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Case Report

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Naso-Orbito-Ethmoid Fibrous Dysplasia, a Rare Case in a Rare Location for Radiologists: A Case Report

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ABSTRACT

Fibrous dysplasia is a non-inherited but congenital disorder which is characterised by the replacement of normal bone by an excessively proliferative cellular fibrous connective tissue intermixed with irregular bony trabeculae. Fibrous dysplasia may be localised to the single bone (monostotic form) or multiple bones (polyostotic form). About 25-30% of the facial dysplasia is associated with cranial or facial involvement. Imaging plays an important role in diagnosis and follow-up in the cases of cranio-facial fibrous dysplasia because of the anatomical complexity of the region. We have described herein a rare case of fibrous dyplasia involving naso-orbito-ethmoid region in radiological perspective.

Keywords
Computed tomography, fibrous dysplasia, naso-orbito-ethmoid

INTRODUCTION

Fibrous dysplasia (FD) was first described by Von Recklinghausen in 1891 and termed by Lichtenstein in 1934.It is the non-inherited but congenital disorder which is characterised by the replacement of normal bone by an excessively proliferative cellular fibrous connective tissue intermixed with irregular bony trabeculae. It is caused by sporadic mutation of alpha subunit of Gs stimulatory protein (GNAS mutation). Fibrous dysplasia may be localised to the single bone (monostotic form) or multiple bones (polyostotic form). Monoostotic form is more common and occur in children and young adults. Monoostotic form never progresses to polyostotic form and spontaneous resolution of fibrous dysplasia does not occur as well. McKune Albright syndrome is associated with extra skeletal manifestations like skin lesions and endocrine abnormalities along with fibrous dysplasia whereas Mazabraud syndrome is associated with intramuscular myxomas. Syndromic form is more associated with polyostotic FD.[1,2,3] We are presenting a case of a 17-year-old male presented with recurrent history of frontal headache.

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CASE PRESENTATION

A 17 year old male presented with progressive frontal headache, facial pain and slight swelling of left orbital region for 6 months duration. He had history of recurrent episodes of nasal obstruction in the past for which he is taking medications on and off. Thus, computed tomography of paranasal sinuses (CT PNS) was advised for further evaluation.

IMAGING FINDINGS

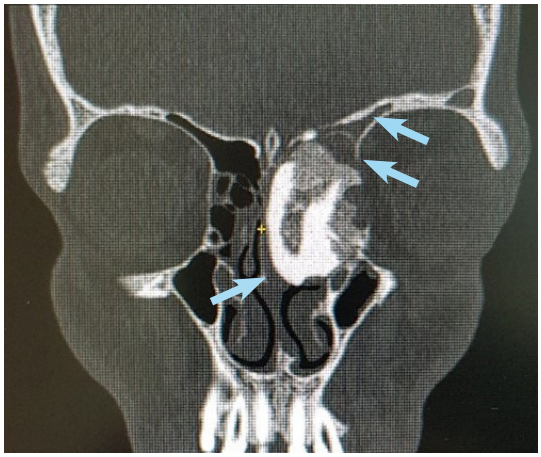

Figure 1. Coronal Plain CT Bone Window shows mixed ground glass and sclerotic expansile lesion seen involving medial wall of left orbit, frontal and ethmoid sinus (yellow arrow). The left ethmoid sinus is completely obliterated. Soft tissue density seen within the left frontal sinus due to obstruction (thin yellow arrow). Left orbit appear slightly deformed (yellow star).

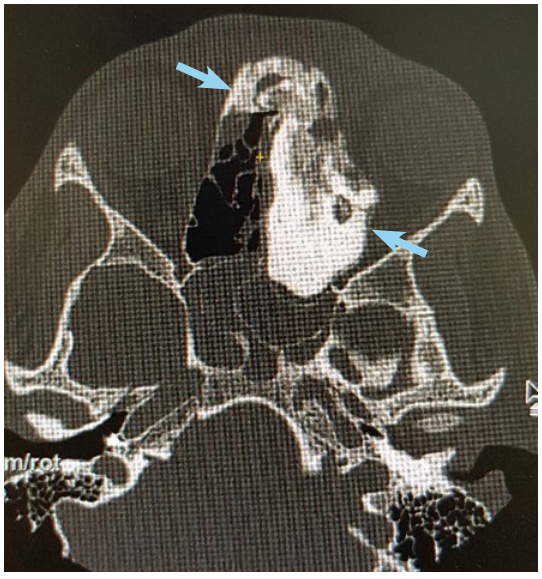

Figure 2. Axial Plain CT Bone window shows large mixed density (ground glass and sclerotic) expansile lesion involving medial wall of left orbit and completely obliterating the left ethmoid sinus. The lesion is extending anteriorly to involve the nasal bone and towards the contralateral side (yellow arrow). Soft tissue density seen within the sphenoid sinus due to obstruction. Mass effect is seen on the medial orbital wall and the medial rectus muscle (thin yellow arrow).

