

Fibrous Dysplasia Of Maxilla: A Case Report

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Abstract

Summary: Fibrous dysplasia is a benign disease of unclear aetiology that is congenital and recurring in nature. It is caused by a problem known as bone modelling, in which the normal bone constantly changes and is replaced by immature fibrous tissue. One or more bones may be impacted by the lesion; the maxilla is the most often afflicted facial bone. Expansile lesions known as fibrous dysplasia lead to issues related to the original location. The most often involved region in the craniofacial bone is the maxilla. In this case report 50 years 50-year-old female patient reported who has maxillary fibrous dysplasia.

Keywords: fibrous dysplasia, congenital, bone.

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1. Introduction

Fibrous Dysplasia (FD) is a benign tumour-like condition of any bone growth that occurs over time and is defined by the substitution of natural bone by abnormal multiplication of cellular fibrous connective tissue mixed with uneven bone trabeculae. ⁽¹⁾ Von Recklinghausen, used the term "generalist fibrous osteitis" to describe a group of clinical diseases that included deformities and bone changes. As a result, Liechtenstein and Jaffe recognized these previously described abnormalities as a well-defined disease and termed it fibrous dysplasia. ⁽²⁾ The cause of this disease is still unknown. ⁽³⁾ It appears to be a rare disease, with an occurrence rate of 1:4000 to 1:10000. ⁽⁴⁾ Fibrous dysplasia affects primarily kids and teenagers, with 75 % occurring well before the age of 30. (highest incidence between 3 and 15 years). ⁽⁵⁾ It affects both men and women equally, and it accounts for roughly 2.5% of all bone lesions and about 7% of all benign bone tumours. ⁽²⁾ Monostotic (including craniofacial), polyostotic, and polyostotic with endocrinopathies are the three types of FD. ⁽³⁾ The monostotic form, which accounts for 80–85% of FD cases, is not a precursor to the polyostotic form. Endocrinopathies affect 3% of polyostotic patients, who have McCune–Albright syndrome. ⁽⁶⁾ The posterior region of the maxilla experiences monostotic bone nearly twice as frequently as the

mandible. ⁽⁷⁾ Swelling and expansion of the craniofacial regions are moderate indications of the condition. However, if the condition is left untreated, it can result in visual loss, hearing loss, airway blockage, anosmia, and numbness. Fibrous dysplasia of the maxilla or mandible can cause permanent teeth to shift, prevent new teeth from erupting, and lead to misaligned teeth. ⁽⁸⁾ Traditional radiography, magnetic resonance imaging, computed tomography scans (CT) and scintigraphy, can all be used to assess it, ⁽⁹⁾ while CT is the preferred method for evaluating craniofacial abnormalities. ⁽¹⁰⁾ When there is aesthetic function or damage caused by the lesion, surgical excision or osteoplasty with cosmetic recontouring is currently suggested; nevertheless, it should be delayed as much as possible due to the risk of relapse in patients where bone formation is vigorous. ^(10, 11) As a result, the goal of this case report is to describe a case of fibrous dysplasia, as well as the clinical, imaging, laboratory, histological, and developmental features of the case.

2. Case Presentation

A 50-year-old female reported to the Department of Oral and Maxillofacial Surgery at Islamic International Dental Hospital with the chief complaint of swelling in the upper right maxillary region for 6 months. The patient was asked to get a CBCT (Cone-beam computer tomography) done (fig 3).

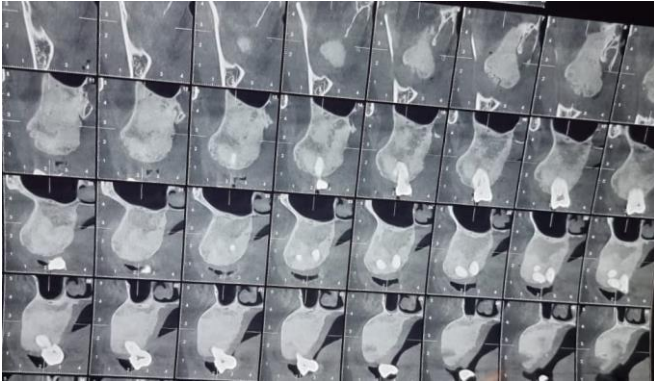


Figure-3 Shows cbct report of the patient

According to the patient, the lesion began insidiously and grew irregularly until it reached its current size (fig 2).



