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A case of isolated craniofacial fibrous dysplasia – Radiologist's perspective

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ABSTRACT

Fibrous dysplasia is a disease characterized by replacement of bone by fibro osseous tissue. FD has four different disease patterns. They are monostotic, polyostotic and McCune–Albright syndrome. Craniofacial pattern of disease occurs in 10–25% of patients with monostotic form and in 50% with polyostotic form. It also occurs as isolated craniofacial form. In isolated variety no extracranial lesions are present. Radiographic diagnosis plays an important role in diagnosis, classification and assessing prognosis of fibrous dysplasia. In this paper we report a case of isolated craniofacial type of fibrous dysplasia in a young female patient involving the maxilla and skull bones with a complete radiographic CT assessment of the extent of the lesion. Temporal bone involvement and bilateral lesion in certain cranial bones are the rare findings noted in this case.

1. Introduction

Fibrous dysplasia is a skeletal developmental anomaly of the bone forming mesenchyme that manifests as a defect in osteoblastic differentiation and maturation. FD is caused due to the activating mutations in the GNAS gene which encodes for the alpha subunit of the signalling G protein, Gs alpha.[1] Following the suggestion of McCune and Bruch (1937), that this disorder is to be considered as a distinct clinical entity, Lichtenstein introduced the term Fibrous dysplasia.[2,3] Fibrous dysplasia is described in terms of three major types, monostotic involving a single bone, polyostotic involving multiple bones and McCune–Albright syndrome, a polyostotic form of fibrous dysplasia that also involves endocrine abnormalities.[4] Craniofacial pattern of disease occurs in 10–25% of patients with monostotic form and in 50% with polyostotic form. It also occurs as isolated craniofacial form. In isolated variety no extracranial lesions are present.[5] Facial region may be affected by a form of fibrous dysplasia that is not strictly monostotic, but may be confined to a single anatomical region. These lesions affect primarily the maxilla and simultaneously cross sutures and enter into the adjacent facial bones. This type of fibrous dysplasia does not meet the precise criteria for the monostotic or polyostotic forms and has been termed craniofacial fibrous dysplasia.[6,7] Though conventional radiographs are needed to evaluate the trabecular pattern, lamina dura and effect on adjacent structures, advanced imaging modalities like CT is essential for assessing the exact extension of the disease. In this paper we report a case of isolated craniofacial type of fibrous dysplasia involving the right maxilla and adjacent cranial bones in a young female patient with a complete radiographic assessment of the extent of the disease. As in most cases of fibrous dysplasia,

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clinical and radiographic features are sufficient to make a diagnosis, a radiologist is to be consulted to give a more informative three dimensional representation of the extent of the lesion. Also radiographs are used as reference tools for comparison in future.

2. Case report

A female patient of age 15 years came with a complaint of asymmetry of face due to a slow growing painless swelling in the right side of her upper jaw for the past one year. Extra oral examination revealed a diffuse swelling in the right side cheek region (Fig. 1). The swelling was bony hard in consistency and nontender. Intra oral examination revealed a swelling of size 3 × 2 cm present obliterating the right buccal sulcus and palatal surface extending from the mesial aspect of 12 to distal aspect of 17 (Fig. 2). The swelling was bony hard in consistency and nontender. A provisional diagnosis of fibro osseous lesion was given. Intra oral periapical radiograph of right anterior and posterior maxillary regions revealed ground glass appearance of trabeculae, with ill defined periphery, blending into the surrounding bone and disappearance of distinct lamina dura (Figs. 3 and 4). Maxillary occlusal view revealed ground glass appearance of trabeculae and expansion with a thinned out buccal cortex (Fig. 5). Panoramic radiograph revealed fine granular opacity involving the right maxillary antrum (Fig. 6). CT axial view (Fig. 7) revealed a homogenously dense lesion involving the right maxilla, obliterating the right maxillary sinus, with parallel thickening of the outer cortical border and a residual air space that has maintained the normal anatomic shape, characteristic feature found in fibrous dysplasia. Involvement of pterygoid plates is also seen.

