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Aneurysmal bone cyst in the clivus, dorsum sella, and sphenoid bone, a case report

Elashry M[1*], Tawfik A[2], Saleh W[3], Galal A[1]
[1] Department of Diagnostic Radiology, Mansoura University, Mansoura, Egypt
[2] Department of Neurosurgery, Mansoura University, Mansoura, Egypt
[3] Department of Oral Medicine and Periodontology, Mansoura University, Mansoura, Egypt

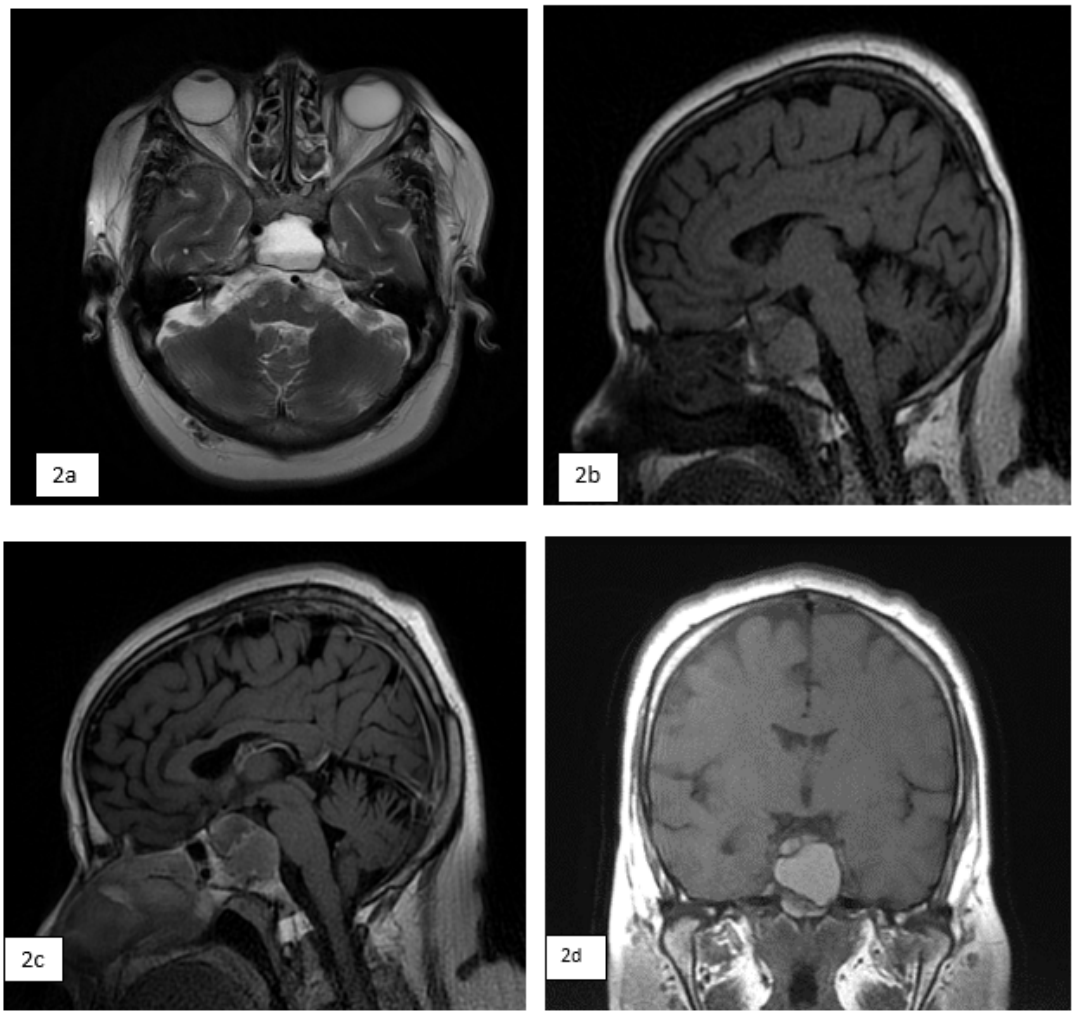
Abstract
Aneurysmal bone cysts are benign bone lesions. The incidence of aneurysmal bone cyst in the skull showed a reported incidence of 1% of all aneurysmal bone cysts. Trauma was assumed as an important etiologic factor in inducing the formation of an aneurysmal bone cyst. However, the exact pathogenesis is not well known. Three reports were found in the literature showing the involvement of the clivus by an aneurysmal bone cyst.
We report a case of 40 years old male who presented with a headache and blurry vision. Radiographic evaluation of the lesion was done by computed tomography, and magnetic resonance imaging. We found the lesion to be an aneurysmal bone cyst that showed a well-demarcated expansible osseous lytic clival mass lesion that extends into the dorsum sella and sphenoid bone.

