Radiological and Histological Observation in a Woman Patient of Fibrous Dysplasia

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ÖZET

Fibröz displazili bir bayan hastanın radyolojik ve histolojik değerlendirilmesi

Fibröz displazi; fibröz dokunun, normal veya tam gelişmemiş kemik dokusu ile yer değiştirmesi veya düzensiz osteoid formasyonu ile karakterize bir hastalıktır. Bu hastalık çoğunlukla kemikle yavaş büyüme gösteren asemptomatik özelliktedir. Olgu sunumunun amacı 49 yaşındaki fibröz displazili bir bayan hastanın tanımlanmasıdır. Hastanın klinik bulguları ile birlikte, panoramik radyografi, konik ışıklı bilgisayarlı tomografi, sintigrafi ve histolojik bulguları da değerlendirilmiştir. Radyolojik ve histolojik muayene dental değerlendirmeler açısından oldukça önemlidir ve klinisyenler klinik ve radyolojik olarak mevcut değişiklikler bakımından dikkatli olmalıdır.

Anahtar sözcükler: Fibröz displazi, konik ışıklı bilgisayarlı tomografi, sintigrafi

ABSTRACT

Radiological and histological observation in a woman patient of fibrous dysplasia

Fibrous dysplasia (FD) is a condition in which normal medullary bone is replaced by an abnormal fibrous connective tissue proliferation in which new, nonmaturating bone is formed. This disease most commonly present as an asymotomatic, slow enlargement of the involved bone. The aim of this report is to present a case of fibrous dysplasia in a 49-year-old female patient. In addition to clinical examination the patient was imaged using panoramic radiography, Cone Beam Tomography, scintigraphy and histological examination. Radiological and histological examinations are important in dental practice and clinicians must be watchful of the presence of clinical and radiological abnormalities.

Key words: Fibrous dysplasia, cone beam tomography, scintigraphy

INTRODUCTION

Fibrous dysplasia (FD) is a noninherited bone disease in which abnormal differentiation of osteoblasts leads to replacement of normal marrow and cancellous bone by immature bone and fibrous stroma. It is often asymptomatic and frequently incidentally detected on radiographs taken for unrelated clinical indications. However, fibrous dysplasia may be complicated by pathologic fracture, and rarely by malignant degeneration (1,2). It can affect one bone (monostotic form), or multiple bones (polyostotic form), and latter may form part of the McCune-Albright syndrome (MAS) or of the Jaffe-Lichtenstein syndrome (JLS). Uncomplicated monostotic lesions are asymptomatic and do not cause significant deformity. The polystotic variety is more severe with the involvement of multiple sites (3).

Gender prevalence of FD is equal. The monostotic form is more common and affects the 20 to 30 years age group: polystotic FD has its onset mainly in children younger than 10 years of age, the lesions grow with the child, stabilize after puberty, and most commonly involve craniofacial bones, ribs, and metaphysis or diaphysis of the proximal femur or tibia. The ratio of occurrence of polyostotic to monostotic FD is 3:7. Sign and symptoms of FD include bone pain, pathological fractures and bone deformities. Serum alkaline phosphatase (ALP) is occasionally elevated, but calcium, parathyroid hormone, 25-hydroxyvitamin D, and 1.25-dihydroxyvitamin D levels in most cases of FD are normal. The craniofacial bones are affected in about 10% of cases of monostotic FD and 50% to 100% of cases of polystotic FD. When only the cranial and facial bones are affected by FD the term craniofacial FD is used. FD of the

jaws affects the maxilla more frequently than the mandible. It is a sporadic condition that results from a postzygotic mutation in the GNAS1 gene (2-4).

HISTOPATHOLOGICAL FEATURES; The typical microscopic findings of FD consist of irregulary shaped trabeculae of immature (woven) bone in a cellular, loosely arranged fibrous stroma. The bone trabeculae are not connected to each other. They often assume curvilinear shapes, which have been likened to Chinese script writing. Serial biopsy specimens in some cases have shown that histopathologically classic fibrous dysplasia of the jaws undergoes progressive maturation to a lesion consisting of lamellar bone in a moderately cellular connective tissue stroma (1,5).

RADIOGRAPHIC FEATURES; The radiographic appearance varies with the stage of maturity of the lesion. In early lesions the area may be radiolucent, becoming more radiopaque as more bone is formed. A mature lesion retains none of the normal architecture of trabecular bone, having replaced it with abnormal bone that produces a "ground glass" or "orange peel" pattern on radiographs. Expansion of the cortical plates and displacement of tooth roots is common. The lamina dura is usually obscured and the cortical plates are thinned (3,5).

