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Research Article

MONOSTOTIC FIBROUS DYSPLASIA AFFECTING MANDIBLE: A RARE CASE REPORT WITH INSIGHT ON ITS PATHOGENESIS

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A B S T R A C T

Fibrous dysplasia is a non-neoplastic skeletal disorder of unknown etiology resulting in defect in bone maturation. It generally has affinity for maxilla over mandible in craniofacial presentation. Hence we present you a case of fibrous dysplasia in a 19 year old female patient affecting the mandible with emphasis on clinical, radiological, gross pathology and histological presentation.

INTRODUCTION

Fibro-osseous lesions are group of disease characterized by replacement of normal bone by fibrous tissue containing a newly formed mineralized substance.[1] Waldron classied fibro-osseous lesions as (I) fibrous dysplasia, (II) Reactive dysplastic lesions (Cemento osseous dysplasia), (III) Fibro-osseous neoplasm (cement-ossifying fibroma).[2] Fibrous dysplasia may be divided into three categories: Monostotic - involvement of a single bone (74%), Polyostotic - involvement of more than one bone (13%) and Craniofacial (13%).[3] Monostotic fibrous dysplasia usually begin as bony swelling during childhood but usually undergoes arrest with maturation of the skeleton.[4]

Case Report

A 19 years old female patient presented with a chief complaint of painful hard swelling in the left lower side of the mandible for the past 2 years. Patient had undergone extraction of tooth (FDI.no.36) two years back. Extra oral examination revealed a bony hard swelling measuring about 3cm in diameter along the left lower border of the mandible. The swelling had grown slowly to present size. Swelling was firm in consistency and tender on palpation. Facial asymmetry was well appreciated and no other skeletal deformity was noted in the patient.(Fig.1-3)

Intraoral examination revealed no expansion of buccal and lingual cortical plates in relation to 36, 37 region which appeared relatively normal with no displacement of teeth.(Fig.4) Incisional biopsy was done a year ago for which a diagnosis of osteoma was made in a private hospital. But the lesion continued to grow and attained the present size, which urged the patient to approach us. Blood investigation revealed normal values of Serum alkaline phosphatase, calcium and serum phosphatae. Orthopantamogram revealed ground glass appearance with downward displacement of left side of mandible in molar region.(Fig.6) The lesion seems to merge with surrounding bone without any demarcation. CT-mandible revealed mild increase in size of left ramus and body of mandible with diffuse sclerotic appearance. Focal lytic area of about 1.3cm in size located inferior to 37 tooth, corresponding to the past biopsy site. (Fig.7-9)

Gross picture of the specimen revealed small fragments of bony spicules.(Fig.5) Histopathological examination showed immature bony trabeculae arranged predominantly in ovoid pattern and few areas showing cuneiform pattern.(Fig.10) Osteoblast rimming was absent in most areas.(Fig.11) Correlating clinical, radiological and histopathologic findings, diagnosis of fibrous dysplasia was confirmed.

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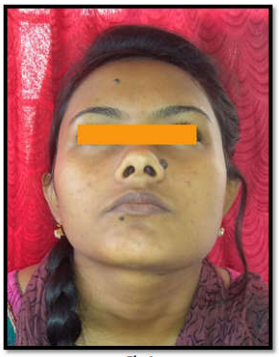

Fig.1

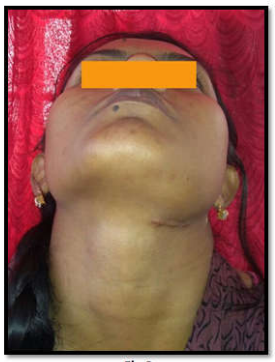

Fig.2

Fig.3

Fig.4

Fig (1,2,3) Extraoral picture showing extent of bony swelling in the left mandible.
Fig 4 Intraoral picture shows firm 37 tooth with relatively normal adjacent structures.

