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Monostotic Fibrous Dysplasia Involving the Mandible: A Case Report

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Abstract

Fibrous dysplasia (FD) is a skeletal developmental anomaly, which is non-hereditary and idiopathic in origin. It is characterized by the replacement of normal bone with the excess proliferation of fibrous tissue in irregular bony trabeculae. Patients might complain of swelling, pain, or numbness on the affected side. The incidence of monostotic FD (MFD) is four times more than that of polyostotic fibrous dysplasia. In MFD, the maxilla is more commonly affected than the mandible. The clinical behavior and rapid progression of FD renders the treatment challenging. The malignant potential is 0.5% for untreated cases. Here, we present a case of FD involving the mandible. The clinical diagnostic approach, different imaging modalities, and histological examination methods for definitive diagnosis have been elaborated.

Keywords

Fibrous dysplasia, Ground glass, Mandible

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Introduction

Fibrous dysplasia (FD) is a type of hamartoma, wherein the medullary bone is replaced by immature and poorly calcified bone.[1,2] FD comprises 2-5% of all bone tumors and 7% of benign tumors.[3,4] FD is caused by an imbalance between osteoblastic and osteoclastic activities.[5,6] Monostotic FD (MFD) is the most common form of FD, and is unilateral in nature.[7] It is more commonly observed in females. With 50% MFDs occurring in the bones of the head and neck.[8] Less than 1% cases of FD show malignant transformation.[8,9] Pain, rapid growth of the lesion, and increased alkaline phosphatase levels are indicators of malignant transformation.[9] Here, we present a case of FD involving the mandible in a female patient who was diagnosed with the help of various imaging modalities, and incisional biopsy confirmed the diagnosis.

Case report

A 24-year-old female Bangladeshi patient reported to the Department of Diagnostic Sciences & Oral Biology, College of Dentistry, King Khalid University with swelling on the lower right side of the face since four years, and pain in the region of swelling since two months. The patient initially noticed a swelling on the lower right side of the mandible, which showed slow growth and progressive nature with no history of prodromal symptoms. Pain followed the swelling, which was gradual in onset, progressive, mild, intermittent, dull in nature, aggravated on chewing solid food, and relieved with medications. The patient consulted a private dental clinic for the swelling. However, she did not experience any relief from pain and swelling. The medical, dental, family, and personal histories of the patient were noncontributory. On general physical examination, the patient was conscious, cooperative, moderately nourished and built, and showed no signs of anemia, icterus, and/or clubbing. All vital signs were within normal limits. On extraoral examination, mild asymmetry of the face was noticed with swelling on the right lower one-third of the face and no signs of impaired vision and/or auditory functions. No pigmentations were observed on the skin or any extraoral surface. Movements of the temporomandibular joints were within normal limits with non-tender muscles of mastication and no enlarged lymph nodes. On examination, a single ill-defined swelling was noticed on the right lower-third of the face measuring 2 × 3 cm. The swelling extended

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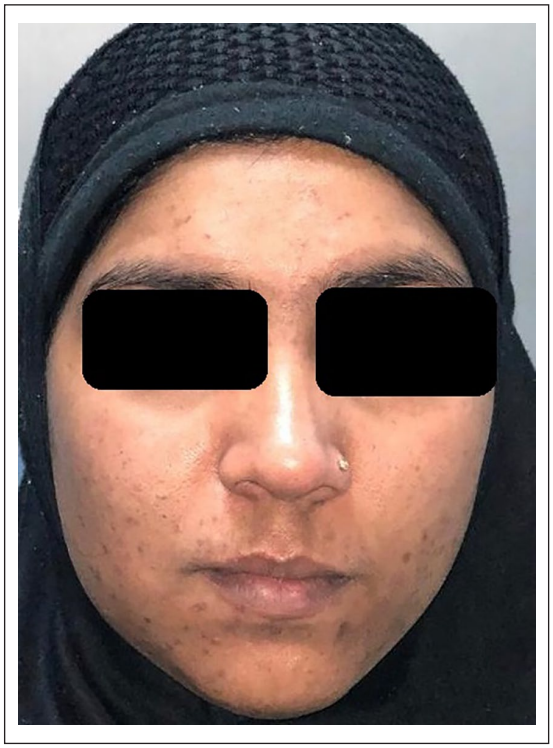

Figure 1. Extraoral photograph.