PREFERRED RADIOGRAPHIC EXAMINATION; Plain radiography is the first-line study. Usually, the diagnosis is straightforward when typical features are present. Computed tomography (CT) scanning may be required to assess complex regions such as the spine, pelvis, chest, and facial skeleton. Bone scintigraphy has a limited role in the detection of subtle pathologic fractures. In fibrous dysplasia, the features on a bone scan are nonspecific for diagnostic purposes. Magnetic resonance imaging (MRI) may be necessary to assess cord compression when the spine is involved (6-8).

The aim of this report is to present a case of fibrous dysplasia in a 49-year-old female patient.

CASE REPORT

A 49-year-old female was referred to the Department of Oral Diagnosis and Radiology, Faculty of Dentistry, Marmara University, with a complaint of radiopaque lesion on the anterior of lower jaw. The clinical features were non-specific on the lingual alveolar mucosa of the anterior region and the patient's medical and dental histories were non-contributory. The overlying mucosa was healthy and did not present any differentiation from surrounding tissue. Lymphadenopathy was not evident. Panoramic radiographs revealed bone involvement of radiolucent lesion on the anterior of mandibula and posterior of maxilla (Figure 1). The area was examined with Cone Beam Tomography (CBCT) imaging (Morita 3D Accuitomo 170, Japan). Many and varied reconstructions could be carried out using CBCT. This reconstruction showed a mixed radiolucent and radiopaque lesion caused by the fibrous displasia in the anterior mandibular and bilateral maxillary region (Figure 2, 3,4,5). The provisional clinical diagnosis were "fibrous dysplasia, hyperparathyroidism or Paget's

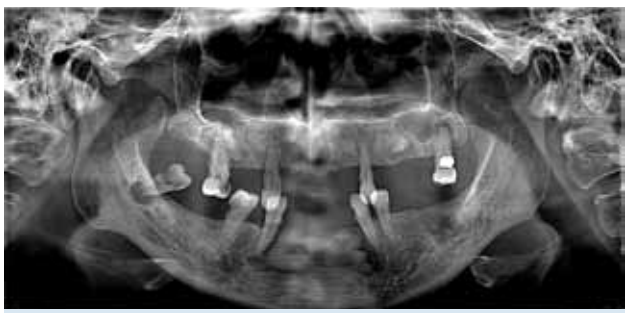

Figure 1: Panoramic radiographs revealed bone involvement of radiolucent lesion on the anterior of mandibula and posterior of maxilla

