Case Report

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Craniofacial Fibrous Dysplasia: A Case Report with Review of Literature

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ABSTRACT

Background: Fibrous dysplasia is a benign self- limiting non-encapsulated bone tumor usually affecting the long bones and craniofacial skeleton. It is characterized by replacement of normal medullary bone by cellular fibrous tissues .There is arrest of bone development in the woven stage with failure to mature into lamellar bone. Thus, bone is structurally inadequate with decreased mechanical quality hence prone to fracture. In the weight bearing bones the main concern is pathologic fractures. However, in the craniofacial skeleton it is the gross asymmetry and disfigurement of the face which is of primary concern to the patient and challenging to the operating surgeon. In this case a debulking procedure with surgical recontouring of zygoma and maxilla was done to create a facial balance.

Keywords: Fibrous dysplasia, De-bulking, Surgical re-contouring.

INTRODUCTION

The onset of fibrous dysplasia occurs in the 1st/2nd decade of life[1].Mono-ostotic variety is 80% more common than polyostotic. In the craniofacial region fibrous dysplasias are more commonly found in the maxilla than mandible[2]. Maxillary lesions may extend to the zygoma. maxillary sinus, floor of the orbit such lesions are typically called craniofacial fibrous dysplasias[3].Proposed pathogenesis is a mutant gene which is a cell surface protein that codes for Gs alpha on chromosome 20[4].Another hypothesis suggests that it is a focal bone expression of a complicated endocrine disturbance[5].Diagnostic aids like CT,MRI, CBCT, bone scan, bone biopsy along with routine radiographs are useful for diagnosis and treatment planning .Classic presentation is a Ground Glass appearance, important fact is that it blends imperceptibly into the surrounding normal bone without a circumscribed border[2]. Histologically, fibrous dysplasia shows fibroblastic tissue with rich cellular whorled pattern with little bone which typically presents as a Chinese Character

CASE HISTORY

A twenty-two-year-old girl reported to the department of oral and maxillofacial surgery with the concern of facial asymmetry and desired cosmetic correction of the same. History revealed that mild asymmetry was noticed by the parents when the patient was about 6yrs of age and it progressed later, they also noticed an accelerated asymmetry around 11 to 15 yrs of age after which it was static. Since the patient was to get married she wanted a cosmetic correction. On extra oral examination there was an obvious facial asymmetry with hardy bony swelling over the right maxilla and zygoma. Inta-orally there was buccal expansion with obliteration of buccal sulcus, overlying mucosa was healthy with no ulceration and no caries or periodontal involvement of teeth was noted. OPG

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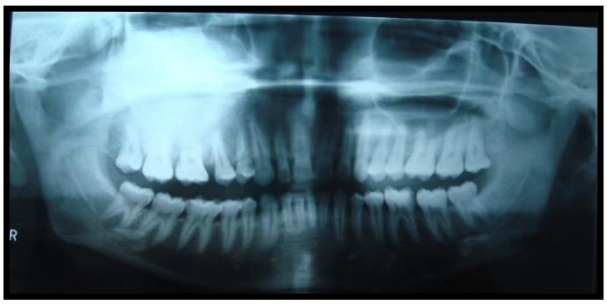

Fig 1: Preoperative OPG showing ground glass appearance ,characteristic of fibrous dysplasia on the right side.

Fig 2: Preoperative CBCT showing fibrous dysplasia on the right side.

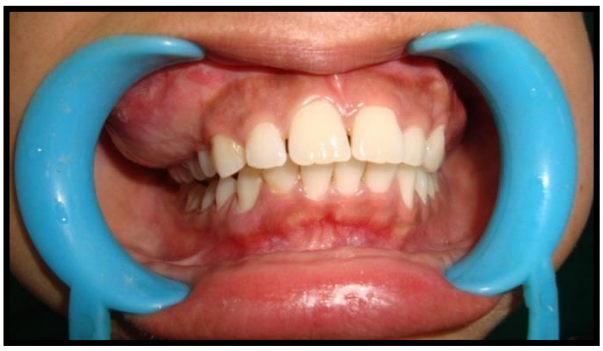

Fig 3: Preoperative intraoral photo showing bony swelling on the right side.

