

International Journal of Radiology and Diagnostic Imaging



E-ISSN: 2664-4444
P-ISSN: 2664-4436
www.radiologypaper.com
IJRDI 2022; 5(2): 31-33
Received: 17-04-2022
Accepted: 20-05-2022

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Craniofacial fibrous dysplasia with aneurysmal bone cyst: A rare coexistence

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DOI: <http://dx.doi.org/10.33545/26644436.2022.v5.i3a.273>

Abstract

Fibrous dysplasia (FD) is a developmental anomaly in which the normal bone marrow is replaced by fibro-osseous tissue. Fibrous dysplasia can affect either single (Monostotic) bone or multiple (Polyostotic) bones. Craniofacial FD is a monostotic FD involving the craniofacial bones. However, CFD is one of the rare forms of FD. Aneurysmal bone cyst (ABC) is a rare non-neoplastic bone lesion. ABCs are expansile lytic lesions composed of numerous blood-filled channels and cystic spaces that mostly involve the long bones and vertebrae and occur rarely in the craniofacial bones. ABCs can occur either as a primary bony lesion or secondary to various pre-existing benign and malignant bone tumors like fibrous dysplasia. Concomitant occurrence of FD and ABC is rare with only few cases reported in the literature. Here, we report on a case of ABC in a 24-year-old woman who had presented with right frontal and facial swelling for over 6 years followed by proptosis of right eye. Computed tomography revealed an expansile lytic lesion with peripheral ground glass densities involving the body and greater wing of sphenoid, lateral wall of orbit, squamous part of temporal bone with associated thinning and rarefaction of the bony walls. Further, MRI of the same patient revealed multiple cystic spaces some of them showing fluid-fluid levels within.

Keywords: Aneurysmal bone cyst (ABC), Fibrous dysplasia (FD), Cranio-facial FD

Introduction

Fibrous dysplasia (FD) is a congenital anomaly manifesting as a localized defect in osteoblastic differentiation and maturation. There is replacement of normal bone with large fibrous stroma and islands of immature woven bone. It represents approximately 7% of all the benign bony tumors and any bone of the skeleton can be affected. It can affect single or multiple bones (monostotic and polyostotic respectively). FD occurring in multiple adjacent craniofacial bones is referred to as monostotic (craniofacial FD) [1, 2]. Aneurysmal bone cyst (ABC) is a non-neoplastic lesion that consists of cystic cavernous cavities without a lining endothelium and filled with blood and occurs commonly in teenagers or young adults. Aneurysmal bone cysts can be primary or occur in association with other abnormalities of the skeleton, particularly fibrous dysplasia, non-ossifying fibroma or chondro-myxoid fibroma, such lesions being described as secondary ABCs [3]. ABCs commonly occur in long bones and spinal column. Reports of secondary ABC occurring in craniofacial FDs are extremely rare in the literature, accounting for less than 20 cases.

Case report

A 24-year-old female, with no significant medical or familial history, presented with right frontal and facial swelling gradually increasing over a period of 6 years. The swelling had exponentially progressed in the last 6 months. The mass was mildly tender and did not show any signs of inflammation. The patient had developed proptosis of right eye in the last three months. Physical and clinical examination revealed a proptotic left eye. She was free of neurological symptoms, and routine laboratory examinations were within normal limits. X-ray skull (Fig 1) was done which showed non-appreciation of greater wing of sphenoid on the right side with ground glass opacities. However, no obvious characterization of the lesion was possible. Patient was planned for a CT (Computed Tomography). CT scan was done in the following month and revealed an expansile lytic lesion with peripheral ground glass densities involving the body and greater wing of sphenoid, lateral wall of orbit, squamous part of temporal bone causing severe thinning and rarefaction of its walls (Fig 2).

On soft tissue window multiple locules with thin hyperdense septations were noted within (Fig 3). There was no evidence of fracture. For further characterization of the lesion MRI (Magnetic Resonance Imaging) of the patient was done which revealed multiple cystic spaces within the lesion, with some of the cystic lesions showing fluid-fluid levels within

(Fig 4). Mild post-contrast enhancement of septae was seen. These imaging appearance on CT and MRI were classical of FD with ABC. Later it was confirmed on histopathology.

