MONOSTOTIC FIBROUS DYSPLASIA OF MAXILLA: A RARE CASE REPORT AND REVIEW OF LITERATURE

Nazish Akhtar [1] , Upender Malik [2] , Vinay Badyal [3] , Nidhi Chitranshi Khare [4]
Senior Lecturer [1,4] , Reader [2] , Post Graduate Student [3]
1, 3- Department of Oral Medicine and Radiology,Seema Dental College and Hospital, Rishikesh, India.
2, 4- Reader, Department of Oral Medicine and Radiology.Teerthankar Mahaveer Dental College & Research Center, Moradabad

Abstract
Fibrous dysplasia is a condition characterized by excessive proliferation of bone forming mesenchymal cells. It can affect one bone (monostotic type) or multiple bones (polyostotic type). It is usually observed in adolescents and young adults and comprises 7% of benign bone tumors. The etiology is not clear but genetic predisposition is suspected. It has a predilection for long bones as well as the craniofacial skeleton. The maxilla is the most commonly affected facial bone, with facial asymmetry being the usual complaint. The diagnosis is based on radiological and histopathological examination. There are different treatment approaches including monitoring, medical treatment or surgery. Once diagnosed, routine follow-up should be done on a yearly basis with x-ray examination. Here we report a case of fibrous dysplasia in a 15 year old male patient on the left side of the face.
Keywords: Fibrous dysplasia, Monostotic, Polyostotic, Fibrous-osseous, Proptosis.

Introduction

Fibro osseous lesion is a commonly used term that includes bone dysplasias, as well as neoplasms and other lesions of bone. In 1891, Fibrous Dysplasia (FD) bone was first described by Von Recklinghausen.[1] Lichtenstein first coined the term fibrous dysplasia in 1938.[2]

Fibrous dysplasia (FD) is a hamartomatous condition in which normal bone and marrow are replaced irregularly arranged and unevenly distributed fibrous tissue and woven bone.

As per the bony involvement, if one bone (monostotic fibrous dysplasia; MFD), multiple bones (polyostotic fibrous dysplasia; PFD) or they may have McCune-Albright syndrome (MAS), which has been classically defined by the triad of polyostotic fibrous dysplasia(PFD), café-au-lait skin macules and endocrinopathies, including among others, precocious puberty.[3]

The condition represents 2.5% of all bone lesions 7% of all benign bone tumors.[4] The disease is with an unknown aetiology, and occurs with an incidence rate of 1:4000-1:10000.[5]

Genetic mutation in the gene that encodes the subunit of a stimulatory G protein (Gsα) located on chromosome20 is thought to be a possible etiology of this condition which is usually an incidental investigatory finding.[6,7]

Histologic features of the disease corresponds to fibrosis with varying degree of simultaneous resorption and repair[5] and the diagnosis is usually be made using radiologic features and histopathological examination.[8]

It has been described by various names in the literature as intraosseous calcifying fibroma, osteomatous cyst, osteitis fibrosa cystica and ossifying fibroma.[9]

Conservative treatment, such as shaving and debridement of the lesion, is often the only thing required if lesions are not severely disfiguring.[8]

In this paper we present a case of 15 year old male patient having fibrous dysplasia on the left side of the maxilla.

Case Report

A 15 year old male reported to the department of Oral Medicine and Radiology with chief complaint of painless swelling on the left side of maxilla since 1 year. The patient noticed the swelling 5 years ago, after trauma, due to hit by a cricket ball on that same area after which it was gradual in onset, initially small in size and later progressed to attain the present size. Medical history and family history of patient was insignificant. No evidence of pigmentations was present on the body.

On extraoral examination, facial asymmetry was evident due to slight fullness in the left malar region. On inspection diffuse swelling was present over the left maxillary region extending from ala of the nose and upto to in front of the left ear. Superiorly from the infraorbital margin upto the left labial commissure. On palpation, swelling was bony hard and non_tender in nature (Figure1).

