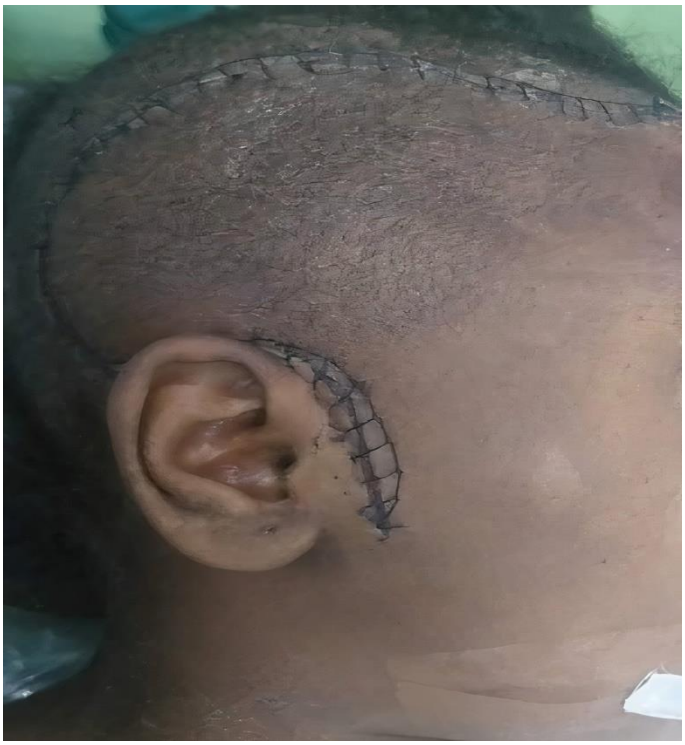


**Figure 8** – Post operative picture after reconstruction with the split rib graft.

## Conclusion

Aneurysmal bone cysts of the skull are unusual lesions but can occur in the different parts of the skull bone. We have presented here a temporal aneurysmal bone cyst that was surgically excised and the defect was reconstructed with an autologous rib graft. The patient did not experience any recurrence or complications justifying the need for surgical excision of these lesions.

## Declarations

### Consent for publication

Patient consented to the publication of this case report in the Millennium Journal of Health.

### Ethical declaration

Informed written consent was obtained from parents/primary caregivers.

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### Competing interest

All authors read and approved the final manuscript. The authors declare that they have no competing interests.

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