Figures

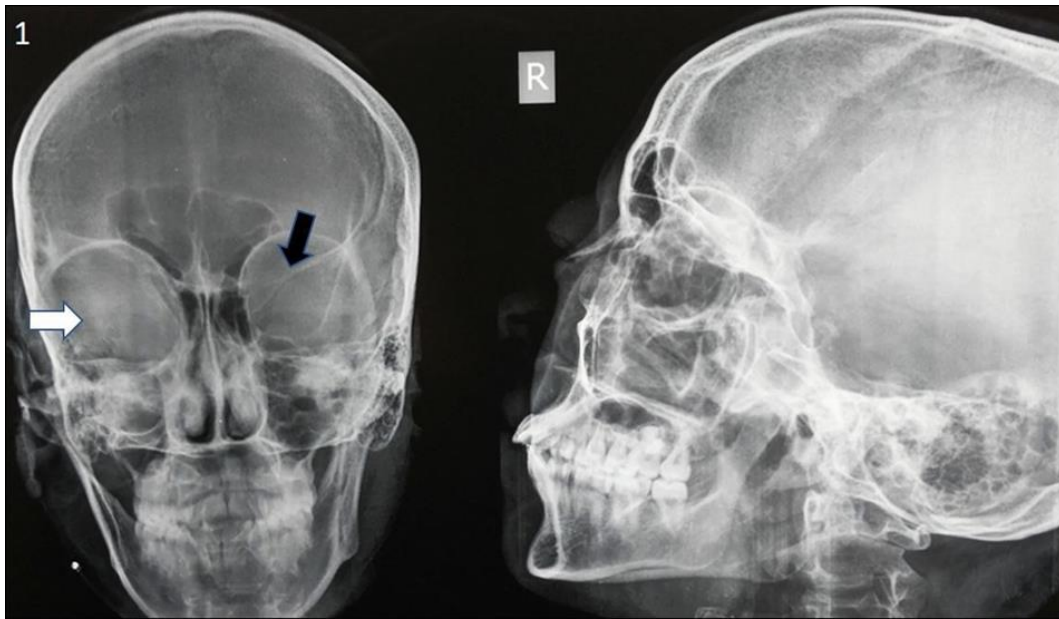


Fig 1: X-ray AP and Lateral view: non-appreciation of greater wing of sphenoid on the right side with ground glass opacities (white arrow). Black arrow depicts the normal greater wing of sphenoid on the left side

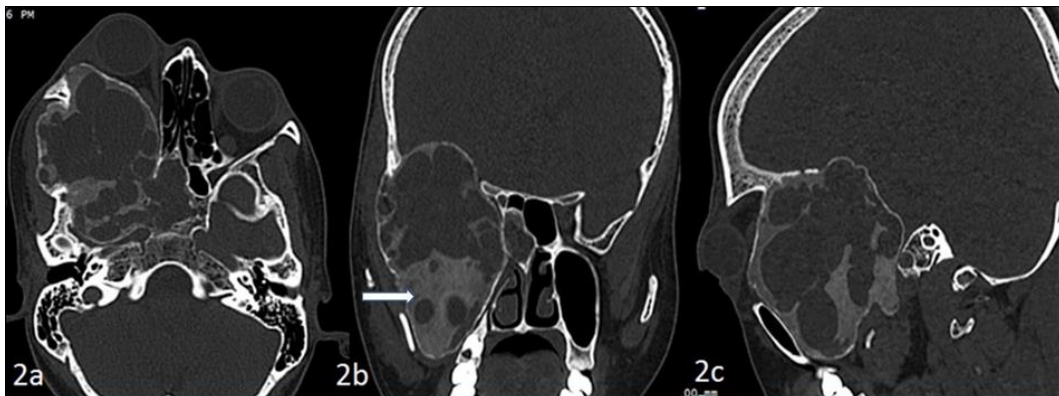


Fig 2: CT bone window axial (2a), coronal (2b) and sagittal (2c). CT reveals an expansile lytic lesion with peripheral ground glass densities (White arrow in 2b) involving the body and greater wing of sphenoid, lateral wall of orbit, squamous part of temporal bone causing severe thinning and rarefaction of its walls. Multiple thin hyperdense septae noted within the lesion.



Fig 3: CT soft tissue window axial (3a), coronal (3b) and sagittal (3c) reveals an expansile lytic lesion with multiple cystic spaces/locules and thin hyperdense septae within.

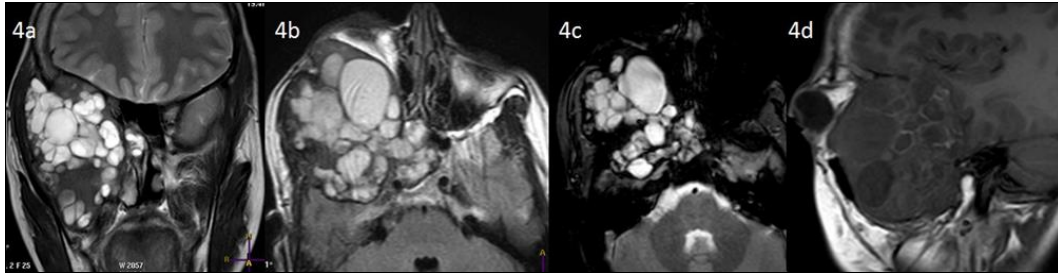


Fig 4: T2 coronal (4a), FLAIR axial (4b), GRE (4c) and T1 post contrast sagittal (4d). MRI depicts multiple cystic spaces within the lesion, some of them showing fluid-fluid levels. Mild post contrast enhancement of septae seen

Discussion

Craniofacial FD, considered a form of monostotic FD accounts for 10-25% of monostotic cases, or may occur as part of polyostotic fibrous dysplasia. The craniofacial bones are affected in up to 50% of polyostotic cases. Occasionally it is seen in the setting of McCune-Albright syndrome [4]. The fibrous tissue in FD undergoes varying degrees of abnormal ossification and show an increase in density, depending on the extent of ossification. This increase in density may be patchy, giving a cotton-wool appearance, or homogenous ground glass appearance [5]. Various theories have been proposed regarding the origin of ABCs including arteriovenous shunts, bleeding due to trauma or bleeding from a pre-existing bony lesion, however the exact pathogenesis remains to be unclear.

Conclusion

Occurrence of FD with ABC is an extremely rare entity. This lesion was first reported by Branch in 1986. Osteoclastoma, osteosarcoma, osteoblastoma and hemangioma were the primary diseases known to be associated with ABC [6]. The report of ABC associated with FD is very rare, and moreover, the occurrence in skull vault is even rarer. Martinez *et al.* [7] found only one case (2.4%) of ABC in 42 patients with FD. Concomitant FD with ABC may progress rapidly and exhibit locally aggressive behavior. Hence early accurate diagnosis of the same on imaging would be helpful in appropriate management of the patient.

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