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Case Report

Features of Monostotic Fibrous Dysplasia in Maxilla using Cone Beam Computed Tomography

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ABSTRACT

Fibrous dysplasia (FD) is a benign nonneoplastic bone disorder in which fibrous tissue replaces normal bone. It is a genetic non-inherited condition caused by a mutation in the GNAS1 gene and is characterized by abnormal proliferation of fibrous tissue interspersed with normal or immature bone. FD causes esthetic disfigurement and loss of function when the maxillo-mandibular region is affected. A dental practitioner can be the first to detect such conditions. We reported a case of a 28-year-old male, with a history of swelling of the left side of the maxilla for 20 years. The present report highlights the clinical, radiographic, and histopathologic features of this condition along with a review of past literature.

Keywords: Cone-beam computed tomography, fibrous dysplasia, maxilla

INTRODUCTION

Fibro-osseous lesions are a diverse group of diseases that are characterized by the replacement of normal bone by fibrous connective tissue. These lesions result from the abnormal development of bone-forming mesenchyme (1). Fibrous dysplasia (FD) is a benign fibro-osseous lesion of unknown etiology, uncertain pathogenesis, and diverse histopathology. It comprises of 2-5% of all bone pathologies. It can be classified as monostotic and polyostotic FD (2). The most common form is the monostotic FD that comprises 70-80% of the lesions (3). The femur, tibia, ribs and facial bones are the most commonly involved structures. The craniofacial bones are involved in 40-60% of the cases (4). We hereby report a case of FD presenting as a swelling of the left maxilla.

CASE PRESENTATION

The patient provided consent for the case report. A 28-year-old male reported to the outpatient department with a chief complaint of swelling in the upper left back region of the face for 20 years. He had noticed the swelling 20 years ago, which was smaller in size at the time but gradually progressed to the present size with associated facial asymmetry. He reported that there was no history of trauma to the region and no history of pain or any associated discharge. The patient had consulted a local doctor for the same but records of the previous visit were unavailable. The medical and dental history was noncontributory.

On extra-oral examination, mild facial asymmetry was detected on the left middle one-third of the face with an ill-defined swelling anteroposteriorly extending from the left ala of the nose to the tragus, and 1 cm below from the left lower eyelid to the ala-tragus line superoinferiorly. The swelling was approximately 4x4 cm in size and was non-tender and bony hard on palpation. No abnormality was associated with the overlying skin (Figure 1a). Intra-oral examination revealed a diffuse swelling in the left maxilla region, extending from the distal aspect of maxillary left lateral incisor to the maxillary tuberosity anteroposteriorly. Expansion of the alveolus and obliteration of the buccal vestibule was noted. The maxillary left first premolar was decayed (Figure 1b, c).

Based on the history and clinical findings, a provisional diagnosis of FD was considered with a differential diagnosis of ossifying fibroma. Intra-oral periapical radiograph revealed loss of lamina dura, narrowing of the periodontal ligament space, altered trabecular pattern with ground glass appearance in the region of maxillary left first premolar to the second molar. Radiolucencies involving the enamel, dentin, and pulp were also noticed concerning maxillary left first premolar, with the bulbous appearance of the apical one-third of the root and an ill-defined periapical radiolucency suggestive of a periapical abscess (Figure 2a). Expansion of the buccal cortical plate was appreciated

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in a lateral occlusal radiograph (Figure 2b). A panoramic radiograph revealed altered trabecular pattern with ground glass appearance from maxillary left lateral incisor to maxillary left third molar region, with obliteration of the floor of the maxillary sinus (Figure 3). CBCT was done to evaluate the extent of the lesion and showed the presence of a homogeneous radiopacity in the left maxilla extending from the midline till the left zygoma with the involvement of the maxillary sinus and expansion of the alveolar process (Figure 4a-c).

The patient's serum alkaline phosphatase level was within normal limits (130 IU/L). The histopathological examination revealed the formation of numerous immature woven bone cells showing a trabecular and curvilinear pattern, with fibro-cellular connective tissue seen interspersed between them (Figure 5). These features were compatible with the clinical diagnosis of FD. The patient was referred to the oral and maxillofacial surgeon for surgical recontouring of the left maxillary alveolus. Root canal therapy was advised for the maxillary left first premolar.

