

## Monostotic Fibrous Dysplasia of Maxilla: A Case Report

Kotya Naik Maloth<sup>1,\*</sup>, Navadeepak Korvipati<sup>2</sup>, Kesidi.Sunitha<sup>3</sup>,  
Mithare Sagmesh<sup>4</sup>

<sup>1,3</sup>Senior Lecturer, Oral Medicine and Radiology,  
Mamata Dental College and Hospital, 5-7-200, Mamata General Hospital Campus,  
Khammam, Telanagana- 507002

<sup>2</sup>Senior Lecturer, Oral Medicine and Radiology,  
SVS Institute of Dental Sciences, Housing Board Colony,  
Yenugonda, Mahbubnagar, Telanagana

<sup>4</sup>Senior Lecturer, Oral Medicine and Radiology,  
HKDE Trust's Dental College and Hospital, Humnabad, India

### \*Corresponding Author

E-mail: dr.kotyanaik.maloth@gmail.com

### Abstract

Fibrous Dysplasia is a non-neoplastic developmental disease of bone, which manifest as a defect in osteoblastic differentiation and maturation. It represents with three forms namely monostotic (single bone involvement), polyostotic (multiple bone involvement) and craniofacial forms. Maxilla is affected most commonly twice as mandible, in the posterior region. This paper describes a case of 18 years old male who presented with swelling on right middle third of the face and intraoral swelling on right posterior maxilla since one year. The diagnosis was based on clinico-radiological and histopathological investigations. The appropriate management was done and he is under followup without any recurrence.

**Keywords:** Craniofacial type, Fibrous lesions, Hamartomatous, Monostotic, polyostotic.

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### Introduction

Fibrous dysplasia is a benign hamartomatous disease of the bone, characterized by replacement of bone with fibrous tissue.<sup>1</sup> Incidence of occurrence is 1:4000 to 1:10000 and it seems to be a rare disease.<sup>2</sup> It affects equally in both the gender, and it represents approx. 2.5% of all bone lesions and about 7% of all benign bone tumors.<sup>3</sup>

The lesion was first described by Von Recklinghausen in 1891 as "Osteitis Fibrosa Generalisata". Later in 1938 Lichtenstein and Jaffe coined the term "Fibrous Dysplasia".<sup>3</sup> Based on the clinical presentation and involvement of the bone, it is divided into three types-Monostotic (single bone involvement), Polyostotic (multiple bone involvement) and craniofacial type. The craniofacial fibrous dysplasia are confined to contiguous bones of the craniofacial skeleton, and it has a slight female predilection.<sup>3</sup> The malignant transformation is rare i.e. 0.4%.<sup>4</sup>

### Case Report

An 18years-old-male patient reported to our department with a chief complaint of swelling on the

upper right back tooth region since one year. Initially the swelling was small like a peanut size and attained the present size with no history of pain. On extra oral examination a solitary diffused oval shaped swelling measuring of size approx. 2x3cms was seen on the right middle 3<sup>rd</sup> of face extending antero-posteriorly 1cm away from the ala of nose and 3cm in front of the right ear, superio-inferiorly at the level of the floor of the orbit, and inferiorly 1cm below the ala tragus line. No visible pulsations were seen. On palpation, there was no local rise in the temperature, non tender, non fluctuant, non compressible and non reducible (Figure 1). On intraoral examination a solitary oval shaped diffuse swelling measuring approx. 4x3 cm was seen on the upper right back tooth region, extending antero-posteriorly from distal aspect of 13, to distal aspect of 17. On palpation it was hard in consistency, non tender, non compressible, non reducible and fixed to the underlying structures (Figure 2). Based on the history & clinical findings a provisional diagnosis of fibrous dysplasia was given with a differential diagnosis of Osteoid osteomas, Cemento-ossifying fibroma, Odontogenic myxoma, Ameloblastoma.

Routine investigations such as hematology, serum calcium and serum alkaline phosphatase levels were performed, which were within normal limits. Intraoral periapical radiograph revealed radiopaque area with cotton-wool appearance (Figure 3), maxillary occlusal radiograph revealed radiopaque area with expansion of buccal cortical plate (Figure 4), Orthopantomogram revealed radiopaque area occupying entire right maxillary sinus with ground-glass appearance (Figure

5), Para-nasal sinus (PNS) view revealed radiopaque area occupying entire right maxillary sinus extending till infra orbital rim (Figure 6). Computed Tomography revealed a mixed radiolucent and radiopaque expansile lesion with patchy calcifications involving right maxillary alveolus and maxillary sinus (Figure7).

