Fibrous Dysplasia of the Maxillary Bone- A Case Report

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Abstract:

Fibrous dysplasia has been regarded as a developmental skeletal disorder characterized by replacement of normal bone with benign cellular fibrous connective tissue. It is a lesion of unknown etiology, uncertain pathogenesis, and diverse histopathology. It result in facial asymmetry, pain, cranial nerve deficiencies, loss of vision or hearing, alterations in breathing, etc. It may involve one or several bones and consists of one or more foci of fibro osseous tissue within the matrix of the affected bone. Here a case of fibrous dysplasia of maxillary bone in a 45 years old female is presented.

Key words:. Fibrous dysplasia, Maxilla, Monostotic form

Introduction

Fibrous dysplasia (FD) is an uncommon developmental bone disease of benign origin. It leads to expansion & replacement of medullary bone by disorganized fibroosseous tissue [1]. It comprises 2.5% of all osseous & 7% of all benign bone tumors [2]. Reed's definition states that fibrous dysplasia is an arrest of bone maturation, woven bone with ossification resulting from metaplasia of a nonspecific fibro osseous type[3]It may present as monostotic or polyostotic,sometimes combined with precocious puberty, endocrine disorders and ''cafe´ au lait'' skin pigmentation termed McCune-Albright syndrome.[4] Gender prevalence of FD is equal. The monostotic form is more common and affects the 20 to 30 years age group: polyostotic FD has its onset mainly in children younger than 10 years of age[5], The ratio of occurrence of polyostotic to monostotic FD is 3:7 [6].

Fibrous dysplasia was first reported by Von Recklinghausen in 1891 & he coined the term *Osteitis Fibrosa Generalisata* [7].

In 1938, Lichtenstein and Jaffe first introduced the term *Fibrous Dysplasia*[8].Mc-Cune and Albright et al during the same period explained the triad of polyostotic fibrous dysplasia, precocious puberty, and areas of cutaneous pigmentation (cafe´-au-lait spots) as the Mc- Cune- Albright syndrome[7].

Fibrous dysplasia is a developmental tumor like sporadic condition that results from a post zygotic mutation in GNAS1 (Guanine Nucleotidebinding Protein -stimulating activity polypeptide 1) gene [7]. GNAS1 gene codes for G protein which stimulates cAMP production in affected tissue; which result in (1) Endocrinal disturbances leading to precocious puberty, hyperthyroidism, growth hormone and cortisole over production (2) Increased proliferation of melanocyte leading to café-au-lait spots (3) Aberrant activity during osteoblasts differentiation, which results in normal medullary bone to be replaced by fibrous tissue and appears radiolucent on the radiograph [2].

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CASE REPORT

This report will present a case of isolated fibrous dysplasia of the maxilla in a 45 years old female. A 45-year-old female presented at the Oral surgery Department, modern dental college and research centre, indore complaining of swelling of upper left maxillary back tooth region & noticed slight disfigurement of left upper side of face (fig. 1).

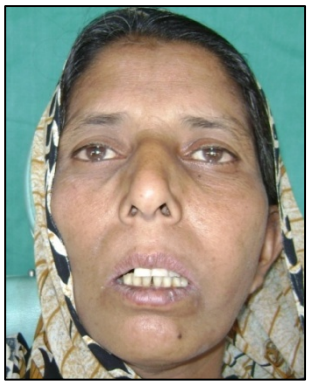

Fig.1: extraoral photograph showing solitary swelling present in the left middle third of the face

There was no family history with similar findings. The general physical examination revealed a moderately built patient with satisfactory vital signs. On extra- oral examination during inspection: Diffuse swelling extending superoinferiorly from 2 cm below the infraorbital rim to the zygomatic buttresses and mediolaterally from 1.5 cm from ala of nose to 3 cm from tragus of ear.was noticed. Oral examination revealed the presence of unilateral symmetrical expansion in the distal part of the alveolar ridge of left upper jaw(fig.2).

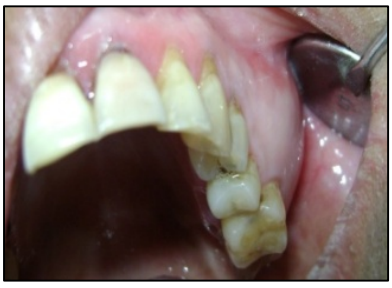

Fig.2: intraoral photograph showing expansion in the left maxilla

The covering and surrounding mucosa was normal in color,. Swelling was hard in consistency, non-tender with smooth texture. No bleeding on palpation was present. Swelling was non-fluctuant & non pulsatile. OPG and waters view shows gross radio-opacity in the maxillary bone from 2nd premolar region to tuberosity region with slight ground glass appearance (fig3,4).

