

Aneurysmal Bone Cyst of the Occipital Bone

Javed Mehboob¹, Uzma Aleem¹ and Asghar H. Asghar²

ABSTRACT

A 13 years boy presented with a painless hard and fixed swelling in occipital region for the last three months. Plain X-ray, CT scan and MRI showed an expansile multi loculated cystic lesion in occipital bone. Histopathological examination revealed it to be an aneurysmal bone cyst. Treatment of choice is surgery. However, radiotherapy may be helpful in incompletely excised lesions.

Key words: Aneurysmal bone cyst. Occipital bone. Radiotherapy.

INTRODUCTION

An aneurysmal bone cyst is a benign expansile osteolytic lesion with a thin wall, containing blood-filled cystic cavities. The term aneurysmal is derived from its radiographic appearance.¹ Aneurysmal bone cyst (ABC) lesion most often affects persons during their second decade of life. It may occur in any bone in the body. Although benign, it can be locally aggressive and cause extensive weakening of bony structure. It impinges significant pressure on the surrounding tissues. Its true etiology and pathophysiology is not yet clear. Treatment of choice is surgery. However, its total surgical removal can be done in one or multiple stages. Prognosis is excellent with complete removal but incomplete removal may affect its cure rate.²

This case report describes this tumor in occipital bone.

CASE REPORT

A 13 years old boy presented with gradually increasing painless hard swelling in the occipital region for the last 3 months. There was no history of trauma. On physical examination, it was a non-tender approximately 6 x 5.5 x 5 cm mass in the left occipital region, which was hard and fixed to the bone having a smooth surface. Skin over the swelling was normal. There was no neurological deficit.

All routine investigations were within normal limits. A plain X-ray skull showed expansile lesion in the occipital bone with internal thin septae of ossification. CT scan revealed expansile lytic lesion in occipital bone with periosteal thinning and cortical break (Figure 1).

Department of Radiology¹/Clinical Oncology², Karachi Institute of Radiotherapy and Nuclear Medicine (KIRAN), Karachi.

Correspondence: Dr. Javed Mehboob, Consultant Radiologist, Karachi Institute of Radiotherapy and Nuclear Medicine (KIRAN) Hospital, Gulzar-e-Hijri, KDA Scheme 33, Off University Road, Karachi.
E-mail: jmehboob69@yahoo.com

Received October 12, 2009; accepted August 17, 2011

It showed internal septation giving a soap-bubble appearance. Intracranial extension of the lesion was exerting pressure over the cerebellum. On T2 weighted images of MRI scan revealed foci of increased signal intensity in occipital bone with loculation and contained multiple fluid-fluid levels (Figure 2). Lesion extended into the cranial cavity and displaced the adjacent

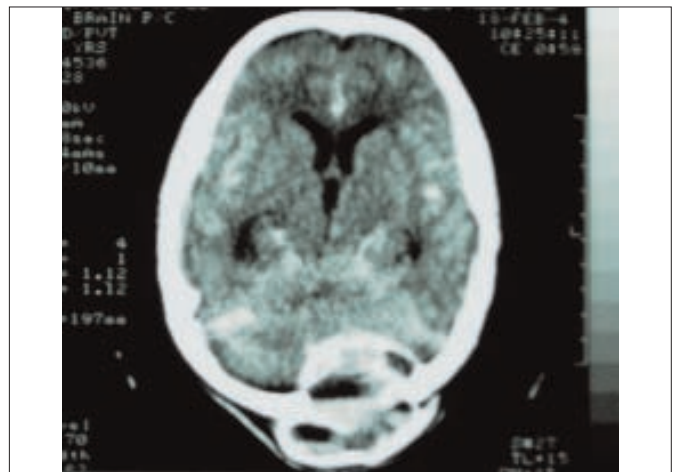


Figure 1: CT scan showing large expansile lytic lesion with internal septation in occipital bone.

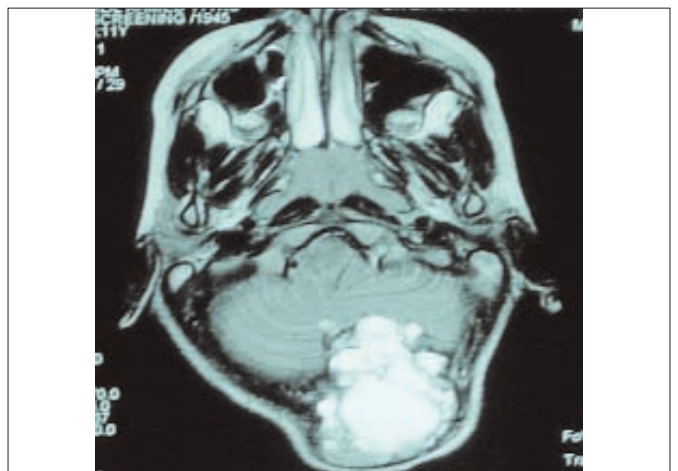


Figure 2: T2 weighted MRI axial image showing hyperintense expansile lesion with internal septation in the occipital bone.

cerebellum and causing compression of the 4th ventricle. Total excision of the tumor was done. The patient was symptom-free in the postoperative period after 6 months and kept under close follow-up.

DISCUSSION

Aneurysmal bone cyst is a rare benign bone lesion. Its occurrence is about 1% of bone tumors. It is blood filled fibrous tumor-like cyst that expands the bone giving it a blow out appearance. It most frequently occurs in second decade of life and can emerge in any bone in the arms, legs, trunk or skull. The vertebrae and knee are the most common sites of occurrence. The metaphysis of the long bones is the usual site of origin.³

An aneurysmal bone cyst is a benign bone tumor but it can be quite destructive locally and has a high propensity for recurrence.

