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## **Fibro-osseous lesion of maxilla. Report of two cases in a family with review of literature**

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### **Abstract**

Fibrous dysplasia is a disturbance of bone metabolism that is classified as a benign fibro-osseous lesion. Fibrous connective tissue containing abnormal bone, replaces normal bone. The etiology of fibrous dysplasia is unknown. The radiographic appearance of the irregularly shaped trabeculae aids in the differential diagnosis. Occurring most commonly in the second decade of life, the lesions of fibrous dysplasia can be surgically recontoured for esthetic or functional purposes once they become dormant.

**Key words:** Fibrous dysplasia, maxilla, ground glass appearance.

## Introduction

The term fibro-osseous lesion (FOL) is a generic designation of a group of jaw disorders characterized by the replacement of bone by a benign connective tissue matrix. Benign fibro-osseous lesions constitute a varied group of lesions with a common histological characteristic: the substitution of normal bone by tissue composed of collagen and fibroblasts, with variable amounts of a mineralized substance that may be bone, cementum or both. These lesions comprise fibrous dysplasia, ossifying fibroma, periapical cemento-osseous dysplasia, focal cemento-osseous dysplasia, florid cemento-osseous dysplasia and cemento-ossifying fibroma.

The relationship between fibrous dysplasia and osteofibroma remains controversial. In 1972 Eversole et al. (1) concluded that fibrous dysplasia and cemento-ossifying fibroma are clinically and radiologically distinct disease entities that nevertheless are not always histologically distinguishable.

Fibrous dysplasia is an uncommon disorder of unknown etiology. It represents a bone developmental disorder, specially a defect in osteoblastic differentiation and maturation (2). Fibrous connective tissue containing abnormal bone replaces normal bone. Bone abnormalities represent a remarkable point in the disease, whereas endocrinopathies, abnormal skin pigmentation and mucosa membrane alterations may also be present. Fibrous dysplasia is a localized abnormality, which can involve one (monostotic) or multiple bones (polyostotic). Most commonly the forms are distributed as follows: 74% monostotic, 13% polyostotic and 13% craniofacial (3). Approximately 3% of the patients have the so-called McCune-Albright syndrome, in which bone involvement is followed by skin lesions and endocrine pathologies. Monostotic fibrous dysplasia, though less serious than polyostotic fibrous dysplasia, is of greater concern to dentist because of the frequency in which the jaws are affected. The diagnosis of fibrous dysplasia is often made in infancy and childhood. The maxilla or mandible may be involved but a predominance of the maxilla has been documented (1). 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.

## Case report

A male patient of 14 years reported with the chief complaint of swelling on the right side of face since 6 months. Initially swelling was of smaller dimensions but gradually has achieved present size. On careful clinical examination, single diffuse non tender hard swelling over right zygomatic area seen. Intraorally expansion of the buccal cortex & obliteration of buccal vestibule seen as seen in the Figure 1.



Fig. 1.

After radiological examination, intra oral periapical radiograph revealed ground glass appearance in the periapical region and displaced roots of 15, 16. Lateral occlusal radiograph revealed expansion of the buccal cortical plate. Water's view revealed radio opacity in the right maxillary sinus region. Computed tomographic scan revealed an expansile mass with calcified matrix in right maxillary sinus region obliterating the entire maxillary sinus region giving an impression of Fibro-osseous lesion of right maxillary bone as seen in the Figure 2.



Fig. 2.

*Second Case Report:* On asking detailed family history, one of his younger male cousins of age 10 years was found to have similar swelling in the right side of upper jaw. He was requested to report to department with his cousin on the next appointment and he reported with his cousin brother. His cousin brother was having swelling on the right maxillary region which was diffuse, non tender and having ill defined margins. Expansion of cortical plate was present both buccally as well as palatally ex-



Fig. 3.

tending from distal to 53 to distal of 16 region as shown in the Figure 3.

Both of them were referred to the Department of Oral and maxillofacial surgery for further treatment, but treatment of both was deferred till the age of 18 years and was instructed for regular follow ups.

## Discussion

Fibro-osseous lesions are the group of disorders in which matrix of the normal bone is replaced by immature matrix of woven bone. As the lesion matures, the fibrous connective tissue is replaced with irregularly patterned trabecular bone. The monostotic form generally occurs during the second decade of life and becomes dormant by the third decade. Hormonal changes, such as those seen in pregnancy, can reactivate a dormant lesion.

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, anosmia, nasal obstruction, epistaxis, epiphora and symptoms mimicking sinusitis(4). With initial development of fibrous dysplasia the patient usually reports facial swellings and asymmetries. Males and females are thought to be affected evenly, although recent research has shown a slight female preponderance. However, McCune-Albright syndrome, a form of polyostotic fibrous dysplasia associated with café-au-lait pigmentation and multiple endocrinopathies such as precocious puberty, pituitary adenoma or hyperthyroidism, almost always affects females.

The lesions of fibrous dysplasia are twice as common in the maxilla as the mandible, and the posterior aspects of the jaw are more frequently affected than the anterior. Treatment usually involves bony recontouring at the affected site to improve esthetics and function. Recurrence is rare in adults, but the lesions can show surprising growth potential if they are surgically altered during their active growth phase(5).

Frequently, the radiologic appearance of fibrous dysplasia is not pathognomonic. The differential diagnosis differs according to the radiologic appearance and extent and site of skeletal involvement.

