

MONOSTOTIC FIBROUS DYSPLASIA OF MAXILLARY BONE: A CASE REPORT**¹Dr. Udupikrishna M. Joshi, ²Dr. Kundan Shah* and ³Dr. Satishkumar G. Patil**¹Professor and HOD, Department of Oral and Maxillofacial Surgery, Hkes S N Dental College, Gulbarga.²Department of Oral and Maxillofacial Surgery.³Professor, Department of Oral and Maxillofacial Surgery, Hkes S N Dental College, Gulbarga..***Corresponding Author: Kundan Shah**

MDS, Department of Oral and Maxillofacial Surgery.

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ABSTRACT

Fibrous dysplasia (FD) is a benign fibro-osseous bone disease of unknown etiology and uncertain pathogenesis. It manifest as a defect in osteoblastic differentiation and maturation. It mainly occurs in two forms namely monostotic (single bone involvement) and polyostotic (multiple bone involvement). Maxillary posterior region is most commonly affected. This article describes a case of 18 years old male who presented with swelling on left middle third of the face and intraoral swelling on left posterior maxilla since five year. The diagnosis was based on clinico-radiological and histopathological investigations. The appropriate surgical management was done and he is under followup without any signs of recurrence.

KEYWORDS: Fibro-osseous, Monostotic, Polyostotic, Maxillary Posterior, Surgical Management.**INTRODUCTION**

Fibrous dysplasia (FD) is a benign hamartomous fibro-osseous pathologic condition characterized by the replacement of bone with fibrous tissue^[1,2] The lesion was first described by Von Recklinghausen in 1891 as "Osteitis Fibrosa Generalisata"³. 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.^[4] Later in 1938 Lichtenstein and Jaffe coined the term "Fibrous Dysplasia"^[3] It is a benign bone disorder of an unknown etiology, uncertain pathogenesis and diverse histopathology.^[5] It is a rare disease with Incidence of occurrence is 1:4000 to 1:10000.^[6] The disease has no sex predilection and it represents approx. 2.5% of all bone lesions and about 7% of all benign bone tumors.^[3]

Fibrous dysplasia is described in terms of three major types: monostotic, involving a single bone; polyostotic, having multiple lesions involving multiple bones; and McCune Albright syndrome, a polyostotic form of fibrous dysplasia that also involves endocrine abnormalities.^[4] In addition to these forms, Jones described hereditary familial form of localized FD which is called cherubism.^[7] The monostotic form is more common and affects 20–30 years of age while the polyostotic form has its onset mainly in children younger than 10 years of age.^[8]

The clinical findings are asymptomatic, but the involved bone's enlargement occurs which causes facial asymmetry.^[9] The complications of the lesions involving

sphenoid, orbital, frontal bones, are proptosis, visual disturbances, facial asymmetry and orbital dystopia.^[10,11] The fifth nerve impairment, hearing loss and seizure disorders have been reported as neurological complications.^[12]

Malignant transformation is rare and occurs in 0.4% of cases of fibrous dysplasia.^[13] Malignancies are almost exclusively osteosarcoma. For unknown reasons, monostotic and craniofacial lesions have the greatest potential for malignant degeneration. Pain, rapid growth of a lesion and a dramatic elevation of alkaline phosphatase may herald malignant transformation.^[4]

In this article we report a case of Monostotic fibrous dysplasia involving left side of maxilla causing bony expansion and facial asymmetry.

CASE REPORT

An 18years-old-male patient reported to our department with a chief complaint of swelling on the upper left back cheek region since five years. Initially the swelling was small in size which gradually increased to attain the present size. The patient doesn't give any history of pain associated with it. On extra oral examination a solitary diffused oval shaped swelling measuring of size approx. 3x2.5 cms was seen on the left middle 3rd of face extending antero-posteriorly 1.5 cm away from the ala of nose and 3cm in front of the tragus of the ear, superior-inferiorly at the level of the floor of the orbit, and inferiorly 1cm below the ala tragus line. No visible pulsations, discharge or changes in the skin over the

lesion were seen. On palpation, there was no local rise in the temperature and the swelling was non tender, non fluctuant, non compressible and non reducible (Fig 1). On intraoral examination a solitary oval shaped diffuse swelling measuring approx. 3.5x2 cm was seen on the upper left back tooth region, extending antero-posteriorly from mesial 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 (Fig 2). Based on the history & clinical findings a provisional diagnosis of fibrous dysplasia was given with a differential diagnosis of Cemento-ossifying fibroma, Odontogenic myxoma and Osteoid osteomas.