Figure-2 Shows intraoral examination of the patient

There was no pain associated with the swelling. The patient revealed no known family history. There was no traumatic history. There was enlargement of the right side of the palate intraorally, as well as extension of the alveolar buccal plate reaching from the upper right canine to the maxillary tuberosity. There was no swelling anywhere else on the body, and there were no café au lait spots. Routine tests were done, including a hemogram, blood calcium, and serum alkaline phosphatase (ALP). All of the values were within acceptable limits. It was a hard swelling on aspiration. An incisional biopsy confirmed that the bone was normal. Lidocaine injection was used as the local anaesthetic during surgery. After a crystal incision from the right maxillary canine to the right maxillary tuberosity, a full-thickness mucoperiosteal flap was raised. A stainless steel drill was used to modify the contour of the alveolar crest, as well as soft-tissue

reduction. For twice-daily use, naproxen sodium (Synflex 550 mg) and amoxicillin/clavulanic acid (Augmentin-BID®, 1,000 mg) were advised. Seven days following surgery, the sutures were removed. The patient had regular follow-up appointments after surgery. Again buccal and palatal extension was noted after a few months. The panoramic radiographs obtained during that moment were found to be nearly identical to those obtained previously. Second surgery was suggested but the patient did not report. The lesion was studied under the microscope and Figure 4 shows the results. Figure 1 shows the proper report of the patient after the lab tests.

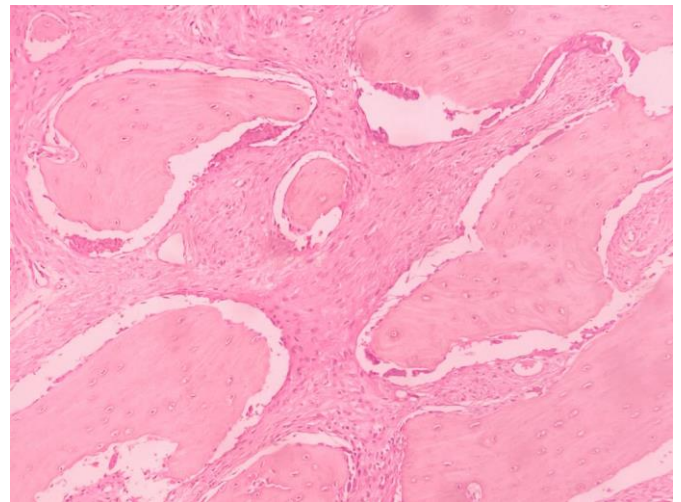


Figure-4 Shows histopathology of the patient

3. Discussion

Fibrous dysplasia is a benign bone disorder characterised by the replacement of bone by fibrous tissue. ⁽¹⁾ The actual cause of fibrous dysplasia is unknown, but it is most likely due to a genetic susceptibility caused by a mutation in the GNAS1 gene (guanine nucleotide-binding protein) on chromosome 20q13 or malformations in AMPc, which can lead to higher emergence of melanocytes and hyperfunction of concerned endocrine organs. ⁽¹²⁾

Both males and females are affected equally, while Singh G et.al found that the average age of incidence was between 3 and 15 years. The patient in this case was 50 years old female.

