Aneurysmal Bone Cyst of Sphenoid Bone– A Case Report

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Summary

Aneurysmal bone cysts of the skull are rare and involvement of sphenoid bone is even less frequent. We present X-ray, CT, MR imaging and histopathologic findings of an aneurismal bone cyst of the sphenoid in a 15-years old female adolescent. Radiological findings of the aneurysmal bone cyst of the skull were highly suggestive of the diagnosis and that were confirmed by histopathologic analysis.

Introduction

Aneurysmal bone cysts (ABCs) are uncommon benign, non-neoplastic cysts of unknown origin. In one third of cases, a history of trauma or a preexisting osseous lesions such as chondroblastoma, Gient Cell Tumour (GCT), osteoblastoma, Non Offifying Fibroma (NOF) or fibrous dysplasia are present[1,2]. Rapid expansion may obliterate the underlying abnormality, leaving behind the blood-filled cavities that are characteristics of ABCs. Grossly, ABCs are multilocuated, expansile and highly vascular osteolytic lesions that often contain blood degradation products. This rare entity commonly occur below 20 yrs. of age with slight female preponderance. ABCs occur in all parts of skeleton, mostly involving the metaphysis of long bone and neural arches of vertebrae[1,2]. Skull involvement is rare, occurring in less than 1% of all aneurysmal bone cysts[3]. Pain and swelling are the most common overall presentation. Plain film & Non Enhanced CT (NECT) scan shows osteolytic lesion surrounded by expanded, thinned, egg shell like cortical bone. A soft tissue mass is often present. Matrix Calcification is absent. Most ABCs are very hypervascular at angiography[4]. MR scan typically demonstrate a lobulated, multiseptated lesion with fluid-fluid levels and blood degradation products. We present X-ray, CT, MR imaging findings in the case of ABC of sphenoid that was confirmed by histopathologic findings.

Case Report

A 15-years old female adolescent had pain and watering of right eye as well as headache and nausea over a period of 03 months. The patient noticed gradual swelling over her

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right orbital region. She had no history of trauma or surgery. Physical examination revealed swelling of right orbital region associated with exopthalmos of eye. Ocular movement was restricted but visual acuity was normal. Fundoscopy was unremarkable. X-ray skull AP and Lateral view showed enlargement of right orbit with destruction of lesser and greater wing of right sphenoid (Fig.-1). On axial and coronal CT scans, a well defined multiloculated mixed density mass with fluid-fluid levels was found in middle cranial fossa involving right temporal & infratemporal fossa, ethmoid & sphenoid sinuses, posterior aspect of nasopharynx, anteriorly extending in to right orbital cavity, superiorly in to right frontal region. Bone destructions was present in lesser and greater wing of sphenoid, ethmoid and orbital plate of maxilla. The mass showed strong enhancement in post contrast scan (Fig-2&3).

TIW, T2W and post gadolinium contrast MR imaging revealed strongly enhancing variable signal intensity lobulated mass with fluid-fluid level within the cystic spaces, consistent with blood degradation products . The mass involved right cavernous sinus, encasing the right Internal Carotide Artery (ICA) (Fig.-4, 5, 6 & 7). Near total removal of the mass was done surgically. Histopathologic analysis revealed gray white soft tissue mass with blood filled cystic spaces. The lesion was made of many osteoclastic giant cells in the background of spindle shaped stromal cells. Some of the cystic spaces were devoid of endothelial lining. Bone formation is seen within the lesion. These findings were consistent with those of an aneurysmal bone cyst.

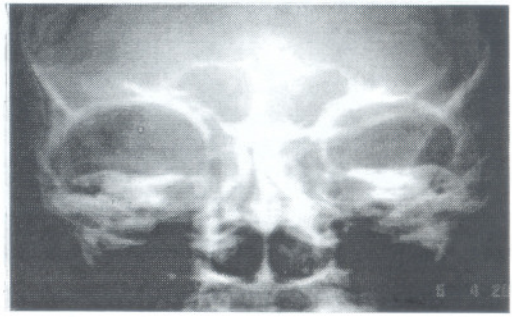

Fig.-1: *Plain x-ray skull P/A view-showing right orbit is enlarged with destruction of lesser and greater wing of sphenoid bone at right side.*

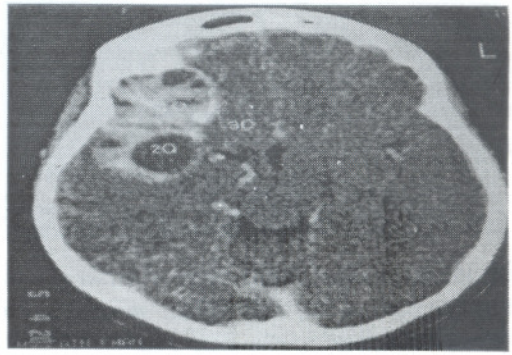

Fig.-2: *Contrast enhanced axial CT scan of brain shows-heterogeneous enhancing mass lesion with cystiç component, few having fluid fluid levels.*

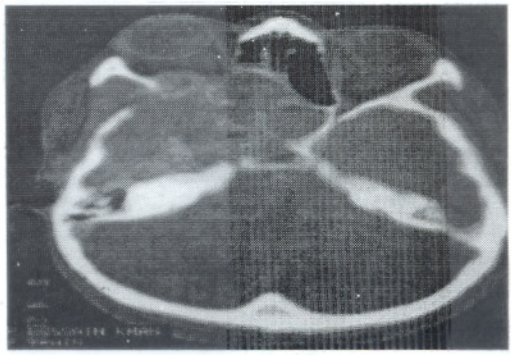

Fig.-3:*Bone window setting of CT scan of brain showing destruction of right greater and lesser wing and part of the sphenoid bone as well as right ethmoid bone.*

