Case Report

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GROUND GLASS ON THE FLOOR: A RARE CASE PRESENTATION OF FIBROUS DYSPLASIA

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ABSTRACT

Fibrous dysplasia (FD) is a benign dysplastic process of distorted osteogenesis frequently affecting the jawbones. It is characterized by replacement of normal bone by an excessive proliferation of cellular fibrous connective tissue intermixed with irregular curvilinear trabeculae of woven bone. Occurring most commonly in the second decade of life, with preponderance for maxillary bone we discuss an unusual case of fibrous dysplasia in the mandible. A 24-year-old female presented with a swelling on the lingual aspect of the mandible extending into the floor of the mouth. Radiological findings of a well-circumscribed radio-opacity were suggestive of ossifying fibroma but the operative findings and histopathological examination confirmed the diagnosis of Fibrous dysplasia.

KEYWORDS: fibrous dysplasia, monostotic, floor of the mouth

INTRODUCTION

Fibrous dysplasia (FD) is a benign dysplastic process of distorted osteogenesis frequently affecting the jawbones that can cause severe deformity, asymmetry and, most devastating of all, blindness.[1]It may involve a single bone (monostotic) or multiple bones (polyostotic).[2] It is a developmental tumour like condition that is characterized by replacement of normal bone by an excessive proliferation of cellular fibrous connective tissue intermixed with irregular curvilinear trabeculae of woven bone.[1] Clinically, the maxilla is affected more commonly than the mandible and lesions are detected in the late first and early second decades without any gender or racial predilection, although recent researches have shown a slight female preponderance.[3] The disease is characterized by painless osseous enlargement with facial asymmetry. The radiographic image of FD varies with the stage of development and amount of bony matrix within the lesion.Early lesions may appear well defined and radiolucent, whereas later lesions may become largely sclerotic. However, classic FD has a ground-glass or orange-peel appearance, with poorly distinguishable borders that appear to blend in with the surrounding, unaffected bone.[1] We report an unusual case of monostotic FD involving the mandible in a 24-year-old female.

CASE REPORT

A 24 year old female patient reported to our outpatient department, with the chief complaint of swelling in lower left back tooth region that had persisted for 1 year. The patient gave a history of a slowly increasing swelling on the lingual aspect of left mandibular region. There was no history of pain or trauma associated with the swelling. Clinical examination revealed a solitary well defined bony hard swelling, oval in shape, measuringapproximately 5cm × 7 cm in dimension, extending anterio-posteriorly from lingual aspect of the right mandibular canine upto

the mesial aspect of the left mandibular second molar and superio-inferiorly from 0.5 cm above the occlusal aspect of the mandibular anterior teeth upto the floor of the mouth. The firm swelling was limited to middle of the floor of the mouth in its medial extent. [Fig I(a),(b)] Occlusal radiograph showed well defined radio opaque lesion in relation to lingual surface of mandible on left side with loss of normal trabecular pattern. [Fig I (c)] Based on the clinical history and radiographic features of the lesion, a benign bone tumor of a slow-growing nature was suspected first. An incisional biopsy was subsequently performed, providing an inconclusive diagnosis of benign fibro-osseous lesion. The complete surgical excision of the mass was done under general anesthesia. A crevicular incision was given extending from distal aspect of lower right 2^{nd} premolar to the distal aspect lower left 2^{nd} molar with short vertical incisions on the right and left side, keeping in mind the presence of lingual vessels and mylohyoid ridge and muscle. The Trapezoidal flap aided in complete reflection on the soft tissue and good access to the mass. The lesion was excised in multiple bits with the help of chisel and mallet. All the osseous spicules present after the removal of the lesion were trimmed of using a contra angle hand piece with round bur No.6 and No.8. Closure was done using interrupted suture technique with absorbable sutures (3-0,Vicryl *Ethicon**) placed at the interdental papillae. All excised bits comprising of both soft and hard tissues were sent for histopathological examination. The gross examinationof the hard tissue specimens revealed firm, grey-white bits with the largest bit measuring 5cm× 1cm × 0.5 cm in size. The masses were surrounded by thin bony cortex like encircling shell. The soft tissue specimen was creamish brown in colour and measured 1 cm × 0.5 cm × 0.5 cm in size. [Fig II (a)]. Histologic analysis of hematoxylin and eosin–stained sections showed disorganised irregularly shaped immature bony trabeculae lacking osteoblastic rimming in a fibrocellular stroma without any mitotic activity. The configuration of these bony trabeculae were observed and resembled Chinese characters. Bony trabeculae showing osteoblastic rimming and basophilic reversal lines were also observed in few focal areas. Lesional bone merging imperceptibly with adjacent normal bone was another evident finding, confirming the diagnosis of Fibrous Dysplasia. [Fig II(b)]

