

## MONOSTOTIC FIBROUS DYSPLASIA – A CASE REPORT

<sup>1</sup> Pournima Godge  
<sup>2</sup> Shubhra Sharma

<sup>1</sup> Reader  
<sup>2</sup> lecturer

<sup>1,2</sup>Terna dental college and hospital, Navi Mumbai, Maharashtra, India.

**ABSTRACT:**

Fibrous dysplasia is a benign fibro-osseous lesion of the bone that commonly affects the jaws with a higher prevalence in the maxilla than the mandible. It is a lesion of unknown etiology, uncertain pathogenesis, and diverse histopathology. Fibrous dysplasia can involve multiple bones (polyostotic) or a single bone (monostotic). It represents about 2 to 5% of all bone tumors and over 7% of all benign tumours. In this article we report the case of a 20 year old patient with monostotic fibrous dysplasia involving the left maxilla.

**KEYWORDS:** Fibrous dysplasia, Fibroosseous lesion, Monostotic

**INTRODUCTION**

Fibrous dysplasia is a disturbance of bone metabolism that is classified as a benign fibro-osseous lesion. It is defined as "a benign lesion, presumably developmental in nature, characterized by the presence of fibrous connective tissue with a characteristic whorled pattern and containing trabeculae of immature non-lamellar bone".<sup>1</sup>

In 1937, McCune and Bruch first suggested that among all of the abnormalities of bone formation, this disorder should have its own place as a distinct clinical entity. The following year, Lichtenstein introduced the term "fibrous dysplasia".<sup>2</sup> In 1942, Lichtenstein and Jaffe', reported different clinical manifestations for fibrous dysplasia that is solitary (monostotic) and multiple (polyostotic) forms.

Monostotic fibrous dysplasia, characterized by involvement of only one bone, is considerably more prevalent than the polyostotic form. The jaws are more commonly affected with maxilla being more frequent than the mandible. In 10% of monostotic fibrous dysplasia and 50% to 100% of polyostotic fibrous dysplasia, there is involvement of the facial and cranial bones.<sup>3</sup>

Fibrous dysplasia is a sporadic condition that results from a postzygotic mutation in the GNAS1 (guanine nucleotide binding protein,  $\alpha$  -stimulating activity polypeptide1) gene.<sup>4</sup>

Males and females are thought to be affected evenly, although recent research has shown a slight female preponderance. Clinically a painless enlargement of the affected bone is the most common presenting symptom. Bulging of the canine fossa and a hyperprominence of the zygomatic process of the maxilla are usual features of maxillary lesions with frequent involvement of maxillary sinus.<sup>5</sup>

Malignant potential of fibrous dysplasia is less than 1%. The radiographic appearance of fibrous dysplasia is extremely variable and depends on the proportion and distribution of fibrous deposition. This report will present a case of isolated fibrous dysplasia of the maxilla in a 20 year old female patient.

**Case report**

A 20 year old female, reported to the Dept of Oral Pathology with a history of swelling on the left side of her face since 6 months.

Extraoral examination revealed a diffuse bony hard swelling on the left side of the face measuring approximately 3x4 cms in size. The lesion extended laterally from the lateral border of the nose upto the posterior border of the masseter. Superiorly the lesion extended upto the inferior border of the orbit causing displacement of the eye. The inferior extension of the lesion was approximately 2 cm below the ala tragus line. There was unilateral bulging of the canine fossa and prominence of the zygomatic process with no apparent changes in the overlying skin. Epiphora was positive. She denied headache, deficit of visual accuracy, otalgia, nasal obstruction and toothache.

A small, slightly tender lymph node could be palpated in the left submandibular region.

Intraoral examination revealed an ill defined swelling obliterating the left buccal vestibule causing expansion of the buccal cortical plate in that area, measuring approximately 3x3.5 cms in size. The extension of the lesion on palate was approaching the midline. Grade II mobility was evident in 23, 24 and 25 although there was

minimal displacement of the teeth. The overlying mucosa was of normal colour and intact as was the rest of the mucosa of oral cavity. The initial diagnosis based on the clinical examination was that of fibrous dysplasia .

Radiographic evaluation (Waters view) revealed diffuse radioopacity approximately 4x4 cms in size involving the maxillary sinus and extending into the inferior border of the orbit, lateral border of the nasal fossa and zygomatic arch . The radioopacity appeared to be more homogeneous and less granular. On the right side of the face, the maxillary sinus, zygomatic arch and maxilla appeared to be normal. Similarly other craniofacial bones and mandible appeared to be unaffected.

A gross surgical recontouring of the maxilla for functional reasons was performed under general anesthesia. Free flaps were raised and an ileac crest grafts were placed.

The histopathological picture of the decalcified sections (H&E) revealed coarse bony trabeculae with osteocytes, absence of osteoblastic rimming around the trabeculae scattered in a cellular fibrous tissue stroma. The trabeculae which were irregular in size and shape rather slender, consisted of immature woven bone, whose margins were not removed with osteoclasts. Microcyst formation due to focal degeneration of the fibrous tissue was seen at places. The lesional bone was seen to fuse directly with the normal bone at the periphery of the lesion so that no capsule or line of demarcation was present. Individual multinucleated giant cells in areas of hemorrhage were seen at one or two places.

