



Fibrous Dysplasia of Maxillary Bone

KEYWORDS

Fibrous Dysplasia, Maxilla, Monostotic form, Ground glass form.

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ABSTRACT *Fibrous Dysplasia is a tumor like condition, characterized by excessive proliferation of cellular fibrous connective tissue intermixed with irregular bony trabeculae with immature bone replacing the normal bone. Seldom, has it caused pain, but, deformities and pathologic fractures are common features. In current study 40 patients diagnosed from the disease were included and were followed up for 5 years post operatively. Group I patients underwent surgical recontouring and Group II patients were treated for enbloc resection of the lesion. Analysis of the study was re done after 5 years for recurrence and esthetic outcome of the treatment. Current study suggests that, surgical recontouring remains a better modality of treatment for monostotic form of Fibrous Dysplasia. However deeper evaluation of the disease and its treatment outcome study should be carried out on multicentric basis and with longer follow up period*

Introduction:

It's a common developmental bone disease of benign origin. It leads to expansion and replacement of medullary bone by disorganized fibrous tissue [1]. It comprises of 2.5% of all osseous and 7% of all benign bone tumors [2]. Male: female ratio is equal [3-5]. It is most commonly found in metaphysiodiaphyseal region of bone i.e. in Ribs 28%, Femur 23%, Tibia and Craniofacial bones 10-25% [4]. Sarcomatous changes are rare [2].

It was first reported by Von Recklinghausen in 1891 and he coined the term Osteitis Fibrosa Generalisata [5-6]. McCune and Albright et al in 1938 explained the triad of Polyostotic Fibrous Dysplasia, Precocious Puberty, areas of cutaneous pigmentations (café – u – lait spots) as the McCune's Albright syndrome, during the same period Lichtenstein and Jaffe first introduced the term Fibrous Dysplasia [5].

It is a developmental tumor like sporadic condition resulting from post zygotic mutation in GNAS1 (Guanine Nucleotide binding protein, a stimulating activity polypeptide 1) [4-5]. This gene codes for G protein, which stimulates cAMP production in affected tissue, which results in 1) Endocrinal Disturbances leading to a) Precocious Puberty, b) Hyper thyroidism, c) growth hormone and cortisol over production. 2) Increased proliferation of Melanocytes, leading to Café' – u – lait spots. 3) Aberrant activity during Osteoblast differentiation, which leads to replacement of normal medullary bone with fibrous tissue, which appears radiolucent on the radiographs [4].

Material and Methods:

40 cases (16 females and 24 females) were evaluated, between the age range of 28 to 47 years) reporting to the oral surgery clinic at Your DentistTM, Bhopal and were diagnosed as suffering from monostotic fibrous dysplasia on the basis of clinico-radiological features. 28 patients had maxillary lesion and 12 patients had mandibular lesion. Patients were randomly segregated into two groups. Group I comprised of patients treated for, corticotomy and surgical recontouring of the lesion, where as group II pa-

tients were treated by enbloc resection of the lesion. Patients were followed up for a period of 5 years. In Group I there were 13 patients with maxillary lesion and 7 patients with mandibular lesion, where as in Group II there were 15 patients with maxillary lesion and 5 patients with mandibular lesion. Post operative evaluation was done on the basis of recurrence of the disease, functional impairment and facial esthetics. A total of Photographic comparative analysis was done with the pre operative clinical photographs and post op photographs at 6 months, 12 months, 24 months and then 60th month post operatively.

Discussion: -

Fibrous Dysplasia is connective tissue disorder of unknown origin, characterized by slow progressive replacement of medullary bone by abnormal, proliferative, isomorphic fibrous tissue which appears radiolucent on a radiograph with classic ground glass appearance [1]. It has four different variants, 1) Monostotic 70%, 2) Polyostotic 30%, Craniofacial form and 4) Cherubism which are rare [3-4]. The range of skeletal involvement varies from an asymptomatic monostotic lesion to polyostotic involvement resulting in functional deficit and esthetic problems. The clinical severity depends on the time of mutation in GNAS1 gene.