Figure 1. a-c. Clinical photograph of the patient. (a) Extra-oral photograph of the patient showing swelling of the middle one-third of the face, (b) Intra-oral photograph of the patient showing diffused swelling in the left maxillary region, (c) Intra-oral photograph of the patient showing diffused swelling in the left maxillary region (palatal view)

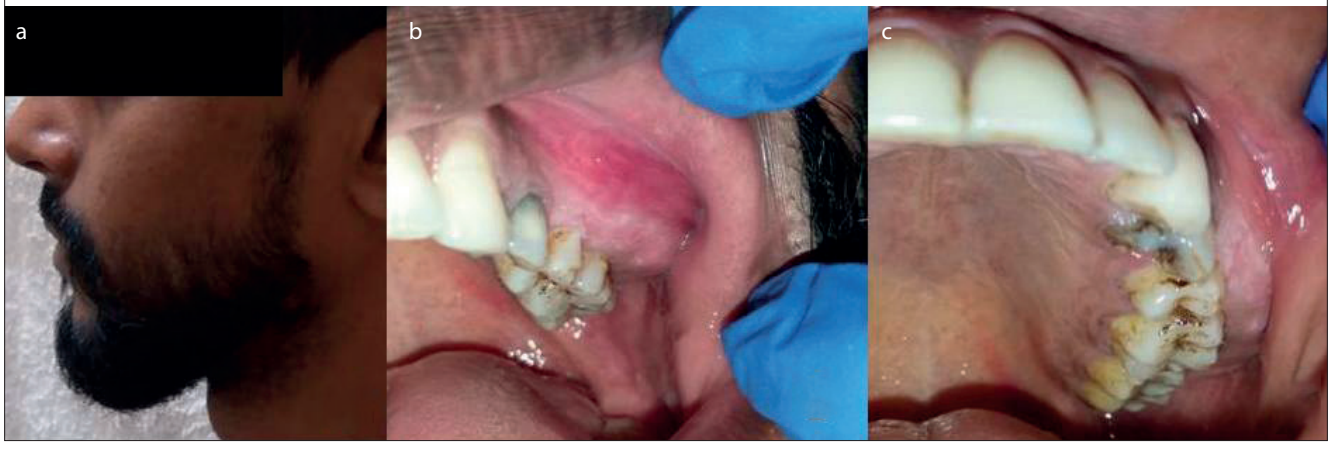
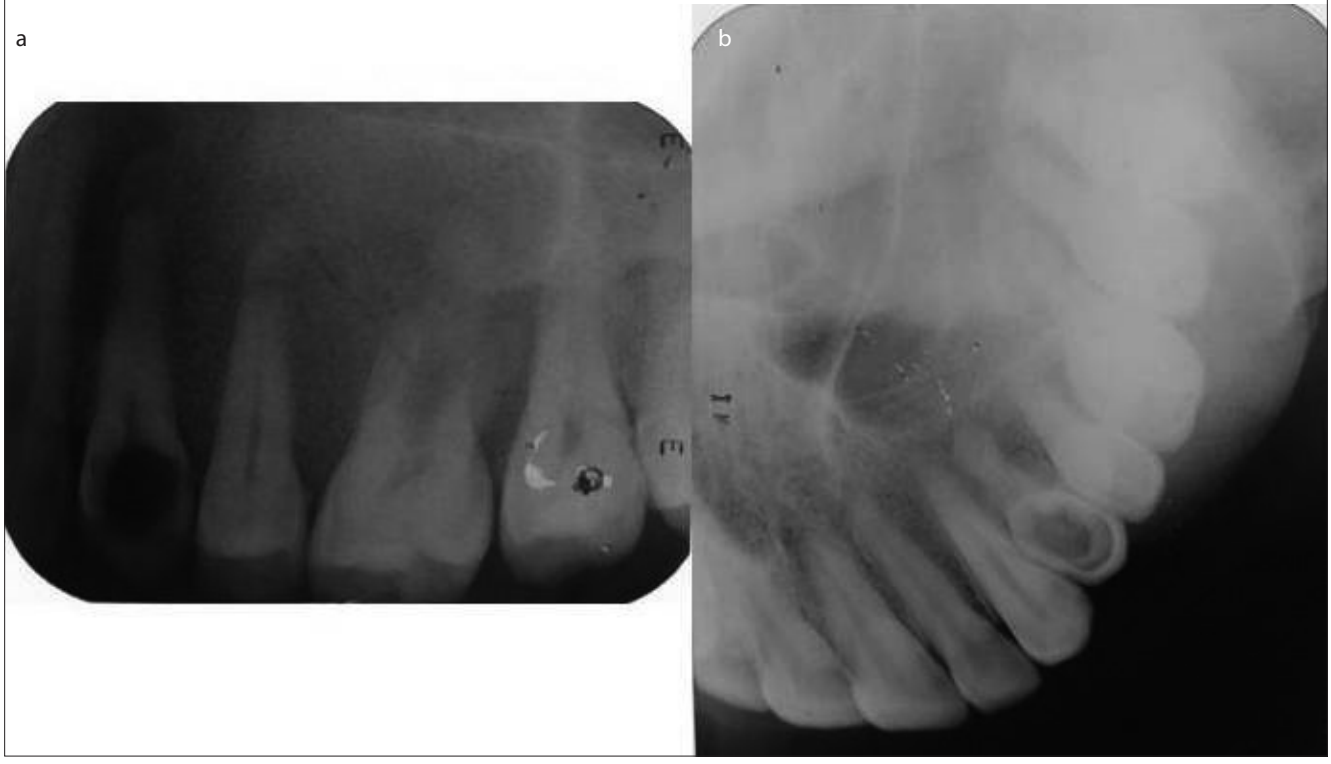