Excisional biopsy was done from right maxilla and the excised specimen (Figure 8) was sent for histopathological examination which revealed a fibro-cellular connective tissue stroma with irregular areas of osseous tissue of varying size and shape, predominantly slender with curvilinear pattern and haphazardly arranged osteocytes in large lacunae. The focal areas exhibit osteoblastic rimming. The intervening connective tissue shows haphazardly arranged fibroblasts and irregular trabeculae along with delicate collagen fibers and extravasated blood cells, suggestive of fibrous dysplasia (Figure 9). Based on clinical, radiological and histopathological findings, a final diagnosis of Monostotic Fibrous Dysplasia was given.



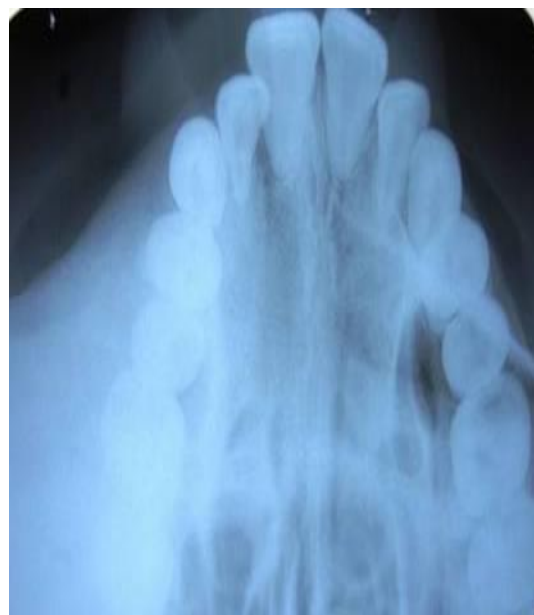
**Fig. 1: Extraoral photograph**



**Fig. 2: Intraoral photograph**



**Fig. 3: IOPA revealed cotton-wool appearance**



**Fig. 4: Maxillary Occlusal radiograph revealed radiopaque area with expansion of right buccal cortical plate**



Fig. 5: OPG revealed radiopaque area occupying entire right maxillary sinus with ground-glass appearance



Fig. 6: PNS-view revealed radiopaque area occupying entire right maxillary sinus

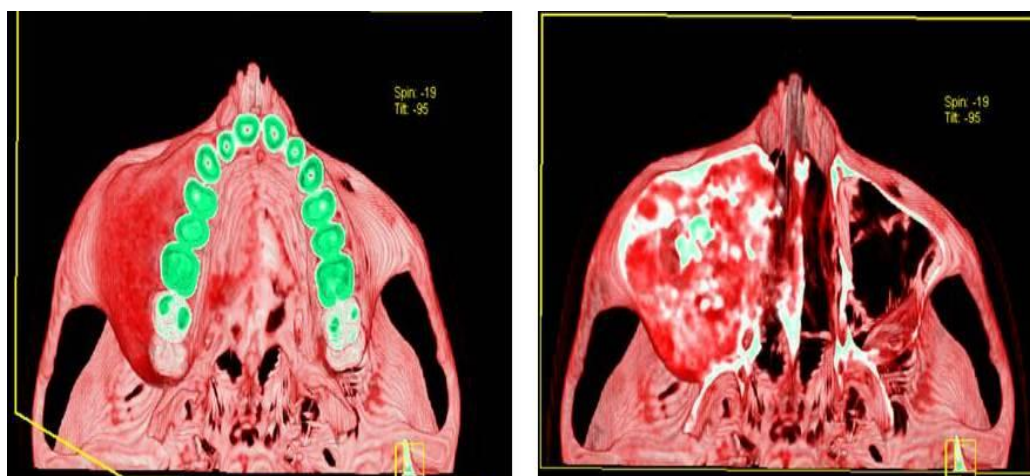
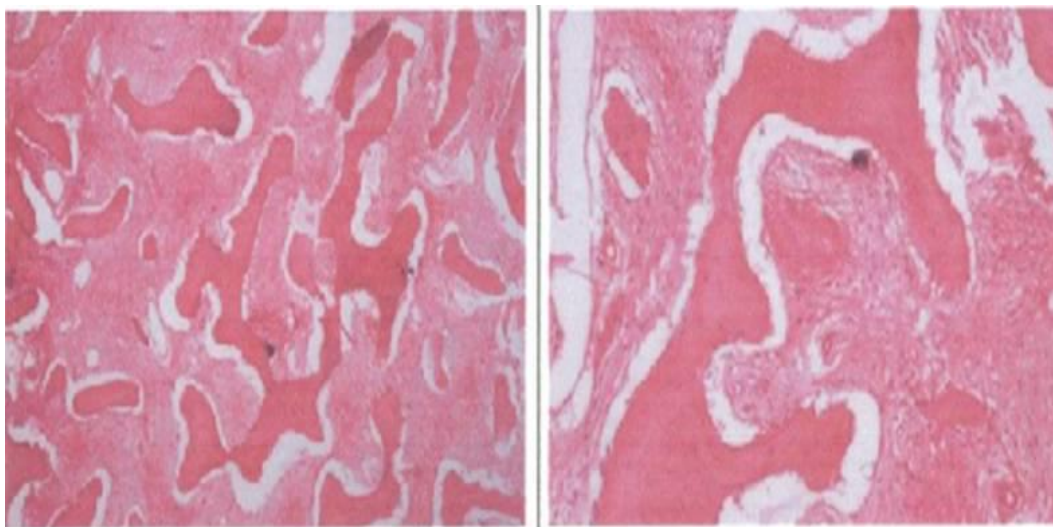