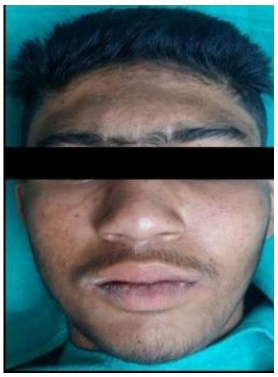

Figure1 : Extra oral photograph

Intraorally, diffuse, roughly elliptical swelling was present measuring 4×2 cm, extending anterio-posteriorly from distal surface of canine upto the distal surface of 27. Obliteration of buccal vestibule was present i.r.t 24, 25, 26, 27 region. On palpation, swelling was bony hard, non-tender with no palpable pulsation. The overlying mucosa appeared normal (Figure 2).

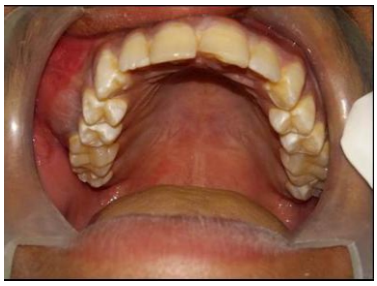

Figure 2: Intra Oral Photograph

Based on clinical finding, provisional diagnosis of fibrous dysplasia of left maxilla was given. Patient was advised investigations as Intra Oral Periapical Radiographs (IOPA), Orthopantomogram (OPG), Para nasal sinus (PNS) view and occlusal radiographs.
Intra Oral Periapical Radiographs showed increased radiopacity with loss of lamina dura in relation to distal surface of 23 upto the distal surface of 27 (Figure 3).

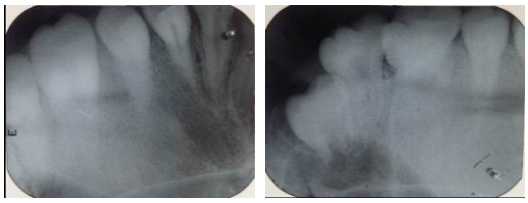

Figure 3 : IOPA revealed loss of lamina dura with increased radiopacity

Orthopantomogram showed gross radiopacity with typical ground glass appearance in the left maxillary bony region extending from canine region upto the 2nd molar region (Figure 4). Para nasal sinus view showed increased radiopacity in left maxillary region with no extension into the maxillary sinus (Figure 5).Occlusal radiograph showed remarkable expansion of left maxillary buccal cortical plate (Figure 6). Blood and biochemical investigations showed alkaline phosphate, serum calcium and serum alkaline phosphorous within normal range.

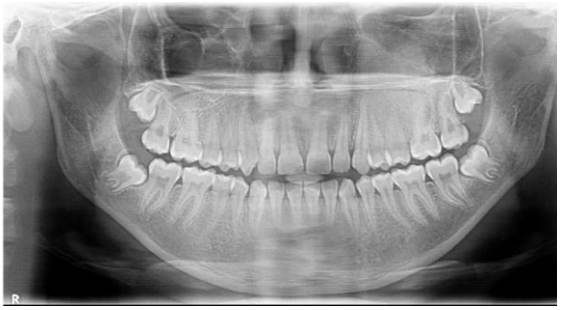

Figure 4: OPG showing the ground glass appearance in the left maxillary region

An incisional biopsy was performed from the left maxillary premolar region by the vestibular approach (Figure 7)

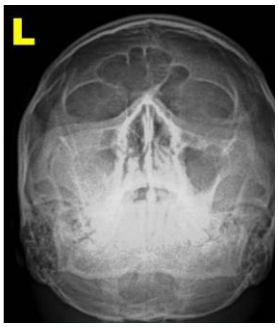

Figure 5: PNS view showing the increased radiopacity in the left maxillary region without extension into the left maxillary sinus

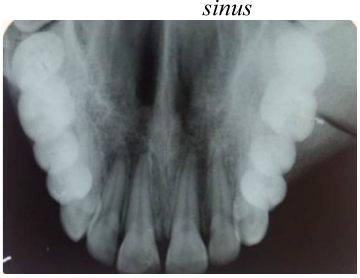

Figure 6: Occlusal radiograph showing the expansion of the left buccal cortical plate