Keywords: aneurysmal bone cysts, clivus, sphenoid bone, Magnetic resonance imaging

Introduction
Aneurysmal bone cysts are well-differentiated benign tumors most noticed commonly in the first and second decade and are more prevalent in females [1, 2]. The aneurysmal bone cyst can be a primary benign tumor or a secondary to trauma, bone malignant tumors or attributable to intraosseous circulatory disturbance [3]. Metaphysis of the long bones are the most common site affected by aneurysmal bone cysts. Other sites may be involved as the flat bones, vertebral column, lumbar and thoracic spine [4-6]. The involvement of the skull base by the aneurysmal bone cysts especially the clivus is rare [7]. Misdiagnosis of the aneurysmal bone cyst is common as the lesion is rare which increases the potential for delaying the appropriate treatment [8].

Case presentation
A 40-year-old male patient presented with chronic headache 2 months ago associated with blurry vision and visual field tapering as he could not see the outer part of his visual field. His lab investigations were within the normal range. He denied any trauma and he was neither smoker nor alcoholic. The medical history is free from any systemic diseases.
The patient was referred from Neurosurgical Department to Diagnostic Imaging Department to perform non-contrast computed tomography (NCCT) of his brain. His brain CT revealed a well-defined space-occupying lesion involving the clivus and sellar region that has fine calcification scattered through its wall and septation seen inside. Fig 1
Then, magnetic resonance imaging (MRI) study of the brain with contrast was done and revealed a lobulated and well-demarcated expansible osseous lytic clival mass lesion that extends into the dorsum sella and sphenoid bone. It measures 3.7 cm x 2.4 cm (craniocaudal x transverse) in diameter. It shows lytic cystic appearance with fluid/fluid leveling, high signal areas on T1 WIs, and iso to high signal on T2 WIs denoting hemorrhagic contents. It shows heterogeneous enhancement on the post-contrast study. Fig 2
This mass is seen posteriorly obliterating the prepontine cistern and smoothly indenting the ventral aspect of the pons, anteriorly displacing the pituitary gland infundibulum and extending into the sphenoidal sinus, superiorly it is also seen indenting the optic chiasm and the inferior margin of the hypothalamic structures including the mamillary body and tuber cinereum.
Then, this mass lesion was surgically excised by neurosurgical team and the excised tissue was sent for histopathological examination. Histopathological analysis showed a multiloculated cystic lesion filled with blood. The wall of cystic spaces consist of loose fibrous tissue, some dense collagen and dense osteoid, inflammatory cells and osteoclast cells. Fig 3

Fig 1: NCCT brain revealed well-defined expansile mass with fluid retained inside. This mass is seen involving the base of the skull region with calcified wall and septation seen its mid porion.



