Research Paper

Volume : 2 | Issue : 2 | Feb 2013 • ISSN No 2277 - 8179

Medical Science

KEYWORDS : Dysplasia, fibro-osseous, curettage, recontouring.

# Extensive Fibrous Dysplasia of Maxilla – A Case Report

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## ABSTRACT

*Fibrous dysplasia is a disease of bone maturation and remodeling. The process in fibrous dysplasia is the replacement of normal medullary bone and cortices into a disorganized fibrous weaker bone. This bone (fibro-osseous bone) is more elastic and structurally weaker than the original bone. The cause of development of fibrous dysplasia is the deletion of a bone maturation protein during embryogenesis and represents about 2.5% of all bone tumors and over 7% of all benign tumors of facial bones. This article presents a case of extensive fibrous dysplasia of the maxilla. The diagnosis was confirmed by imaging studies such as OPG and CT scan and by histopathological examination. Curettage and surgical recontouring was performed as a mode of treatment.*

## INTRODUCTION

Fibrous dysplasia is an asymptomatic non neoplastic disease of bone in which normal trabeculae like osseous structure are present. In 1937, McCune and Bruch first suggested that among all of the abnormalities of bone disorders, the fibrous dysplasia should have its own place as a distinct clinical entity. Later, Lichtenstein introduced the term fibrous dysplasia as an arrest of bone maturation in which woven bone with ossification results from metaplasia of a non specific fibro-osseous type. Fibrous dysplasia is not a true neoplasm because it is self limiting. It begins its development as a fibrous replacement of the medullary bone which is gradually replaced by metaplastic woven bone that eventually matures into dense lamellar bone. Fibrous dysplasia is a disease of bone maturation and remodeling in which the resultant fibro-osseous bone is more elastic and structurally weaker than the original bone due to the abnormal growth process [1]. This lesion is related to a mutation in the gene that encodes the submit of a stimulating G protein (GS) located as chromosome 20. Due to this mutation, there is a substitution of the cysteine or the histedonian acids of the genomic DNA in the osteoblastic cells by another amino acid, arginine[3]. As a result, the osteoblastic cells will elaborate a fibrous tissue in the bone marrow instead of normal bone. Fibrous dysplasia affects the facial bones in about 30% of the patients. It represents about 2.5% of all bone tumors and over 7% of all benign tumors. The condition develops in children and teenagers primarily with few if any cases beginning after the age of 25 years [4]. The maxilla is affected more often than the mandible. Teeth are often displaced, rotated or malaligned resulting into severe malocclusion. The affected area presents as an asymptomatic diffuse expansion of the cortices. Fibrous dysplasia can be described in three major types – monostotic fibrous dysplasia, polyostotic fibrous dysplasia and craniofacial fibrous dysplasia. Monostotic fibrous dysplasia involves a single bone and accounts for 70% of fibrous dysplasia cases [7]. Polyostotic fibrous dysplasia involves several skeletal sites occurs as part of the McCune Albright syndrome. It features also skin pigmentation and endocrine dysfunctions [5]. When polyostotic form of fibrous dysplasia occurs in absence of endocrine disturbance, it has been formed Jaffe-Lichtenstein type of polyostotic fibrous dysplasia. Craniofacial fibrous dysplasia involves two or more bones of the jaw-midface skull complex in continuity. This type of fibrous dysplasia is seen relatively often in dental and oral and maxillofacial practices. It is thought to be a monostotic fibrous dysplasia of the maxilla, yet it often involves the zygoma, sphenoid, temporal bone, nasal chonchae and clivus. Alkaline phosphatase may be raised in up to 30% of patients with polyostotic fibrous dysplasia, and a dramatic rise may herald malignant degeneration. Malignant degeneration occurs in less than 1% of cases of fibrous dysplasia. The disease is self-limiting but grossly disfiguring lesions may need to be excised. Treatment is surgical recontouring for cosmetic and functional achievements [2].