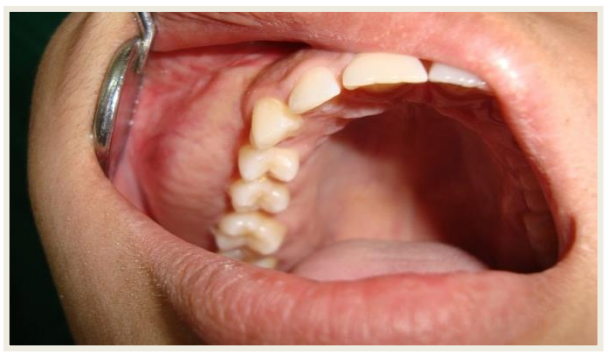

Fig 4: Preoperative intraoral photo showing bony swelling on the right side.

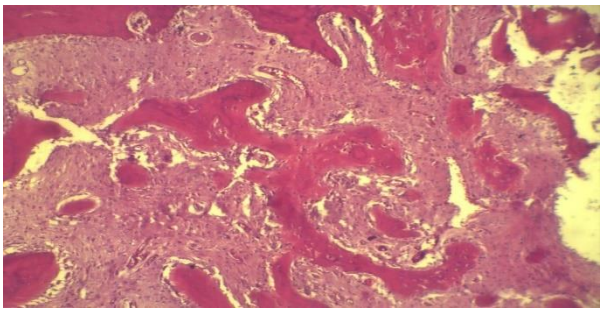

Fig 5: Histpathologic examination of the excised specimen showing characteristic" Chinese letter patter n"of fibrous dysplasia-10X.

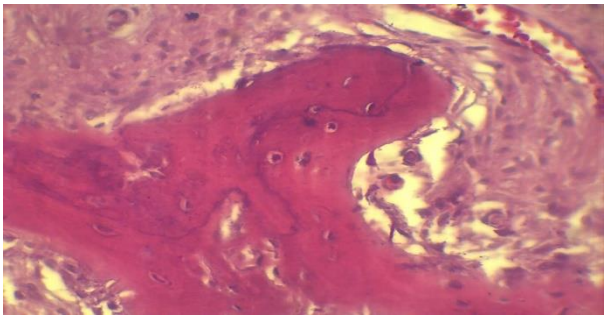

Fig 6: Histpathologic examination of the excised specimen Characteristic "Chinese letter pattern" seen showing bony trabeculae -40X

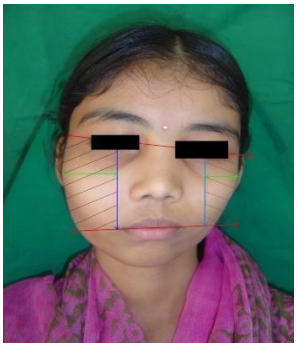

Fig 7: Preoperative extra- oral, frontal view.

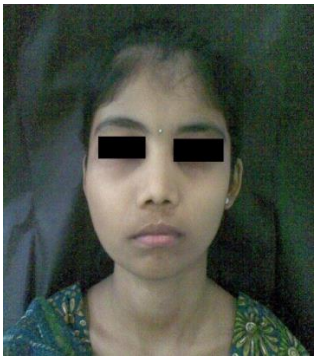

Fig 8: Postoperative extra- oral, frontal view

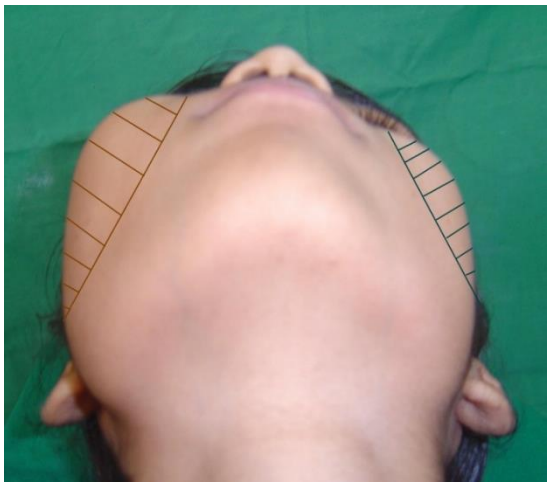

Fig 9: Preoperative extra- oral ,worms view.