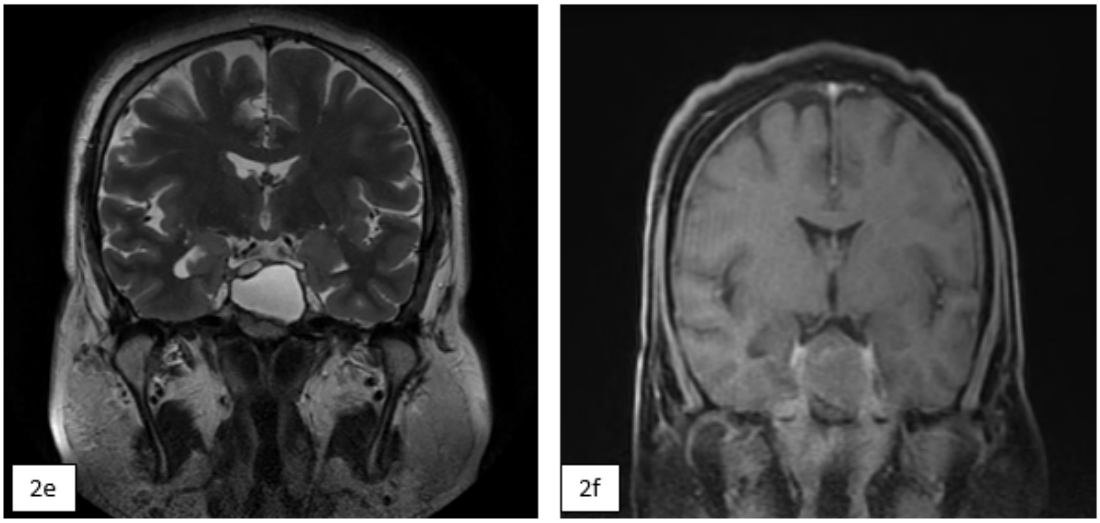
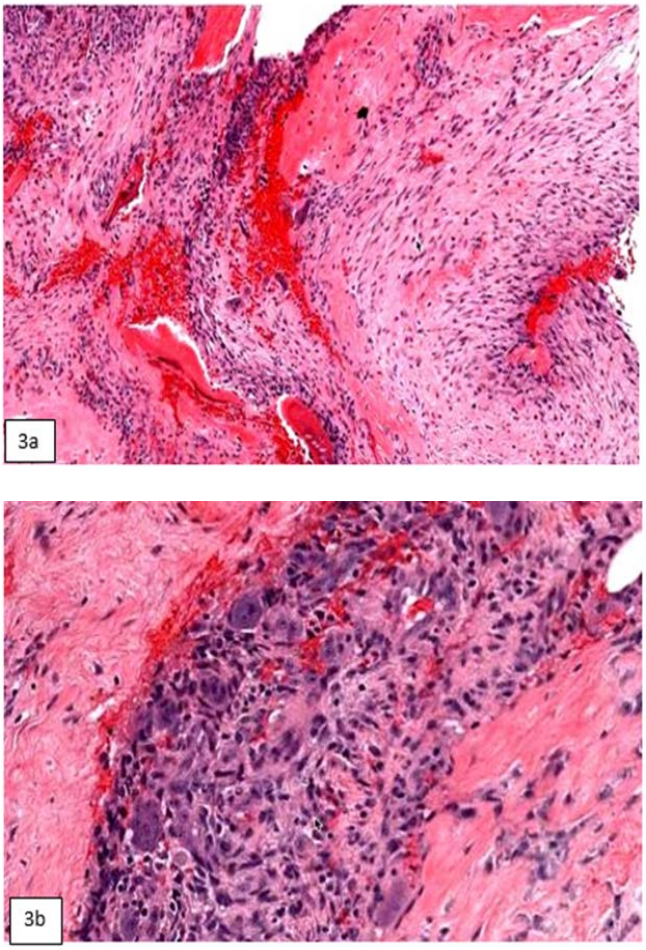


Fig 2: (A) Axial T2-weighted image of the skull base showed a multilocular fluid/fluid mass lesion which is a good radiological sign for diagnosis of an aneurysmal bone cyst. (B) Sagittal T1-weighted images. (C) Sagittal T1-weighted, the post-contrast image reveals heterogeneous enhancement of the solid components of the skull base lesion. (D) Coronal T1-weighted image. (E) Coronal T2 –weighted image reveals fluid/fluid level of the lesion indicating hemorrhagic content. (F) Coronal T1-weighted, fat sat image and post-contrast image. Note that hyperdense calcification noticed in the lesion at NCCT Brain (figure 1) turned to be of low signal in T1 and T2- weighted. The para sellar structures including the cavernous sinuses appear intact. Normal size and configuration of the ventricular system. There is no obvious intra axial mass lesion or extra-axial fluid collection.