In monostotic fibrous dysplasia, a small lytic lesion may be radiographically indistinguishable from a simple bone cyst. Both may cause mild expansile remodeling of the affected bone. The finding of a thick peripheral rind surrounding the lesion favors the diagnosis of fibrous dysplasia over bone cyst.

Differential diagnosis of the initial radiolucent stage must include: central ossifying fibroma (COF), central giant cell granuloma (CGCG), aneurysmal bone cyst, osteomyelitis and early fibro-osseous lesions.(6). Because these lesions represent a variety of disease processes with different behaviors, including infection and endocrine dysfunction, prompt diagnosis incorporating clinical, radiographic and, occasionally, histologic findings, is essential.

COF is a benign neoplasm that commonly has a radiographic and histological appearance similar to that of fibrous dysplasia. A mixture of lamellar and woven bone is typically seen. Often, there are also scattered ovoid calcifications that resemble cementum. In contrast to fibrous dysplasia, a well-defined capsule occasionally surrounds the lesion. Radiographically, a well-defined margin is consistent with COF, whereas the margins of fibrous dysplasia tend to merge with the surrounding normal bone. On occasion, a sclerotic border, absent from fibrous dysplasia, is also seen in COF. COF occurs mostly in the third and fourth decades of life, whereas fibrous dysplasia is most often discovered in the second decade. COF is more common in the mandible, tends to occur in anterior regions and is smaller in size, whereas fibrous dysplasia is more common in the posterior maxilla, and the lesions tend to be larger. Both lesions tend to expand the bone cortex.

Differentiation of these two lesions is critical because the treatment protocols are quite different. COF, although benign, must be enucleated due to its potential to recur. Fibrous dysplasia is generally self-limiting and does not require treatment except for cosmetic reasons, pain, discomfort or impaired function(3). If undertaken, the treatment, consisting of recontouring or resection, should be postponed until after cessation of skeletal growth, since early treatment may accelerate growth of the lesion.

Early-stage fibrous dysplasia should also be differentiated from CGCG, which more commonly appears in the anterior mandible, results in generally painless expansion of bone and appears radiographically as unilocular or multilocular radiolucent defects with well-delineated, noncorticated margins.

Fibrous dysplasia may also mimic Paget's disease of bone on clinical examination, particularly if a patient

with fibrous dysplasia does not present until later in life. In addition to the predilection of Paget's disease for an older population, certain radiographic and clinical features help to distinguish this lesion from other radiographically similar lesions. These features include thickening of the cortices, cotton wool appearance of the involved bone and increased blood levels of alkaline phosphatase. The most useful clinical feature for distinguishing Paget's disease from fibrous dysplasia is that the former tends to occur bilaterally in the jaws, whereas the latter affects only one side.

Although osteomyelitis demonstrates sequestra in the later stages, it may resemble fibrous dysplasia in the early stages, especially if there is associated swelling. Usually, inflammatory signs and the presence of draining sinus tracts are indicative of osteomyelitis. Periosteal new bone, manifesting as one or more laminations occurring parallel to the outline of the jaw, often occurs in osteomyelitis affecting young patients and is a useful indicator of the condition. Osteomyelitis may occur secondary to odontogenic infections of pulpal origin, although hematogenous spread from distant sites has also been reported. Once the offending tooth has been treated, the lesion often resolves spontaneously.

Fibrous dysplasia can often be differentiated from osteosarcoma on the basis of radiographic appearance. The radiographic features of osteosarcoma are orthoradial striations, destruction of cortices with an outgrowth of the soft tissue component, generalized widening of the periodontal ligament spaces and destruction of the lamina dura (7). Previously, MacDonald Jankowski D (8) evaluated the features of fibrous dysplasia by systematic review of 31 reports and 788 cases. All cases were confirmed fibro-osseous lesions histopathologically. Fibrous dysplasia was found to affect males and females equally, but it was 50% more prevalent in the maxilla. The mean age at first presentation was 24 years, and the greatest frequency occurred in the second decade; in this group, males accounted for 63% of cases. The primary symptom in 90% cases was swelling, including deformation of the jaw. All cases displayed buccolingual expansion; all mandibular cases exhibited downward displacement of the lower border of the mandible; and almost all maxillary cases involved the maxillary antrum.

Valentini et al (9) studied the results of conservative and surgical treatment for craniomaxillofacial fibrous dysplasia in 68 patients from 1980 to 2002. Sixty-one patients had radical excision, 6 received conservative treatment, and 1 patient with mandibular involvement received radical excision and immediate reconstruction with a free fibula flap. The authors concluded that in most cases of monostotic or monofocal fibrous dysplasia of the craniofacial region, surgical techniques allow an aggressive but definitive treatment with good functional

and aesthetic results. The authors noted that they perform radical treatment even in cases involving the maxilla and mandible and prefer a conservative approach only in polyostotic cases and cases of McCune-Albright syndrome.

Fibro-osseous lesions are jaw disorders of benign nature and of unknown etiology. The dentist interest lies in the fact that these disorders may affect facial bones, causing deformities and dysfunctions. Diagnosis of fibro osseous lesions although a technical challenge for dentist, but dental surgeon can diagnose with careful clinical and radiological evaluation and differentially diagnosed from other lesions having similar clinical and radiological picture.

Although in our cases, there was no compelling indication to seek a biopsy, any sudden change in the clinical presentation or behavior of the lesion might warrant further investigation.

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