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ABSTRACT

Fibro-osseous lesions of the craniofacial complex are represented by a variety of disease processes that are characterized by pathologic ossifications and calcifications in association with a hypercellular fibroblastic marrow element. They are a diverse group of processes that characterized by replacement of normal bone by fibrous tissue containing a newly formed mineralized product. The commonly included fibro-osseous lesions are fibrous dysplasia (FD), cemento-osseous dysplasia and ossifying fibroma. Ossifying fibroma is a true benign neoplasm, whereas Fibrous dysplasia is a genetic developmental anomaly of the bone-forming mesenchyme with a defect in osteoblastic differentiation and maturation that leads to a replacement of normal bony tissue by fibrous tissue of variable cellularity and immature woven bone. The main histologic differences between the two lesions lie in the absence of a capsule and the presence of more immature bone without osteoblastic activity in fibrous dysplasia. Cemento-osseous dysplasia is the most common fibro-osseous lesion affecting the jaw bones.

KEYWORDS: Bone, Fibrous dysplasia, Ossifying fibroma, Cemento-osseous dysplasia

INTRODUCTION

Fibro-osseous lesions (FOL) are a poorly defined group of lesions affecting the jaws and craniofacial bones. Fibro-osseous lesions (FOL) of the oral and maxillofacial region represent a rare, benign group of lesions that share similar clinical, radiological, and histopathological features.

They are characterized by progressive, variable replacement of healthy bone tissue in the jaw by fibrous connective tissue containing varying amounts of mineralized substances that include bone, osteoid, and cementum-like material.

All are characterized by the replacement of bone by cellular fibrous tissue containing foci of mineralization that vary in amount and appearance.

The concept of fibro-osseous lesions has evolved over the last several decades and now includes two major entities: fibrous dysplasia and ossifying fibroma.

The less common lesions include florid osseous dysplasia, periapical dysplasia, focal sclerosing osteomyelitis, proliferative periostitis of Garre, and osteitis deformans.

Fibrous dysplasia is a typically benign bone lesion characterized by intramedullary fibro-osseous proliferation secondary to altered osteogenesis.

First introduced by Lichtenstein and Jaffe in 1942 and originally termed Jaffe-Lichtenstein syndrome, fibrous dysplasia can occur in monostotic form (single bone) or polyostotic form (multiple bones).

The etiology of fibrous dysplasia has been linked with a missense mutation in the GNAS1 gene on chromosome 20. Abnormal proliferation of fibrous tissue ensues following an activating mutation.

Fibrous dysplasia has histologic elements of immature collagen and immature bone trabeculae forming a fibrocellular matrix. Trabeculae are not rimmed by osteoblasts secondary to osteoblast maturation arrest and histologic transition from normal to abnormal bone is usually abrupt.

Ossifying fibroma, which is considered a benign mesenchymal odontogenic tumour of the jaws, is a type of fibro-osseous lesion characterized by slow growth and proliferation of fibrous cellular stroma

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containing osteoid, bone or cementum-like tissue.

CASE REPORT

A 21 year old female patient name saroj sunita, residing at Ahmedabad, belonging to middle socio-economic class, came to the department of oral medicine and radiology with chief complaint of swelling in upper right back tooth region since 7-8 years. Patient was relatively asymptomatic before 7-8 years, then she noticed swelling in upper right back tooth region (in relation to 14, 15, 16, 17) since 8 years. Patient has no past medical and no past dental history. Patient has no history of habit.

EXTRAORAL

No abnormality detected.



INTRAORAL

INSPECTION and PALPATION

Dome shape swelling seen on upper right back tooth region

Swelling is non tender, hard in consistency, fixed to irt 14 15 16 17. Swelling starting from mesial side of 14 to 18 underlying mucosa.

Extending posteriorly up to distal side of 17 and superiorly

No associated discharge present. from maxillary vestibule, going inferiorly up to occlusal level.

Surrounding structure appears normal.

Displacement irt 15 16 seen.

Lymphnodes are non tender and non palpable.