Figure 2. a, b. Intra-oral periapical radiograph (IOPA) and left lateral maxillary occlusal radiograph. (a) IOPA showing an altered trabecular pattern with ground glass appearance along with caries and periapical pathology for maxillary left first premolar, (b) Left lateral occlusal radiograph showing expansion of the buccal cortical plate



DISCUSSION

Fibrous dysplasia is a nonneoplastic condition caused by mutations of the GNAS-1 gene that results in the inhibition of differentiation and proliferation of bone-forming cells. This leads to the replacement of normal bone by fibrous tissue and woven bone (5). The incidence varies from 1:4000 to 1:10,000 (6). It was initially called "osteitis fibrosa generalisata" by Van Recklinghausen in 1891 and was renamed "fibrous dysplasia" by Lichenstein in 1938 (7). The disease is classified into three forms: monostotic, polyostotic, and craniofacial.

The monostotic form comprises about 80% of these lesions, mostly seen in the second and third decade of life. Involvement of facial bones is seen in 10-27% of cases of monostotic FD (6). It is a self-limiting condition in most cases and reaches a state of dormancy by adolescence. Monostotic FD has a predilection for males and affects the maxilla more frequently than the mandible. It manifests clinically as a slow-growing painless mass, leading to facial asymmetry, as seen in the present case. Headache is one of the common symptoms associated with FD when it involves the maxilla, the orbit, and the sinus (5). The patient can develop other symptoms due to the growth of the mass and the compression of the adjacent structures. Hypertelorism, visual impairment, and blindness are seen in association with orbital involvement. Symptoms of neuralgia and sinusitis may also be present (8).

Malocclusion and facial disfigurement is a common feature associated with FD. Other associated dental disorders are enamel hypoplasia, dentin dysplasia, taurodontic pulp, odontoma, tooth displacement, and high caries index (9). In our patient, decay involving pulp and the associated periapical abscess was noticed for the maxillary left first premolar.

Fibrous dysplasia shows a vast variety of radiographic appearances, which depends on the stage of diagnosis of the lesion. The most common manifestations include ground glass appearance, chalky pattern, and cystic pattern (10). The present case showed

Figure 3. Panoramic radiograph showing an altered trabecular pattern with ground glass appearance from maxillary left lateral incisor to maxillary left third molar region, with obliteration of the floor of the maxillary sinus

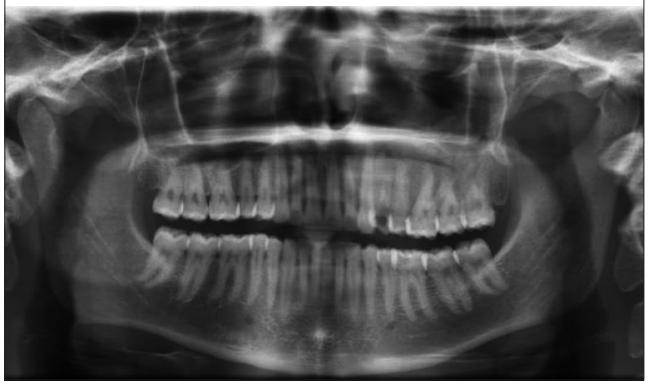


Figure 5. Histopathological picture showing the formation of numerous immature woven bone cells in a trabecular and curvilinear pattern

