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Unusual Case of Craniofacial Fibrous Dysplasia

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Abstract

Craniofacial fibrous dysplasia is a rare bone disorder characterized by abnormal growth and deformity. We present a case report of a patient with craniofacial fibrous dysplasia, highlighting its radiological features, clinical presentation, and management. The patient demonstrated characteristic ground-glass appearance on CT scans and expansile lesions with sclerotic margins. Surgical intervention and rehabilitation resulted in improved symptom management and quality of life. This case report contributes to the understanding of craniofacial fibrous dysplasia and emphasizes the importance of multidisciplinary approach in diagnosis and treatment.

Keywords: Fibrous dysplasia, Craniofacial fibrous dysplasia, paranasal sinuses, CT, MRI.

Introduction

Fibrous dysplasia (FD) is a developmental benign medullary fibro-osseous process characterized by the failure to form mature lamellar bone and arrest as woven bone. It can be in a monostotic form involving only one bone or a polyostotic form involving multiple bones.

Classically, fibrous dysplasia lesions are well circumscribed, intramedullary and cause expansion of bone. Although endosteal scalloping may be present, they always maintain a smooth cortical contour. Lesions show varying degrees of hazy density with a ground-glass quality, although some may appear almost completely radiolucent or sclerotic. Hereby we report a case of pediatric male patient with unusual presentation of fibrous dysplasia extensively involving paranasal sinuses.

Case History

A 9 year old male child presented to paediatric OPD with complaints of an abnormal, large swelling over left cheek, associated with gradually progressing breathlessness, painless protrusion of left eye and headache since 1 year

On clinical examination, a nontender diffuse swelling was noted over left cheek causing left sided facial deformity causing bony prominence over left cheek. There was reddish soft tissue mass in left nasal cavity lying anterior to left inferior turbinate. The child had mucopurulent discharge through nose, depressed nasal bridge and deviation of nasal septum to right. Rest of the systemic evaluation did not reveal any other abnormality. Vitals were within normal range

MRI brain + orbit and CECT PNS was advised to evaluate morphology and extent of the mass lesion. Biopsy was planned for histopathological diagnosis.

Diagnostic Evaluation with imaging

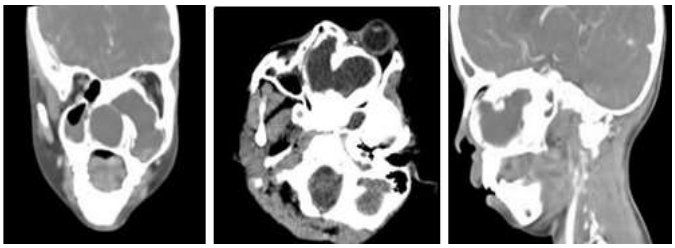

Figure 1:

The CECT PNS showing a large homogenous, hypodense, space occupying mass lesion arising from left maxillary sinus and invading left nasal cavity with no abnormal post contrast enhancement, associated with thickening of bony walls of the left maxillary sinus, widening of its osteomeatal unit and significant deviation of nasal septum to right. Significant anterolateral protrusion of left globe seen. Polypoid mucosal thickening in rest of the sinuses.

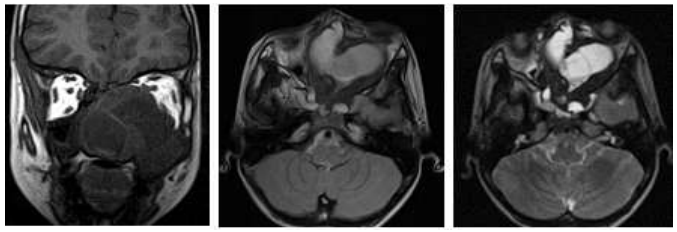

Figure 2:

On MRI images the lesion shows isointense signal on T1W images, hyperintense signal on T2W. It is multiloculated with few internal septations and fluid-fluid levels.

Discussion

Craniofacial fibrous dysplasia is considered a form of monostotic fibrous dysplasia despite multiple cranial bones involvement (10-25% of monostotic cases), or craniofacial involvement may occur as part of polyostotic fibrous dysplasia.

The maxillary, sphenoid, ethmoid, mandible being anterior craniofacial bones, are more frequently involved than more lateral or posterior bones. Extracranial involvement is rare.

Clinical presentation is usually due to cosmetic deformation like cranial or facial asymmetry or because of the mass effect on cranial structures causing nasal stuffiness, proptosis and visual impairment/unilateral blindness.

The affected bones demonstrate a variety of radiographic features like expansile blistering/bubbling cystic skull vault lesions which commonly cross sutures or sclerotic skull base with widened diploic space and obliteration of paranasal sinuses, classical homogeneous ground glass appearance is seen on CT, although mixed lucencies and sclerosis are also common.

Instead of conventional radiography, CT (Computed Tomography) and MRI (Magnetic Resonance Imaging) scans are commonly used to diagnose and evaluate craniofacial fibrous dysplasia.

Key CT Findings include

Homogenous ground-glass appearance is a characteristic feature of fibrous dysplasia, representing a mixture of fibrous and osseous tissue due to failure of formation of mature lamellar bone.

Lesions may expand the affected bone, causing cortical thinning and deformation. In craniofacial lesion the expansion of affected bone results in pressure effect on adjacent structures. It may result in narrowing of neural and vascular foramina if skull base is involved.

Lesions may have sclerotic borders, indicating the interface between the lesion and normal bone.

Key MRI Findings include

- T1-weighted images show hypointense signal due to the presence of fibrous tissue.
- Lesions may appear hyperintense on T2-weighted images due to presence of the cystic or fibrous components.
- Post gadolinium contrast enhancement may variable.

This case presented with unusual feature of internal septation with fluid-fluid levels mimicking aneurysmal bone cyst of maxilla and left maxillary sinus involvement with extension into the left nasal cavity mimicking chronic mucocele.

General imaging differential considerations include: cemento-ossifying fibroma, Paget's disease, intraosseous meningioma of skull vault.

Conclusion

In conclusion, this case report highlights the characteristic CT and MRI features of craniofacial fibrous dysplasia, The CT scan played a crucial role in diagnosing and assessing the extent of the disease, which is essential for guiding treatment decisions and predicting potential complications. The MRI findings provided valuable information on the extent of disease involvement, particularly in mass effect seen on the globe, optic nerve and rest of the soft tissues. The excellent soft-tissue contrast and multiplanar capabilities of MRI allowed for precise evaluation of the lesion's relationship with critical structures, facilitating surgical planning and management. This case highlights the complementary role of CT and MRI in the assessment of craniofacial fibrous dysplasia.

The patient's clinical presentation and imaging findings are consistent with craniofacial fibrous dysplasia, a rare and complex bone disorder requiring multidisciplinary management.

This case report contributes to the existing literature on this condition and emphasizes the importance of radiological evaluation in its diagnosis and treatment."

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