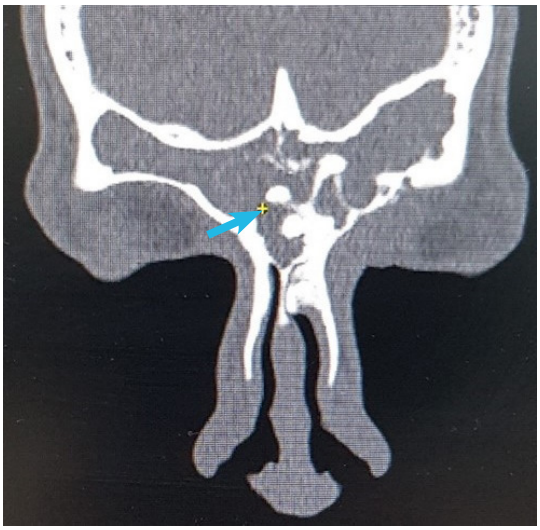

Figure 3: Coronal plain CT shows bilateral frontal sinuses are filled with soft tissue density/fluid (yellow arrow) due to obstruction of ostium due to mass effect of the lesion.

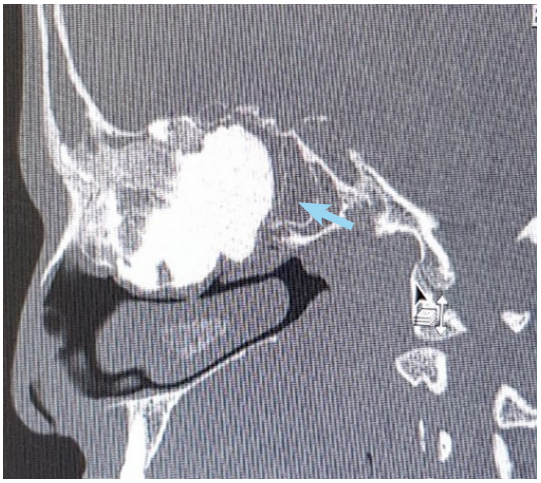

Figure Image 4: Sagittal plain CT shows large mixed density (ground glass and sclerotic) expansile lesion completely obliterating the left ethmoid sinus. Sphenoid and frontal sinuses are filled with soft tissue density due to obstruction (yellow arrow).

DISCUSSION

Most common sites of occurrence are the proximal femur, humerus, tibia, ribs and craniofacial bones.

About 25-30% of the facial dysplasia is associated with cranial or facial involvement. Term cranio-facial dysplasia is given to fibrous dysplasia involving the cranial or facial bones rather than monoostotic or polyostotic because of the involvement of the adjacent bones like nasal, ethmoid, orbital, zygomatic or sphenoids. Most common symptom of cranio-facial dysplasia is the facial asymmetry with swelling of the involved site or rarely due to the mass effect of the lesion with compression of the optic nerve in the form of blurring of vision, hearing loss due to the lesion in temporal bone, displacement of teeth due to the jaw lesions or nasal obstruction due to the lesions in the nasal bone.[4] The term "leontiasis ossea" is the rare term of craniofacial disease involving polyostotic facial and frontal skull bones resembling the lions face. Cherubism is the form of cranio-facial dysplasia involving mandible and maxilla.[5]

Conventional radiographs and CT reveal the characteristics of fibrous dysplasia. The three patterns are seen with FD. They are the ground glass pattern (most common), homogenously dense or sclerotic pattern and the cystic pattern. Most common is the intramedullary expansile ground glass opacity lesion causing endosteal scalloping in the long bones. CT and MRI are useful for the evaluation of the extra skeletal soft tissue component and the complications as well as follow-up. FD shows low to intermediate signal intensity in T1 with heterogeneous signal intensity in T2 and heterogeneous enhancement in contrast images. CT and X-ray is more important in diagnosis of fibrous dysplasia rather than MRI.[3-5]

In majority of cases of fibrous dysplasia, the lesions stabilise after the period of puberty or skeletal maturation. However, some deformity may exist. Surgery may be indicated in the cases with functional or aesthetic complications. It is sometimes associated with aneurysmal bone cyst, simple bone cyst or central cell granuloma. Complications might be pain, secondary fractures or progression into malignancy. The common malignancies include fibrosarcoma, osteosarcoma and malignant fibrous hisitiocytoma.[6,7]

CONCLUSION

Fibrous dysplasia can present in rare locations. Therefore, imaging plays an important role in diagnosis and follow-up in the cases of cranio-facial fibrous dysplasia because of the anatomical complexity of the region. Many patients undergo multiple imaging during the process, so radiation exposure should be kept as low as possible.

DIFFERENTIAL DIAGNOSES

1. Frontal osteomyelitis
2. Frontal sinusitis
3. Malignancy involving ethmoid sinus

CONSENT

Written informed consent was obtained from the patient for publication of this case report.

FINANCIAL SUPPORT

The author(s) did not receive any financial support for the research and/or publication of this article.

CONFLICT OF INTEREST

The author(s) declare that they do not have any conflicts of interest with respect to the research, authorship, and/or publication of this article.

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