Figure 2. Intraoral photograph showing expansion of the buccal cortical plate on the right side.

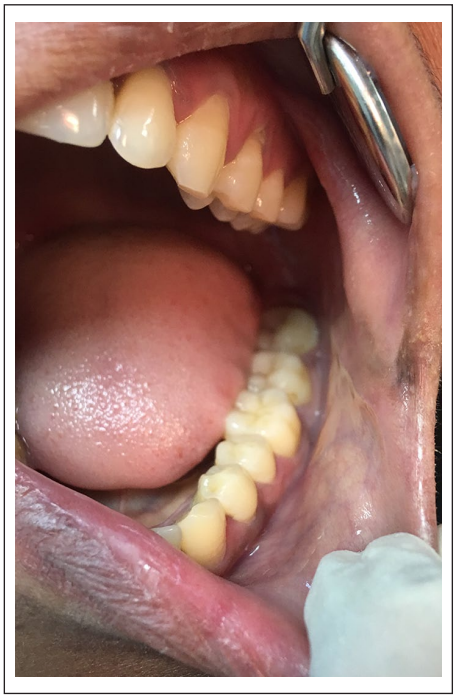

Figure 3. Intraoral photograph showing no expansion of the buccal cortical plate on the contralateral side.

anteriorly from the parasymphysis region to the ramus of the mandible posteriorly. No signs of inflammation were observed on the swelling. On palpation, the swelling was hard in consistency, non-tender, and immobile, with signs of paresthesia on the affected side (Figure 1). Intraoral examination showed expansion of the buccal cortical plate, extending from the mandibular right first premolar to the mandibular right second molar, which was hard in consistency, non-compressible, and non-tender. The mucosa over the buccal cortical plate showed no signs of crackling (Figure 2). No pain on percussion and no tooth mobility were evident on the affected site. The buccal cortical plate on the other side did not show any evidence of expansion (Figure 3). After analyzing the patient's data, a provisional diagnosis of MFD was made, and differential diagnoses of hyperparathyroidism and Albright's syndrome were considered. The patient was referred for further laboratory investigations where her complete blood count and serum calcium, alkaline phosphatase, T_3, T_4, and thyroid stimulating hormone levels were within normal limits. On examination of the panoramic radiograph of the patient (Figure 4), loss of trabecular bone pattern (step-ladder pattern) with mixed radiolucent and radiopaque lesions were observed on the right side of the mandible accompanied by obliteration of the mandibular canal. An intraoral periapical radiograph (Figure 5) showed typical "ground-glass appearance" of the trabecular bone with generalized loss of the lamina dura and narrowing of the periodontal ligament space. Occlusal radiographs revealed expansion of the buccal cortical plate (Figure 6) with mild expansion of the lingual cortical plate in the right second and third molar regions. An incisional biopsy was advised on the affected side, and histopathological analysis showed the presence of bone trabaculae interspersed with fibrous stroma. Detailed analysis under high magnification revealed the presence of bone trabaculae with entrapped osteocytes and osteoblastic lining,

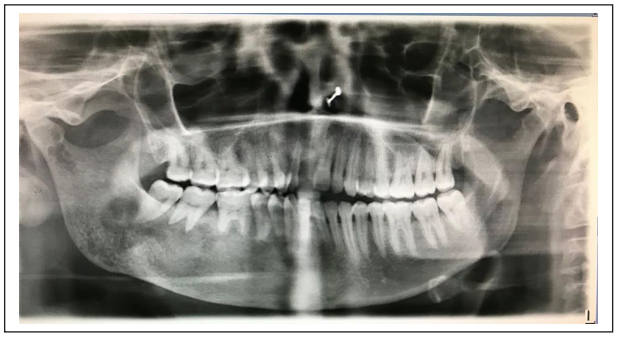

Figure 4. Panoramic radiograph showing mixed radiopaque and radiolucent lesion on the right side of the mandible.