DISCUSSION

Fibrous dysplasia (FD) is a benign skeletal disorder, first described by Lichtenstein in 1938

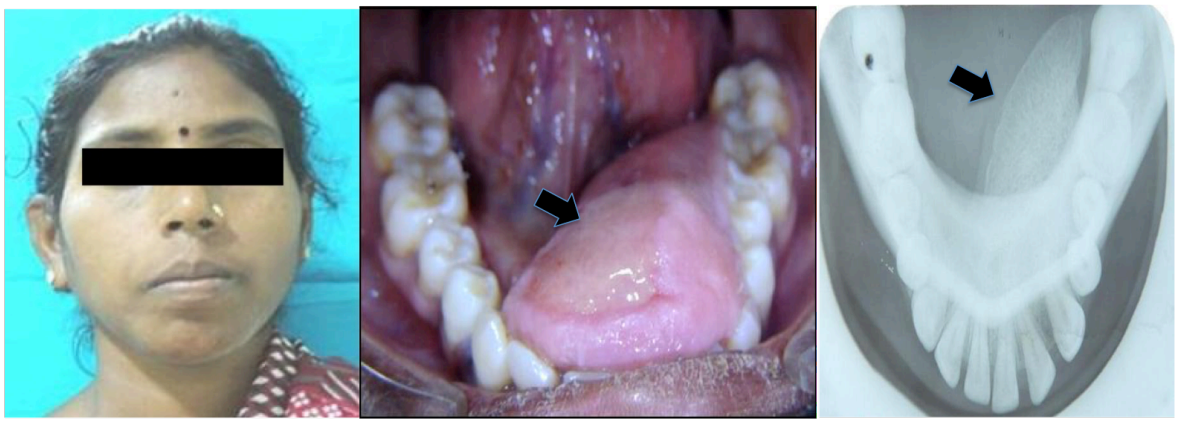

Figure 1 :I (a)Extraoral picture depicting no facial Asymmetry, (b) Intraoral picture showing the swelling on lingual aspect of the mandible extending into the floor of the mouth, (c) occlusal radiograph showing the radio-opacity on the lingual aspect- black arrow

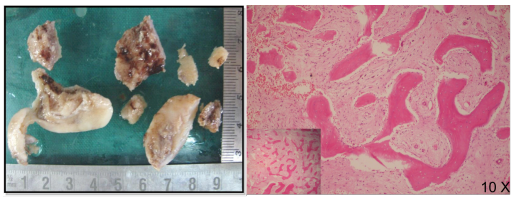

Fig II (a) gross examination showing multiple bits of hard and soft tissue received, (b) H & E stained section of the specimen showing chinese letter shaped immature bone trabeculae in a band of fibrocellular stroma-4 X and 10 X.

