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Case Report

The histological artistry of fibrous dysplasia: A case report

Sanpreet Singh Sachdev[1,*], **Tabita Joy Chettiankandy**[1], **Manisha Ahire Sardar**[1], **Ashish Sarda**[2], **Ashwini Chhapane**[1]

[1]*Dept. of Oral Pathology and Microbiology, Government Dental College & Hospital, Mumbai, Maharashtra, India*
[2]*Dept. of Oral and Maxillofacial Surgery, Government Dental College & Hospital, Mumbai, Maharashtra, India*

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ABSTRACT

Fibrous dysplasia (FD) is a fibro-osseous lesion characterized by the replacement of normal medullary bone by proliferative fibrous connective tissue. It comprises only 2% of all benign tumors and tumor-like processes of the bone. Craniofacial FD can occur in monostotic or polyostotic forms, and in conjunction with other endocrinal abnormalities. The monostotic FD is of particular importance for dental professionals owing to its propensity to occur in the jawbones. Differentiating FD from other fibro-osseous lesions such as ossifying fibroma and cemento-osseous dysplasia requires a fair amount of expertise, but is much crucial as the treatment plan differs for each. Clinicodemographic and radiological features are equally important in differentiating fibro-osseous lesions from one another. The present case report describes craniofacial monostotic FD in the anterior mandibular region of a 13-year-old Indian male.

1. Introduction

The fibro-osseous lesions (FOLs) are the craniofacial bones are a group of lesions that comprise reactive, neoplastic, developmental, and dysplastic pathologic processes.[1] The lesions included under the rubric of FOLs closely resemble each other in clinical, radiographic, and particularly, histopathological aspects, thereby making differentiation between them a challenging task. These include fibrous dysplasia (FD), ossifying fibroma (OF), Cemento-osseous dysplasia (COD), segmental odontomaxillary dysplasia (SOD), and familial gigantiform fibroma.[2]

FD is characterized by the replacement of normal medullary bone by proliferative fibrous connective tissue. The resultant bone consists of underdeveloped, inadequately calcified irregular trabeculae.[3] The term was initially coined by Lichenstein and since then many descriptive terms have been used for the lesion.[4] These range from a developmental anomaly to hamartoma to a benign tumor-like process.[5–7] Considering the incidence rate, FD comprises only 2% of all benign tumors and tumor-like processes of the bone.[8]

FD may affect a single bone (monostotic) or multiple bones (polyostotic). In the case of involvement of craniofacial bones, it is referred to as craniofacial FD, which is generally polyostotic (more than 75% cases) and seldom monostotic (10-25% cases).[7] Although much less alarming in terms of severity of symptoms, monostotic craniofacial FD is of great interest to dental professionals, owing to its relatively higher predilection to occur in the jawbones.[9]

The present case report comprises one such case of monostotic craniofacial FD occurring in the mandible of an Indian adolescent male.

* *Corresponding author*.
E-mail address: sunpreetss@yahoo.in (S. S. Sachdev).

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2. Case Report

A 13-year-old male complained of swelling in the left mandibular anterior region since one year. No remarkable findings were noted on extraoral examination. Intraorally, ill-defined intra-osseous swelling extending from the mandibular permanent left central incisor to the mandibular left deciduous second molar region was present.(Figure 1) The size of the lesion was approximately 5 x 4 cm. Buccoversion of the deciduous mandibular left first molar and permanent maxillary left canine was noted. The lesion was firm on palpation.

Orthopantomogram showed an ill-defined radiolucent lesion involving the mandible from the permanent left mandibular central incisor to the second molar region.(Figure 2) All the permanent teeth were present except for the mandibular left second premolar. Permanent maxillary canines of both sides and mandibular left first molar (distoangular) and second molars (mesioangular) were impacted. Dilaceration was noted apically with the roots of the impacted permanent mandibular first molar.

