DOI: 10.7860/JCDR/2022/55151.16553

Case Report

Radiology Section

An Unusual Site of Aneurysmal Bone Cyst in the Ethmoid Sinus- A Case Report

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ABSTRACT

Aneurysmal bone cyst is a rare non neoplastic tumour of bones. It is a cystic vascular lesion causing local destruction and invasion of adjacent structures with rapid invasion. Therefore, it requires early intervention. It typically involves long bones of limbs, ribs of thoracic cage, pelvic bones, and vertebrae in axial skeleton. Rarely it occurs in the head and neck region where mandible is found to be the most common site. It has been reported to arise from paranasal sinuses. However, ethmoid sinus is a very rare site. The rarity of aneurysmal bone cyst in such a location, involvement of adjacent structures, and recurrences makes it more interesting for radiologists. Here, the authors present an unusual site of aneurysmal bone cyst in a 22-year-old female involving the right ethmoid sinus with its typical Computed Tomography (CT) and Magnetic Resonance Imaging (MRI) features.

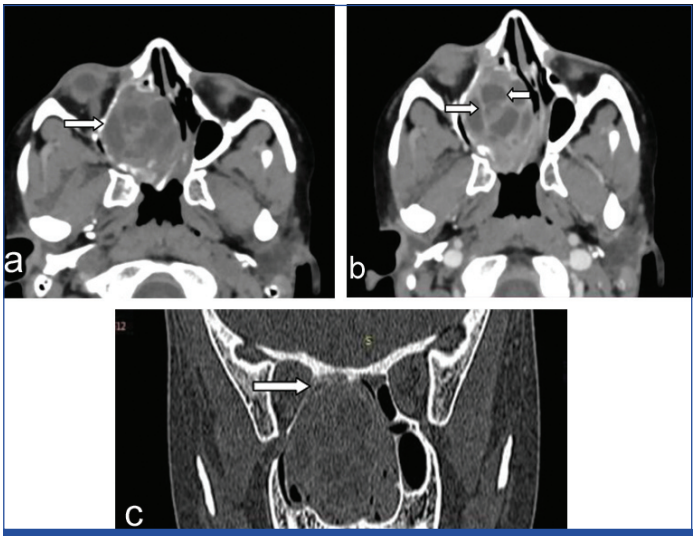
Keywords: Benign, Bone tumour, Paranasal sinus, Rare site

CASE REPORT

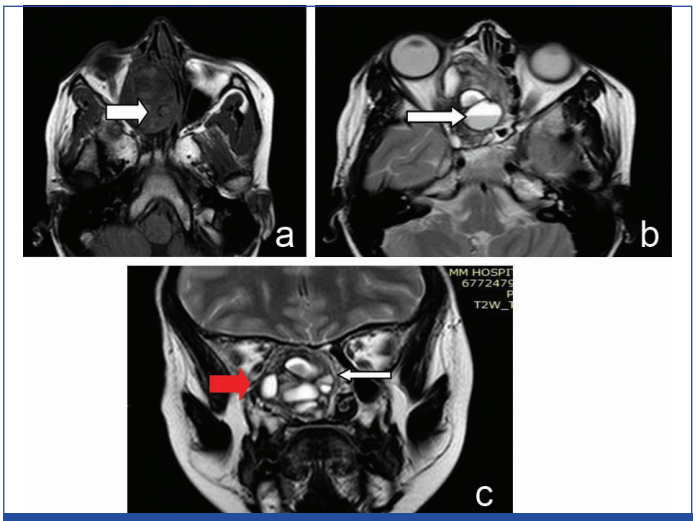
A case of 22-year-old female who came to the Ear, Nose and Throat Outpatient Department, with complaints of insidious onset of headache for last one year confined to the right frontal region and was associated with nose pain. The patient also complained of right sided nasal obstruction for 6-7 months, which was insidious in onset and gradually progressive. The patient also complained of nasal bleed from last 2-3 months on the right side. There was a history of intermittent nonpurulent watery discharge from the right eye for last 2-3 months. There was no history of trauma, fever, weight loss or loss of appetite. The systemic examinations were normal. On Diagnostic Nasal Endoscopy (DNE), right sided nasal cavity showed a reddish fleshy soft tissue mass lateral to the inferior turbinate, completely filling it. The surface of the mass was smooth and was non tender. Left nasal cavity showed deviated nasal septum with spur touching the inferior turbinate. There was mild axial proptosis on the right side. Routine investigations like blood profile, serum electrolytes level and urine examination were normal.