Intraoral periapical radiograph was taken which revealed radiopaque area in relation to 25,26 and 27 (Fig 3), maxillary occlusal radiograph revealed radiopaque area with expansion of buccal and lingual cortical plates (Fig 4), Orthopantomogram revealed radiopaque area occupying entire left maxilla showing cotton wool appearance (Fig 5). Computed Tomography reveals a ground glass opacity in left maxillary sinus with bony

expansion. Lesion extending superiorly till floor of the left orbit, laterally involving zygoma, inferiorly the alveolar margin on left side and extending till last molar. The lesion approximately measures 4.6x4.5x3.8 cms (Fig 6). Based on the radiographic findings a diagnosis of fibrous dysplasia of left maxilla was given.

Routine blood and urine investigations were performed, which were within the normal limits. After taking the informed consent from the patient. Bony recontouring was done of left maxilla (Fig 7) and the excised specimen was sent for histopathological examination which revealed a fibro-cellular connective tissue stroma with irregular areas of osseous tissue of varying shape and size with predominantly arranged osteocytes in lacunae. The focal areas exhibits 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 (Fig 8). Based on clinical, radiological and histopathological findings, a final diagnosis of Monostotic Fibrous Dysplasia was given.

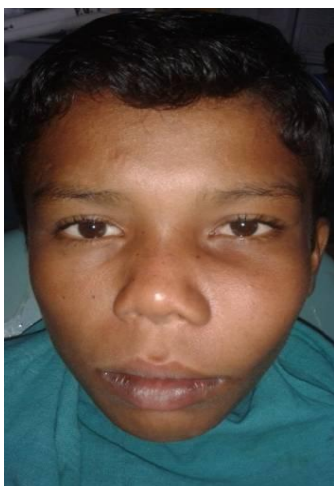


Figure 1: Preoperative extraoral profile showing swelling on the left cheek.



Figure 2 : Preoperative profile showing swelling intraorally on maxillary left posterior region.



Figure 3 : Intraoral periapical radiograph reveals radiopaque area in relation to 25,26 and 27.



Figure 4: Maxillary occlusal radiograph reveals radiopaque area with expansion of buccal and lingual cortical plates.



Figure 5 : Orthopantomogram revealed radiopaque area occupying entire left maxilla.



Figure 6 : Computed Tomography reveals a ground glass opacity in left maxillary sinus with bony expansion and the extent of lesion.



Figure 7 : Intraoperative picture demonstrate bony recontouring wrt left maxilla.

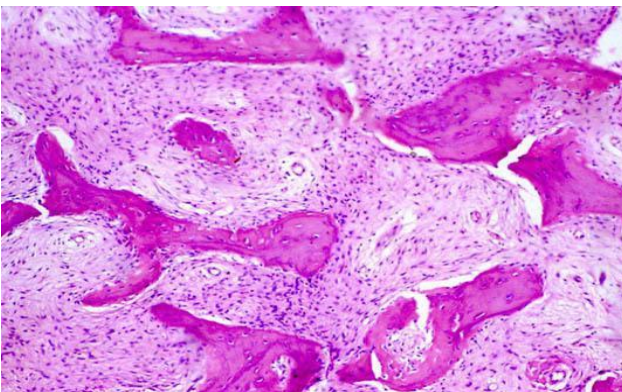


Figure 8 : Histopathological examination of the excised specimen.

DISCUSSION

Fibrous dysplasia is defined as “a benign lesion, presumably developmental in nature, characterised by the presence of fibrous connective tissue with a characteristic whorled pattern and containing trabeculae of immature nonlamellar bone”.^[14] Eversole defines fibrous dysplasia of craniofacial bones as “a benign, nonneoplastic intramedullary cellular proliferation of fibroblasts, with formation of irregular trabeculae of bone or ovoid calcifications that shows indistinct, nonencapsulated borders”.^[15]

Fibrous dysplasia is a hamartomatous condition or disorder of bone metabolism^[16] with an uncertain etiology and is probably a genetic predisposition.^[17] Fibrous dysplasia shows no sex predilection except for Mc Cune Albright syndrome which affects females almost exclusively and mean age of occurrence as reported by Zimmerman^[18] was 27 years in 69 patients. The polyostotic variant usually affects children younger than 10 years, whereas monostotic variant affects individuals in the second and third decade, as in the present case patient was 18 years old.^[19] Fibrous dysplasia affects maxilla twice as common than mandible, and is usually seen in the posterior region according to Abdulai et al.^[20] which was in concordance to our case where left posterior maxilla was involved.