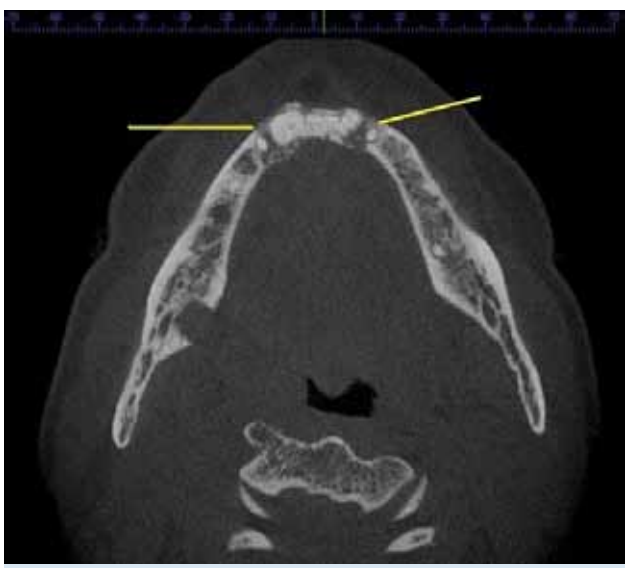

Figure 2: Axial view of the lesion

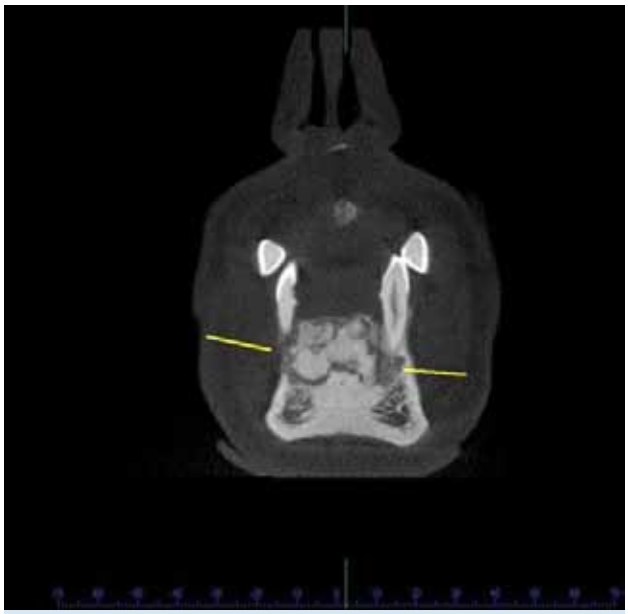

Figure 3: Coronal view of the patient showing lesion in the mandibula

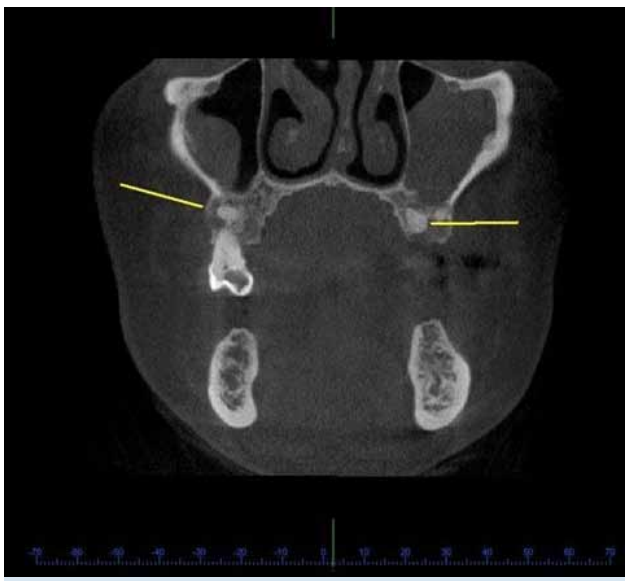

Figure 4: Coronal view of the patient showing lesion in the maxilla

disease ". An incisional biopsy was performed on the lingual alveolar mucosa of the anterior region under local anaesthesia at the Department of Oral and Maxillofacial Surgery and the specimen was sent for histopathological examination to the Department of Oncologic Cytology and Tumor Pathology, Institute of Oncology, Istanbul University. Histopathologic features revealed primitive mesenchymal cells consisting the fibrous tissue, and new

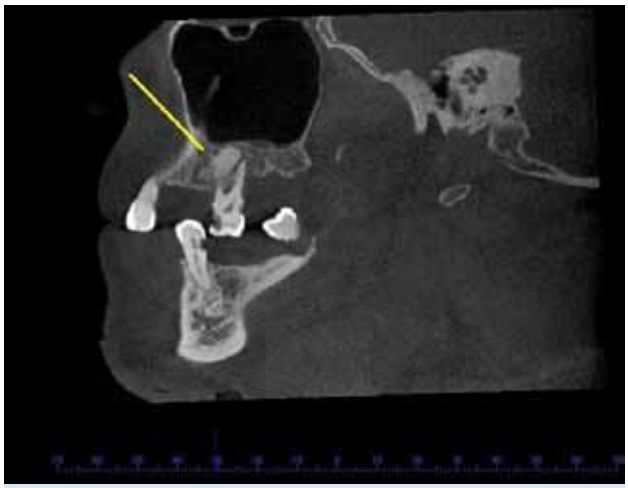

Figure 5: Sagital view of the patient

bone formation ending as numerous bone lamella of different shapes within this fibrous tissue. Some of these bone lamellae join with each other in order to make larger areas of bone (Figure 6 a, b). The histopathologic diagnosis was fibrous dysplasia. Therefore, the area was examined with bone scintigraphy to evaluate any body bone involvement. A bone scan showed increased uptake in the anterior region of the mandibula and bilateral posterior region of the maxilla (Figure 7). Serum ALP, calcium, parathyroid hormone, 25-hydroxyvitamin D, and 1.25-dihydroxyvitamin D levels were normal. The patient's operation was planned under local anaesthetic with intravenous sedation.

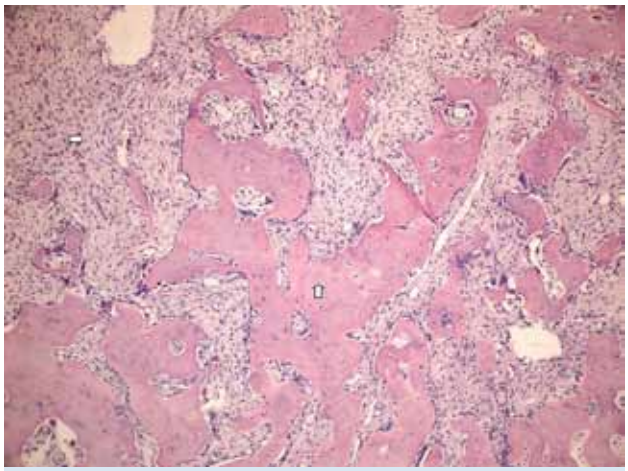

Figure 6a: Histopathologic appearance of fibrous dysplasia of the lower alveoler jaw bone. Hematoxylene-Eosin, x100⇨Fibrous tissue ⇧bone.

