POLYOSTOTIC FIBROUS DYSPLASIA—A case report

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ABSTRACT

Fibrous dysplasia, a benign fibro osseous condition involving one or more bones of the cranial and extra cranial skeleton, consists of non-encapsulated lesion which shows replacement of a normal bone by cellular fibrous tissue containing islands of metastatic bone (Edwards 1984). It has been classified by W.H.O. as developmental in origin. Fibrous dysplasia of bone is one of the most perplexing diseases of the osseous tissue. It is a lesion of unknown etiology, uncertain pathogenesis and diverse histopathology. A case report of fibrous dysplasia is presented here and discussed in detail.

KEY WORDS: ***Fibrous dysplasia, fibro-osseous, Histo pathology***

CASE REPORT

A patient by 20 years old age attended the department with the complaint of hard growth on the right side of the upper jaw since 6 months.

He observed small painless growth on the right side of the Maxilla which was slowly growing not associated with pain. At the same time he also observed growth on the chest which was first observed by his friends, it was also slowly growing. Both lesions were asymptomatic. On general examination patient's skin is completely normal without any pigmentation. No endocrinal dysfunction is observed. Patient's face is slightly asymmetrical due to the growth on the right side of the maxilla(Fig 1.). Patient's oral hygiene is good with full set of teeth. Growth was extending mesially from right upper canine to distally up to second molar. The size of the lesion is approximately 4 X 2.5 cm. In superio-inferiorly extending from cervical level of first molar to beyond the buccal sulcus. Growth is irregular in shape and bony hard in consistency. The Gingiva over the lesion is normal (Fig 2.) There is another lesion present on the right side of the chest on second rib extending up to the fourth rib, medially from mediastinum to 1 Cm. mesial to mid clavicular live. Provisionally it is diagnosed as POLYOSTOTIC FIBROUS DYSPLASIA. Differential Diagnosis included - OSTEOMA, OSSIFYING FIBROMA. Investigations like IOPA, occlusal view of right maxilla, PNS view of the skull, PA view of the chest, Serum Calcium, Serum Alkaline Phasphatase. Biopsy is taken from right buccal region. Radiographic reports of IOPA and occlusal view showed ground glass pattern(Fig.3). PA view of the chest exhibited RadioOpacity at the second rib. Serum Alkaline Phasphatase was slightly raised. Biopsy report confirmed the diagnosis as fibrous dysplasia

DISCUSSION:

Fibrous dysplasia usually affects young people and tends to become static after puberty. However without treatment or careful management of diagnosed cases severe skeleton deformity can result. There are two main clinical types of this disease [1-5].

1) Monostotic form involving only one bone.
2) Polyostotic form involving many bones which can be sub-classified as:
   a) Jaffe's type in which several bones of the skeleton are involved, pigmented lesions of the skin (café-au-lait spots).
   b) Albright's syndrome a polyostotic form accompanied by pigmented skin lesions and endocrine dysfunction presenting as precocious puberty in females and sometimes other anomalies (Albright etal, 1937)[4,5].

PATHOLOGY & PATHOGENESIS:

Histopathological appearance can vary according to the relative maturity of the lesion are area of the lesion being examined. It consists of well vascularised, cellular fibrous connective tissue containing irregular trabeculae of immature bone sometimes having a Chinese character appearance. The bone in some lesions or parts of the lesions may show evidence of osteoclastic activity that are lined by osteoblasts, a feature suggesting of active phase of the disease. In contract moth eaten

spicules devoid of osteoblastic activity which can be seen in some lesions may represent a burned out phase[3]. Other morphological features of Calcification (cementum bodies are globular calcification) as seen in ossifying fibroma and minute basophilic and laminated calcifications may be seen. This feature is not pathognomic and diagosis requires correlation of histopathological radiographic and clinic features

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The pathogenesis of this disease is poorly understood. Schlumburger (1946)[1] suggested that infection or trauma might play a role in some cases. But it is possible that there is genetic predisposition to this defect involving osteogenic mesenchymal tissue. Greco and Steiner (1996) examined fibrous dysplasia cases and suggested that abnormal osteoblastic maturation of the bone forming mesenchyme has one of the most important alterations in this condition. Osteosarcoma develops in 1% of patients with Albright's syndrome.