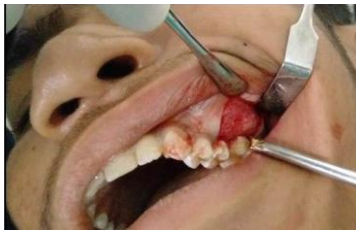

Figure 7: Incisional biopsy was performed taking the vestibular approach

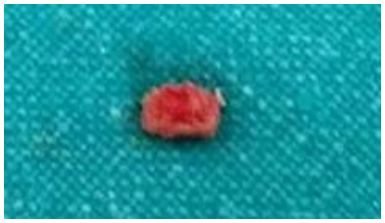

Figure 8: Specimen sent for histopathological

and the specimen was taken (Figure 8) and sent for histopathological examination, that showed presence of trabeculae of demineralized bone arranged in a Chinese letter pattern. Bone trabeculae were linned by osteoblast and showed the presence of resting lines, the marrow spaces in between were hypocellular with few chronic inflammatory cells in a fibrous connective tissue stroma(Figure 9).
Based on clinical, radiological and histopathological investigations- a final diagnosis of Monostotic fibrous dysplasia was given involving the left maxilla.Surgical recontouring was done under LA in the department of Oral and Maxillofacial Surgery, the patient is still under follow up with no recurrence.

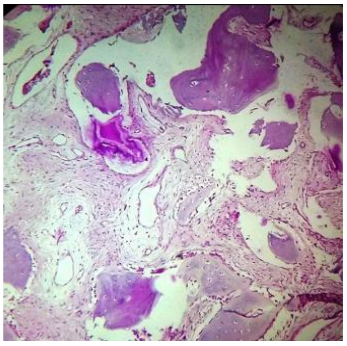

Figure 9: Histopathological picture revealed the Chinese letter pattern in fibrous connective tissue stroma

Discussion

Fibrous dysplasia is a slowly progressive, expansile, benign bony disorder of unknown etiology in which the normal bone is replaced by an abnormal fibrous-osseous tissue.[9] It is a hamartomatous condition or disorder of bone metabolism.[10]

Fibrous dysplasia although usually diagnosed in first three decades of life with gender equality, they progress gradually and halt after attaining maturity and may sometimes regress in few cases [5] Common bones affected include femur, tibia, fibula, ribs and facial bones with involvement of maxilla almost twice as often as the mandible, frequenting the posterior region with unilateral presentation.[2]

Although of uncertain etiology, sporadic, postzygotic mutation of GNAS1 gene found on chromosome 20q13 is responsible for the formation of the alpha subunit of stimulating G-proteins. The mutation leads to activation adenylate cyclase and increases intracellular concentrations of cAMP resulting in abnormal osteoblast differentiation and production of dysplastic bone. It also stimulates release of several cytokines (mainly interleukin-6) leading to normal osteoclasts congregation and increased bone resorption.[1]

Fibrous dysplasia is classified according to the number of affected bones along with presence or absence of extra-skeletal abnormalities. The monostotic form affects only one bone and constitutes 70-80% of the cases. The polyostotic form, involves several bones and can be divided into three subtypes: craniofacial, in which only the craniofacial complex are involved including the mandible and the maxilla; Lichtensteinjaffe, in which in addition to the polyostotic involvement there are cafe au lait pigmentations; Albright's Syndrome, characterized by the affection of several bones, cafe au lait in the skin and endocrine abnormalities with precocious puberty and development of secondary sexual characters in girls before the age of three. The polyostotic form is present in 20-30% of the cases.[11]

Although asymptomatic, the signs and symptoms of the condition depends on the location of the lesion(s) and the effect on the adjacent structure as the tumor progresses.[11].The lesion progresses in a gradual, painless manner leading to enlargement of the involved bone or bones causing facial asymmetry. Other symptoms in craniofacial type are due to constriction of cranial foramina or obliteration of bony cavities and include anosmia, orbital dystopia, diplopia, proptosis, blindness, epiphora, strabismus, facial paralysis, hearing loss, tinnitus, etc.[4]