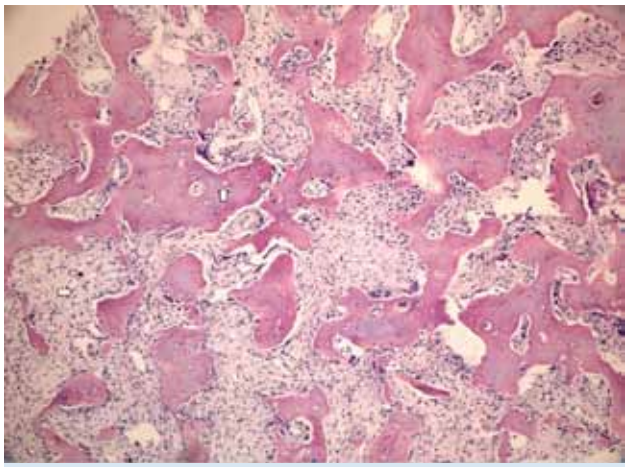

Figure 6b: Histopathologic appearance of fibrous dysplasia of the same case. Hematoxylene-Eosin, x100. ⇨Fibrous tissue, ⇧ bone. Primitive mesenchymal cells consisting the fibrous tissue, and new bone formation ending as numerous bone lamella of different shapes within this fibrous tissue. Some of these bone lamellae join with each other in order to make larger areas of bone as in Figure 6b.

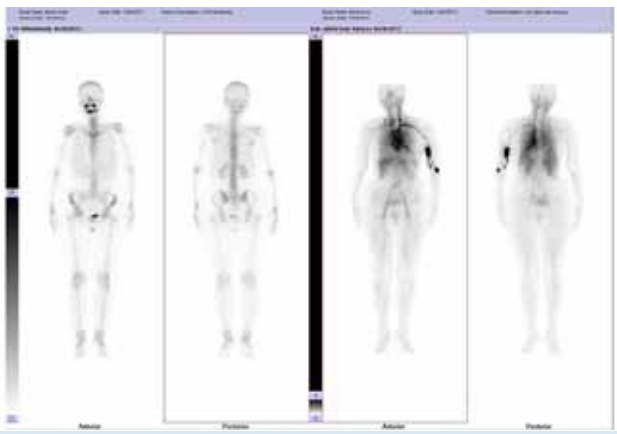

Figure 7: A bone scan showed increased uptake in the anterior region of mandibula and bilateral posterior region of maxilla.

DISCUSSION

Fibrous dysplasia is a non neoplastic developmental hamartous disease of the bone. It is often found childhood and usually stabilises in adulthood. Craninofacial involvement is found in about 10% of cases of monostotic FD. This monostotic type affects the calvaria, the skull base, the zygoma, the maxilla and the mandible. In 50% of cases the maxilla is more often involved than the mandible (9-12).

Although fibrous dysplasia is a benign condition, in an unfavourable location it can cause considerable diagnostic and therapeutic problems (13,14). In most cases the radiographic characteristics of fibrous dysplasia and the clinical information are sufficient to allow the practitioner to make a diagnosis without a biopsy (1,12). There are reports of exaggerated growth from stimulation of a lesion during surgical intervetion in young patients (4). A consultation with a dental radiologist is advisable. The radiologist may supplement the examination with CT, which can give a more accurate, three-dimensional representation of the extent of the lesion and can serve as a precise baseline study for future comparisons, as it was done in our case. It is reasonable to continue occasional monitoring of the lesion or ask the patient to report any changes.

The three general approaches for the treatment of FD involve monitoring, conservative or radical surgery. Currently there are no uniformly accepted guidelines for treatment of FD; the general step involves frequent follow up examinations unless the patient does not show any symptoms. Surgical treatment becomes necessary whenever clinical symptoms occur (12). The maxillary region and also other regions of the facial skeleton such as the orbital region, the cranial base, the mandibular or the zygomatic bone can present special problems to the surgeon because of their anatomical relationships to important structures (15). With most lesions, growth is complete at skeletal maturation; therefore orthodontic treatment and cosmetic surgery may be delayed until this time. In a few studies, patients with primarily polyostotic disease have been effectively managed with bisphosphonate therapy. However, further studies are needed for adequate assessment of the risks and benefits of bisphosphonate therapy (13,14).

CONCLUSION

Fibrous dysplasia is common benign bone disease existing in monostotic and polyostotic forms. The imaging features of FD are characteristic (panoramic view, CT,CBCT), although not specific, and depend on the underlying histopathology of a given lesion. Knowledge of the various appearances and associations of fibrous dysplasia is important to ensure the accurate diagnosis and appropriate management of FD.

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