



## Maxillary Fibrous Dysplasia with review of literature

### Oral Pathology

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### ABSTRACT

Fibrous dysplasia (FD) is a benign fibro-osseous bone ailment. It is characterised by the conversion of normal bone by disproportionate proliferation of cellular fibrous connective tissue which is gradually restored by bone or osteoid. The clinical characteristics are pain, deformities and pathological fractures. It has three types, monostotic, polyostotic and craniofacial form. Polyostotic forms are associated with hyperpigmentation and endocrinological pathology. FD is usually observed in adolescents and young adults and comprises 7% of benign bone tumors. FD has a predilection for long bones plus craniofacial skeleton. The etiology of FD is not clear but predisposition of genetic etiology is suspected. The diagnosis of FD is based on radiological and histopathological examination. There are various treatment approaches including monitoring, surgical recontouring for esthetic and functional purposes. We are presenting a case of fibrous dysplasia of the maxilla on left side with clinical, radiological and histopathological features

### KEYWORDS:

Fibrous Dysplasia, monostotic, Polyostotic, craniofacial

### Case Report:

A 20 year old male patient reported with the complaint of swelling in his upper left back region of the jaw since last 6 years. He was apparently all right 6 years back, he first noticed slight asymmetry of left cheek region; slowly the swelling increased in size in next three years and was latent about 3 years. There is no history of trauma in the past. There is no relevant past dental, medical and family history and all vital signs were in normal limits. Patient appeared to be a well built nourished young man with no history of any systemic diseases.

On extra oral examination facial asymmetry detected on left side due to an obvious swelling seen over the body of zygoma; 1 cm below left infra-orbital margin up to the left corner of the mouth. Left side submandibular lymph node is palpable, nontender, mobile, and soft to firm. [Fig.1] Intra-orally, inspection findings showed smooth swelling was; extending from 23 to 28 teeth region; measuring about 1 x 2 cm in size; overlying mucosa appeared normal; causing the vestibular obliteration palpation, swelling is non tender; smooth; bony hard in consistency; fixed to underlying structure. There was no paresthesia over the distribution of infra-orbital nerve [Fig. 2].

In Radiographic investigations standard maxillary occlusal radiograph showed haziness and opacification over the left maxilla [ground glass appearance]; extending from 23 to 28 region; left lacrimal canal cannot be traceable [Fig.3]. In Panoramic radiograph, there was homogenous radiopacity with distinct margins blending with the adjacent normal bone, extending from 23 to 28 region, pushing the floor of maxillary sinus upwards on left side [Fig. 4]. Reformatted CT scan view showed hyperdense area covering entire maxillary sinus on left side [Fig.5] Biopsy specimen [Fig.6] revealed uniformly distributed curvilinear trabeculae in a fibrocellular stroma with numerous fibroblasts and a few blood vessels [Fig. 7].

Fibrous dysplasia and ossifying fibroma were considered as differential diagnosis. In the treatment plan surgical management was planned under general anesthesia. Surgical recontouring was performed and enucleation of the lesion was done.

The histopathological report revealed the lesional tissue, composed of a highly cellular fibrous stroma interspersed with numerous curvilinear delicate bony trabeculae of woven bone devoid of any osteoblastic

rimming. There are a few darkly stain reversal lines, however no resorptive bays or osteoclasts are evident. Lesion does not show any inflammatory cell infiltrate. In conjunction with the age of the patient and clinical and radiographic findings histopathologic diagnosis of fibrous dysplasia was made [fig. 6]. One year post operative follow up showed good prognosis with no facial asymmetry.

### Discussion:

Fibrous dysplasia (FD) of bone is a skeletal disorder characterized by widespread propagation of fibrous tissue in bone marrow. This may go ahead to osteolytic lesions, fractures and deformations. The disease is constituent for 2% of osseous tumors [1]. FD was first described by Von Recklinghausen in 1891 and coined the term 'Osteitis Fibrosa Generalisata' [2]. In 1938, Lichtenstein and Jaffe were first to commence the term "Fibrous dysplasia" [3]. McCune and Albright illustrated a syndrome "McCune-Albright syndrome" in 1937 [4].

