Case Report

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Fibrous Dysplasia of Maxilla: A Case Report

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ABSTRACT

Background: Fibrous Dysplasia(FD) is a benign bone lesion characterized by replacement of normal bone by fibrous connective tissue, and its diagnosis is based on clinical, radiological and histological findings. FD is commonly seen in children and by adulthood it usually becomes quiescent. Here a case of FD in right maxilla is reported in 12 year old female patient.

Keywords: Fibrous Dysplasia, fibro osseous lesions.

INTRODUCTION

Fibrous dysplasia (FD) is a benign disorder of bone caused by post zygotic, activating mutations of the GNAS gene that results in inhibition of the differentiation and proliferation of bone forming stromal cells and lead to the replacement of normal bone and marrow by fibrous tissue and woven bone.(1)There are two primary categories of the disease: monostotic FD, that involves only one bone and represents 70% of cases. In polyostotic form, multiple bones are affected ,can be subdivided in three types: craniofacial, in which only the bones of craniofacial complex are involved ,Lichtenstein – Jaffe, which has similar involvement of multiple bones of skeleton but has in addition "café-au-lait" spots in the skin; Albright syndrome, characterized by involvement of multiple bones, "café-au-lait" spot and endocrine impairment .(1,2).In addition to these forms, hereditary familial form of localized FD called cherubism is described.(3)

The clinical findings are asymptomatic ,involved bone enlargement causes facial asymmetry, loss of teeth and facial deformity.(4,5) If craniomaxillofacial bones are affected by FD, due to megacranium, the face of the patient is referred to lion face.(4)The complications of the lesions involving sphenoid, orbital, frontal bones are proptosis, visual disturbance, facial asymmetry and orbital dystopia. The fifth nerve impairment, hearing loss and seizure disorders have been reported as neurological complications.(6)

In this case report, we aim to document clinical, radiological and histopathological features of a patient with monostotic fibrous dysplasia in right maxilla.

CASE REPORT

A 12 year old female patient was referred to AMC dental college with chief complaint of swelling in right anterior maxilla since 2 years. Intraoral examination revealed expansion of alveolar crest in right maxillary canine to molar area. There was no history of loosening of teeth. The swelling was non-tender and hard in consistency with normal overlying mucosa. The swelling measured approximately 3x2 cm in size and was roughy oval in shape. Differential diagnosis of this bony hard swelling was considered ossifying fibroma, osteoma, paget's disease and cement osseous dysplasia.

Occlusal view of radiograph of right maxilla revealed expansion of buccal cortical plates with increased trabeculation making lesion opaquer and more mottled in appearance. An incisional biopsy was performed which microscopically revealed a fibrous connective tissue containing slender, irregular trabecula of woven bone with Chinese letter configuration suggestive of Fibrous dysplasia.

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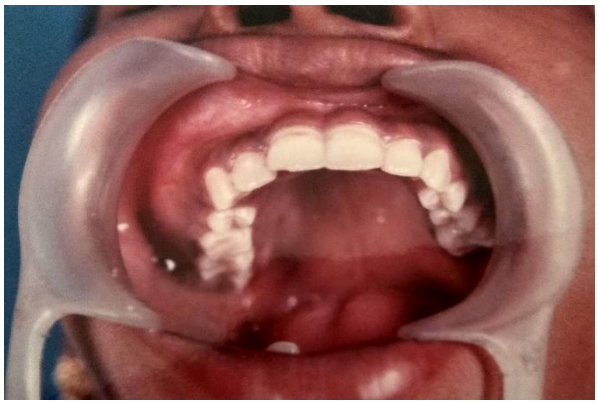

Fig 1: Intraoral view showing bulging of the right maxillary bone involving buccal cortical plate.