The association of fibrous dysplasia and soft tissue tumors also is described in literature and has been termed Mazabraud's syndrome. Many endocrinal abnormalities associated with the condition includes hyperthyroidism, Cushing's disease, precautious puberty, gigantism, thyromegaly, hypophosphatemia, hyperprolactinemia and neurofibromatosis type-II.[11]

There is considerable variation in the microscopic appearance but the histopathological features are similar in all three types of fibrous dysplasia and are viewed as benign fibroblastic tissue, arranged in a loose, whorledpattern interspersed with spicules of woven bone with typicalosteoblastic rimming embedded in fibrous tissue [4] characteristically resembling C-shaped or Chinese characters shaped trabaculae, (described as "Chinese letters" or "alphabet soup"), consisting of coarse woven bone instead of well-organized lamellar bone.[5]

Radiographically, fibrous dysplasia vary depending on the stage of development and quantity of bony matrix within the lesion. The lesion is more radiolucent and well-defined initially which gradually changes to a mottled, ill-defined radiopacity in the later stages. The radiological appearance corresponds to a thin bony cortex with well-defined borders and ground glass appearance. Three distinct patterns have been described by Panda *et al.,* The pagetoid appearance on Computed Tomography imaging is characterized by bone expansion and scattered islands of bone formation in a low-attenuation field. The sclerotic type has a homogeneous appearance with a ground-glass appearance. The cystic type appears as a well-defined low-attenuation lesion with a sclerotic margin.[4]

Panoramic radiographs showed involvement and extension of the jaws well [12] Computed tomography (CT) is a better radiological tool, especially for assessing the extent of the tumor in cases of suspected optic canal involvement. Fibrous dysplasia has characteristic appearances on Computed tomography and consists of three varieties: ground-glasspattern, homogeneously dense pattern and cystic variety [13]

The Magnetic Resonance Imaging characteristics of fibrous dysplasia are variable, typically showing signalintensity that is intermediate to low on T1-weighted images, intermediate to high on T2-weighted images, and heterogeneous enhancement after administration of gadolinium [13] Bone scan uses radioactive tracers (usually Tc-99), which are injected into the bloodstream. The damaged parts of bones (hyperactive) take up more of the tracers, which are seen as hot spots. Biochemical tests are used to detect Bone Turnover Markers (BTMs) like Serum Alkaline phosphatase, monitoring the levels of various hormones if indicated.[5]

Treatment of bony lesions includes surgical and nonsurgical therapies. [10] There are no uniformly accepted guidelines for treatment of this disease, but the three general approaches involve monitoring, medical management, or surgery.[9]

Certain cases may be monitored for disease activity and progression. Depending on the location of the lesions, age of the patient, and the patients views towards surgery, serial clinical examinations may be the best option.[9] Only monitoring the condition with periodic X-rays is required. Patientsshould be warned for any sign of vision or hearingdisturbances and any disturbing obstructive symptoms
in nasal airway. Orthodontic support and braces may be needed to manage mal-occlusion or misaligned tooth.[5]

Biphosphonates are used in cases where surgery cannot be performed.[6] Bisphosphonates inhibits the activity of osteoclasts, cells that dissolve bone.[5]

Surgical approaches are aimed towards stable occlusion, facial aesthetics and evasion of post-operative relapse.[6] In children, the surgical procedure should be delayed,till the lesion tends to become static.[9] Surgical treatment in young-aged minor cases and biopsy with minor bony osteoplasty at affected site are adequate. In more severe cases complete excision with graft reconstruction can be done.[4] Conservative surgery involves a remodelling procedure. This can however result in recurrence, especially during the growth period.[4]

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.[11,14,15] Usually the prognosis is good in monoostotic involvement but becomes poorer with polyostotic forms of the disorder.

Conclusion

The fibrous dysplasia is significant for the dentists as the dentist may be the first one to diagnose the condition and make the patient aware of it. Moreover, in craniofacial type of the disease inolvment of jaws are a most prominent feature and the patient might confuse it with a dental ailment.

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Corresponding Author
Dr.Upender Malik
Reader
Department of Oral Medicine & Radiology
TMDCRC, Moradabad
Email: upender_malik@yahoo.co.in

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