PROVISIONAL DIAGNOSIS: FIBROUS DYSPLASIA

DIFFERENTIAL DIAGNOSIS

OSSIFYING FIBROMA

AMELOBLASTOMA

INVESTIGATION

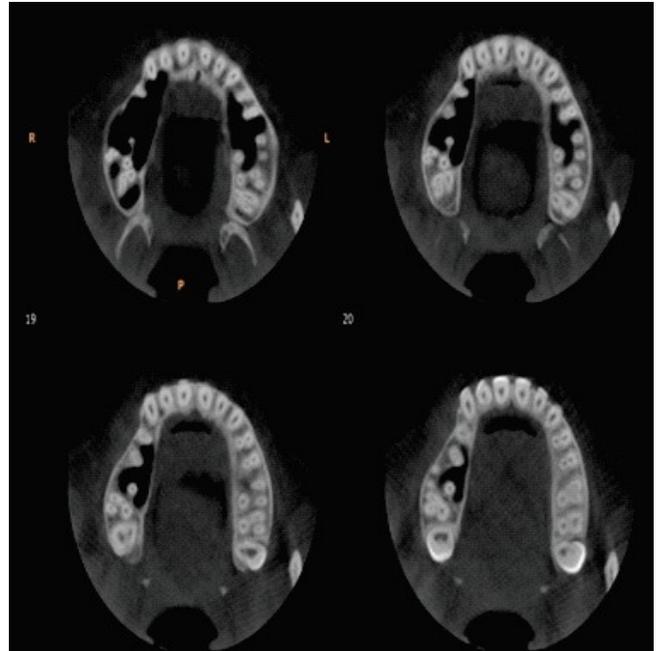
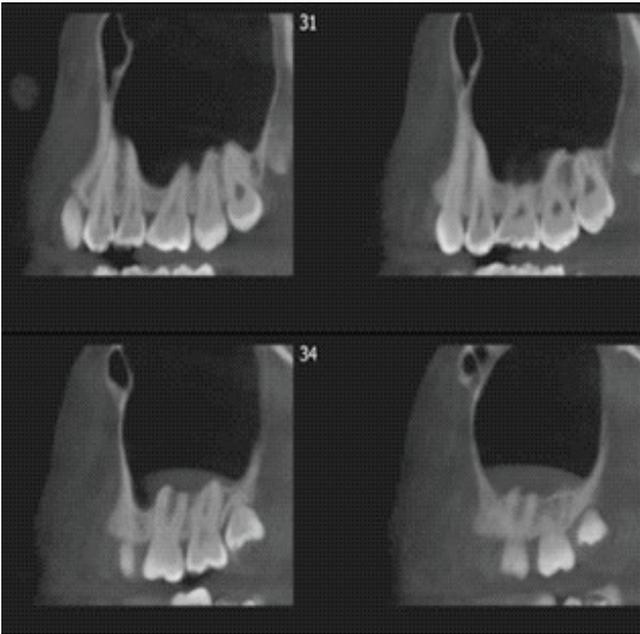
1. OPG
2. CBCT
3. Biopsy

1. OPG



An orthopantomograph showed normal condylar process, coronoid process, and temporomandibular joint. Maxillary sinus on right side appears larger as compare to left side. All teeth are present, and 18, 28, 38, 48 are impacted. Dilaceration seen irt mesial root of 38. Disto-angular teeth irt 18 and 28. Displacement of teeth seen irt 14, 15. Unilateral bony expansion with intact buccal cortex is seen in upper right tooth back region. A radiolucent lesion with irregular margin is seen in upper right premolar. Lesion extending from upper right canine region to 1st molar region.

2. CBCT



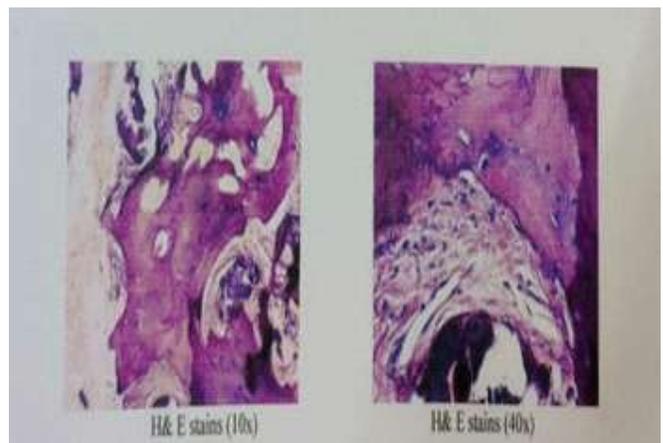
An axial view of maxilla CBCT shows mesial of 15 to distal of 17. Lingual cortical plate thinning seen in same region. Lesion appears to be mixed of radiolucency and radiopacity. Border is Poorly defined.

Radiopacity Seen in inferior 1/3^r of maxillary sinus. Maxillary Sinus appears larger in size

3. HISTOPATHOLOGY REPORT

Macroscopic Examination Microscopic Examination:

Given H&E stained section shows, fibrous tissue, calcification and mature lamellar bone without osteoblastic rim



DISCUSSION

The FOL of the jaws comprise a diverse, interesting, and challenging group of conditions that pose difficulties in classification and treatment. Common to all is the replacement of normal bone by a tissue composed of collagen fibers and fibroblasts that contain varying amounts of mineralized substance, which may be bony or cementum-like in appearance. The case presented here also showed the similar histopathological picture. The differentiating factor was the clinical and radiographical examination which helped us to reach to a conclusive diagnosis. Various studies showed female predilection, first and second decade and clinical site is maxillary posterior in fibrous dysplasia. The clinical examination revealed a classic fusiform expansion of maxillary ridge, related to fibrous dysplasia. The gender predominance, the clinical site and the features on inspection and palpation are all directed towards fibrous dysplasia. The radiographic examination revealed a radio-opaque swelling involving both buccal and palatal cortices and maxillary antral floor too. As evident on the CBCT, the lesion does not have any well defined extension, distinguishing the present lesion from cement-ossifying fibroma having a well defined extension. Conclusive diagnosis of any FOL is always based on clinical, radiographical and histopathological examinations. Based on all the key points, the conclusive diagnosis of this patient is FIBROUS DYSPLASIA.

CONCLUSION

FOLs of the maxillofacial bones make up a diverse collection of disorders that include

neoplastic and non-neoplastic and hereditary and non-hereditary conditions. FOLs of the jaw have been under frequent renaming and reclassification due to their varied features.

FOLs share many histopathological features like the replacement of the normal bone with fibrous connective tissue which is sometime interspersed with mineralized products such as osteoid, psammoma body mature bone or cementum like calcifications. A wide knowledge on the molecular biology behind this group of lesions is essential to understand the differing radiological pattern exhibited by them. Hence, the definitive diagnosis of FOLs requires correlation with the clinical and radiological findings.

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