## CASE REPORT

A 32 years old female patient attended the department of oral surgery with a chief complaint of swelling in the left side of face. Swelling was slowly progressing for the last 4 years. The swelling was causing problem with facial esthetics and difficulty in mastication. There was no past medical history and family history with similar findings of swelling. The extra oral clinical examination revealed hard swelling of about 4 cm. in size on the left side of maxilla extending from alla of the nose to the cheek area “Fig 1about here”. Intraorally the swelling is bulging in the left maxillary alveolar process with the ulceration of mucosa. There was displacement of posterior teeth “Fig 2 about here”. On palpation the lesion was non tender, hard in consistency, expansion of buccal cortical plate from canine region to second molar region. OPG and CT showed radiolucent shadow in the first premolar to second molar region. CT scan revealed a radiolucent radiopaque shadow in the posterior part of the left maxilla “Fig 3,4 about here”. Preoperatively synthetic salmon calcitonin injections in doses ranging from 50 to100 IU three times weekly for a period of 3-months were given subcutaneously to prevent excessive bleeding during operation and to induce bone formation by its anti-osteoclastic actions. The mass was removed under LA by curettage and surgical excision. The mucoperiosteal flap was raised extending from central incisor to molar region of the same side. A hard bony mass was excised by curettes “Fig 5,6 about here”. The wound was decontaminated with normal Saliva and betadine solution. Carnoy’s solution was applied as a fixative to fix the lesion. This was used to prevent any recurrence of the lesion by the necrosis of the remaining lesion.Primary closure was done with vicryl 4-0 interrupted suture. The mass was sent for histopathological examination to confirm the diagnosis. The histopathological report showed areas of cellular fibrous connective tissue and immature metaplastic bone with a woven pattern. The lesion blends with the surrounding normal bone and cortical plates “Fig-7about here”. This confirmed the diagnosis of fibrous dysplasia. Follow up of the lesion was done for one and half years for any recurrence. No recurrence was observed “Fig 8,9 about here”.

## DISCUSSION

Fibrous dysplasia is a disease of bone in which the normal medullary bone and cortices are replaced by a disorganized form of woven bone. It is caused by the deletion of a bone matura-

tion protein during embryogenesis. There is no hereditary involvement. Fibrous dysplasia is not considered a true neoplasm, because it is self-limiting. It begins as a form of replacement of the medullary bone and cortices which is replaced by a disorganized forms woven bone [1]. Fibrous dysplasia is a benign bone disorder of unknown etiology, uncertain pathogenesis and diverse histopathology. All forms of fibrous dysplasia result from a defect in bone maturation that begins in the embryo. Some believe that in the histodifferentiation phase of the embryo, a genetic mutation or a deletion occurs in the gene that encodes for the intracytoplasmic transducer protein required for bone maturation. Consequently all the daughter cells of the original aberrant cell lacks that signal transducer and therefore will be able to produce only fibrous dysplastic bone. If the genetic defect occurs early in embryonic development, a large number of daughter cells are affected. When such early term-altered cell migrates into several skeletal sites, they produce polyostotic fibrous dysplasia. If the genetic defect occurs in an even earlier phase of embryonic development, the original cell may produce daughter cells of divergent differentiation. Some that will migrate into bone primordia and some into endocrine gland primordial, this produce either McCune – Albright Syndrome in the Jaffee-Lichtenstein type of polyostotic fibrous dysplasia [2]. In about 3% of the cases fibrous dysplasia clinically it is in the beginning hidden, having no clear symptoms and often the diagnosis is difficult. Other 97% of the patients have diverse symptoms depending on location, swelling, deformation and presence of pain [8]. Monostotic fibrous dysplasia gives rise to a bony swelling caused by a poorly & circumscribed area of fibro-osseous proliferation. Polyostotic fibrous dysplasia is rare but shows histologically similar lesions in several bones. Polyostotic fibrous dysplasia involves two or more noncontiguous bones usually unilateral. This type of is less common than monostotic fibrous dysplasia and may involve the skull, jaws or facial bone together with ribs, long bones or pelvic. Two syndromes are isolated with polyostotic fibrous dysplasia. Mc Cune Albright Syndrome comprises polyostotic fibrous dysplasia with concomitant melanotic pigmentations called cafe-all-lait muscles and endocrine abnormalities. The most common of the endocrine abnormalities is precocious puberty. In polyostotic fibrous dysplasia sudden increase in the level of alkaline phosphates is one of the indications for malignant transformation and for that reason its amount should be periodically observed [9]. The patient is advised to visit regularly for the increase in size of the lesion and changes in the level of alkaline phosphates. Histological findings are the main criteria for the diagnosis of the lesion. The normal bone is replaced by a generally loose, cellular fibrous tissue which is composed of haphazardly arranged, variably shaped trabeculae of woven bone containing numerous osteocytes. Aggregates of multinucleated giant cells may be presents. By adulthood, the fibrous dysplasia of the jaws may show maturation which is characterized by formation of lamellar bone and parallel arrangement of the trabeculae. There is the bone which may be same as what normal bone. The treatment of fibrous dysplasia is the correction of disfigurement and function by surgical excision of the affected bone tissue [10].