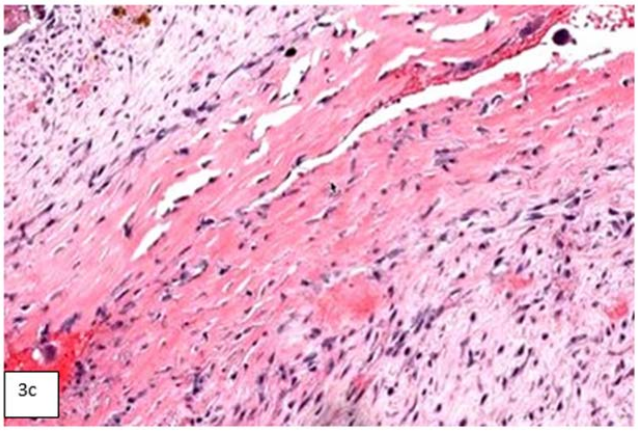


Fig 3: Histopathology of aneurysmal bone cyst. Aneurysmal bone cyst consist of hemorrhagic spaces of blood-filled cavities (A) (Magnification 10X), multinucleated osteoclast-like giant cells (B) (Magnification 20X), Loose fibrous tissue forming the wall of the cystic spaces (C) (Magnification 20 **X).**

Discussion

The concept of an aneurysmal bone cyst was introduced by Jaffe [9]. Clinically, the aneurysmal bone cyst is a benign condition that doesn't usually require surgical removal. aneurysmal bone cysts of the sphenoid bone and the base of the skull result in different symptoms as ptosis, exophthalmos, headache, swelling, loss of vision, nasal obstruction, and sinus congestion [10]. The etiology of aneurysmal bone cysts is unknown, it may be detected as a primary benign tumor in bone or may be correlated with other pathological bony abnormalities as osteosarcoma, giant cell tumors, trauma, fibrous dysplasia, chondroblastoma, and chondroid myxoma [11]. In addition, the local hemodynamic changes may induce the formation of an aneurysmal bone cyst [12, 13].

Multiple imaging modalities can be used in the diagnosis of an aneurysmal bone cyst. The appearance of aneurysmal bone cysts in the plain films shows a cystic lesion with a thin shell of periosteal bone formation however, the extent as well as the soft tissue extension of the aneurysmal bone cyst can be defined by CT scan, particularly in the skull. Furthermore, a CT scan can show multiple small fluid levels which characterize the sedimentation of RBCs within the blood-filled cavities while they are considered important features of an aneurysmal bone cyst on a CT scan [14].

Magnetic resonance imaging (MRI) can detect the fluid-fluid level with heterogeneous signal intensity on both T1W1 and T2W1 [15]. MRI is the best modality to demonstrate the fluid levels in aneurysmal bone cysts which are indicative signs of the presence of hemorrhage with sedimentation. Their signal intensity may be increased on T1WI due to methemoglobin. Contrast-enhanced MR imaging reveals internal septa within the lesion. MRI reveals a low signal intensity rim around the lesion with thickened, intact periosteal membrane [16, 17]. The typical appearance of ABC in our case shows fluid-fluid levels by MRI.

Few reports were found in the literature showing the involvement of the clivus by an aneurysmal bone cyst. Rajput et al reported the detection of an aneurysmal bone cyst in 45 years old female. The lesion was found in the clivus as well as the anterior and posterior arches of the atlas and C2 vertebral body [7]. Furthermore, Ustabasioglu et al detected the ABC in a case of a 21-year-old male that was misdiagnosed as chondroma. According to the contrast-enhanced MRI findings which revealed a lobulated and well-demarcated expansile clivus and sphenoid, the patient underwent surgery. After surgical excision and histopathological analysis, the lesion was diagnosed as an aneurysmal bone cyst. The final diagnosis of the aneurysmal bone cyst was considered based on the imaging findings, and histopathological evaluation which demonstrated numerous blood-filled cystic cavities separated by thin fibrous septa and numerous multinucleated giant cells [18].

Our case is unique as the lesion developed in a male patient of 40 years old while the aneurysmal bone cyst is common to be found in females age younger than 30 years old [1, 2]. In addition, the epicenter of the lesion is found in the clivus with extension into the sphenoid sinus and the dorsum sellae. While the other reports found the aneurysmal bone cyst in the clivus and sphenoid [18] and the clivus and C1C2 vertebrae [7].

The basic treatment of the aneurysmal bone cyst is the curative surgical excision [19]. Despite the aneurysmal bone cyst is a benign lesion, it may undergo a rapid course causing compression of the adjacent structures or pathological fracture of bones [1]. Malignant transformation of the aneurysmal bone cyst was reported in the literature in the absence of the history of irradiation [20].

Conflict of Interest

The authors declare that they have no conflict of interest".

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