Polyostotic form: -

It is seen when the mutation of the gene occurs at 6th intra uterine week, as multiple bones being involved. It commonly involves skull and facial bone, pelvis, spine, shoulder girdles etc. initial symptoms are bone pain and spontaneous fractures. Femur shows Shepherd's Crook deformity [4]. It is again sub classified into Jaffe's type and Albright's syndrome. Both types have variable degree of bone involvement with Café' – u – lait spots. Albright type has additional endocrinal disturbances of varying degree [4-7]. Polyostotic Fibrous Dysplasia with soft tissue diseases is also called Mazabroud Syndrome [7].

Monostotic form: -

if mutation occurs in post natal life, progeny of that mutated cells remain confined to one site resulting in Fibrous Dysplasia affecting a single bone. The incidence of this disease is about 70%, which may involve rib, tibia, femur

and craniofacial bones [1-2, 4]. Any bone may be affected, but monostotic are never being reported to transform into Polyostotic forms [4]. The lesions remain asymptomatic until they are accidentally found or are noticed when it causes deformity causing enlargement and distortion of bone.

Craniofacial form:

-50-100% of the patients with Polyostotic and 10% of the patient with monostotic form suffer from craniofacial disfigurement [1, 5]. Maxilla is more commonly involved than Mandible [1]. When maxilla is affected it may involve zygomatic and sphenoid bones. Involvement of frontal, sphenoid, naso-ethmoidal bone may lead to nasal and sinus obstruction with sinusitis [1]. Hypertelorism, cranial asymmetry, facial deformity, visual impairment, exophthalmos and blindness may occur due to involvement of orbital and parietal bone [4]. Malignant changes with Fibrous Dysplasia may lead to Osteosarcomas, Chondrosarcomas, Malignant Fibro Histiocytomas and Ewing's sarcomas [1]. Association of Ameloblastoma, Cystic degeneration, Angiosarcomas, Frontal sinus mucocele has also been reported [1, 2, 8, 9]. It gets stabilized after puberty [1]. Treatment is surgery. When only teeth bearing area is involved conservative treatment is bone recontouring. Use of Calcitonin and Pamidronate is also been reported.

According to Hunter AG et al [11] maxilla is more commonly affected than mandible, which is accordance to the current study, but males are affected more than females in our study where as feal predilection is more in study conducted by Hunter et al. According to Anthony Pogrel [13] esthetic recon-touring of the bone should be the mainstay of treatment which is in accordance to our result, where as according to Chen YR et al [12] surgical procedure should be aggressive and patient should undergo resection of the lesion to ensure no recurrence, which in author's opinion should be reserved for recurrent lesions only. Though the treatment modality suggested by Chen YR is for poly-ostotic form of FD and therefore may not coincide with the result of current study done on patients with mono-ostotic form of the disease.

Result:

Total of 40 patients in group I and II were evaluated for the treatment outcome of corticotomy and surgical recontouring compared to enbloc resection of the lesion respectively. Male is to female ration in current study is 2.3:1, and maxilla to mandible ratio is 2.6:1. Mean age of occurrence was 32 years. Evaluation was done on the basis of recurrence of the disease, functional impairment and facial esthetics. According to the p test applied for comparison of both the groups, p value in Group I and II is >0.1, and therefore is insignificant in terms of recurrence. However, p value is <0.1 and therefore significant in Group II for functional impairment and facial esthetics.

Conclusion: -

Fibrous Dysplasia may manifest as monostotic or Polyostotic form. Diagnosis of Polyostotic form is easier due to involvement of multiple extra skeletal bones. Monostotic form is common in jaws. Fibrous Dysplasia is a tumor like developmental disorder with minimal chances of malignant transformation. Primary treatment modality remains bone recontouring done comfortably by surgical shaving of the bone [10]. In the current study of 40 patients were diagnosed on clinicoradiological basis, suffering from mono-ostotic form of FD. Sex ratio was 2.3:1 with male predilection and incidence of maxillary involvement to be more than mandible. In lieu of 5 years post operative recurrence,

functional morbidity and facial esthetics, corticotomy with surgical recon-touring appears to be better modality of treating the mono-ostotic FD. Nevertheless conclusion of the study should be farther evaluated on multicentre basis with more sample size.

Figures and Legends



1. Pre Op Orthopentamograph of Left Maxillary Fibrous Dysplasia



2. Surgical Recontouring of Left Maxillary Fibrous Dysplasia

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