

Case Report**Fibrous Dysplasia of The Jaw Bones: Clinical, Radiographical and Histopathological Features. Report of Two Cases.****Dr. Jyothi Mahadesh¹, Dr. Charan Gowda², Dr. Laxmi Devi³, Dr. Kokila G⁴.**¹Prof. and Head, ²Reader, ³Senior Lecturer, ⁴Professor Department of Oral Pathology, Sri Siddhartha Dental College, Tumkur, Karnataka.**Abstract:**

Fibrous dysplasia is a developmental benign bone lesion characterised by the replacement of normal bone by excessive proliferation of cellular fibrous connective tissue which is slowly replaced by bone, osteoid or cementum like material. It causes bone pain, deformities and pathological fractures. It may either be monostotic or polyostotic. Here we present two cases of fibrous dysplasia affecting mandible and maxilla(involving the sinus) respectively.

Key words:- Fibrous Dysplasia, Monostotic, Polyostotic, Craniofacial

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Introduction

FD is a disturbance of bone metabolism that is classified as a benign fibro-osseous lesion. The fibrous connective tissue containing abnormal bone replaces normal bone⁽¹⁾.

Fibrous dysplasia (FD) is a sporadic benign skeletal disorder that can affect one bone (monostotic form) or multiple bones (polyostotic form). Gender prevalence of FD is equal. 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. Signs and symptoms of FD include bone pain, pathological fractures and bone deformities. Laboratory findings include elevated serum alkaline phosphatase rarely, but calcium, parathyroid hormone, 25 hydroxy vitamin D, 1,25-dihydroxyvitamin D levels in most cases are normal. Malignant transformation is rare and is usually precipitated by radiation therapy⁽²⁾.

Etiology is a post zygomatic , somatic mutation of GNAS gene which encodes the α Subunit of the ubiquitous stimulatory G protein⁽³⁾.

Case report 1.

A 20 year old male patient reported to the department of maxillofacial surgery at Sri Siddhartha Dental College with the chief complaint of swelling in the lower back teeth region. The clinical examination revealed a diffuse bony hard swelling with no tenderness in lower left 1/3rd of the face measuring approximately 2X3cm, extending antero-posteriorly from angle of mandible to the para-symphysis region, superio-inferiorly from 2cm below ala tragal line to 0.5cm medially to the lower border of mandible.

CT scan revealed a mixed radiolucent and radiopaque lesion in left mandible from canine to third molar with diffused borders(fig1). An incisional biopsy was made and the tissue was sent to the department of oral and maxillofacial pathology for histopathological examination.

After decalcification the histopathology revealed. A highly cellular connective tissue stroma resembling fibroblasts that are spindle shaped, plump and benign along with some osteoid like tissue with osteocytes but mostly not lined by osteoblasts in various shapes. Along with this extravasated RBCs also seen(fig2). A diagnosis of fibro-osseous lesion was made. Since cemento-osseous fibroma and other fibro-osseous lesions have similar histopathology, after correlating with the clinical features and radiographical features a diagnosis of Fibrous Dysplasia was made.

The lesion was surgically excised as there was erosion of lingual cortical plate and the specimen was submitted for histopathological examination. The histopathology confirmed the diagnosis of fibrous dysplasia. The case was negative for both the syndromes.

The follow-up is being done since one year and so far no recurrence is seen. patient is healthy and fit

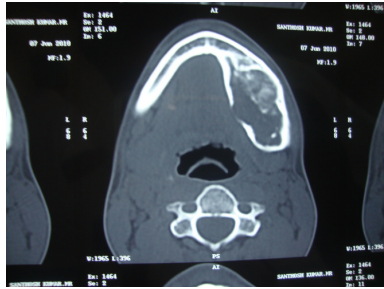


Fig 1:- showing mixed radiolucent and radiopacities with lingual cortical Erosion.

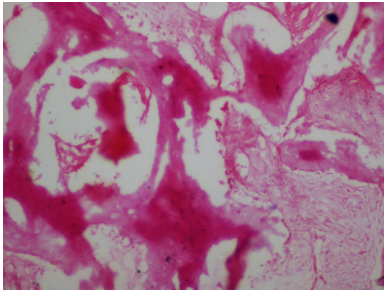


Fig-2 showing bony trabeculae and hypercellular fibrous Connective tissue stroma.

Case report 2.

A 24 year old male patient reported with the chief complaint of swelling in the right side of the face since 2 years. Clinical examination revealed a facial asymmetry on right mid face region in the maxilla measuring 5X3 cm extending supero-inferiorly from right infra orbital margin till upper alveolar region and antero-posteriorly from right canine fossa to the right zygomatic arch. The swelling was bony, firm and well defined. The skin over the swelling was normal. CT scan revealed an expansile bony lesion with ground

glass appearance in right maxillary sinus including the alveolar process and zygomatic process (fig3). The clinical impression was Fibro-osseous lesion. An incisional biopsy was made from a representative area and was sent for histopathological examination to the department of oral and maxillofacial pathology.

After decalcification, section revealed a connective stroma which was cellular, fibrous with plump cells. It was intermixed with bone of varying sizes and shapes. Under low magnification Chinese letter pattern could be appreciated (fig4). Features were suggestive of Fibro-Osseous lesion. After clinical features and radiological features were correlated it was diagnosed as Fibrous Dysplasia. Surgical recontouring was done for the patient.

The follow-up is being done regularly since 6 months and so far no recurrence is seen. Patient is healthy and fit.