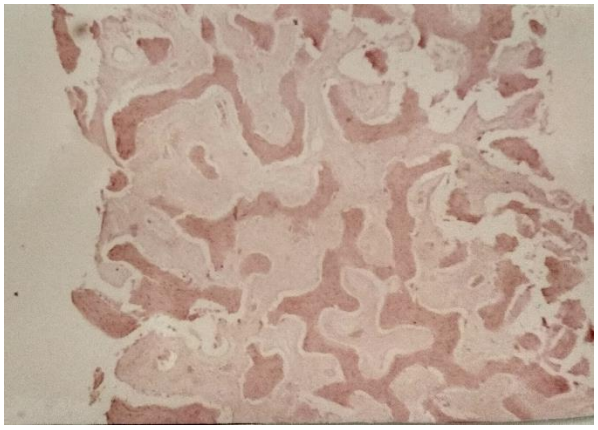

Fig 2: H & E stained section shows irregular trabeculae of woven bone of Chinese character, shape in background fibrous stroma.

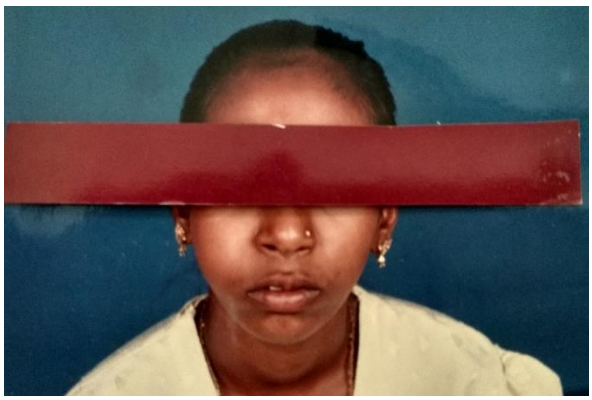

Fig 3: Extra oral view showing swelling in right maxillary region.

Blood and biochemical investigations were done which showed alkaline phosphatase, serum calcium and serum phosphorus within normal range. Based on history, clinical, radiological and histopathological examination and investigations the case was diagnosed as fibrous dysplasia of right maxilla. Since there are no symptoms and evidence of progression, mere presence of FD in craniofacial region itself is not indication for treatment. Hence treatment was deferred for this patient and is under regular follow up.

DISCUSSION

The fibro osseous lesions(FOL) are characterized by replacement of normal bone tissue by tissue containing a mineralized newly formed material called osteoid due to the similarity to normal bone. The FOLs are classified as cement-osseous dysplasia, fibrous dysplasia, ossifying fibroma and paget's disease. The differential diagnosis between these lesions are based on clinical, radiographic and histopathological examination.(1)Generally younger age of onset and its unilateral distribution allows FD to be readily differentiated from Paget's disease which affects older patients and is frequently bilateral(7)

The main differential diagnosis should be made in relation to cemento-ossifyi ng fibroma presenting similar histologic characteristic of FD. Nevertheless, the first one presents a radiographic image encapsulated with radiolucent halo delimiting the lesion unlike FD.(8) The FD can be divided in to three variants: monoostotic, polyostotic and craniofacial. Despite that, some authors suggest five subgroups, as presented: Group I-monostotic; Group II-polyostotic; Group III –polyostotic associated with Jaffe syndrome; Group IV-polyostotic associated with McCune Albright syndrome; and Group V-craniofacial limited to maxillofacial complex.(9)

The clinical consequences of the disease will depend on when and at what age of cell proliferation mutation occurred. Alteration during embryonic development will lead to polyostotic FD McCune Albright syndrome whereas monostotic FD happens due to mutation in postnatal life.(10)

FD has a predilection to arise in women in the first two decades of life and stabilizes after puberty.(1)In contrast, other authors stated that the monostotic form shows an equal gender distribution.(11)

The sign and symptoms observed in the case presented are corroborant with those described in

the literature, such as: predilection for women, in second decade of life, increased volume without ulceration, hard on palpation, asympatomatic, unilateral, located in maxilla without involvement of adjacent bones.

CONCLUSION

Lesions with unilateral maxillary involvement and ground glass radiographic appearance should signify FD to clinicians. Moreover, 50-90% of craniofacial FDs are polyostotic ,patient presenting with even single bone involvement must be assessed to identify the involvement of other bones or for the presence of endocrine disorders.

CONFLICT OF INTEREST

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

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