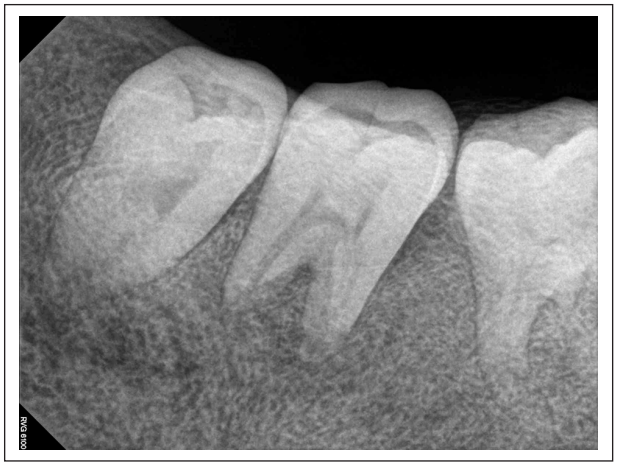

Figure 5. Intraoral periapical radiograph showing ground glass appearance.

which were suggestive of FD (Figures 7 and 8). The patient was referred to an advanced center for further investigations such as computed tomography and magnetic resonance imaging, as they were unavailable in our institution and for surgical evaluation.

Discussion

Diagnosis of FD in 36.3% cases is challenging, as no characteristic symptoms are evident, and 63.6% patients complain of non-specific symptoms such as pain and/or swelling.[10] MFD affects the maxilla more frequently than the mandible.[11] However, in our patient, the mandible was affected and the maxilla was spared. Signs and symptoms often differ based on the location of the tumor. Patients may complain of facial deformity, visual alterations, nasal congestion, pain, and/or auditory disabilities. Our patient complained of pain and swelling on the affected site of the jaw. Most tumors appear in the premolar region and extend to the third molar region; the anterior area is the least affected. Similar findings were observed in our patient. The preferred diagnostic approach for

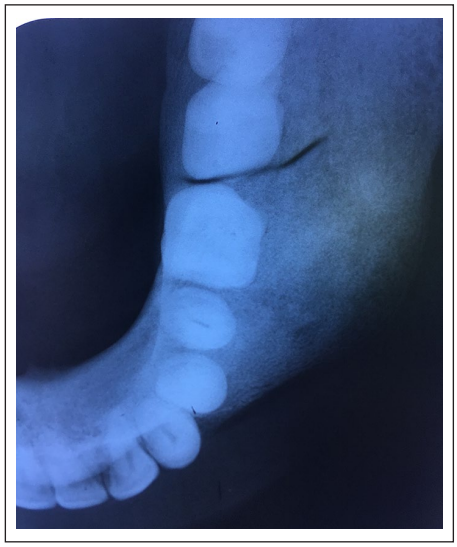

Figure 6. Expansion of the buccal cortical plate seen on occlusal radiograph.

