A CASE REPORT SERIES ON FIBROUS DYSPLASIA

Dr. Bhavika Vhatkar
Associate Professor, Department of Oral Medicine and Radiology D.Y Patil School of Dentistry, Navi Mumbai
bhavika.vhatkar@dypatil.edu

Dr. Sandeep Pagare
Professor, Department of Oral Medicine and Radiology D.Y Patil School of Dentistry, Navi Mumbai
sandeep.pagare@dypatil.edu

Dr. Naveen Kumar Shetty
Professor, Department of Oral Medicine and Radiology D.Y Patil School of Dentistry, Navi Mumbai
nksrad@rediffmail.com

Dr. Abhishek Mukherjee
MDS III year, Department of Oral Medicine and Radiology D.Y Patil School of Dentistry, Navi Mumbai
Eabhishekmukherjee20@gmail.com

Dr. Janice Pinto
MDS III year, Department of Oral Medicine and Radiology D.Y Patil School of Dentistry, Navi Mumbai
janicepinto3214@gmail.com

Dr. Stutee Beriwal
MDS II year, Department of Oral Medicine and Radiology D.Y Patil School of Dentistry, Navi Mumbai
stuteeberiwal@gmail.com

ABSTRACT
Fibrous dysplasia is a benign lesion, congenital, recurrent character and etiology unknown, caused by a bone modeling disorder with constant alterations of the normal bone being replaced by immature fibrous tissue. It is classified according to the number of affected bones, and its association to endocrine alterations, that is, monostotic (80–85%), polyostotic forms (20–30%), and Albright's disease. They may involve one or more bones being the maxilla the facial bone more affected. Fibro-osseous lesions diverse group process which includes developmental, dysplastic, neoplastic. We report a series of three cases of polyostotic fibrous dysplasia in a male patient two in maxilla and one in mandibular complaining of increased volume on the right side of the face and lower border of the mandible. In this case, Computed Tomography (CT) and Orthopantomograph (opg) was the main radiographic examination to demonstrate the extension and radiodensity that the lesion assumes in the craniofacial bones, being therefore of fundamental importance in the surgical planning and in the longitudinal follow-up of the operated patient. The diagnosis was confirmed later by anatopathology and the treatment of choice was osteoplasty. This article
also aims to review the main clinical, radiological & differential diagnosis Fibrous dysplasia treatment.

Keywords: Polyostotic maxillary fibrous dysplasia, Fibrous lesions.

INTRODUCTION
Fibrous dysplasia (FD) is an uncommon non-hereditary, developmental anomaly of the bone that occurs due to defect in osteoblastic differentiation and maturation. Lichtenstein originally described it in 1938 and by Lichtenstein and Jaffe in 1942.
In 1891, Von Recklinghausen denominated de "generalist fibrous osteitis" pathological conditions that characterized deformities and bone alterations. Clinically it is classified as monostotic or polyostotic, the first being focal, limited to a single bone and the second, multifocal, involving several bones simultaneously.
As polyostotic FD is a very rare condition occurring only in 20–25% of patients, therefore the aim of this article is to report such a rare series of case of polyostotic FD in three young patients and add it to the literature.

Case Report

A 24-year-old male patient reported to Oral Medicine Department with the chief complaint of diffused swelling on the left side of the face since 6 month. Initially the swelling was small and gradually increased till present size. No medical history was associated. On clinical examination, unilateral extra oral diffuse swelling of approximately 3 cm × 4 cm in dimension was seen in (Figure 1). The swelling was seen increasing in size, which extended from 1cm corner of the mouth and crossing the inferior border of mandible. Anterior posterior swelling was crossing the midline of the right side and posteriorly extending to the right body of mandible. Teeth surrounding the lesion were vital. Provisional diagnosis of dentigerous cyst, Ossifying fibroma & Ameloblastoma was made.
Intraorally swelling is seen in left mandibular molar region approximately 1×1cm extending from distal aspect of 35 till distal aspect of 36 with expansion of buccal and lingual cortical plates as seen in [Figure 2].