According to Abdulai et al., ⁽¹³⁾ fibrous dysplasia infects the maxilla double as often as the mandible and is most commonly observed in the posterior region, as

it was in the present case. The polyostotic version typically affects youngsters under the age of ten, whereas the monostotic form typically affects people in their second and third decades, ⁽¹⁴⁾ as the patient in this case was 50 years old. The indications of FD differ depending on which bones are damaged; however, the most frequent sign is a slow-growing, painless swelling ⁽¹⁵⁾. Other craniofacial bone-related symptoms of FD include facial deformities, pain, headache, and vision or olfactory impairment. ⁽¹⁶⁾



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Patient name: Mrs. Muneer	Accession no: D-20-215
MRN: 03-177240	Procedure date: 31-12-2020
Age: 50-years	Report date: 14-1-2021
Gender: Female	Referring doctor: Dr. Kinza

Clinical History: Swelling in right buccal bone for 1 year.

Gross: The specimen is received in a single formalin container, properly labeled with the site of biopsy (maxilla), and the type of procedure (excisional biopsy). The specimen consists of a single piece of hard tissue, tan-grey in color, approximately 2 x 1 x 1 cms in dimensions. The tissue is decalcified before being submitted for processing as a single block.

Histology: Sections show a decalcified specimen consisting of trabeculae of vital bone. The intertrabecular area is composed of fibrovascular connective tissue.

Diagnosis:
CONSISTENT WITH BUCCAL MAXILLARY EXOSTOSIS

Dr. Amber Kiyani (BDS, MS) Fellow-American Academy of Oral and Maxillofacial Pathology Diplomate-American Board of Oral and Maxillofacial Pathology Assistant Professor, Department of Oral Diagnosis and Medicine.	Dr. Nadia Zaib (BDS, M.Phil), Associate Professor, Oral Pathology
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Figure-1 Report of the Patient from the Lab

As seen in this study, the majority of FD lesions in the craniofacial region are unilateral ⁽¹⁷⁾. On radiographs, our patient's FD had a ground-glass appearance. The stage of the illness mostly determines the appearance of FD, with early-stage lesions being more apparent and late-stage lesions being sclerotic, radiolucent, or mottled. ⁽¹⁸⁾ Histologically, fibrous dysplasia has an essentially fibrous element with replicating fibroblasts, and an osseous portion with irregularly shaped osteoid trabeculae with Chinese character pattern or jigsaw

puzzle form, as well as juvenile woven bone in the fibrous stroma. ⁽¹⁹⁾ There are no universally acknowledged management guidelines for this condition. Observation, medicinal therapy, and surgical therapy are the suggested treatment choices. For minor asymptomatic lesions that are cosmetically acceptable to the patient. ⁽⁷⁾ Observation is the cornerstone of treatment. Medical therapy is not commonly used to treat fibrous dysplasia, but a few medicines, such as bisphosphonates and Pamidronate (60 mg/day intravenous route), can inhibit osteoclastic activity. Patients with low serum calcium levels were advised to take calcitonin, vitamin D, and calcium supplements. ⁽⁷⁾ Surgical therapy, aimed at repairing (remodelling) or avoiding functional disability and obtaining normal facial aesthetics, remains the standard treatment for extensive lesions. ⁽⁷⁾ Remodeling was carried out in this circumstance. When it occurs in adults, fibrous dysplasia has a low recurrence incidence, ranging from 15% to 20%. However, it is more common during the growing stage. The major sign for detecting recurring lesions is an increase in serum alkaline phosphatase (ALP) levels. ⁽²⁰⁾

5. Conclusion

Fibrous dysplasia is an extremely uncommon bone disorder that causes abnormal fibrous tissue to replace normal osseous tissue. Following initial observations acquired from a routine X-ray, CBCT provides an instructive modality for further confirming the presence and severity of the disease.

CONFLICTS OF INTEREST- None

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Potential competing interests: None to report

Contributions:

K.A, Z.R, G.R - Conception of study

K.A, H.B, M.F - Experimentation/Study Conduction

Z.R, G.R - Analysis/Interpretation/Discussion

Z.R, G.R - Manuscript Writing

K.A, Z.R, G.R, H.B, M.F - Critical Review

K.A, Z.R, H.B, M.F - Facilitation and Material analysis

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