

Aneurysmal Bone Cyst Of The Temporal Bone - A Pathological Diagnostic Dilemma

M SALEK, M ISLAM

Citation

M SALEK, M ISLAM. *Aneurysmal Bone Cyst Of The Temporal Bone - A Pathological Diagnostic Dilemma*. The Internet Journal of Neurosurgery. 2013 Volume 9 Number 1.

Abstract

Aneurysmal bone cyst (ABC) is a benign osteolytic bone neoplasm characterized by blood filled spaces separated by fibrous septa. The tumor can have an occurrence anywhere that there is bone. It occurs mostly in extremity bones, whereas in skull and mandible bones its incidence is about 4%. In this article an unusual case of aneurysmal bone cyst in the right temporal region with histopathological diagnostic dilemma has been reported.

INTRODUCTION

Aneurysmal bone cysts are expansile, lytic, bone lesions consisting of multiple thin walled, cystic cavities containing blood. Aneurysmal bone cyst is a rare and benign lesion of the skull with obscure pathogenesis.¹ Jaffe and Lichtenstein coined the term in 1942, to describe a peculiar bone lesion with a vascular lining and characteristic “soap bubble” radiological picture of expanded bone.⁶ Aneurysmal bone cyst is a rare lesion of the skull with obscure pathogenesis.⁸ Approximately, 36 to 50% of all aneurysmal bone cysts are found near one end of long bone and 25% involve the vertebrae, hyoid, mandible and odontoid.^{18,23} Sixty-three cases of aneurysmal bone cysts of skull have been reported in the literature, of which 11 were of temporal bone.¹⁵ An unusual case of aneurysmal bone cyst in the right temporal region is reported here.

CASE REPORT

An 18 year old boy was admitted with headache, right earache and gradually increasing painless swelling in the right temporal region for 3 months. There was no history of trauma. On examination there was a non-tender ill-defined mass, in the right temporal region; which was firm to hard and fixed to bone with smooth surface. Skin over the swelling was normal. There was no neurological deficit. All routine investigations were within normal limits.

Figure 1

X-ray skull revealed a radiolucent area in the right temporal bone.

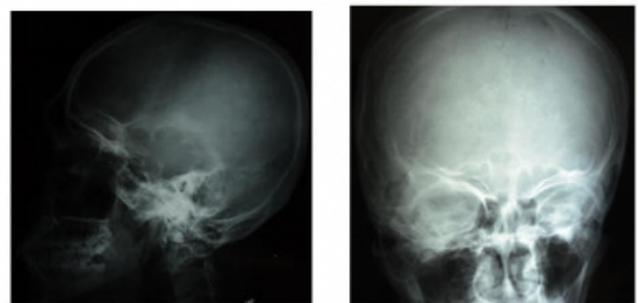


Figure 2

CT scan head (plain and enhanced) showed extra axial heterogeneous mixed density mass in the right temporal region, taking irregular variegated enhancement. The mass showed medial extension compressing left temporal lobe. There was erosion of petrous part of temporal bone.

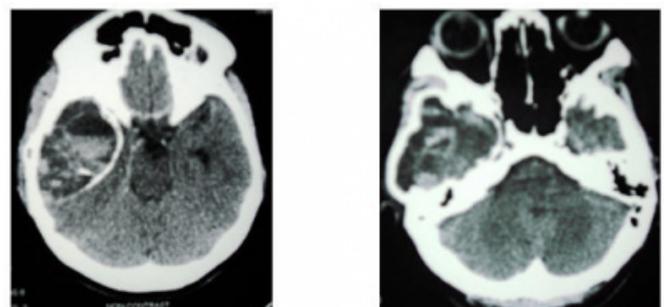
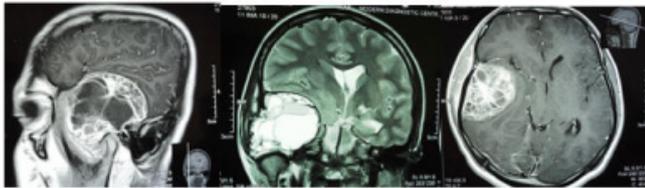


Figure 3

MRI showed large extra axial T1 intermediate signal intensity with small hyperintense areas and T2- FLAIR hyperintense mass (about 06cmx07cm) in the right temporal region. No significant perifocal oedema. The mass shows multiple fluid-fluid levels. Overlying extracranial soft tissues show swelling and altered signal intensity. Involvement of overlying calvari.



On post gado T1W1, the mass shows heterogenous contrast enhancement. The overlying bone and extra calvarial soft tissues also show moderate contrast enhancement.