Based on clinical findings, provisional diagnosis of fibro-osseous lesion was made. Differential diagnosis such as Ossifying fibroma, Paget's disease, and focal cementosseous dysplasia were considered for bony hard swelling. Furthermore, the patient was advised investigations such as complete hemogram, serological investigations such as serum calcium and phosphorous, alkaline phosphatase level, orthopantomogram (OPG), CT scan, and incisional biopsy. Complete hemogram showed normal parameters and serological investigations were within normal limits.

OPG showed expansion of bone and thinning of outer cortex. Abnormal trabecular pattern with irregular shape, displacement of teeth, mixed radiolucent and radiopaque pattern, and ground glass to orange peel appearance and cystoid appearance was also seen [Figure 3].
CT scan revealed expansion of mandible and bulging out of lesions. Mandibular lesions were seen encroaching the tooth roots and maxillary lesions were seen to be occupying maxillary sinus. Generalized calvarial and facial bone thickening was noted [Figure 4].

A 23-year-old male patient reported to the Department of Oral Medicine and Radiology with the chief complaint of swelling seen on left side of the face since last 4yrs extra orally it was noticed initially it was small and then gradually increased to present size approx. 1 x 2cm. Superior inferiorly swelling is extending 2cm above to inferior border of mandible and inferiorly submandibular and submental region. On palpation, the consistency was bony hard and nontender and there was no local rise of temperature [Figure 1]. Overlying mucosa appeared normal, firm, and was no tender. Based on clinical finding provisional diagnosis of central giant cell granuloma, ossifying fibroma was made.
Intraorally swelling was seen from left mandibular alveolar bone from 33 to 36 region approximately 2× 4cm with expansion of buccal and lingual cortical plates and obliteration of the buccal vestibule.[Figure2].

Opg shows diffuse radio opaque region in 31 to 36 area .Large radiopaque region seen in left parasymphaseal region. Radiopacity seen in 36 Change in bony architecture. Lamina dura is indistinct.

Ct scan for coronal section shows large mixed lesion crossing the midline showing buccal and lingual cortical expansion. Diffuse radiopacity seen in most of the lesion along with large radiolucency space in left molar ramus of mandible. There is expansion of buccal and lingua cortical plates and change in bony architecture. Whereas in Axial section shows diffuse radiopacity crossing the midline extending from left molar ramus to right parasymphysseal region .Expansion of buccal and lingual cortical plates is seen in this section.

8yr old male patient reported to Oral medicine and radiology department with a chief complaint of Swelling seen in right upper side on the face since last 6months. Extraorally it was noticed a diffused swelling seen on right zygomatic region approx 4 x 5cm Extending from infra orbital region to tragus area. Swelling is not crossing the midline.On palpation, the consistency was bony hard and non tender and there was no local rise of temperature
[Figure 1]. Overlying mucosa appeared normal, firm, and was no tender. Based on clinical finding provisional diagnosis of Aneurysmal bone cyst & pagets disease, ossifying fibroma was made.

Intraorally swelling is seen in right maxillary canine region of approximately 3×4cm of buccal and lingual plate’s expansion seen.

Ct scan of coronal section shows dense radiopacity of right maxilla region extending superiorly and obliterating maxillary sinus and involving alveolar part of maxilla crossing the midline. A large lobulated mass measuring approx. 6.3cm×5.4cm with internal body and soft tissues densities is noted expanding maxillary antrum.