Polyostotic fibrous dysplasia[3]:

The first recognized case of polyostotic fibrous dysplasia with skin and endocrine lesions was in 1922 by well. Since then various cases have been described.

a) fibrous dysplasia involving a variable number of bones, where most of the skeletal bones are normal and are accompanied by pigmented lesions of the skin or café-au-lait spots known as jaffe's type.

b) Fibrous dysplasia of a more severe type involving nearly all bones in the skeleton accompanied by pigmented skin lesions and in addition having endocrinal disturbances of varying types known as " Albright's type ".

Warrick in 1973 compiled and evaluated following endocrinal disorders with polyostotic fibrous dysplasia, like: Sexual precocity, accelerated skeletal growth with premature fusion of epiphyses, Goiter and hyperparathyroidism, Cushing's syndrome, Gynaecomastia, Diabetes mellitus, Acromegaly, Hyperparathyroidism – caused by abnormality of parathyroid gland.

Clinical features It usually manifests early in life. Recurrent bone pain is the most common skeletal symptoms. Bowing and thickening of long bones with leg length discrepancies were common. Deformities such as coxavara, shaephar's crook, deformity of femur, bowing of the tibia, Harvison's groove and intrapelvic protrusions of acetabula were also seen. Spontaneous pathologic fractures were a common compilation. Café-au-lait spots on the skin were seen-most commonly seen on the abdomen, back and thighs. These lesions were due to irregular melanin pigmentation.

In cases of fibrous dysplasia with endocrine dysfunction (Albright's type) precocious puberty was seen in females. Cases of vaginal bleeding with advent of menarche during 4th month of life have been seen. Early development of breasts along with variety of disturbances involving various endocrine glands should also been noted. Bones of the face and skull are frequently involved causing an obvious facial asymmetry. Universal finding of a bony, non-tender swelling, obstruction or displacement of adjacent structures is often seen. Oral manifestations in the form of expansion and deformities of maxilla and mandible along with altered oral pigmentation are seen. Roots were often displaced along with loss of lamina dura around the teeth. Less frequently multiple jaw lesions with facial asymmetry and titled occlusal plane caused a grotesque appearance.

Clinical course and progression:

1. Extension of existing lesions.
2. Appearance of new lesions.
3. Increasing the deformity of the involved bones.

Roentogenographic features:

The clinical basic behind the reontgenographic changes are due to rarefaction of the medullary portion of the bone. Irregular bony trabeculature along with variations in cortical thickness.

Psuedo De. Orange or Ground Glass appearance : alternating areas of granular density and lucency, giving a radiographic appearance resembling the rind of an orange, most common type – 40% of cases.