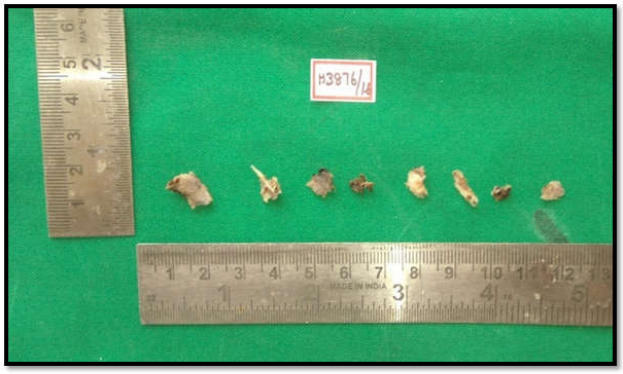

Fig. 5

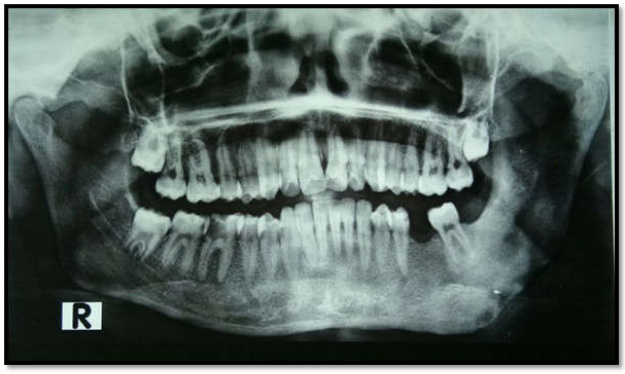

Fig.6

Fig 5 Gross picture showing tiny fragments of bone.
Fig 6 OPG shows radioapaque changes in relation to 37 along with bulging of lower border of left mandible.

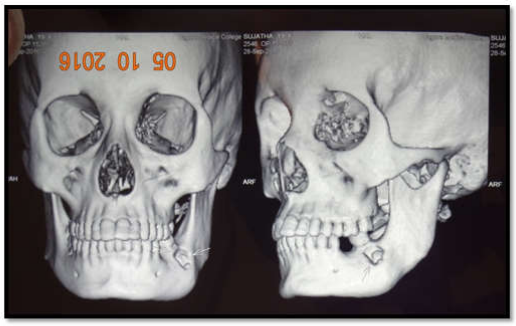

Fig.7

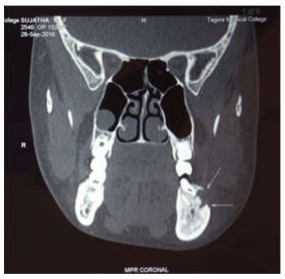

Fig.8

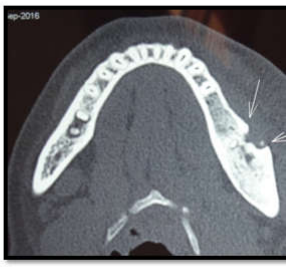

Fig.9

Fig 7,8,9 CT Scan- left ramus and body of mandible shows diffuse sclerotic appearancewith mild increased size (arrows – indicate focal lytic lesion of about 1.3 cms in size at the inferior aspect of left second molar area)

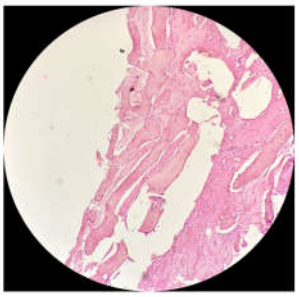

Fig.10

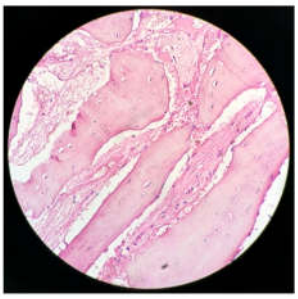

Fig.11

Fig 10(4x magnification)) histopathological picture shows immature bony trabeculae surrounded by fibrous stroma

Fig 11(40x magnification) histopathological picture shows bony trabeculae predominantly devoid of osteoblastic rimming.