DISCUSSION
FD is commonly a benign lesion in which irregularly distributed spicules of bone lie in cellular fibrous stromain pathologic condition of the bone which fibrous tissues. The lesion is believed to be hamartomatous developmental abnormality of bone gradually expand and replace the normal bone. The disease usually begins in childhood and progresses throughout puberty and adolescence, and then becomes dormant in early adult life. About 75% of the cases are found under the age of 30 years. Malignant transformation is rare0.05% and is usually seen only in polyostotic cases. Following the ribs and long bones,
craniofacial bones are the second most common site of involvement and comprise 25% of the cases. Fibrous dysplasia of the para nasal sinuses is very rare. Most commonly, fibrous dysplasia is asymptomatic until there is encroachment upon adjacent vital structures. Facial asymmetry is the most common sign of fibrous dysplasia in the

Head and neck followed by pain, ocular proptosis and neurological changes. The lesions of fibrous dysplasia are twice as common in maxilla as compared to mandible, and the posterior aspects of the jaw are more frequently affected than the anterior. The craniofacial form of fibrous dysplasia can be diffuse and may involve multiple bones. When the anatomic spaces and foramina are constricted because of encroachment of the lesions, the patient may experience a variety of symptoms, including headaches, loss of vision, proptosis, diplopia, loss of hearing, anosmianasal obstruction, epistaxis, epiphora and symptoms mimicking sinusitis.

The series of three cases describes here with first patient with the swelling present on left side of maxilla on intra oral examination, swelling is seen in left mandibular molar region extending from distal aspect of 35 till distal aspect of 36 with expansion of buccal and lingual cortical plates as seen. In differential diagnosis, periapical cyst was considered because of carious primary first molar, Ossifying fibroma & Fibrous dysplasia was considered. In second case swelling in the patient seen on left side of the face since which was seen noticed during 4th year intraoral examination swelling was seen from left mandibular alveolar bone from 33 to 36 region with expansion of buccal and lingual cortical plates and obliteration of the buccal vestibule, Whereas in third pedo patient swelling was seen on right upper side of maxilla since, Intraorally swelling is seen in right maxillary canine region of buccal and lingual plate’s expansion seen.

All the above listed differential diagnosis was eliminated once the ground glass appearance of the bone was noticed on the radiograph. A neurysmal bone cyst was included in third patient where lesion was seen in maxilla rest two lesions was in mandible.

The radiographic appearance revealed increased radiopacity of bone with “Ground glass” appearance in the region of right maxilla& left mandibular. CT scan revealed sclerosis with bony expansion with few cystic changes. Also, it revealed typical “Ground glass” appearance involving multiplebones of the cranium. The trabeculae pattern appeared thin and irregular in size. After the final diagnosis patient was referred to department of Oral Maxillofacial surgery. The surgeon’s were of the opinion that they would do the surgery, only after completion of patient’s growth spurt in 8year old male patient.
Therefore, patient was advised periodic recall visits at regular intervals to record the bony changes and involvement of other vital structures.

Surgical treatment of fibrous dysplasia consists of either conservative shaving/contouring or radical excision with immediate reconstruction. The choice of surgical option depends on several factors like site of involvement, rate of growth, aesthetic disturbance, functional disruption, and patient preference, general health of the patient, surgeon’s experience and the availability of a Multi-disciplinary team (neurosurgeon, ophthalmologist, orthodontist and endocrinologist). The multi-disciplinary approach becomes even more important in polyostotic fibrous dysplasia.

CONCLUSION
The mere presence of FD of the craniofacial region is not in itself an indication for treatment. Small solitary lesions may remain asymptomatic for long periods. Thus for this present case report, treatment was deferred and is under regular follow up.

Declaration of patient consent
The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

REFERENCES
1. Marcelo Domingues Pinto1, Gustavo Gaffrée Braz2, Roberto Gomes Santos3, Diego Polido4, Marcelo Domingues Pinto1, Gustavo Gaffrée Braz2, Roberto Gomes Santos3, Diego Polido4, Jonathan Rodopiano Paixão1, Hugo Solé de Melo1 and Roberto Almeida Elias5Fibrous Dysplasia in Maxillary Bone: Case ReportDOI: 10.23937/2469-5734/15100
