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Craniofacial Fibrous Dysplasia: A Case Report

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Abstract: Fibrous dysplasia is a Benign Fibro-Osseous disorder, characterized by fibrous connective tissue containing abnormal bone which replaces normal bone. It represents 2.5% of all bone lession and 7% of all benign tumors. It is a lesion of unknown etiology, uncertain pathogenesis, and diverse histopathology. Fibrous dysplasia can involve multiple bones (Polyostotic) or a single bone (Monostotic). Most commonly it affects younger age groups, with a higher prevalence in the Maxilla than the Mandible. Skull involvement is only about 15% with the majority of it being Monostatic. Here we report a case of craniofacial fibrous dysplasia in a 40-year-old female patient whose diagnosis was based on clinical findings and radiographic imaging.
Keywords: Computed tomography, Craniofacial, Fibro Osseous Lesion, Fibrous Dysplasia, Maxilla.

INTRODUCTION

Fibrous dysplasia is a tumour like lesion of the bone. It is self-limiting, is not encapsulated and is characterized by replacement of normal bone with cellular fibrous connective tissue [3], which contains irregular trubaculae of immature, nonlamellar, metaplastic bone. With an incidence of 1:4000-1:10,000 it is a rare disease. It represents 2.5% of all bone lession and 7% of all benign tumors.

CASE REPORT

A 40-year-old female presented with a swelling on the right side of the face for past 12 years. Initially the swelling was small in size, but it gradually and slowly progressed over the years to attain the present size. There was no history of trauma, fever and similar swelling elsewhere in the body, with a non-contributory past medical or dental and family history. There was no history of headache, visual disturbance, earache, nasal obstruction or nasal discharge.

On General Examination, the patient was of normal build and height with obvious facial asymmetry on the right side. On **Extraoral** (Figure 1) examination, a diffuse, bony hard swelling was present on the right side of the maxillary region, obliterating the nasolabial groove, measuring approximately 4cm X 6cm in size. With normal and intact overlying skin, Regional lymph nodes were not palpable. On **Intraoral** (Figure 2) examination, there was expansion of the right maxillary alveolar process and the mass over the hard palate measured 2x2cm is size. The overlying mucosa was normal in colour and was intact. On Palpation, the swelling was firm to hard in consistency and non-tender. Rest of the oral cavity appeared normal except the periodontal status of the teeth was poor.

Fig-1: Extraoral examination

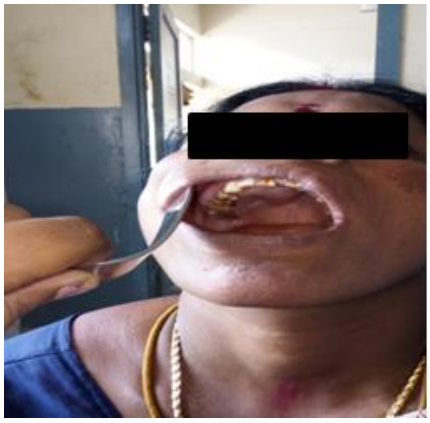

Fig-2: Intraoral examination

On the basis of the history and clinical examination, a provisional diagnosis of Benign osseous neoplasm was made, most propably a Fibro-Osseous lesion. Routine investigations were all within normal limits.

Computered tomography (axial and coronal section**)** scan revealed a diffuse, well defined, fibro osseous lesion with [7] ground glass matrix appearance involving the right maxilla upto gingival margin, all walls of right maxillary sinus, both pterygoid plates, greater and lesser wings of sphenoid and squamous part of temporal bone on right side, basisphenoid and posterior nasal septum.

Based on the history of long duration and asymptomatic nature of the lesion, clinical features and imaging features, showing typical ground-glass appearance with involvement of adjacent bones, a provisional diagnosis of Fibrous Dysplasia(Craniofacial type) was made. Due to its asymptomatic nature, patient was not willing for further treatment, so a regular follow-up was advised.