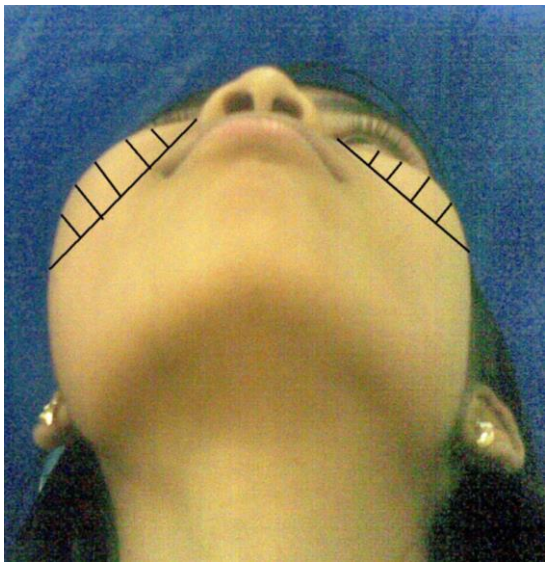

Fig 10: Postoperative extra- oral ,worms view

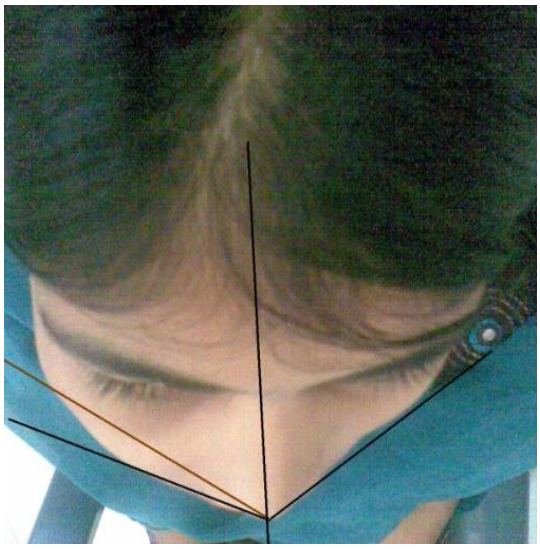

Fig 11: Preoperative extra oral ,birds view

Fig 12: Postoperative extra- oral, birds view

revealed a typical 'Ground Glass' appearance with obliteration of maxillary sinus. CT revealed peaud'orange appearance with maxillary sinus 3-D CT showed the involvement of the zygoma as well and revealed the obvious facial deformity. A bone biopsy was done which confirmed that the lesion was fibrous dysplasia. Since facial asymmetry was the primary concern clinically as well as from the patient point of view. De-bulking and surgical re-contouring was decided under G.A through an intraoral approach .No abnormal bleeding was encountered during the surgical procedure although there was excessive oozing as is expected in such cases .Blood was not transfused .The recovery was uneventful .

DISCUSSION

Lichenstein and Jaffe in 1942 coined the term fibrous dysplasia. In this disease since bone development ceases at woven stage mature lamellar bone is not formed such immature bone has poor mechanical quality and is prone to fracture. However, in the craniofacial region it is the facial asymmetry which is of major concern. There are two types of fibrous dysplasias, mono-ostotic and polyostotic. In the craniofacial region fibrous dysplasias are more commonly found in the maxilla than mandible and 80% are of mono-ostotic variety. Mc-Cune Albright syndrome is an example of polyostotic fibrous dysplasia with Café-au-lait spots and endocrine disturbances like hyperthyroidism, precocious puberty etc[6]. Current medical

management of fibrous dysplasia includes the use of Bisphosphonates with calcium and V it D which is based on the rationale that Biophosphonates decrease trabecular breakdown and increase bone mass[7,8,9,10,11].They have been the mainstay in medical management of fibrous dysplasias. Pamidronate(2nd generation bisphosphonate) intravenous 60mg/day i.v. for 3 days every 6 months combined with calcium 500-1500mg/day &Vit. D 800-1200 IU/day appear to be useful for reducing bone pain[8,9]. In craniofacial fibrous dysplasia's, debulking is the primary treatment which is not aimed at removing the dysplastic bone completely but re-contouring the bone to an acceptable contour. In most cases dysplasia stabilizes and stops enlarging with maturation of facial skeleton[12].There is some role for biochemical markers like Serum alkaline phosphatase and Urinary hydroxyproline to monitor bone turnover in the management of FD[13,14]. Considering the sclerotic and avascular nature of mature dysplasia osteomyelitis has also been reported. Osteosarcoma probably due to radiation has been reported in 0.4% cases and carries poor prognosis[1516,17]. Blindness or reduced visual acuity have also been reported due to the possible decrease in the diameter of the optic canal by dysplastic bone in which nerve decompression becomes necessary[18]. In this case report bone shaving and re-contouring the maxilla and zygoma was done purely to enhance and improve the aesthetics and give a balance to the face, going by the history of the patient around pre-puberty and at puberty there was bone growth on the affected side which resulted in facial asymmetry. Since the deformity had already established and the patient was in post pubertal stage without any bone pain, medical line of treatment was not considered and surgical intervention was done. There is a three year follow-up of the patient.