On skiagram of Paranasal Sinus (PNS), a lesion of mixed radiodensity was seen in the region of ethmoid air cells on the right side. Contrast Enhanced Computed Tomography (CECT) of PNS and orbit was done on 128 slice multidetector spiral Computed Tomography (CT) (Philips Ingenuity). It showed a large rounded relatively well-circumscribed multi-loculated expansile cystic lesion involving right-sided ethmoidal air cells and extending into the right nasal cavity [Table/Fig-1a,b]. Postcontrast scan showed moderate peripheral enhancement as well as enhancement of internal septae. Fluid-fluid levels were also seen within it. Posteriorly, the lesion caused widening of the sphenoethmoidal recess with extension of mass into the sphenoid sinus. Laterally, the lesion was causing scalloping of the medial wall of the orbit with mass effect appreciated on the right medial rectus muscle. No frank bony destruction was seen. Nasal septum was severely deviated to the left side with marked narrowing of the left nasal cavity [Table/Fig-1c]. Lower part of the right nasolacrimal duct was also blocked.

Subsequently, Magnetic Resonance Imaging (MRI) was done, which also showed expansile multi-loculated cystic lesion with fluid-fluid levels of different signal intensities on T2W sequences. On T1W sequences, hyperintense areas were seen within the fluid, suggesting haemorrhage [Table/Fig-2a,b]. In T2W coronal, similar findings lateral deviation of nasal septum toward left side and hypoplastic right maxillary sinus [Table/Fig-2c]. Hence, based on clinical and



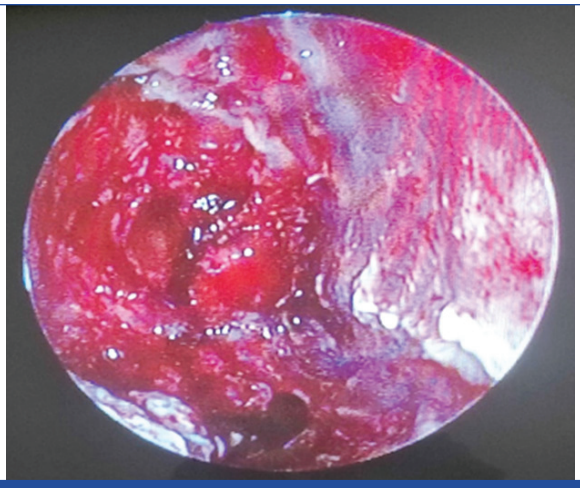
[Table/Fig-1]: a) Axial NCCT: Multiloculated cystic expansile lesion in right ethmoid sinus and nasal cavity (block white arrow); b) Axial Post Contrast CT: shows enhancement of septae (block white arrows); c) Coronal bone CT: Remodelling, expansions and subtle erosions of surrounding bones.



[Table/Fig-2]: a) T1W axial: showing multilocated lesion in right nasal cavity predominately appearing hypointense with few hyperintense areas representing blood (white arrow); b) T2W axial: showing multiloculated cystic lesion appearing hyperintense and showing fluid-fluid levels of different signal intensities (white arrow); c) T2W coronal: shows multiloculated lesion in right nasal cavity with lateral deviation of nasal septum toward left side (white arrow) and hypoplastic right maxillary sinus (red arrow).