FD is an uncommon disorder of unknown etiology. It represents a bone developmental disorder, specially a defect in osteoblastic differentiation and maturation [5]. Fibrous connective tissue containing abnormal bone replaces normal bone. FD is a localized abnormality, which can involve one bone (monostotic) or multiple bones (polyostotic). Most commonly the forms are distributed as follows: 74% monostotic, 13% polyostotic and 13% craniofacial [6]. Approximately 3% of the patients have the McCune-Albright syndrome, in which polyostotic FD is followed by skin lesions and endocrine pathologies. Monostotic FD, though less serious than polyostotic FD, is of greater concern to dentist because of the incidence in which the jaws are affected. The diagnosis of FD is often made in infancy and childhood. The maxilla or mandible may be involved but a predominance of the maxilla has been documented [1,5,6]. Males are less often affected than females. The deformity of the jaw results from a progressively slow growing painless swelling, but growth often slows or become arrested at a time coinciding with the onset of puberty [7]. Here, we report a case of fibrous dysplasia without any syndromic manifestations.

FD is a sporadic benign skeletal disorder that can involve one bone i.e. monostotic form and or multiple bones i.e. polyostotic form. Polyostotic form may be a part of the McCune-Albright syndrome (MAS) or of the Jaffe-Lichtenstein syndrome (JLS).

JLS is characterized by polyostotic FD and café-au-lait pigmented skin lesions, while MAS has the additional features of hyperfunctional endocrinopathies manifesting as precocious puberty, hyperthyroidism or acromegaly. Polyostotic FD with soft tissue myxomas is called Mazabraud syndrome [1]. Third form of FD is craniofacial form in which involvement of FD occurs in nearly 100% of polyostotic and 30% of monostotic forms [7]. The lesions of FD are twice as common in the maxilla as the mandible, and the posterior regions of the jaw are more frequently affected than the anterior, involvement of ethmoid, sphenoid, frontal and temporal bones are uncommon. These lesions cause expansion, thickening, and sclerosis of the involved bones with resultant facial asymmetry, visual complications, hearing disturbances, and tooth displacement depending upon the bone involvement. The onset of FD occurs during early life, usually in late childhood or early adolescence. It is common in 2nd decade of life. The polyostotic form usually is seen in children younger than 10 years, whereas monostotic form usually is found in slightly older age group [8] Ozek et al. [9] reported in the series of 16 patients with FD of the craniomaxillofacial bones, 1 patient was in his first decade, 11 patients were in their second decade, 3 patients were in their third decade whereas 1 patient in the series was in his fourth decade when the symptoms occurred. Our patient was in his third decade, hence our case supported to the case of Ozek et al. Polyostotic FD having slight female predilection while monostotic FD occurs equally in both males and females [10]. The lesion grows very slowly, causing expansion of the involved bone and giving a non-tender facial asymmetry of variable degree. Ganapathy N [11] reported a similar case of 23 year old male presented with a painless enlargement of maxilla.

Further he stated that premolar-molar area is the most common site and also in polyostotic FD the frequency of occurrence female to male is 3:1 ratio.

As a rule of thumb, the lesion usually stops growing when skeletal growth ceases, but cases of continual enlargement have been reported. Laboratory assay of serum calcium and phosphate levels of affected patients are normal, but alkaline phosphatase level may be slightly elevated. On the contrary, lack of an elevation should not rule out fibrous dysplasia [12].

FD is a developmental dysplastic disorder of bone in which the normal bone matrix is replaced by fibroblastic proliferation. Lesions characterized by irregular trabeculae of partially calcified osteoid [12]. The etiology of fibrous dysplasia has been linked with a mutation in the GNAS1 gene that encodes the alpha subunit of the stimulatory G protein-coupled receptor, G $\alpha$ s, and is located at chromosome 20q13.2–13.3 [13]. As per the current belief FD results from a defect in bone maturation that begins in the embryo. The classical division of FD into monostotic, polyostotic and craniofacial forms may reflect the timing of the mutation and by this means, the initial size of the mass of FD precursor cells. The polyostotic form may arise in fetal life while the monostotic form may arise postnatal. This correlates with the confirmation that the monostotic form is not a precursor of the polyostotic form. Thus FD may reveal a programmed field effect of abnormal osseous development in congenitally predisposed bone matrix. This may justify the fusiform expansion of affected bone [1,14].

Radiographic features of FD vary depending on the amount of bony and fibrous matrix within the lesion and have been sub-divided into three different patterns: pagetoid type 56%, sclerotic type 23% and the radiolucent type 21%. Our patient had sclerotic type of lesions; which is the commonest form of involvement seen in facial bones and bones of the base of skull. The lytic and pagetoid types usually occupy the calvarial bones [15]. Early FD of craniofacial bones is radiolucent with either ill defined or well defined borders, and may be unilocular or multilocular.