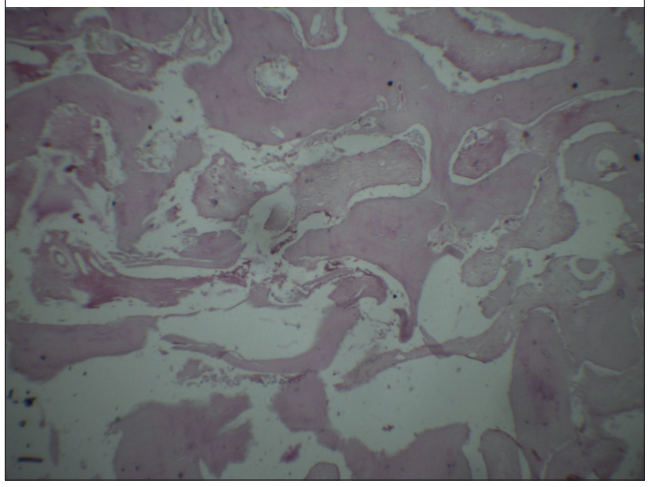
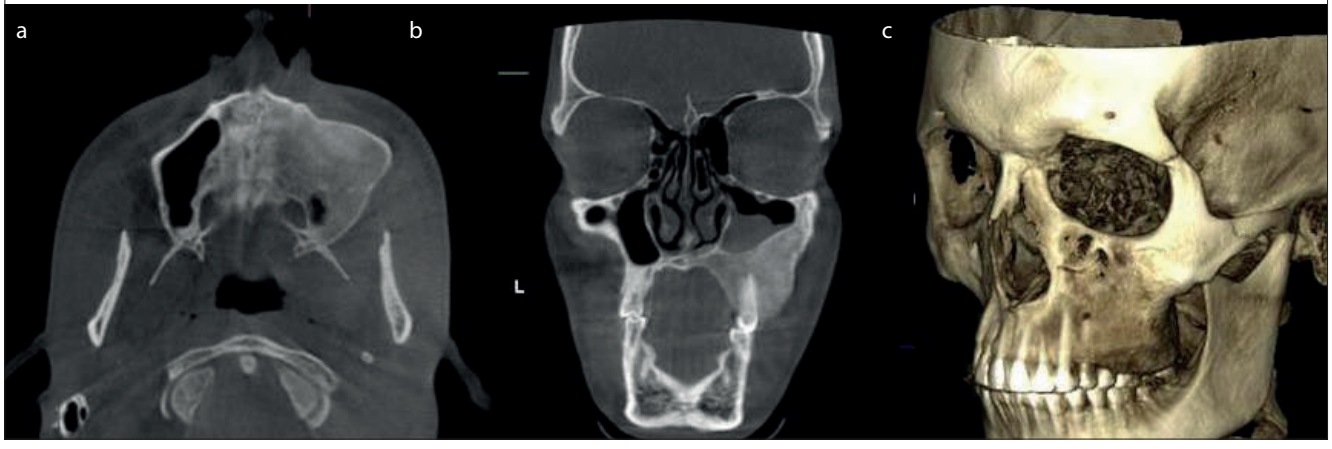


Figure 4. a-c. Cone beam computed tomography (a: axial section, b: coronal section, c: 3D reconstruction) showing the presence of a homogeneous radiopacity in the left maxilla extending from the midline anteriorly till the left zygoma posteriorly with the involvement of the maxillary sinus superiorly and expansion of the alveolar process



the presence of ill-defined radiopacity with the characteristic ground glass appearance in conventional imaging. CBCT helps to evaluate the extent of the lesions. The radiographic features of FD are not pathognomic (9). Histopathological examination is considered as the gold standard, which shows the presence of low to moderate cellular fibrous stroma surrounding irregular trabeculae of woven bone, commonly referred to Chinese letter characters. Osteoblastic rimming is characteristically absent (11).

Laboratory investigations include the estimation of serum calcium and alkaline phosphatase levels which can be elevated in certain cases (8). The present case showed normal serum alkaline phosphatase level.

Management of patients with FD involving maxilla and mandible who require dental extractions, dental implants, root canal therapy, and orthodontic therapy is challenging for the dentist, but it is possible to carry out routine dental care in FD patients. However, research and evidence are needed to address issues such as healing after tooth extraction, aggravation of FD lesion after surgical treatment, and successful orthodontic therapy due to poor quality of bone. Further research in patients with FD involving maxilla and mandible regarding the above-mentioned issues can help in better understanding of treatment outcomes (9).

Treatment modalities of FD vary according to the area of involvement of the craniofacial skeleton, function, and esthetics. Surgical approaches for skeletal deformities include two types: conservative and radical. Conservative method includes osseous contouring, which has to be performed periodically until the lesion achieves a static phase. Radical therapy includes the complete excision of the lesion with reconstruction (12). Medical management with the help of bisphosphonate therapy and calcitonin have also been mentioned in the literature (13, 14). The surgical management of FD has a high recurrence rate of 15-20% (15), hence, patients have to be kept on a long-term follow-up schedule.

CONCLUSION

Fibrous dysplasia is a benign lesion that causes esthetic disfigurement and loss of function when the maxillo-mandibular region is affected. A dental practitioner can be the first to detect such conditions. Thorough knowledge and careful clinical and radiographic examinations are required for proper diagnosis and management. Further management of other dental problems in the same region as the FD poses a challenge to dentists. The present report highlights the clinical, radiographic, and histopathologic features of this condition along with a literature review.

Informed Consent: Verbal informed consent was obtained from patient who participated in this case.

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