



ANEURYSMAL BONE CYST

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ABSTRACT

An aneurysmal bone cyst in frontal region is rare. They are benign skull tumors and may also extend intracranially. Total surgical removal is done, whenever possible, in one or in multiple stages. Prognosis is excellent provided complete removal is obtained. An unusual case of aneurysmal bone cyst of frontal region is reported.

KEYWORDS: Aneurysmal bone cyst, Cranioplasty, Benign skull tumors.

INTRODUCTION

Aneurysmal bone cyst is a rare and benign lesion of the skull with ambiguous pathogenesis. It may occur in any part of the skeleton. Long bones and vertebrae are the most common sites, and the skull is affected rarely.^[27] The afflicted bone becomes expanded and ballooned with many communicating cavities enclosing venous blood. Although most of the spaces are filled with unclotted venous blood under great pressure, some may contain sanguinous or even clear fluid. It usually affects the young, 90 percent being less than 20 years of age. It grows rapidly. When the skull is affected, signs and symptoms are related to the location of the lesion. An unusual case of aneurysmal bone cyst in the right frontal bone is reported here.

CASE REPORT

A 28 year old Asian male, presented with a history of a swelling noted on right side of top of skull following a trivial trauma 3 years ago. The swelling was painless and small in the beginning. It grew very slowly over the next three years to the present size. Occasional mild pain was experienced in the swelling at times. Patient experienced mild to moderate global headaches from time to time which were relieved by common pain killers. There was no discharge noted from the swelling anytime. The swelling was consistent and gradually progressive with no waxing and waning in the size. There were no other swellings complained over skull or body. There were no associated complaints like fever, weight loss, loss of appetite etc. The male had no significant past or familial histories and was on no regular medications for any other illnesses or for this swelling. As the growing size was a

concern, he consulted a local family doctor who advised a CT scan and subsequently the patient was referred to a neurosurgery consultation.

On examination, the patient was a healthy looking adult male of 28 years, well oriented to time, place and person, with normal vital signs. The general examination was unremarkable and there were no skin stigmata. On examination of the swelling, a 4x2x2.5 size swelling was noted on right high parietal parasagittal location just abutting the midline with indistinct margins, merging of the edges into the surrounding calvarium. It was soft in the center and hard in periphery. It was immobile and fixed to the skull. It was non tender. The skin over the swelling was free and normal in color and texture. There was no discharge from the swelling. There was a distinct cough impulse over the swelling but no pulsation was felt. The swelling was non translucent. The veins of the scalp over the swelling and the surroundings were not engorged. The temperature of the swelling was slightly higher than the surrounding. There were no other swelling felt over the skull or any other bones or any other parts on the body. Central Nervous system reveals no abnormality. Other systemic examination also revealed no abnormalities.

The patient was investigated with a CT scan and a MRI scans. The CT scan revealed a focal lucent destructive lesion involving the right parietal bone and adjacent coronal suture and frontal bones with a narrow zone of transition. It measured about 32x20mm. MRI scan revealed a focal multiloculated expansile lesion in the right parietal bone near bregma, possibly of secondary

aneurysmal bone cyst; primary lesion may be fibrous dysplasia. Histopathological report revealed aneurysmal bone cyst of the frontoparietal skull. His blood reports were within the normal limits, serum electrolytes were also normal and HBSAg and HIV is non-reactive. The patient was operated with craniotomy involving the right parietal bone, extending in the left parietal bone with enmass excision of the cyst with cranioplasty. Postoperative changes with extra cranial CSF collection in the right fronto-parietal region were noted on the CT scan.

DISCUSSION

The incidence of an aneurysmal bone cyst in the skull varies from 2.5 to 6% of all aneurysm bone cysts.^{[6],[8],[9],[10]} They usually present as scalp mass,^{[7],[9],[10],[11]} Occasionally, they may present as intracranial space occupying lesion^[12] or cerebral hemorrhage.^[5] Age of presentation varies, but they usually present in the second or third decade, with equal sex distribution^{[4],[14]} In 1942, Jaffe and Lichtenstein^[13] introduced the concept of aneurysmal bone cyst as a lesion having a characteristic radiological appearance of ballooned out distension of the periosteum which is usually outlined by a paper thin subperiosteal bone shell which in turn is overlined by a region of disintegrated cortex. Trauma has been put forward as an important etiological factor.^{[15],[16]} Edling^[1] regarded aneurysmal bone cyst as one of the manifestation of solitary dysfibroplasia of bone, suggesting a defect in the development of the epiphyseal plate but it doesn't explain its occurrence in the mature bone. Lichtenstein^[2] suggested that it could result from a local circulatory disturbance, because of sudden vascular occlusion of venous drainage of that segment of the bone or development of an arterio-venous shunt. This results in progressive blood spaces in the medulla, which leads to gradual distension of the bone with atrophy. Jaffe^[5] 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^{[3],[18],[23]} and include unicameral bone cyst, non-ossifying fibroma, giant cell tumor, chondroblastoma, fibrous dysplasia, osteofibrous dysplasia of campanacci, 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.^[20] The aneurysmal bone cysts which do not show fluid levels on CT scan are often non-homogeneous and resemble some giant cell tumor.^[20] MRI also shows fluid levels, particularly in T1WI. 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.^[21] Pathologically, the cysts contain multiple fluid filled cavities separated by multiple septa and lined by multinucleated giant cells. These large spaces filled with blood contain no any endothelial lining, but are rather delimited by cells with the morphology, ultrastructural and immunohistochemical features of fibroblasts, myofibroblasts and histiocytes. An additional feature showing a particular degenerated calcifying fibromyxoid tissue was reported by Rosai.^[23] The risk of recurrence is increased with an increase of mitotic figures.^[26]

In some cases, surgical biopsy has in itself been curative.^[19] Total excision has been recommended as an ideal treatment for aneurysmal cyst of skull.^[18] The tendency for recurrence is related with young age of the patient, size of the lesion, presence of mitosis and incomplete surgical removal. Simple curettage is associated with high recurrence rates varying from 21% to 50%.^[14] Radiotherapy has been advocated for deeply situated lesions of the base of the skull, dural involvement or where subtotal excision is done but its effect is unclear.^{[5],[24]} The suggested dose ranges from 600 to 3000 rads.^{[14],[25]} Radiotherapy is however contraindicated in the treatment of aneurysmal bone cyst associated with fibrous dysplasia as there are increased chances of malignant transformation in them.^[22]

CONCLUSION

Highlights the fact that aneurysmal bone cysts are benign skull tumors which can be treated completely by excision of the whole cystic lesion. The prognosis is extremely good if treated properly.

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