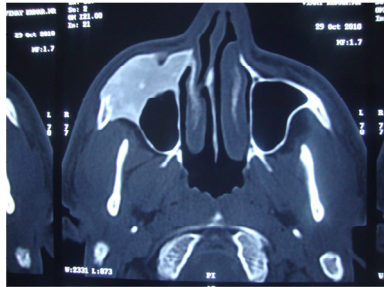


Fig 3:- showing ground glass appearance.

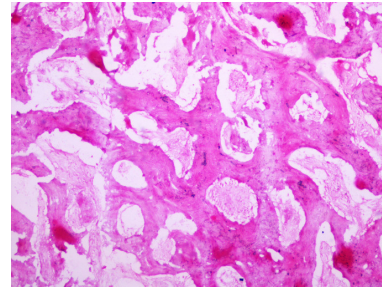


Fig 4:- showing delicate bony trabeculae and cellular connective tissue.

Table 1. Summary of cases of Fibrous Dysplasia in Sri Siddhartha Dental College And Hospital Tumkur.

Case	Sex	Age (years)	Site	Radiographic appearance
1	Male	20	Left mandible	Mixed radiolucent/radio opaque appearance
2	Male	24	Right Maxillary sinus + zygomatic processes + alveolar	Ground glass appearance

Discussion:-

Fibrous dysplasia is defined as a benign osseous disease characterised by a process of normal bone resorption, followed by an abnormal proliferation of a disorganised fibro-osseous tissue. It represents about 7% of all benign osseous tumors and may affect any bone ⁽⁴⁾.

The etiology of F.D is a genetic non-inherited condition caused by mutation in the gene GNAS on chromosome 20, that encodes the alpha subunit of the stimulatory G protein – coupled receptor, GSa.

The activating mutations occur post zygomatically , replacing the arginine residue amino acid with either a cystein or a histidine amino acid. The mutation selectively inhibits GTPase activity, resulting in constitutive stimulation of AMP-protein Kinase A intracellular signal transduction pathways.

This mutated protein coupled receptor complex autonomous function in bone through parathyroid hormone receptor, in skin through melanocyte-stimulating hormone receptor, in ovaries through the follicle-

stimulating hormone receptor and in thyroid and the pituitary gland , through the thyroid and growth hormone receptors respectively⁽²⁾.

FD is classified depending on the number of affected bones and the presence or absence of extra-skeletal abnormalities. The monostotic form affects only one bone and corresponds to 70-80% of F.D cases. The polyostotic form corresponds to 20-30% of cases. The craniofacial type in which only the craniofacial complex are involved including the jaw and maxilla. The craniofacial bones are more affected in the polyostotic (50-100%) than in the monostotic form (20%)⁽⁴⁾.

The other subtypes of polyostotic F.D show café-au-lait pigmented skin lesion and endocraniopathies, when they are called Jaffe-Lichtenstein syndrome and McCune-Albright syndrome.

The presenting symptoms in both our cases was an increasing swelling of the jaw bone that was disfiguring and intraorally the swelling was bony hard and the overlying mucosa appeared normal

and the swelling was asymptomatic. Both the cases were in the third decade of life and were males. In case 1 the swelling was in mandible while in case 2 the lesion involved the maxilla and the craniofacial complex. The age group corresponded with the other studies.

The radiographic appearance in F.D varies greatly depending upon the stage into 3 types,

Type I- a small unilocular or multilocular radiolucency with a well circumscribed border containing a network of fine bone trabeculae.

Type II- similar but with increased trabeculation rendering a more opaque and mottled appearance.

Type III- the lesion is quite opaque with many delicate trabeculae giving a ground glass or Peau d'orange appearance to the lesion. This type is not well circumscribed but blends with the normal bone⁽⁵⁾.

In our cases, case 1 showed mixed radiopaque /radiolucent lesion with diffused border blending with the normal bone anteriorly.

Case 2 showed an expansile bony lesion with ground glass appearance in the maxillary sinus as well as the alveolar and zygomatic processes with the border merging with normal bone. Both the cases had type III radiographic presentation.

Based on the histopathology alone it is virtually impossible to differentiate between fibrous dysplasia and ossifying fibroma. For this reason X-ray studies are indispensable for the interpretation of fibro-osseous lesions of jaws. ⁽⁶⁾

The histopathology shows considerable variation. The lesion may be a fibrous one made up of proliferating fibroblasts in a compact stroma of interlacing collagen fibres. Irregular trabeculae of bone are scattered through out the lesion with no definite pattern of arrangement. Some of these trabeculae may show Chinese letter pattern. The trabeculae may be coarse woven bone or lamellar bone which over time will mature especially the craniofacial type ⁽⁵⁾.

Therefore for both the cases , the clinical features radiographic features and histopathological features were correlated to come to a final decision of fibrous dysplasia.

Conclusion:-

The fibrous dysplasia is significant for the dentists because it may affect the facial, cranial and jaw bones leading to many deformities and dysfunctions. The cells of fibrous dysplasia are committed osteogenic cells with impaired capacity to form normal bone. The mutated protein not only affects osteoblasts but can also affect various hormone receptors leading to endocrinopathies and café -au-lait spots. Both our cases did not show any skin pigmentations or endocrinopathies. Malignant transformation occurs infrequently with reported frequencies ranging from 0.4-4%. Both our cases have not shown any malignant changes till date and the follow up is still on going.

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Corresponding Author:-

Dr. Jyothi Mahadesh,

Professor and Head

Department Of Oral &
Maxillofacial Pathology

Sri Siddhartha Dental College &
Hospital

Agalakote, Tumkur-572107

Email id:

jyothi.desh@gmail.com