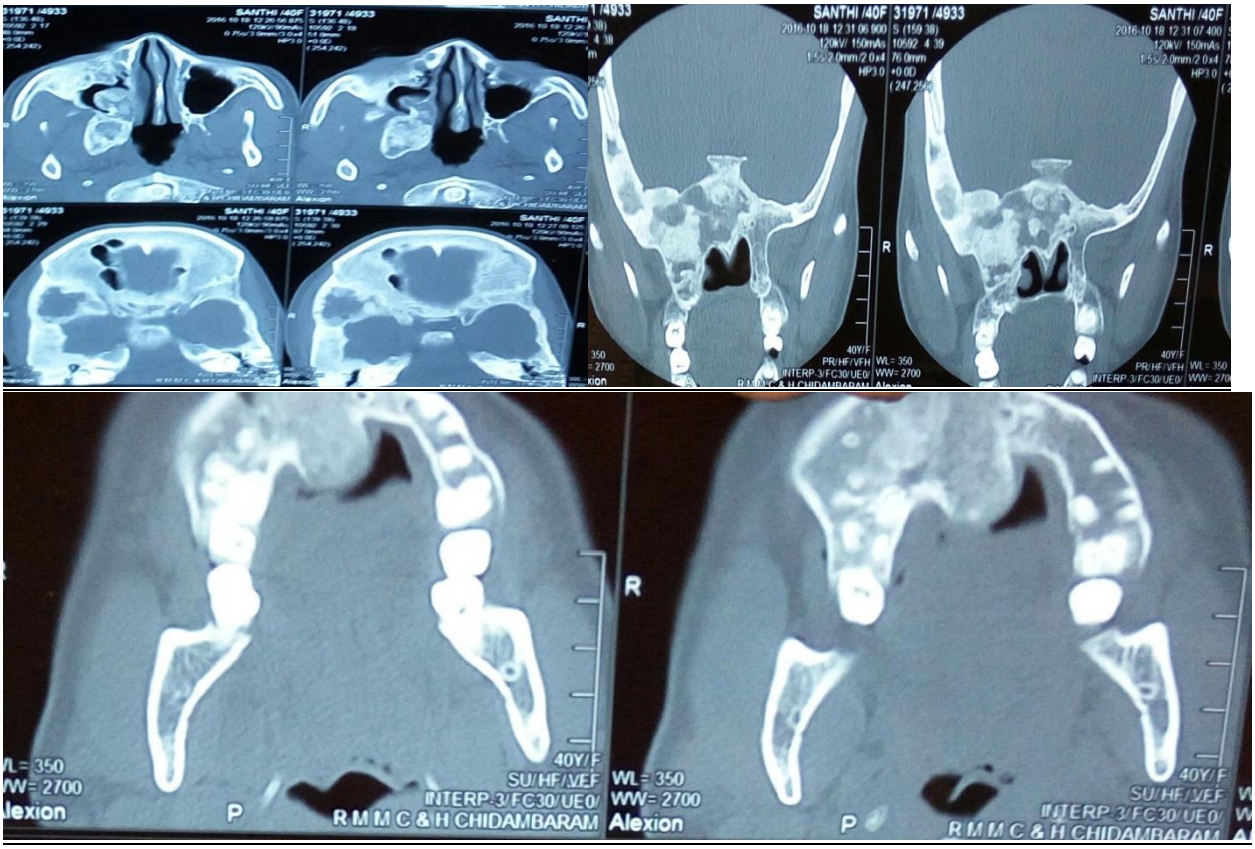

Fig-3: CT Scan (both axial and coronal section) scan revealed a diffuse, well defined, fibro osseous lesion with ground glass matrix appearance involving the right maxilla upto gingival margin, all walls of right maxillary sinus, both pterygoid plates, greater and lesser wings of sphenoid and squamous part of temporal bone on right side, basisphenoid and posterior nasal septum.

DISCUSSION

Von Recklinghausen [1] first described the condition in 1891 in a patient with skeletal deformities and coined the term “Osteitis Fibrosa Generalisata”. It was renamed “Fibrous Dysplasia” in 1938 by Lichtenstein. Perhaps the most accurate term to describe Fibrous Dysplasia is “Fibro-Osseous Dysplasia” [4, 5] or “Fibrous Osteodysplasia”. The etiology although unkown different hypotheses have been postulated. The disease may develop at an early age, progress actively during childhood and stabilize in adulthood. In the majority of cases, Fibrous Dysplasia is diagnosed

before the second decade of life, rarely after the fifth decade. This disease is much more frequent in white populations than black and the Monostotic form is found more often in females as in our case.

Clinically [2], three types of fibrous dysplasia are differentiated:

Monostotic, Polyostotic, McCune-Albright syndrome, which present as a combination of Polyostotic Fibrous Dysplasia, skin hyperpigmentation and endocrine dysfunction. It occurs in one of 30-40 cases of Fibrous Dyplasia [6].

Fibrous dysplasia can occur in both types of bones, enchondral and membranous. In addition to these two entities, another presentation is "Craniofacial Fibrous Dysplasia" where the lesions are confined to contiguous bones of the craniofacial skeleton. Craniofacial Dysplasia cannot be truly categorized as Monostotic because of the possibility of the involvement of multiple adjacent bones of the craniofacial skeleton but they are usually not involved. Between 50 to 100% of patient with Polyostotic disease will have craniofacial involvement, whereas only 10% with Monostotic lesions will have involvement of these structures.

In the head and neck region, maxilla is involved more frequently as in our case than the mandible, especially the posterior aspects of the jaws. Clinically, Craniofacial Fibrous Dysplasia generally presents as a gradually increasing, asymptomatic, bony hard and non-tender swelling with intact overlying surface. Since the condition is asymptomatic, patients seek treatment only when severe facial deformity or functional disturbances occur.

Diagnosis of Polyostotic Fibrous Dyplasia is generally based on clinical symptoms and radiological imaging. In contrast, the Monostotic Fibrous Dysplasia requires bone biopsy.

Recommended treatment options can be divided into 4 categories:
1.observation , 2.Medical therapy, 3.Surgical remodeling, 4.Radiacl excision and reconstruction.

In 1990 Chen and Noordhoff proposed a treatment algorithm for the management of Cranio Maxillofacial Fibrous Dysplasia incorporating aggressive, radical surgery for the resection of diseased tissue. For this algorithm, they proposed that the head and face could be divided into 4 zones based on the esthetic and functional consequences of the disease at each of these sites and the unique anatomic considerations for operating in each area.

Zone 1 [1] represents the fronto-orbito-malar regions of the face. These are esthetically critical and can be adequately reconstructed with simple bone grafting techniques after reconstruction. For this region, they recommended radical excision and reconstruction.

Zone 2 refers to the hair bearing scalp. It is not typically an aesthetic concern, and as such, intervention is optional for the patient.

Zone 3 refers to the central skull base including the sphenoid, pterygoid, petrous temporal bone, and mastoid. Given the difficulty in obtaining surgical access to these areas, the authors recommended observation of lesions in this region.

Zone 4 comprises the tooth bearing portions of the skull, the maxilla and mandible. The authors recommended conservative management, given the difficulty in reconstructing defects in this region.

CONCLUSION

The Fibrous Dysplasia affecting the craniofacial region is a distinctive entity that can in most cases be treated by conservative recontouring. The procedure is preferably indicated after the active growth phase has ceased. A long term follow up of these patients is mandatory considering the probable flare up and continuous growth of the lesion. We are reporting this case due to its rarity.

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