Fig. 7: computed tomography revealed a mixed radiolucent and radiopaque expansile lesion with patchy calcifications



**Fig. 8: Excised specimen**



**10X**

**40X**

**Fig. 9: Histopathological picture revealed irregularly shaped trabeculae of osteoid showing Chinese character pattern or jigsaw puzzle in the fibrous stroma.**

### Discussion

Fibrous dysplasia is a common benign fibroosseous lesion characterized by replacement of normal bone by excessive proliferation of cellular fibrous connective tissue intermixed with irregular bony trabeculae.<sup>5</sup> The exact etiology of fibrous dysplasia is uncertain and is probably a genetic predisposition, by mutation in *GNAS1* gene (guanine nucleotide binding protein), found on chromosome 20q13 or abnormalities in *AMPC* which may result in increased proliferation of melanocytes causing cafeu-lait spots and hyperfunction of affected endocrine organs.<sup>6,7</sup>

Both the genders are equally affected, although the mean age of occurrence was 27 years in 69 patients as reported by Zimmerman.<sup>8</sup> In the present case patient was male and was 18 years old.

According to Abdulai et al.,<sup>9</sup> fibrous dysplasia affects maxilla twice as common than mandible, and is usually seen in the posterior region, as in the present case right posterior maxilla was involved. The polyostotic variant usually affects children younger than 10 years, whereas monostotic variant affects individuals in the second and third decade<sup>10</sup>, as in the present case patient was 18 years old.

The clinical presentation of the fibrous dysplasia shows asymptomatic diffuse swelling in the affected region, causing aesthetic impairment and deformities. As the lesion progresses the clinical symptoms occurs, such as visual disturbances, proptosis, orbital dystopia, nasal malfunction, dental problems and sensory disturbances in the affected regions.<sup>3</sup>

The radiographic appearance of FD lesions is extremely unique and will vary depending on the stage of development and quantity of bone matrix within the lesion. Early lesions may be more radiolucent than mature lesions and in rare cases may appear to have granular internal septa, giving the internal aspect a multilocular appearance. The abnormal trabeculae usually shorter, thinner, irregularly shaped and more numerous than normal trabeculae. This creates a variable radiopaque pattern, it may have a granular appearance ('ground-glass' appearance), a pattern resembling the surface of an orange (orange-peel), a wispy arrangement (cotton wool), swirling pattern (fingerprint) or an amorphous, dense pattern.<sup>10</sup> Prapayasatok et al<sup>11</sup> reported a case of, a rare radiographic 'sunray' appearance of the lesion. In the present case, the panoramic radiograph revealed a 'ground-glass' appearance and intraoral periapical radiograph revealed a "cotton-wool appearance" of the affected area.

Histopathology of fibrous dysplasia constitutes essentially fibrous component with proliferating fibroblasts and the osseous component includes irregularly shaped trabeculae of osteoid showing Chinese character pattern or jigsaw puzzle appearance along with immature woven bone in the fibrous stroma.<sup>6</sup>

There are no uniformly accepted guidelines for the management of this disease. The recommended treatment options are observation, medical therapy and surgical therapy. Observation is the mainstay of treatment modality for small asymptomatic lesions that are esthetically acceptable to the patient.<sup>3</sup> Medical therapy does not play a prominent role in the management of the fibrous dysplasia, however few drugs like bisphosphonates, Pamidronate (60 mg/day intravenous route) reduces osteoclastic activity. The calcitonin, Vitamin-D and calcium supplements were recommended for patients with low serum calcium levels.<sup>3</sup>

Surgical therapy remains the mainstay of treatment for large lesions, directed at correcting (remodelling) or preventing functional deficits and achieving normal facial esthetics. In the present case remodelling was done.<sup>3</sup> The recurrence rate of fibrous dysplasia is rare i.e 15% to 20%, when it occurs in adults. But it is more common in growth period. Increased serum alkaline

phosphatase (ALP) levels, is the main indicator for detection of recurrent lesions.<sup>12,13</sup>

## Conclusion

Fibrous Dysplasia is a most common fibroosseous lesion occurring in younger individuals causing cosmetic and functional problems. Many treatment modalities have been recommended though still unsatisfactory, as the small lesions that are esthetically acceptable to the patient kept under observation till the puberty, surgical intervention is done after the active growth phase has ceased.

**Conflict of Interest: None**

**Source of Support: Nil**

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