As the lesions mature, the bony defects acquire a mixed radiolucent/radiopaque appearance, and long-established FD exhibits mottled radiopaque patterns often described as ground glass appearance, orange peel appearance or fingerprints, with ill defined borders blending into the normal adjacent bone [16].

Prapayastok et al. [17] reported a case of 19 year old woman which was seen a rare radiographic 'sunray' appearance. In this presented case, the standard maxillary occlusal radiograph revealed a 'groundglass' appearance of the affected area.

Differential diagnosis of FD includes ossifying fibroma, CGCG, paget's disease, osteomyelitis, osteosarcoma. Recurrence is rare in adults, but the lesions can show surprising growth potential if they are surgically altered during their active growth phase [8]. The conservative surgery and unsuccessful removal of the lesion is one of the causes of increased risk of recurrence. Patients with craniofacial FD have the risk of recurrence ranged from 15 to 20% [18]. Concentration of serum alkaline phosphatase (ALP) may be important marker for detection of the recurrence of the lesion. The patients who had FD, have higher ALP, this may be a reliable marker for estimating tumor progress and a sudden rise in ALP was correlated with the reoccurrence of FD by Park et al. [19] Even if the prognosis of FD is usually very good, malignant transformation might occur rarely (1% average, 4% in McCune-Albright), with almost all cases being of sarcomatous origin [20]. Previous radiation and spontaneous degeneration may be the reason of malignant transformation. Malignant changes with fibrous dysplasia include Osteosarcoma, Fibrosarcoma, Chondrosarcoma, Malignant fibrous histiocytoma & Ewings sarcoma [21].

The surgical treatment of FD ranges from biopsy specimens to modeling osteotomies, bi-maxillary osteotomies and calvarial remodeling. The aim of the surgical therapy for FD is to prevent pathological fractures, control the pain and to reduce bone deformities [20]. Surgical recontouring was performed in our case.

### Conclusion:

The monostotic FD has an asymptomatic course, with a slow growth and stabilization after puberty, and only needs follow up. The polyostotic type has a progressive behaviour and associates with complications and recurrence. FD is a tumor like developmental disorder with minimal chances of malignancies. Delay in diagnosis is believed to be the most likely reason for the malignant transformation in Polyostotic FD

Figure 1: Extraoral picture showing diffuse swelling seen 1 cm below left infra-orbital margin up to the left corner of the mouth.

Figure 2: Intraoral picture showing diffuse swelling extending from 23 to 28. vertically obliterated buccal vestibule.

Figure 3: Standard maxillary occlusal radiograph showing "ground glass appearance".

Figure 4: OPG showing uniform radiopacity seen with 23,24,25,26,27 with distinct margins.

Figure 5: Reformatted CT Scan view showing hyperdense areas covering entire maxillary sinus on left side.

Figure 6: Gross picture showing multiple bits of hard bony mass.

Figure 7: H&E Section showing curvilinear bony trabeculae in fibrous connective tissue

Figure 1



Figure 2



Figure 3

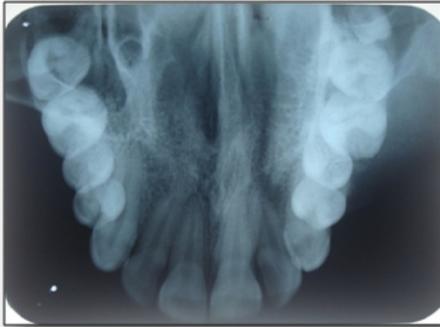


Figure 4



Figure 5

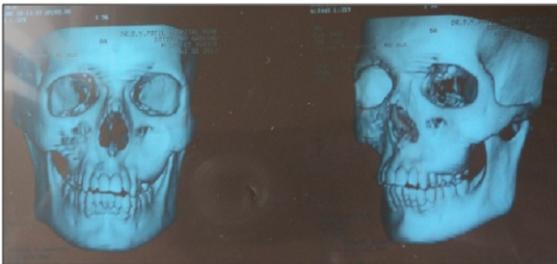
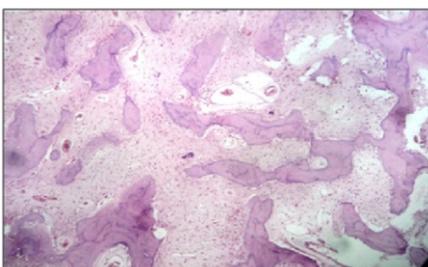


Figure 6



Figure 7



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