and Lichtenstein and Jaffe in 1942.The term was earlier described as osteitis fibrosa disseminate.[4] The mutation at the GNAS gene encoding for the α -subunit of a signal transducing G protein is regarded as the etiologic cause of FD. This mutation triggers a cascade of the enzymatic reactions and causes incomplete differentiation of marrow stromal cells into abnormal osteoblasts which results in a fibrotic bone matrix.[5] This genetically based sporadic disease of bone may present as monostotic form in 70–80% of cases and polyostotic in 20–30% of cases.[6] But the incidence of craniofacial involvement in fibrous dysplasia has been reported as 10–25% in the monostotic form and 50–90% in polyostotic form. The polyostotic form is often associated with endocrinopathies manifesting as a part of Jaffe Lichenstein , McCune Albright , Mazabraud Syndrome.[2,3] The monostotic forms restricted to the bones of the cranio-facial skeleton are usually classified as 'craniofacial fibrous dysplasia' because of the suture crossing character of the pathology in this region.[2] Monostotic fibrous dysplasia is characterized by a swelling resulting from a poorly delimited area of fibro-osseous proliferation. Though less serious than polyostotic fibrous dysplasia, it is of greater concern to the dentist because of the frequency in which the jaws are affected. Presentation is most often during the 2nd and 3rd decades with bulk of cases diagnosed before the age of 30 but cases can be detected throughout adult life. The mean age of occurrence in the 69 patients reported by Zimmerman and his associates was 27 years, while in the 53 patients with craniofacial fibrous dysplasia reported by Gardner and Halpert, the mean age was 34 years.[7] Observed mean age at diagnosis ranges from 25 to 28 years in large series and 24 years in a systematic review.[8] The sexes are equally affected in monostotic disease but with a female predilection reported in more generalized forms of disease. The maxilla or mandible may be involved but a predominance of the maxilla has been documented.[9] In the case presented, the patient was a 24 year old female with a monostotic lesion involving the lingual aspect of mandible, contrary to the reviewed literature is a rare occurrence. The radiographic appearance of fibrous dysplasia differs with the stage of development and ossification. It is described as poorly defined lesion that merges with adjacent bone and radiolucent at initial stages which become increasingly radiopaque finally changing to largely sclerotic matured forms. Classically fibrous dysplasia is characterised by ground-glass or orange-peel appearance which blends with normal surrounding bone.[3] The lesion in our patient presented as well defined radio-opacity, correlating with the stage of maturating calcifications in a fibrous stroma. The typical histologic features are delicate and irregularly shaped trabeculae of immature bone with no osteoblastic rimming, enmeshed within a fibrous stroma giving a "Chinese letter" pattern.In older or mature lesions there may be lamellar bone with mature trabeculae arranged in elongated parallel fashion.[3] In our case numerous bony trabeculae arranged in chinese letter pattern with fibrocellular and cytologically uniform connective tissue stroma and absence of osteoblastic rimming were seen which confirmed the diagnosis. Another challenge diagnosing the case was differentiating it from ossifying fibroma because of the overlap between fibrous dysplasia and ossifying fibroma at a radiological and histological level.[2] But distinguishing FD from ossifying fibroma is important since surgical treatment of the two lesions is different. Despite the overlap in radiological feature of a well defined radio-opacity, the lesion was excised in multiple bits and could not be separated easily from the surrounding bone.This characteristic feature of fibrous dysplasia where, the lesional bone merged imperceptibly with cortex helped us distinguish it from ossifying fibroma.[3] The existing guidelines for treatment of fibrous dysplasia aren't universally accepted. Lesions that are not symptomatic, do not progress and do not cause deformities or functional impairment should simply be monitored. Surgery is indicated

for confirmatory biopsy, correction of a deformity, prevention of pathological fracture and/or eradication of symptomatic lesions.Radiation should not be employed as there is a risk of sarcomatous transformation of 0.4% that increases to over 40% following radiotherapy.[10]

CONCLUSION

It is important to integrate all the clinical, radiographic and pathologic features to diagnose monostotic FD, despite its unusual location in the lingual aspect of the mandibular bone.

CONSENT

Written informed consent was acquired from the patient for publication of this case report and any accompanying images.

CONFLICT OF INTEREST & SOURCE OF FUNDING

The author declares that there is no source of funding and there is no conflict of interest among all authors.

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