CONFLICT OF INTEREST

No potential conflict of interest relevant to this article was reported.

REFERENCES

1) Waldron C A, Giansanti J S: Benign Fibro-osseous lesions of the jaws. Part I:Oral Surg Oral Med Oral Path 1973:35:190-210.

2) Daffner RH, Kirks DR, Gehweiler JA, Heaston DK. Computed tomography of fibrous dysplasia. Am J Roentgenol 1982; 139: 943–948.

3) Camilleri A E: Craniofacial fibrous dysplasias. J LaryngolOtol 1991:105:662-666

4) Marie P J, de Pollak C, Chanson P, Lomri A. Increased proliferation of osteoblastic cells expressing the activity of Gs alpha mutation in mono-ostotic fibrous dysplasia. A J Pathol 1997:150:1059-1069

5) Shenker A, Weinstein LS, Sweet DE, Spiegel A M: An activating Gs alpha mutataion is present in fibrous dysplasia of the bone in the Mc-Cune Albright Syndrome. J Clin Endocrinol Metab 1994;79:750-755

6) Chapurlat R D,Orcel P. Fibrous dysplasia of bone and McCune–Albright syndrome

7) Best Practice & Research. Clinical Rheumatology. Vol. 22, No. 1, pp. 55–69, 2008

8) Liens D, Delmas PD & Meunier PJ. Long-term effects of intravenous pamidronate in fibrous dysplasia ofbone. Lancet 1994; 343: 953–954

9) Dimeglio LA, PediatrEndocrinol Rev. Bisphosphonates therapy for fibrous dysplasia.2007 Aug;4Suppl 4:440-5.

10) Chapurlat RD. J Bone Miner Res. 2006 Dec;21 Suppl 2:114-119.

11) Barnes L, Peel RL, Verbin RS, Goodman MA, Appel BN. Diseases of the bones and joints. In: Barnes L, editor. Surgical pathology of the head and neck. New York: Marcel Dekker; 1985. 883–1044.

12) Barnes L, Peel RL, Verbin RS, Goodman MA, Appel BN. Diseases of the bones and joints. In: Barnes L, editor. Surgical pathology of the head and neck. New York: Marcel Dekker; 1985. 883–1044.

13) Kos M, Luczak K Godzinski J, Klempous J. Treatment of monostotic fibrous dysplasia with pamidronate. J Cranio maxillofac Surg. 2004;32:10-5.

14) Plotkin H, Rauch F, Zeitlin L, Munns C, Travers R, Glorieux FH. Effect of pamidronate treatment in children with polyostotic fibrous dysplasia of bone. J Clin Endocrinol Metab 2003;88:4569-75.

15) Petrikowshi C T, Pharoh M J, Lee L, Grac MG. Radiographic differences of osteosarcoma, myelitic fibrous dysplasia of the jaws Oral Surg Oral Med Oral Path Oral Rad 1995:80:744-750

16) Schwartz DT & Alpert M. The malignant transformation of fibrous dysplasia. The American Journal of the Medical Sciences 1964; 247: 1–20.

17) Yabut SM, Kenan S, Sissons HA & Lewis MM. Malignant transformation of fibrous dysplasia: a case report and review of the literature. Clinical Orthopaedics and Related Research 1988; 228: 281–289.

18) Lee JS, Fitzgibbon E, Butman JA et al. Normal vision despite narrowing of the optic canal in fibrous dysplasia. The New England Journal of Medicine 2002; 347: 1670–1676.