Thus based on the clinical, radiological and histological findings the patient could be diagnosed as having Monostotic Fibrous dysplasia of left maxilla .

## Discussion

Fibrous dysplasia is a developmental tumor like condition that is characterized by replacement of normal bone by an excessive proliferation of cellular fibrous connective tissue intermixed with irregular bony trabeculae<sup>6</sup>.

It is classified by the number of affected bones and the presence or not of extra-skeletal abnormalities. The monostotic form affects only one bone and corresponds to 70-80% of the FD cases. The polyostotic form, in which several bones are affected, may be divided into three subtypes: craniofacial, in which only the craniofacial complex is involved including the mandible and the maxilla ; Lichtenstein-Jaffe, in addition to the involvement of several skeletal bones there are café au lait

pigmentations; Albright's Syndrome, characterized by the involvement of several bones, café au lait pigmentations in the skin and multiple endocrinopathies with a remark for the early adolescence in girls. The polyostotic form corresponds to 20-30% of the cases<sup>7</sup>.

The etiology of this abnormal growth process is related to a mutation in the gene that encodes the sub unit of a stimulatory G protein (G $\alpha$ ) located on chromosome 20. As a consequence of this mutation, there is a substitution of the cysteine or the histidine—amino acids of the genomic DNA in the osteoblastic cells—by another amino acid, arginine. Consequently, the osteoblastic cells elaborate a fibrous tissue in the bone marrow instead of normal bone<sup>4,8</sup>.

With initial development of fibrous dysplasia the patient usually reports facial swellings and asymmetries. In a systematic review of previous studies of fibrous dysplasia, McDonald-Jankowski determined that FD mostly occurs in females, the maxilla is the most common facial bone affected and the posterior aspects of the jaw are more frequently involved than the anterior<sup>9</sup>.

The differential diagnosis of fibrous dysplasias includes lesions like ossifying fibroma, Paget's disease, Diffuse sclerosing osteomyelitis etc.

Chronic ossifying fibroma ( COF ) is a benign neoplasm that commonly has a radiographic and histological appearance similar to that of fibrous dysplasia.<sup>10</sup> Tissue sections of COF show a cellular or sclerotic fibrous connective tissue stroma containing numerous osseous trabeculae of various sizes associated with prominent osteoblasts.<sup>11</sup> A mixture of lamellar and woven bone is typically seen. Often, there are scattered ovoid calcifications that resemble cementum.

Fibrous dysplasia may also mimic Paget's disease of bone . Certain radiographic and clinical features like bilateral occurrence of Paget's disease in jaws , thickening of the cortices, cotton wool appearance of the involved bone and increased blood levels of alkaline phosphatase helps distinguishing it from FD .<sup>12</sup> Histologically, Paget's disease exhibits many osseous trabeculae with prominent reversal lines showing simultaneous osteoblastic and osteoclastic activity. The affected bone resides within a well-vascularized fibrous connective tissue stroma.

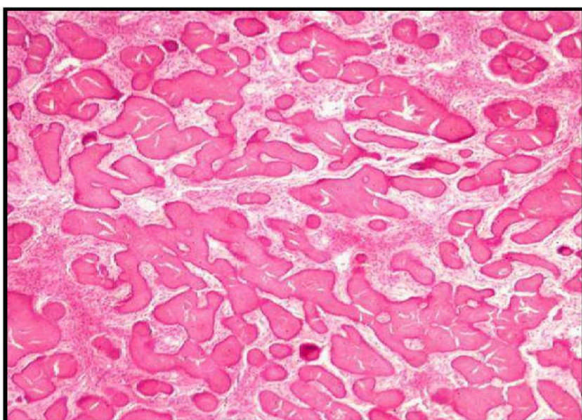
Diffuse sclerosing osteomyelitis (DSO) is a sequela of chronic jaw infection and inflammation. It presents radiographically as an ill-defined radiopacity, often encompassing large areas of bone, which may exhibit small radiolucent zones.



**Fig.1. Diffuse extraoral swelling in the left maxilla, approximately 3x4 cms in size.**



**Fig.2. Waters view - showing diffuse radioopacity approximately 4x4 cms in size in the left maxilla**



**Fig.3. H & E stained section suggestive of Fibrous dysplasia**

Histologically, DSO demonstrates sclerotic bone showing alternating areas of apposition and resorption. Between the bone trabeculae lies fibrous connective tissue infiltrated by chronic inflammatory cells<sup>13</sup>.

The main factors that guide the FD approach are the presence and the intensity of the symptoms, the tumor location and the patient's age. The main indications for surgical treatment of FD are the presence of significant clinical symptoms and the control of large aesthetic deformities.<sup>2, 9</sup> Orbital involvement with loss of vision is one of the most severe but relatively uncommon complications of fibrous dysplasia.

Biphosphonates are used in cases when an intervention is necessary but the surgery cannot be performed.<sup>14</sup> Some authors suggest applying calcitonin in combination with surgical treatment.<sup>15</sup> Usually the prognosis is good although the bad outcomes occur more frequently among young patients or those with polyostotic forms of the disorder. Radiotherapy is contra-indicated not only because the tumor is radioresistant but also because of the probable increase of the capacity for the dysplasia sarcomatous transformation.