A right temporal question mark skin flap was raised with temporalis muscle. The underlined temporal bone was thinned out with erosion of petrous and greater wing of sphenoid. The tumor was highly vascular had well defined capsule and was soft in consistency. Dark coloured blood was found from the cyst. The squamous part of temporal bone was thinned out and temporal dura was adherent to it, which remained intact. Inferiorly, tumor mass was extending upto the floor of middle cranial fossa and posteriorly to the petrous bone. Gross total excision of the tumor was done. Postoperative recovery was uneventful.

Histopathological examinations from different histopathologists revealed:

1. Specimen showed bone and soft tissue. These contain many spaces filled up with blood. Some of the spaces are empty. These spaces are lined by plump cells. The stroma contains osteoid. In some areas, calcification seen. In other areas, haemosiderin laden macrophages are seen. A good number of osteoclastic giant cells are seen. The stromal cells are spindle shaped. Features are compatible with aneurysmal bone cyst.

2. Sections made from submitted specimen showed meningotheial cells arranged in whirled pattern. Focal areas of haemorrhage and bony tissue are also seen. Findings are consistent with meningioma.

DISCUSSION

This case report has special emphasis on histopathology reports which brought into a diagnostic dilemma. This may be related with the sample from the specimen containing bony component as well as meningeal component. But the patient has been treated and folloed up as a case of

aneurysmal bone cyst.

Aneurysmal bone cyst is a benign, non-neoplastic lesion that presents most frequently under the age of 20 years.^{18, 5, 21} The involvement of the skull is rare. The incidence of aneurysmal bone cyst in the skull varies from 3 to 6% of all aneurysm bone cysts.^{3, 13, 17} It usually presents as scalp mass.^{9, 4, 14}

Occasionally, they may present as intracranial space occupying lesion⁷ or cerebral hemorrhage.¹⁴ Age of presentation varies, but they usually present in the second or third decade, with equal sex distribution.^{23, 15} In 1942, Jaffe and Lichtenstein⁶ introduced the concept of aneurysmal bone cyst as a lesion with characteristic radiological appearance of ballooned out distension of the periosteum, usually outlined by a paper thin subperiosteal bone shell which is overlined by a region of disintegrated cortex. Trauma has been put forward as an important etiological factor.²² Edling⁸ regarded aneurysmal bone cyst as one of the manifestation of solitary dysfibroplasia of bone, suggesting a defect in development of the epiphyseal plate but it does not explain its occurrence in the mature bone. Lichtenstein¹⁸ suggested that it could result from local circulatory disturbance, because of sudden vascular occlusion of venous drainage of that segment of bone or development of an arterio-venous shunt. This results in progressive blood spaces in the medulla, which lead to gradual distension of the bone with atrophy. Jaffe¹¹ reported that a pre-existing lesion of bone may initiate an osseous A-V fistula.

Aneurysmal bone cyst in coexistence with other lesions of bone have been reported^{18, 2, 20} and include unicameral bone cyst, non-ossifying fibroma, gaint cell tumor, chondroblastoma, fibrous dysplasia, osteofibrous dysplasia of campanacci, fibrous histiocytoma osteoblastoma and cartilaginous hematoma of chest wall of infants.

CT scan is superior to plain radiology in defining extent and soft tissue extension of an aneurysmal bone cyst, particularly in the skull. Multiple small fluid levels are important characteristics of aneurysmal bone cyst on CT scan, which represents sedimentation of red blood cells within blood filled cavities.¹⁰ MRI also shows fluid levels, particularly in T1W1.¹² Other findings include prolonged relaxation time, complete delineation of the margin of the lesion by a rim of low intensity signal and internal septation creating cystic cavities where wall contains diverticulum like projections.

Pathologically, these cysts contain multiple fluid filled cavities separated by multiple septa lined by multinucleated

giant cells. These large spaces filled with blood do not have an endothelial lining, but are rather delimited by cells with the morphology, ultrastructural and immunohistochemical features of fibroblasts, myofibroblasts and histiocytes.

Aneurysmal additional feature of a peculiar degenerated calcifying fibromyxoid tissue reported by Rosai.²⁰ The risk of recurrence is increased with an increase of mitotic figures.

The treatment option of choice for these lesions is total excision when possible, which is curative^{7, 14} but difficult to accomplish in cases where the skull base is involved.¹⁶ In these cases, partial resection and curettage of the inner contents may suffice to halt progression of the disease or even cause regression, but there may be recurrence. As these lesions are non-neoplastic and benign, the use of radiation therapy is not recommended, although it is reported in the literature.¹⁹ There is no role for chemotherapy.