DISCUSSION

Fibrous dysplasia is a fibro osseous lesion characterized by defect in osteoblastic differentiation and maturation caused by GNAS 1 gene mutation.[5] It is characterized by substitution of arginine (at codon 201) for cysteine or histidine resulting in the constitutive activation of the GSα-cyclic adenosine monophosphate (cAMP) signalling pathway with subsequent overproduction of cAMP by the affected cells. In bones, increased concentration of cAMP in cells of osteoblastic lineage leads to increased proliferation and abnormal differentiation of preosteoblasts, which explains their spindle cell morphology and failure to form a mature bone.[6]

Based on the number of bones involved, clinically fibrous dysplasia is usually typed as monostotic and polyostotic. In the case of monostotic type, the ribs, femur, tibia and maxilla are the bones most commonly involved.[7] Mandible was affected only in 10% of cases with pre-molar and molar region being frequently involved.[8]

Most cases are reported in first two decades of life with equal predilection for both males and females.[9] Asymptomatic unilateral slowly progressing bone expansion is the usual clinical presentation.[10] Bone expansion is usually fusiform type[11] with teeth staying firm.[4] In case of mandible, the expansion may cause bulging of lower border and displacement of mandibular canal superiorly.[12]

The characteristic radiographic finding of fibrous dysplasia is "ground-glass or orange peel appearance" with opacification, which is most identifiable on intraoral radiographs.[13] It is formed by superimposition of amyriad of poorly calcified bone trabeculae arranged in a disorganized pattern interspersed with areas of radiolucency.[14] Change of lamina dura to the abnormal bone pattern, and narrowing of the periodontal ligament space forms the primary distinguishing features of fibrous dysplasia. Lesions of craniofacial region are poorly defined and more radioapaque compared to that of axial skeleton.[11]

Most of the surgical tissue is obtained by curettage. The specimen has a distinct gritty feeling reflecting the osteoid trabeculae inherent in the lesion, which is specific for fibrous dyaplasia.[15] Blood investigation usually shows normal value of serum calcium, serum phosphorus and serum alkaline phosphatase.[16]

On histopathological examination, fibrous dysplasia typically demonstrates a monotonous pattern throughout the lesion rather than being a haphazard mixture of woven bone, lamellar bone, and spheroid particles.[1] The fine branching, curvilinear bony trabeculae of woven bone (which have been likened to Chinese script writing) show little evidence of osteoblast rimming.[17] Craniofacial lesion will mature over a period of time and demonstrate lamellar bone.[11] This is particularly true in specimens from older patients.[1] In these mature forms, the typical Chinese letter pattern of bony trabeculae may be lost and instead, elongated and parallel bony trabeculae will be prominent,[5] whereas in our case it is seen at very young age.

The primary differential diagnosis considered for fibrous dysplasia of the jaws is cemento-ossifying fibroma.[14] Others lesions considered are cemento-osseous dysplasia, chronic osteomyelitis, diffuse sclerosing osteomyelitis, Paget's disease and malignant tumors of bone.[12]

Fibrous dysplasia characteristically stabilizes, after puberty although a slow advance may be noted into adulthood.[1] Small lesions may require no treatment other than biopsy confirmation and periodic follow-up. Large lesions that have caused cosmetic or functional deformity may be treated by various methods like curettage and packing with cancellous bone grafts.[18] In case of mandible, the treatment is always directed towards correction of asymmetries and prevention of functional problem by conservative surgery, without considering the size of the lesion.[19]

CONCLUSIONS

Monostotic type of fibrous dysplasia affecting mandibular region are very rare, making clinician difficult to differentiate it from other bony lesions with similar presentations. Hence its prudent for a clinician to have proper knowledge about clinical appearance, radiological changes, gross pathology, histological features and laboratory findings of this disease and being a fibro-osseous lesion its diagnosis is must be based on conglomerate of above information.

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