**CONCLUSION**

Fibrous dysplasia represents a variety of disease processes with different behaviours, including infection and endocrine dysfunction resulting in a need for prompt diagnosis incorporating clinical, radiographic and histologic findings. Isolated cases of fibrous dysplasia in maxillo mandibular region are rare and can be difficult to differentiate from other benign and malignant bone disorders. The general dental practitioner can be the first to detect such conditions especially when the only affected areas are in maxillo-mandibular region. So sufficient knowledge on this condition is important for the proper diagnosis, treatment and prevention of further complications.

**References:**

1. Lionel G, Deborah C: Fibro-osseous Diseases oral and Maxillofacial Surgery Clinics of North America: Surgical Pathology, Nov: 551-561,1997.
2. Cholakova R., P. Kanasirska, N. Kanasirski, Iv. Chenchev, A. Dinkova. Fibrous dysplasia in maxillomandibulo region -ACase reportJournal of IMAB - Annual Proceeding (Scientific Papers) vol. 16, book 4, 2010.
3. Ricalde P, Horswell BB Craniofacial fibrous dysplasia of the fronto-orbital region: a case series and literature review. J Oral Maxillofac Surg. 2001 Feb;59(2):157-67; discussion 167-8 PMID.

[doi:10.1053/joms.2001.20487](https://doi.org/10.1053/joms.2001.20487)

PMid:11213984

4. Sakamoto A, Oda Y, Iwamoto Y, Tsuneyoshi M. A comparative study of fibrous dysplasia and osteofibrous dysplasia with regard to Gs alpha mutation at the Arg201 codon: polymerase chain reaction-restriction fragment length polymorphism analysis of paraffin embedded tissues. *J Mol Diagn.* 2000 May; 2(2): 67-72.  
[doi:10.1016/S1525-1578\(10\)60618-6](https://doi.org/10.1016/S1525-1578(10)60618-6)
5. Zimmerman DC, Dahlin Dc, Stafne EC, Fibrous dysplasia of the maxilla and mandible oral surg Oral Med Oral Pathol 1958; 11(1):55-68.
6. Neville Damm Allen Bouquot ; Textbook on Oral and Maxillofacial pathology, second edition.
7. Tinoco P, Pereira JCO, Filho RCL, Silva FCB, Ruela KP .Fibrous dysplasia of maxillary sinus. *Arq. Int. otorrinolaringol.* 2009; 13(2):214-217.
8. Sargin H, Gozu H, Bircan R, Sargin M, Avsar M, Ekinci G, Yayla A, Gulec I, Bozbuga M, Cirakoglu B, Tanakol R. A case of McCune-Albright syndrome associated with Gs alpha mutation in the bone tissue. *Endocr J.* 2006 Feb; 53(1):35-4410. PMid:16543670  
[doi:10.1507/endocrj.53.35](https://doi.org/10.1507/endocrj.53.35)
9. McDonald-Jankowski D. Fibrous dysplasia in the jaws of a Hong- Kong population: radiographic presentation and systematic review. *Dentomaxillofac Radiol* 1999; 28(4):195–202.  
[doi:10.1038/sj.dmf.4600440](https://doi.org/10.1038/sj.dmf.4600440)
10. Differential diagnosis in fibro-osseous lesions of facial bones: report of a case of ossifying fibroma and review of literature. *Acta Otorhinolaryngol Ital* 2002; 22(5):295–300. PMid:12510342
11. Slootweg PJ. Maxillofacial fibro-osseous lesions: classification and differential diagnosis. *Semin Diagn Pathol* 1996; 13(2):104–121. White SC, Pharoah MJ. PMid:8734416
12. Oral radiology. Principles and interpretation. 5th ed. St. Louis (MO): Mosby Inc; 2004. p. 485–515.
13. Groot RH, van Merkesteyn JP, Bras J. Diffuse sclerosing osteomyelitis and florid osseous dysplasia. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod* 1996; 81(3):333–42.  
[doi:10.1016/S1079-2104\(96\)80334-9](https://doi.org/10.1016/S1079-2104(96)80334-9)
14. Parekh SG, Donthineni-Rao R, Ricchetti E, Lackman RD. Fibrous dysplasia. *J Am Acad Orthop Surg.* 2004 Sep-Oct; 12(5):305-13. PMid:15469225
15. Yasuoka T, Takagi N, Hatakeyama D, Yokoyama K. Fibrous dysplasia in the maxilla: possible mechanism of bone remodeling by calcitonin treatment. *Oral Oncol.* 2003 Apr; 39(3):301-5.  
[doi:10.1016/S1368-8375\(02\)00037-4](https://doi.org/10.1016/S1368-8375(02)00037-4)

#### Corresponding Author

#### Dr. Pournima Godge

Reader,  
Terna Dental College,  
Navi Mumbai  
Maharashtra, India.  
Contact no.- 09867315341  
E-mail address-  
pournimagodge@rediffmail.com