The osseous contouring surgery is to be performed only in adulthood (ages between 18 to 21 years). Surgery should be avoided during the active period of expansion of bone. Resection is not indicated in monostotic fibrous dysplasia of the jaw. Some surgeons suggest applying calcitonin in combination with surgical treatment.Calcitonin treatment causes bone calcification leading to reduction in bleeding during bone remodeling [11].

**CONCLUSION**

Fibrous dysplasia of jaws or facial bones is rare and is difficult to differentiate from other benign and malignant bone disorders. Histological features, clinical and radiographies findings are imperative for the final diagnosis of the lesion Monostotic fibrous dysplasia and craniofacial fibrous are self-limiting but grossly disfiguring esthetics and function may require surgical excision. Lesions should not be treated by radiotherapy due to the risk of a malignancy in later years of life.

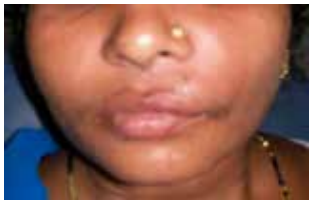

Figure- 1: Preoperative Front View of the Patient

Figure- 2: Preoperative Intra Oral View of the Lesion

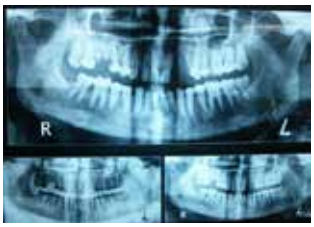


Figure- 3: OPG showing radiolucency in the region of Anterior Maxilla of the Left Side

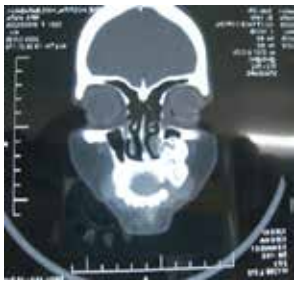

Figure- 4a: Coronal Section CT Scan the Lesion

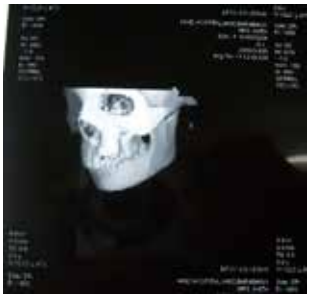

Figure- 4b

Figure 4a & b: 3D CT Scan Showing Mixed Radiolucency – Radiopacity on the Left Anterior part of Maxilla

Figure- 5: Surgical Exposure of the Lesion

Volume : 2 | Issue : 2 | Feb 2013 • ISSN No 2277 - 8179

Figure- 6: Total Excision of the mass with control of hemorrhage

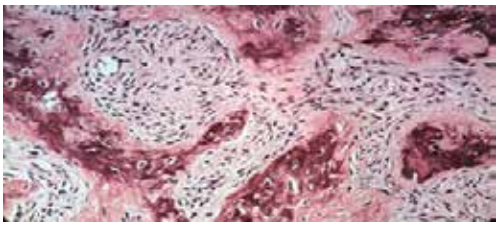

Figure- 7: Histopathological Picture of the Mass showing Immature Woven Bone with Fibrous Tissue

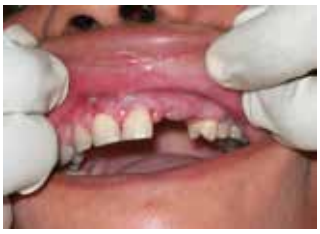

Figure- 8: Postoperative Intra oral view of the Patient

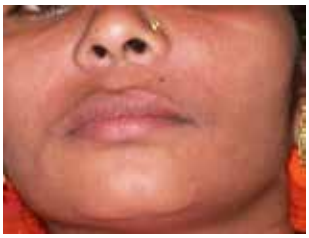

Figure- 9: Postoperative Front View of the Patient

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