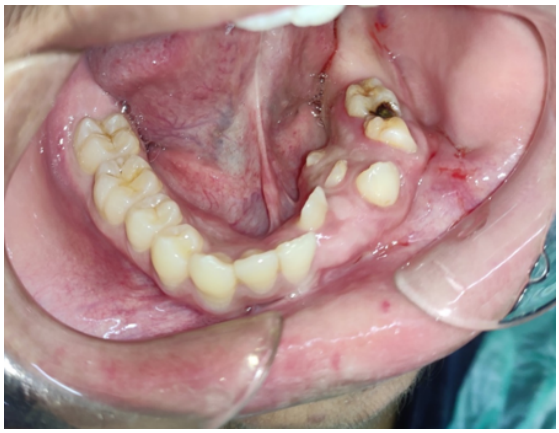

Fig. 1: Ill-defined intra-osseous welling extending from the permanent mandibular left central incisor to the deciduous second molar region.

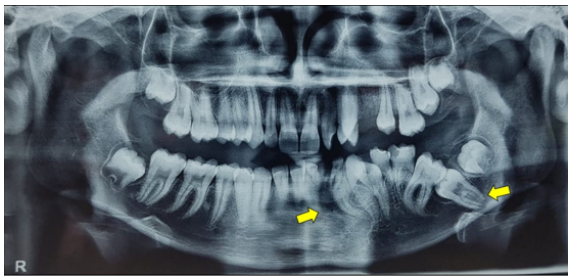

Fig. 2: Ill-defined radiolucent lesion extending from the permanent mandibular left central incisor to the deciduous second molar.

An incisional biopsy from the lesional area along with a rim of peripheral tissue was obtained. The tissue was decalcified in 2% nitric acid for three days and subjected to routine histological processing. Microscopically, thin trabeculae exhibiting a typical 'Chinese-letter' pattern were noted without any demarcating zone from the peripheral trabeculae. (Figure 3) The curvilinear trabeculae were in a background of fibrocellular connective tissue stroma. Most of the trabeculae consisted of osteocytes within the lacunae but were devoid of osteoblastic rimming. Prominent resting and reversal lines could be noted. The interface between the trabeculae and stroma exhibited 'brush borders' and also clefting in some areas. (Figure 4)

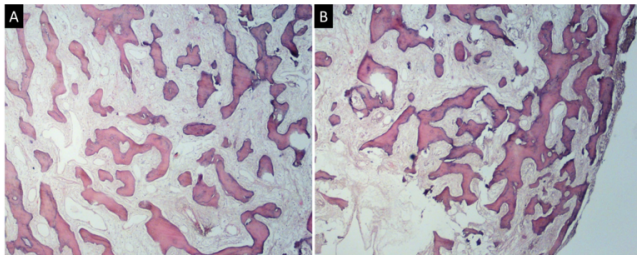

Fig. 3: A: Curvilinear trabeculae exhibiting typical 'Chinese letter' pattern; **B:** Absence of a definite transition zone between the trabeculae of the lesional area and the peripheral bone. (H and E, Original magnification x100)

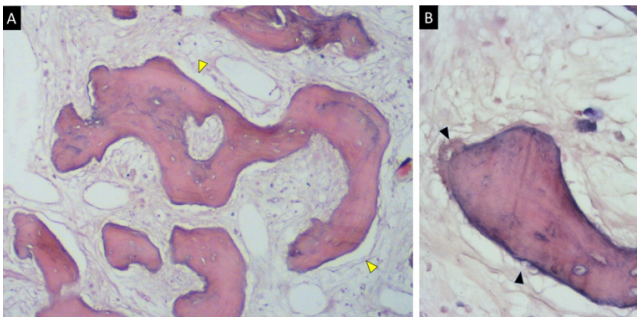

Fig. 4: A: Oseeous trabeculae with osteocytes in the lacunae, and prominent resting and reversal lines. Note the clefting at the bone—stroma interface (yellow arrowheads). (H and E, Original magnification x100); **B:** High power view showing the classic 'brush border' of the trabeculae (black arrowheads). (H and E, Original magnification x400)

A final diagnosis of Fibrous dysplasia was imparted. Surgical re-contouring was performed and the patient is free of symptoms after a one-year follow-up.