Mirra supported a concept, which stated, “when a single bone (monostotic) is affected, it probably represents a forme fruste of the more severe form (polyostotic).” He stated that the craniofacial bones affected by fibrous dysplasia are in the following order that is frontal > sphenoid > ethmoid > maxilla > mandible > zygoma > parietal > occipital > temporal.^[21]

The clinical presentation of the fibrous dysplasia shows asymptomatic diffuse swelling in the affected region, causing aesthetic impairment and deformities. The similar findings were seen in our case which involved left maxilla. 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^[3], such clinical manifestations were not seen in our case.

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. The density and trabecular pattern of FD lesions is variable. 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, resembling the small fragments of a shattered windshield), a pattern resembling the surface of an orange (peau d’orange), a wispy arrangement (cotton wool), or an amorphous,

dense pattern. A distinctive characteristic is the organization of the abnormal trabeculae into a swirling pattern similar to a fingerprint.^[19] Prapayasatok *et al.*^[22] reported a case which was seen a rare radiographic 'sunray' appearance in 19-year-old woman. In this presented case, the panoramic radiography revealed a 'cotton wool appearance' and CT scan reveals "ground glass appearance" of the affected area which correlated the existing literature.

In most cases, the clinical and radiographic findings are sufficient to diagnose without a biopsy^[19] unless any sudden change in the clinical presentation or behavior of the lesion might warrant further investigation. Even in our case, there was no compelling indication to seek a biopsy as the radiographic and clinical findings were sufficient to diagnose the case.

Histologically, fibrous dysplasia is composed of fibrous tissue with randomly oriented bony trabeculae that are thought to be formed by osseous metaplasia of the fibrous stroma.^[23] The fibrous stroma is usually of low cellularity and contains variable amounts of myxoid material to dense collagenous matrix.^[24,25] The fibroblasts usually have plump, ovoid nuclei, although elongated, narrow nuclei may also be seen. A storiform pattern of the fibroblasts may also be identified.^[26] The osseous trabeculae are composed of immature woven bone and are typically not lined by osteoblasts, although focal osteoblastic rimming may be seen. Osteoclasts are frequently observed, especially on the concave side of the trabeculae. The outline of the trabeculae varies from solid, round islands to short, irregular, curvilinear or serpiginous shapes, giving the characteristic "Chinese character" or "alphabet soup" appearance.^[26] The ratio of fibrous tissue to bone ranges from fields that are totally fibrous to those filled with dysplastic trabeculae. Multiple, delicate capillaries are found throughout the lesion and, when injured, incite a giant-cell reactive process. Similar histopathological findings like fibrocellular connective tissue stroma with irregular areas of osseous tissue of varying shape and size with predominantly arranged osteocytes in lacunae. The focal areas exhibiting osteoblastic rimming. The intervening connective tissue shows haphazardly arranged fibroblasts and irregular trabeculae along with delicate collagen fibers and extravasated blood cells were seen in our case.

There are no accepted protocol for the management of fibrous dysplasia. The recommended treatment options are wait and watch, medical therapy and surgical therapy. Observation is the treatment modality for small asymptomatic lesions that are esthetically acceptable to the patient^[3], until puberty. Medical therapy does not play an important 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.^[3]

Surgical therapy remains the mainstay of treatment for large lesions which are esthetically unacceptable or is associated with any secondary changes, where surgical recountouring or excision of the lesion is done to prevent functional deficits and achieve normal facial esthetics.^[3] In the present case surgical recountouring of the lesion was done. 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.^[26,27]

CONCLUSION

Fibrous Dysplasia is a most common fibroosseous lesion occurring in younger individuals causing cosmetic and functional problems. It may manifests as monostotic or polyostotic form. Diagnosis of polyostotic form is easier due to extra-skeletal involvement. Monostotic form is common in the jaw. It is a tumor like developmental disorder with minimal chances of malignancies. Treatment of such lesion ranges from observation to medical therapy and then to the surgical therapy. Surgical therapy is usually undertaken after the growth phase has completed and the lesion warrants aesthetic or functional corrections. In the case presented here the surgical therapy was undertaken mainly because of aesthetic concern of the patient.

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