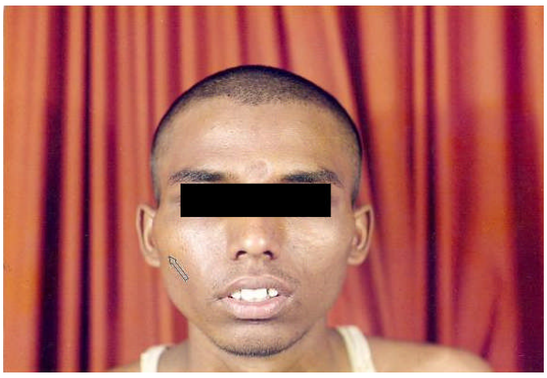

Fig 1. **Asymmetry of the face due to swelling on the right side of the face.**

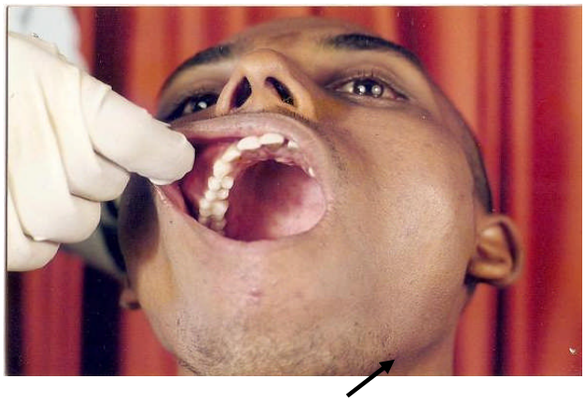

Fig 2. **Intraoral view of the lesion.**

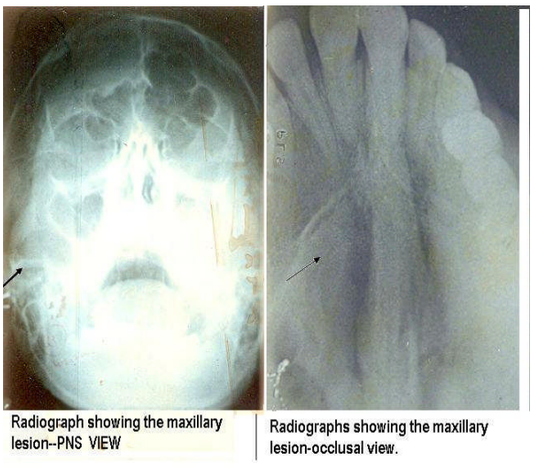

Fig 3 . **Radiographs of the lesion.**

2. **Whorled plaque like**: The matrix of the well circumscribed expansive lesion was composed of plaques of amorphous material of radio density intermediate between bone and soft tissue which, on close examination, are seen to be arranged in a whorled, onion peel, or whirl pool pattern and is seen in 20% of cases

3. **Diffuse sclerotic type**: This type presented as a homogeneously dense area of involvement, with no clear of lesion and normal bone. These lesions have varied shapes and sizes and are usually extensively seen in 16% of cases.

4. **Cyst like**: on close examination faint ground glass matrix due to mineralization in fibrous dysplasia.

5. **Pagetoid type**: In this radiologic type of lesion the affected area of bone was markedly expanded and showed alternating areas of radiopacity and radiolucency, such as seen in Paget's disease of bone.

6. **Chalky type**: The matrix consisted of amorphous radiodense material with sharply, marginated lesions containing whorled plaque like densities – rare type.

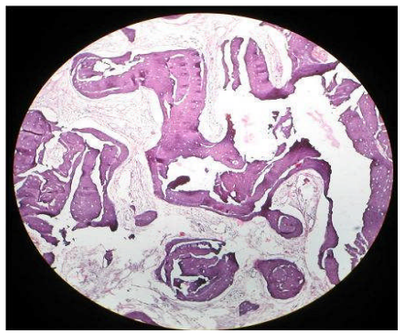

Fig.4. Histopathological section of the lesion

Monostotic fibrous dysplasia[3]

Monostotic variety of craniofacial region typically presents lesions without a clearly defined boundary. Periphery of the lesion often blends into normal bone and hence it is difficult to clearly delineate the lesion.

Unlike polyostotic variety, monostotic variety is diagnosed only after conducting biopsy.

Histological features: (Fig.4)

Macroscopic: Fresh tissues from an area of fibrous dysplasia appear reddish grey to grey in color. Tissue is firm and gritty and easily cuts with a scalpel. Cut surface is flat and non-compressible and on palpation sensation in similar to that of rubbing a fine grade of sand paper.

Microscopic: Bone formed in fibrous tissue is coarse with minimal collagen production. Woven bone is arranged randomly with few osteoblasts on the bony trabeculae. Areas of calcification along with resorption cavities containing osteoblasts and few giant cells seen. Although resorption and formation of bone seen, it did not result in mature lamellar bone formation. This is characteristic difference between reparative bone and fibrous dysplasia histologically. Cement line as separate the different generation of boner; however bony trabeculae are not oriented along stress lines. Cortical thinning is evident and multiple islands of cartilage with fluid filled cysts may be seen.

Biochemical findings: serum alkaline phosphates may be slightly raised due to increased bone activity.

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CONCLUSION

Fibro-osseous lesions appear in the jaw bones with a variety of clinical and radiographic characteristics. The fibro-osseous nature of the lesion can only be confirmed by biopsy. As it has varied radiographic appearance, a fibro-osseous lesion must be considered in the differential diagnosis of almost all radiolucent, radiopaque or radiopaque-radiolucent lesions of the jaw bones. Conversely many serious bone diseases and lesions are manifested radiographically with characteristic “fibro-osseous” bone changes. Even clinically asymptomatic lesions that show radiographic changes should have a biopsy. But differentiation between the various fibro-osseous lesions i.e., fibrous dysplasia, ossifying fibroma and cementifying fibroma is difficult on the biopsy results alone. A review of the clinical and radiographic features together with the histopathologic changes and surgical findings will allow for a definitive diagnosis.

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