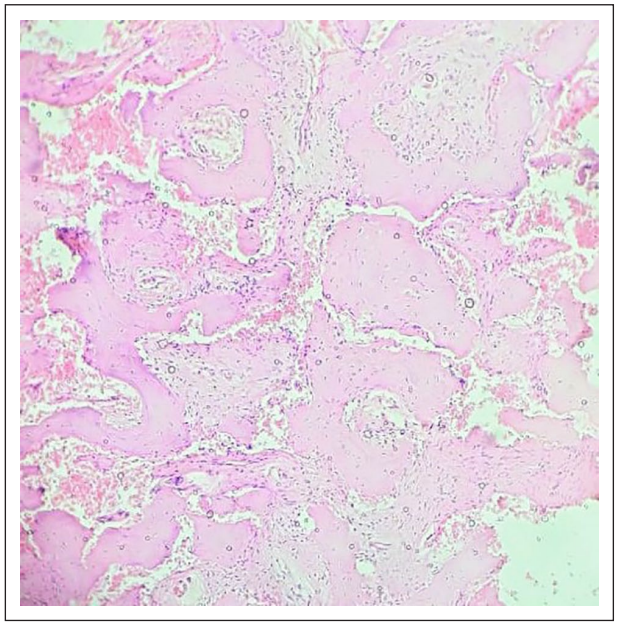

Figure 7. Histological picture shows trabecula of bone interspersed with a fibrous stroma

such bony lesions is a 3D imaging modality. Considering the socioeconomic background of our patient and the accessibility in our institution, 3D imaging modalities were not advised for our patient. The most common radiographic finding in FD is the ground-glass appearance, which was observed in our patient's radiograph. The periapical radiographs of the involved side often show thinning of the periodontal ligament space with an irregular or loss of lamina dura accompanied by abnormal bone pattern. Similar findings were observed in our patient. To establish the diagnosis of FD, patient's history,

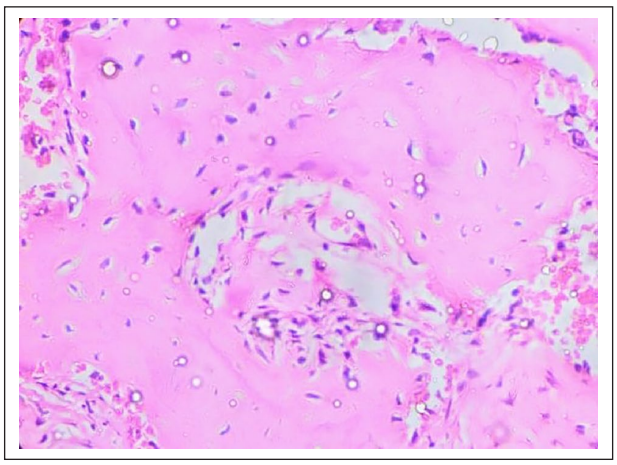

Figure 8. High-power magnification shows trabecula of bone with entrapped osteocytes and osteoblastic lining.

thorough examination, and radiographic assessment are often adequate.[11] Alarming signs for malignant transformation in FD are increased alkaline phosphatase levels.[11,12] Therefore, its levels should be periodically monitored in such patients. The patient's alkaline phosphate levels were within normal limits, and she was advised alkaline phosphatase assessment every six months. The aim of treatment should be correction of the deformity caused by the tumor for adequate esthetics. Surgical excision of the tumor including bone is a successful treatment modality. However, it leads to considerable functional and esthetic deficits as well as long-term postoperative complications. Other non-surgical treatment modalities such as bisphosphonates have been suggested.[13] They reduce the osteoclastic activity bound to bone surfaces. Its use in adults has shown promising results in controlling FD-induced pain.[14] However, their long-term use should be limited, as they can lead to bone necrosis and are contraindicated in pregnant women. The role of RANK ligand inhibitors (denosumab) in reducing pain and growth should be evaluated.[15] Malignant potential is high in patients with polyostotic FD compared to those with MFD. However, our patient was followed every six months for the early detection of malignant changes, if any.

Conclusion

A single confined case of FD in either the maxilla or mandible is rare. The differentiation of these benign bone disorders from malignant ones is difficult. To establish a diagnosis of FD, adequate patient history, thorough examination, and radiographic assessment are often sufficient. Each case is different with peculiar symptoms and unique clinical findings. Therefore, the management of this condition must be pertinent to the site of involvement.

Declaration of conflicting interests

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Ethical approval

Our institution does not require ethical approval for reporting individual cases or case series.

Informed consent

Written informed consent was obtained from the patient for their anonymized information to be published in this article.

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