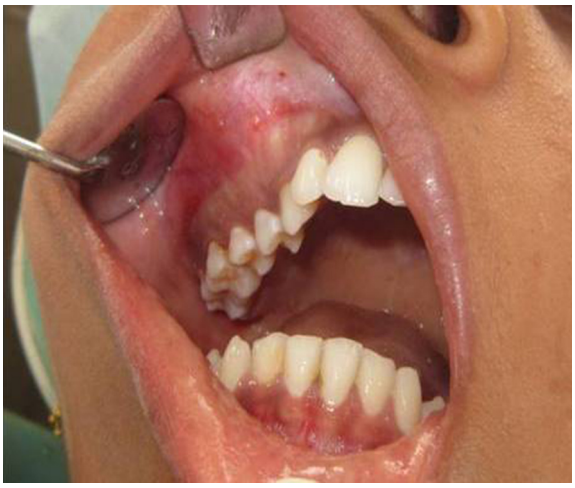

Fig. 2 – Preoperative intra oral view.

Further axial sections (Figs. 8–10) and coronal sections (Figs. 11 and 12) revealed obliteration of the sphenoid sinus, squamous portion of right temporal bone, posterior ethmoidal air cells and frontal bone. Complete skeletal analysis was done to rule out extracranial involvement. Biochemical analysis revealed an elevated serum alkaline phosphatase level (987 U/L – ref value (60–306 U/L)) and normal serum calcium and phosphorous levels. A working diagnosis of Fibrous dysplasia – craniofacial type was given as the lesion involved multiple bones of the craniofacial region only with no extracranial involvement. Histopathology revealed irregularly shaped bony trabeculae of varied sizes in a fibrocellular stroma (Fig. 13) which correlated with the clinical diagnosis. As the

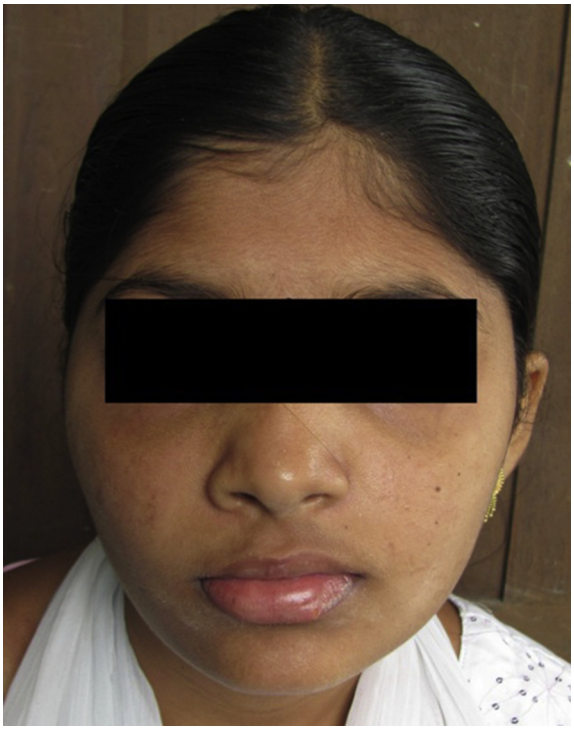

Fig. 1 – Preoperative extra oral view.

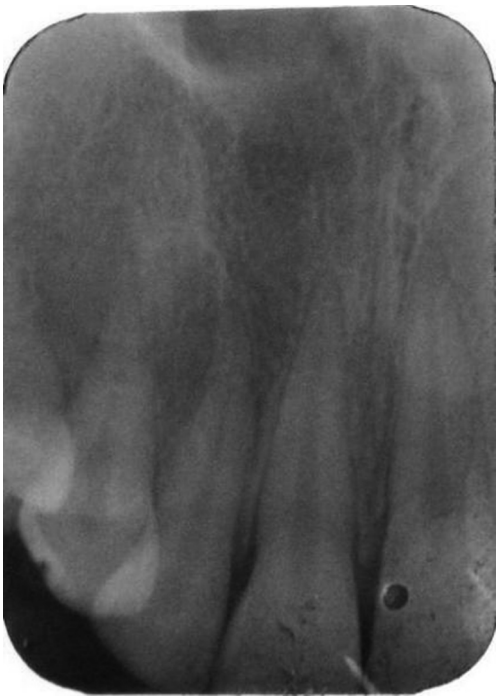

Fig. 3 – IOPA – ground glass appearance.