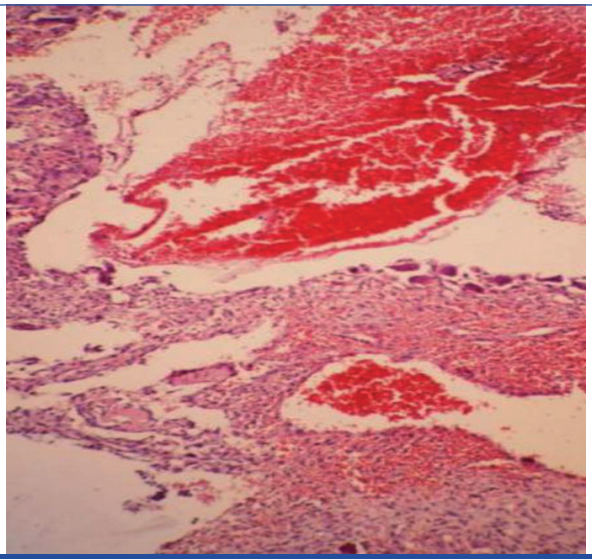
radiological findings, a provisional diagnosis of Aneurysmal Bone Cyst (ABC) was made.

The patient was operated via Functional Endoscopic Sinus Surgery (FESS) procedure under general anaesthesia. Intraoperative findings showed mass lesion obliterating the right nasal cavity and extending upto the posterior extent of inferior turbinate. Middle turbinate was eroded. The mass was attached to the septum and was pushing it towards left side. The entire mass was removed and the specimen was sent for histopathological examination [Table/Fig-3].



[Table/Fig-3]: Operative picture of mass.

Biopsy: The mass was composed of fibrous walled channels or spaces containing blood. No elastic fibres or smooth muscle tissue was demonstrable in the walls of these vascular spaces. Evidence of haemorrhage and associated foci of giant cells were irregularly distributed in the fibrous septae. At the periphery of the lesion, osteoid and subperiosteal bone formation was present. Multiple sections of the tumour were similar and no other definable bone tumour was identified [Table/Fig-4].



[Table/Fig-4]: Histopathological slide of lesion: showing blood filled spaces separated by septae containing giant cells and fibroblasts (10x magnification, H&E staining).

However, presence of giant cells may pose a difficulty in differentiating these lesions with other histologically similar lesions such as giant cell reparative granuloma, giant cell tumours, haemorrhagic cysts, and fibrous dysplasia. The most important feature to differentiate it from histologically similar lesions is lining of blood filled spaces. However, if the pathologist fails to appreciate this lining of blood filled spaces, then diagnostic error can occur [1].

Hence, on the basis of radiological, histological and operative findings, the diagnosis of right ethmoidal ABC was made. The patient was discharged uneventful, regularly followed-up after endoscopic resection for a period of three months and there were no signs of any residual/recurrent disease.

DISCUSSION

In 1942 Jaffe HL and Lichtenstein L, first described ABC. They described the lesion as thin-walled lesion that typically involve the bones of pelvic and spine region with presence of a large hole underneath that contained fluid blood [2].

The ABC term is a misnomer, as neither these lesions are truly aneurysmal in origin nor these lesions are truly cystic in histopathology and there was no endothelial wall. The word “aneurysm” in ABC was used to describe the typical blowout distension of contour of surrounding involved bones. The word “cyst” was used to describe the blood filled cavities and spaces that lack endothelial lining. Instead, these are benign expansile lesions that are present within the bone. They form cavities underneath which are filled with blood. These cavities are lined by proliferative type of fibroblasts, giant cells and also by the trabecular bone [3].

Aneurysmal Bone Cyst is mostly found during the childhood and adolescent phase of life. The median age of occurrence of ABC is 13 years. About 90% of the cases of ABC are seen before the age of 30 years. There is female predilection of ABC, with ratio of male to female 1:2 [4].

Pathogenesis of ABC is obscure. Historically, it was believed that ABC resulted from the increase in venous pressure that resulted in extravasation of the cellular and the blood contents into cyst-like voids in the bone [3]. Secondary origin of ABC from pre-existing lesions like giant cell tumour, fibrous dysplasia, osteoblastoma, osteosarcoma and unicameral bone cyst are well documented in the literature like Mortaji HE et al., has documented a recent case report on ABC of ethmoid sinus secondary to fibrous dysplasia [5]. More recent work showed that identification of a genetic driver-a translocation-induced up-regulation of the Ubiquitin-specific Protease USP6 (Tre2) gene-defined at least a subset of ABCs to be a primary neoplasm [3].