3. Discussion

FD is relatively rare in the craniofacial region (20% cases) and generally exhibits a female predilection.[10] Monostotic FD more frequently affects the maxilla than the mandible.[9] Most of the lesions affect the premolar region or areas posterior to it. The anterior mandible is seldom affected. This makes the present case quite unusual in terms of

clinicodemographic profile, wherein the lesion occurred in the anterior mandible without a noticeable involvement of the maxilla, in an adolescent male.

Radiographically, FD exhibits a characteristic 'ground glass' matrix, which is smooth, homogenous, and eccentric in the alveolar bone. While the radiological appearance of typical FD is mixed, the spectrum also includes a percentage of homogenous dense sclerotic (23%), and radiolucent (21%).[11] The poorly defined margins along with histopathological confirmation are currently considered the gold standard for the diagnosis of FD.[12]

Histopathologically, the differential diagnosis of FD comprises the other FOLs that closely resemble each other which include ossifying fibroma and cemento-osseous dysplasia. These FOLs can only be differentiated from one another by taking into account the clinicodemographic and radiological findings.[2]

While both FD and OF affect adolescents and young adults, differentiating between the two is important because while the former is a reactive phenomenon, the latter is a true neoplastic phenomenon warranting surgical removal.[13] Radiographically, OF is well-defined with a sclerotic rim, indicating a confined intra-osseous benign mass.[11] Histopathologically, OF shows a distinct 'transitional zone' between the lesional area and the normal bone.[14] In the present case, the lesion was ill-defined without a sclerotic border. Furthermore, the osseous trabeculae of the lesional area and peripheral normal bone blended indistinctly without a definite zone of transition.

COD tends to affect patients of relatively older age groups, usually 30-40 years.[14] Histopathologically, spherules of cementum-like deposits are noted, that fuse to form ginger root-like trabecular masses.[13] The present case depicted classic thin curvilinear trabeculae without evidence of any cementum-like material. The trabeculae of COD are also typically devoid of the 'brush border' at the bone-stroma interface. SOD was only recently included in the official classification of head and neck tumors.[2] It characteristically involves the maxilla and generally presents with additional extragnathic dermal features.

Generally, conservative treatment or correction of the deformity is deemed sufficient for controlling FD.[5–7] Surgical resection is considered for lesions with aggressive behavior or in case of the presence of symptoms. The pathogenesis of FD involved somatic activating mutations of the GNAS-1 gene.[15] The protein product, stimulatory G protein – α amplifies Interleukin-6 production, which results in an imbalance in the osteoblastic progenitor cell and osteoclastic cell activity.[16,17] It is believed that this imbalance wanes or even completely ceases after puberty when bone maturation is completed. Only rarely do the lesions continue progressing in old age; in such cases, surgical excision of the demineralized bone followed by osteoplasty may be necessary.[18]

The selection of surgical modality for treatment depends on the age, willingness, and financial ability of the patients. The treatment approach should be tailored considering these factors for each individual case subjective to the clinician's judgment and expertise. In the present case, the patient being in the pubertal age group, with the absence of symptoms and monostotic form of the disease, a conservative treatment plan was adopted.

About 1% of cases of FD may transform into a malignant lesion. Such cases generally have associated symptoms such as pain, rapid growth, or ulceration. Elevated serum alkaline phosphatase levels (AP) levels are considered a reliable indicator marker for the malignant transformation of FD.[5] In the present case, the patient did not experience any symptoms and their serum calcium and AP levels were also within the normal range. Nevertheless, long-term follow-up is crucial for patients with FD even in the absence of these 'red flags.' A re-assessment of the serum AP levels and periodic six-monthly follow-ups were advised to the patient.

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None.

5. Conflicts of Interest

There is no conflict of interest.

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Author biography

Sanpreet Singh Sachdev, Post Graduate https://orcid.org/0000-0001-7655-8180

Tabita Joy Chettiankandy, Professor & HOD https://orcid.org/0000-0002-6839-6959

Manisha Ahire Sardar, Associate Professor https://orcid.org/0000-0003-2003-1544

Ashish Sarda, Post Graduate https://orcid.org/0000-0002-1503-5009

Ashwini Chhapane, Post Graduate https://orcid.org/0000-0003-2706-8307

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