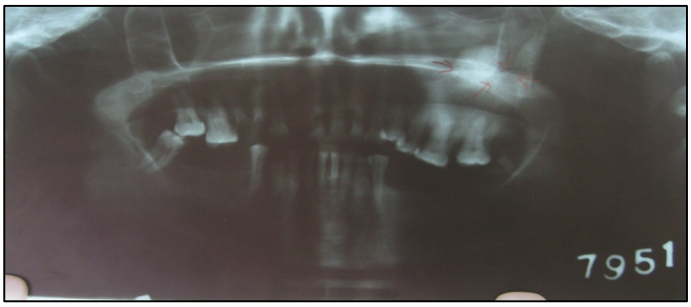

Figure-3: OPG shows diffuse radiopacity in left maxilla

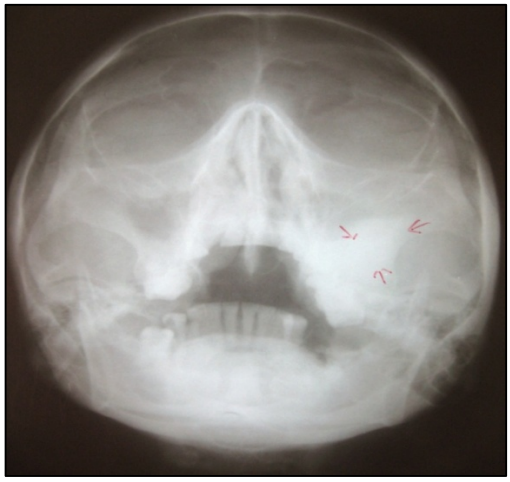

Figure-4: PNS view shows diffuse radiopacity in left maxilla

SURGICAL PROCEDURE

Transnasal intubation was done and a crevicular incision along gingival sulcus was made extending from lateral incisor to tuberosity. A vertical releasing incision was made i.r.t. canine and full thickness mucoperiosteal flap was elevated exposing the buccal cortical plate of the maxillary bone.(fig.5)

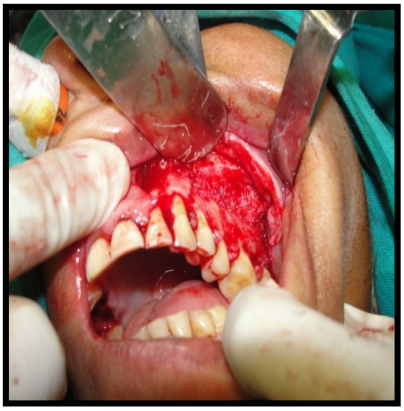

Fig. 5- exposure of lesion

Using osteotome a large part of buccal cortical plate was removed from canine to tuberosity region & bone re-countered with large flame shaped tungsten carbide bur.(fig.6,7) Wound was closed in one layer using interdental sutures. Post-operative period was uneventful.

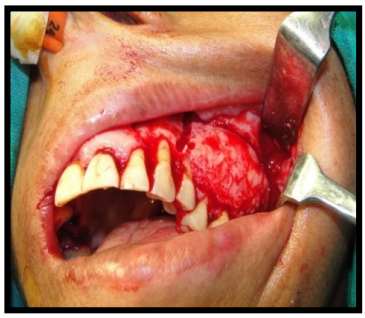

Fig.6. Removal of buccal cortical plate and recontouring

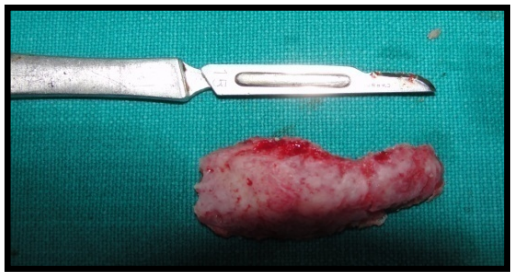

Fig.7- osteotomized segment

Histopathological examination reveals numerous trabecular of course of woven bone of irregular in shape extending into fibrous tissue having white osteoid seams suggesting of Monostotic Fibrous Dysplasia with left maxilla(fig 8).

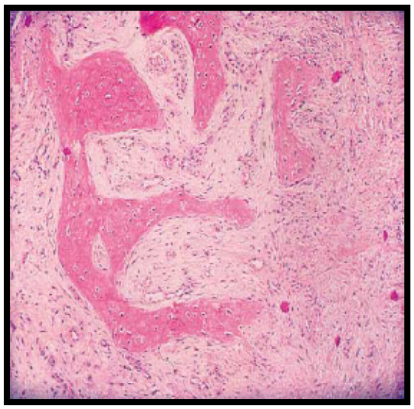

Fig.8- histopathological photograph

DISCUSSION

It is a benign bone disorder of an unknown etiology, uncertain pathogenesis and diverse histopathology.

Fibrous dysplasia has 4 different disease patterns

Monostotic (70%),

Polyostotic (30%),

Craniofacial form

The incidence of FD in 45 yrs as seen in this case can be explained by the fact that age of onset and symptoms vary considerably from patient to patient & the onset is often so gradual that the patients seek medical attention only when they become aware of symptoms or signs, causing a late diagnosis. The exact etiology of FD is unknown.

Another possible explaination can be related to endocrinal disturbances. It has been seen that hormonal changes, can reactivate a dormant lesion. Thus, in this case as the patient was approaching her menopausal age, changes in hormones might have reactivated the previously unnoticed lesion. The choice of surgical option depends on several factors: site of involvement, rate of growth, aesthetic

disturbance, functional disruption, patient preference, general health of the patient & surgeon's experience. Surgical Protocol for management as given by Chen YR et al

Zone 1- includes the fronto-orbital, zygomatic and upper maxillary regions;

Zone 2- represents the hair-bearing cranium;

Zone 3- is the central cranial base;

Zone 4- includes the teeth-bearing regions of the maxillary alveolus and mandible.

For lesions in zone 1- total excision of the dysplastic bone is recommended. For lesions in zones 2, 3 and 4, conservative excision or shaving has been proposed.[9]

Medical treatment has a role in the management of FD. The use of pamidronate, which inhibits the resorptive activity of osteoclasts may be an alternative to surgical interventiont.[10] unfortunately there are no objective methods to assess or predict the outcome of treatment, especially medical treatment.

CONCLUSION

FD is a benign disease that has the potential to cause significant cosmetic and functional disturbance. Surgical intervention in patient's with fibrous dysplasia is required to correct functional impairments or significant cosmetic deformities.

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