CONCLUSION

The histopathology of this temporal region mass – aneurysmal bone cyst vs meningioma remains a diagnostic dilemma in the final diagnosis. But the patient has been managed as aneurysmal bone cyst of temporal bone. His postoperative recovery is uneventful without any recurrence in subsequent follow up.

References

1. Edling NPG: Is the aneurysmal bone cyst a true pathological entity? *Cancer*; 1965; 18: 1127-1130
2. Biesecker JL, Marcove RC, Huvos AG, Miké V: Aneurysmal bone cyst- A clinicopathologic study of 66 cases. *Cancer*; 1970; 26: 615-625.
3. Bhende YM, Kothare SN: Aneurysmal bone cyst- A case report. *Indian Med Gaz*; 1950; 85: 544-546.
4. Burns-cox CJ, Higgins AT: Aneurysmal bone cyst of the frontal bone. *J Bone Joint Surg*; 1969; 51B: 344-345.
5. Calliauw L, Roels H, Caemaert J: Aneurysmal bone cysts in the cranial vault and base of skull. *Surg Neurol*; 1985; 23:193-198
6. Jaffe HL, Lichtenstein L: Solitary unicameral bone cyst with emphasis on the roentgen picture, the pathologic appearance and the pathogenesis. *Arch Surg*; 1942; 44:1004-1025
7. Costantini FE, Irad G, Benedetti A: Aneurysmal bone cyst as an intracranial space occupying lesion. *J Neurosurg*; 1966; 25: 205-207.
8. Edling NPG: Is the aneurysmal bone cyst a true pathological entity? *Cancer*; 1965; 18: 1127-1130.
9. Giovanni L, Maurizio F, Mario S: Angiography and computerized tomography in the diagnosis of aneurysmal bone cyst of the skull, Case report. *J Neurosurg*; 1980; 53: 113-116.
10. Hudson TM: Fluid levels in aneurysmal bone cyst - A CT feature. *Am J Radiol*; 1984; 142: 1001-1004.
11. Jaffe HL: Discussion following a paper by W.F. Donaldson Jr. Aneurysmal bone cyst. *J Bone Joint Surg*; 1962; 44A: 25-40.
12. Javier B, Donald CS, Micheal L: Aneurysmal bone cyst: MR imaging at 1.5 T. *Radiology*; 1986; 158: 689-690.
13. Jeremiah BS: Aneurysmal bone cyst of the temporal bone. *J International Coll surgery*; 1965; 43: 179-183.
14. Keuskamp PA, Horouptian DS, Fein JM: Aneurysmal bone cyst of the temporal bone presenting as a spontaneous intracerebral hemorrhage - Case report. *Neurosurgery*; 1980; 7:166-170.
15. Kumar R, Mukherjee KK: Aneurysmal bone cyst, of the skull - Report of three cases. *Br J Neurosurg*, 1999; 13: 82-84.
16. Levy RA, Quint DJ, Devaney KO: Unusual tumours of the skull Base - A pictorial essay. *Can Assoc Radiol*; 1996; 47:434-443
17. Lichtenstein L: Aneurysmal bone cyst- Further observations. *Cancer*, 1953; 6: 1228-1237.
18. Lichtenstein L: Aneurysmal bone cyst - Observations of fifty cases. *J Bone Joint Surg*; 1957; 39A: 873-882.
19. Probost C: Clivus C2 aneurysmal bone cyst - Contribution to problem of surgical treatment, radiotherapy and individually determined growth trends. *Neurochirurgie*; 1990; 33: 195.
20. Rosai J: *Ackerman's surgical pathology*. CV Mosby Co. London; 1989.
21. Rutter DJ, Van Rijseel THG, Velde EA: (1977) Aneurysmal bone cyst - A clinicopathological study of 105 cases. *Cancer*; 1977; 39: 2231-2239.
22. Thompson PC: (1954) Subperiosteal giant cell tumor ossifying subperiosteal hematoma aneurysmal bone cyst *J Bone Joint Surg*; 1954; 36A: 281-306.
23. Tillman BP, Dahlin DC, Lipscomb PR, Stewart JR: (1968) Aneurysmal bone cyst - An analysis of ninety five cases. *Mayo Clin Proc*; 1968; 43: 478-495.

Author Information

MAA SALEK, MBBS, MCPS, FCPS, MRCS

Division of Neurosurgery, National University Hospital Singapore
Singapore

MA ISLAM, MBBS, MS

Department of Neurosurgery, Combined Military Hospital
Bangladesh, India