In this article we are presenting a rare case of ABC centred on an unusual site i.e., in the ethmoid paranasal sinus with extensions as described above. The radiological, surgical, and pathological findings were compatible with ABC. Aneurysmal Bone Cysts can be either primary or secondary. Secondary ABCs are related to fibrous dysplasia, non-ossifying fibroma, giant cell tumours, chondroblastoma, osteoblastoma, and osteosarcoma [5]. Very few ABC cases in the sinuses were reported uptill now [6,7]. Only 3-12% of the ABC cases are seen in the head and neck region. In the head and neck region, mandible is the most common site [3]. Involvement of maxilla by ABC is more rare [6]. The ABCs of ethmoidal bone are very rare [5]. Aneurysmal bone cyst is a rapidly expanding benign bony mass that is multicystic in appearance and causes local destruction [6].

Characteristic imaging features of ABCs are seen on CT and MRI. During angiography, the blood supply to these vascular lesions can occasionally reveal arteriovenous shunts [7]. In CT scan- erosions of the adjacent bone with no signs of frank destruction, cortical thinning and presence of ridges in the walls of the bone with fluid-fluid levels are seen. In MRI images, well-defined lesion showing altered signal appearing heterogeneous on T1W images with hyperintense areas (representing blood) with hypointense capsule, and a homogeneous increase in signal intensity on T2W images with fluid-fluid levels is observed. Haemorrhagic fluid content and septated appearance (like in this case) is the characteristic feature of ABC [5,8,9]. The present case had typical CT and MRI findings. Treatment options considered for ABCs include sclerotherapy, embolisation, radiotherapy, simple curettage, surgical excision, or a combination of methods. Surgical excision is the first treatment of choice, and there is a 10 to 30% recurrence rate [9]. On pathology, it shows blood filled channels lined by thin layer of endothelial

cells. It is surrounded by connective tissues containing giant cells. Macrophages and new bone formation is also seen within the stromal matrix. The presence of multiple giant cells sometimes make it difficult to differentiate from other histological similar lesions such as osteoclastoma, giant cell reparative, fibrous dysplasia ossifying haematoma [1,4]. Radiological differentials of such lesions include giant cell tumour, giant cell reparative granuloma, fibrous dysplasia and other vascular tumours like haemangioma. Giant cell tumour is usually seen in older age group usually >30 years. Giant cell reparative granuloma is always preceded by a history of trauma. Other vascular tumours like haemangioma are basically soft tissue tumours and do not show fluid-fluid levels. Fibrous dysplasia is a bony tumour and does not show fluid-fluid levels. Aneurysmal Bone Cysts characteristically show fluid-fluid level on CT and MRI, thus differentiating them from other similar lesions. Therefore, radiological diagnosis should be preferred.

CONCLUSION(S)

It was concluded that paranasal sinuses are a very rare site for the origin of ABCs. But the specific age group and typical radiological imaging findings of multiloculated cystic expansile lesion with fluid-fluid levels showing haemorrhage on MRI helps us to reach the diagnosis. Histologically, it can be sometimes confused with other giant cell containing tumours. Hence, the radiological diagnosis should be preferred.

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PLAGIARISM CHECKING METHODS: [Jain H et al.]

- Plagiarism X-checker: Feb 14, 2022
- Manual Googling: Apr 02, 2022
- iThenticate Software: Apr 12, 2022 (21%)

ETYMOLOGY: Author Origin

AUTHOR DECLARATION:

- Financial or Other Competing Interests: None
- Was informed consent obtained from the subjects involved in the study? Yes
- For any images presented appropriate consent has been obtained from the subjects. Yes

Date of Submission: **Feb 12, 2022**
Date of Peer Review: **Mar 13, 2022**
Date of Acceptance: **Apr 04, 2022**
Date of Publishing: **Jul 01, 2022**