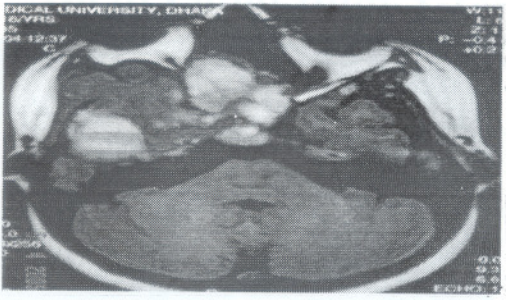

Fig.-4: *Noncontrast axial Tl W MR image of brain shows- mixed intensity mass lesion having fluid fluid levels.*

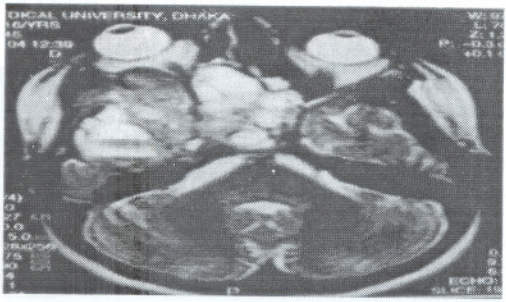

Fig.-5: *Axial T2W MR image of brain shows heterogeneous hyperintensity mass lesion having fluid fluid levels.*

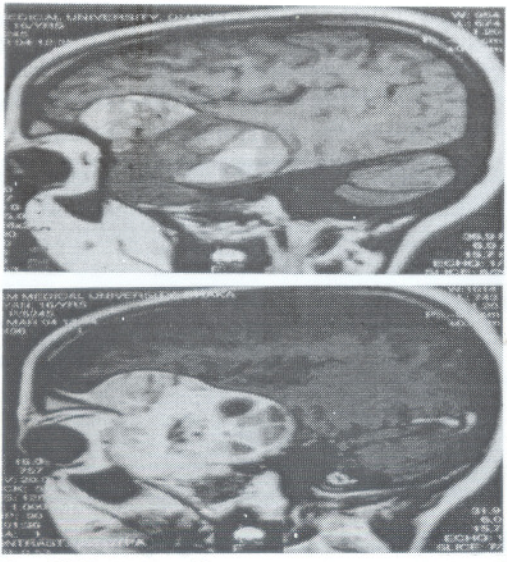

Fig.-6&7: *Pre and post contrast parasagittal MR images of brain shows heterogeneous strong enhancement of the lesion.*

Discussion

ABCs were first described in 1942 as "peculiar blood-containing cysts of large size"[5]. They are composed of blood-filled, anastomosing cavernous spaces, separated by cystlike walls. The exact etiology is not clear. However, the ABC is considered to be the result of a specific pathophysiologic changes, which is probably caused by trauma or an anomalous vascular process. The lesion is a component of, or arises within a preexisting bone tumor in about one third of cases. An ABC can arise from a preexisting chondroblastoma, a chondromyxoid fibroma, an osteoblastoma, a giant cell tumor or fibrous dysplasia. Less frequently, it results from some malignant tumours, such as osteosarcoma, chondrosarcoma and hemangioendothelioma[1,2].

ABC rarely involves the skull and can occur in any part of it. Sizes of the frontal ABCs were reported within 4-6 cm in diameter[6].

Radiological findings reflect the pathophysiologic properties of an ABC. Initially, well-defined osteolysis and periosteal elevation are present. The lesion grows rapidly & causes progressive destruction. Radiographs typically show an eccentric lesion with an expanded, remodeled blown out or ballooned bony contour of the bone. The expanded contour is the result of bone production by the periosteum. Lesions frequently have a delicate trabeculated appearance[1]. At histologic examination, these lesions are large, septated sinusoids filled with blood and lined by endothelium and multinucleated giant cells[7,8]. Haemorrhage of variable age within the cysts

are present and the degradation of blood products causes fluid levels.

CT is superior in the evaluation of the lesions located in the regions that cannot be assessed well by radiography. CT scan can clearly demonstrate the extent of mass and bone destruction. Fluid levels are observed is 35% of the cases, and dependent layer show increased attenuation[1].

On MR images, the hypointense rim surrounding the lesion is an important finding and suggest a benign process. This rim is composed of fibrous tissue[1,8,9]. A fluid-fluid level is another finding and is more readily seen on MR images than on CT scans[1]. Fluid-fluid level caused by the degradation of blood product strongly suggest the presence of ABC[7] but this finding has been reported to occur in association with various condition and is not pathognomic. Chondroblastoma, osteosarcoma, giant cell tumor, Fibrous displasia, osteoblastoma and tumor calcinosis also show fluid-fluid level[10,11]. Although it is not specific, its presence indicates an ABC if other suggestive feature exist[12]. Another important MR imaging finding is the presence of small cysts produced from larger cysts, these are called diverticula[7,8].

Considering the age, clinical features and radiological findings of our patient, the radiological diagnosis was suggestive of aneurysmal bone cyst. Fluid-fluid level, diverticula and enhancement of fibrous tissue in MR images were considered typical of an ABC.

To the best of our knowledge, this is the first case report of ABC involving tile sphenoid bone of skull from Bangladesh.

ABC should be considered in rapidly growing calvarial masses in adolescents. CT scan and MR images showing strongly enhancing well defined loculated soft tissue mass having fluid-fluid level suggest the presence of ABC. Especially the MR imaging is important in the evaluation of neighbouring structures before surgical intervention.

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