The incidence of aneurysmal bone cyst in the skull and mandible varies from 3 to 6% of all aneurysm bone cysts. It usually presents as scalp mass but may present as an intracranial space occupying lesion or cerebral hemorrhage. A majority of aneurysmal bone cysts occur in the temporal and very rare in occipital bones.⁴

Exact pathogenesis of aneurysmal bone cyst is unknown. The concept of aneurysmal bone cysts arising to preceding benign or malignant lesions is now generally accepted. The original lesion can be identified in only one-third of cases. The most common precursor lesion is giant cell tumor (19-39%) followed by osteoblastoma, angioma and chondroblastoma. Less common are fibrous dysplasia, non-ossifying fibroma, chondromyxoid fibroma, unicameral bone cyst, fibrous histiocytoma, eosinophilic granuloma and osteosarcoma.⁵

In 1942, Jaffe and Lichtenstein introduced the concept of aneurysmal bone cyst as a lesion with characteristic radiological appearance of ballooned out distension of the periosteum. It is usually outlined by a paper-thin subperiosteal bone shell which is overlined by a region of disintegrated cortex.

On plain film, aneurysmal bone cyst is normally placed eccentrically in the metaphysis and appears osteolytic. The periosteum is elevated and cortex is eroded to a thin margin. Expansile nature of the lesion is often reflected by a "blow-out" or "soap bubble" appearance. In vault bone the lesion originates within the diploic space causing expansion and displacement of the inner and outer tables of the skull. CT and MRI showed multiloculation of the cyst. These investigations are useful to conform the diagnosis by demonstrating the characteristic fluid level within the cyst. The content of this is partly cystic and partly solid with the presence of fluid levels in the most cases. The solid component of the tumor usually enhances with contrast injection. MRI

can also confirm the multiple fluid levels and non-homogeneity of the lesion because of the septa and variability in breakdown of blood products. Presence of paramagnetic blood breakdown products give rise to fluid levels of varying signal intensities ranging from very bright signal (extracellular methemoglobin) on T2 weighted imaging to a very low signal (intracellular deoxyhaemoglobin, cellular debris or haemosiderin).⁶

Histologically the lesion is characterized by the presence of multiple, filled with venous blood and lined by spindle shaped fibroblasts and with scattered multinucleated giant cells and stromal cells.⁷

Treatment of aneurysmal bone cyst will be one or the combination of many procedures.

Curettage / bone grafting is the most common form of treatment for an aneurysmal bone cyst and has a 20-40% recurrence rate.³ In this procedure, the cyst is completely scraped out, chance of recurrence is high so it must scrape aggressively and the remaining cavity is then packed with donor bone tissue (allograft), bone chips taken from another bone (autograft) or other materials.⁸

Marginal or wide excision of the host bone is the surgical procedure of choice. Wide excision is recommended when the cyst is located in a bone considered expendable, such as the ribs or fibula.⁸

Due to the risk of recurrence associated with these cysts, adjuvant therapies, such as cryotherapy, a surgical freezing of the cyst are sometimes considered. Cryotherapy is associated with complication such as fracture of the bone, nerve injury and other, so is not widely accepted as standard in many institutions. Radiotherapy is used only if other means of treatment have failed.

Treatment of choice for aneurysmal bone cyst of skull is total excision of lesion. Due to vascular nature of the cyst there is some potential risk of bleeding during surgery. So pre-operative arterial embolization may reduce the risk of bleeding.^{4,5} However, surgical biopsy has itself been curative. Tendency for recurrence is related with young age of the patient, size of lesion, presence of mitosis and incomplete surgical removal. Radiotherapy has been advocated for deep situated lesions at the base of skull, dural involvement or where subtotal excision has been done but its effect is unclear.⁶ There is no role for chemotherapy.

REFERENCES

1. Ameli NO, Abbassioun K, Azod A, Saleh H. Aneurysmal bone cyst of the skull. *Can J Neurol Sci* 1984; **11**:466-71.
2. Kumar S, Retnam TM, Krishnamoorthy T, Parameswaran S, Nair S, Bhattacharya RN, *et al.* Intracranial aneurysmal bone cyst manifesting as a cerebellar mass. *Neurol India* 2003; **51**: 121-3.

3. Calliauw L, Roels H, Caemaert J. Aneurysmal bone cysts in the cranial vault and base of skull. *Surg Neurol* 1985; **23**:193-8.
4. Keuskamp PA, Horoupian DS, Fein JM. Aneurysmal bone cyst of the temporal bone presenting as a spontaneous intracerebral hemorrhage: case report. *Neurosurgery* 1980; **7**:166-70.
5. Lichtenstein L. Aneurysmal bone cyst; observations on fifty cases. *J Bone Joint Surg Am* 1957; **39-A**:873-82.
6. Zimmer WD, Berquist TH, Sim FH, Wold LE, Pritchard DJ, Shives TC, *et al.* Magnetic resonance imaging of aneurysmal bone cyst. *Mayo Clin Proc* 1984; **59**:633-6.
7. Cataltepe O, Inci S, Ozcan OE, Sa_lam S, Erben A. Aneurysmal bone cyst of the frontal bone. *Surg Neurol* 1990; **33**:391-4.
8. Wheeless CR. Wheeless' textbook of orthopaedics [Online]. Durham: Duke University Medical Centre; 2011. Available from: <http://www.wheelessonline.com/>
9. Probst C. [Clivus C2 aneurysmal bone cyst: contribution to problems of surgical treatment, radiotherapy and individually determined growth trends]. *Neurochirurgia (Stuttg)* 1990; **33**:195-8. German.

