OLGU SUNUMU CASE REPORT

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Fibrous Dysplasia in Mandibular Posterior Region: Report of Two Cases

Mandibula Posterior Bölgede Fibröz Displazi: İki Olgu Sunumu

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ABSTRACT Fibrous dysplasia (FD) is a fibro-osseous lesion of the bones. Although the exact is unknown, it has recently been reported that somatic mutations are effective. This mutation causes focal congenital failure in bone formation and hardness. Visual and functional changes are seen as a result of this mutation. These are 3 types of FD; monostatic, polyostatic, craniofacial. This report describes 2 cases of monostotic FD in mandibular bone. The clinical features, radiological and histopathological findings and treatment have been evaluated.

Keywords: Fibrous dysplasia of bone; fibrous dysplasia; monostotic; mandible

ÖZET Fibröz displazi (FD), kemiklerin fibro-osseöz bir lezyonudur. Kesin nedeni bilinmemekle birlikte son zamanlarda nedenin somatik mutasyon olduğu bildirilmiştir. Bu mutasyon kemik oluşumu ve sertliğinde fokal konjenital yetersizliğe yol açar. Bu mutasyon sonucunda görsel ve fonksiyonel değişiklikler görülür. FD'nin 3 alt tipi; monostotik, poliostotik ve kraniyofasiyaldir. Bu rapor, mandibular kemikte 2 monostotik FD vakasını tanımlamaktadır. Klinik özellikleri, radyolojik ve histopatolojik bulguları ve tedavisi değerlendirildi.

Anahtar Kelimeler: Kemiğin fibröz displazisi; fibröz displazi; monostotik; mandibula

Fibrous dysplasia (FD) is the replacement of normal bone marrow by abnormal new fibrous connective tissue.[1] Due to the fibrous connective tissue, the mechanical strength of the bone decreases, causing bone pain and pathological fractures.[2] Mutation in the guanine nucleotide receptor on the cell surface (alpha stimulating 1 gene located at chromosome 20q132) causes FD.[3-5] The increase in the activity of G protein increases the proliferation and abnormal differentiation of osteoblast progenitor cells. Studies have established a link between the G_S α mutation and the increased production of interleukin-6 stromal cells, which increase osteoclast activity.[6,7] Therefore, the pathogenesis of FD causes bone formation and destruction.[8] FD usually occurs in the first and second decades of life and progresses slowly and asymptomatically. Women are affected 2 times more than men. The maxilla is more affected than the mandible, and the monostatic type is usually seen.[3] Facial deformity, hearing problem, visual change, nasal obstruction, pain, paresthesia and malocclusion may develop in areas with abnormal bone formation as a result of FD.[9] Treatment may not be necessary in most cases which are moderate or mild. Surgery may be indicated if the bone deformity is huge and causes compression of tissues. Bisphosphonates have recently been reported to use as a medical therapy in severe cases as they can reduce increased bone resorption.[10] This case report presents the treatment of 2 monostotic craniofacial FD in the mandible region by surgical remodulation and conservatively.

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CASE REPORT

Two patients referred to the Oral and Maxillofacial Surgery Department of Dental Faculty of Ankara University with a painless swelling on the mandible with no systemic history and trauma. Findings of these 52- and 39-years old female patients were very close to each other. Facial asymmetry were observed on extraoral examination. Both of the patients had a solid, painless swelling causing an increase in hemi mandibular body size. There were no other asymmetry in other parts of their body. There were asymmetry in the hemimandible on intraoral examination. Both patients had tooth loss in the area with FD. The alveolar crest were enlarged in the buccal and occlusal planes. No malocclusion was observed but there was local mucosal irritation on the edentulous alveolar crest. A panoramic X-ray and 3D tomography were taken. X-ray showed an enlarged left mandible in one patient and enlarged right mandible in the other patient with change in pattern of the cancellous bone (Figure 1). The cone beam computed tomography scan showed a radio dense mass with ground-glass appearance involving the hemi mandibular bone and its expansion caused facial asymmetry. No cortical defect was observed except from the bony expansion in tomographic examination (Figure 2). Investigations such as hemogram, serum calcium, vitamin D and serum alkaline phosphatase and parathormone were performed. All parameters were close to normal limits. An incisional biopsy was performed (Figure 3, Figure 4). As a result of the histological examination, a lesion characterized by small trabecules with irregular lamellation, cement-bone-like, and spindle cellular, fibroblastic stroma with continuity with the host bone is observed (Figure 5). As a result of clinical, radiological and histological findings, the lesions were diagnosed as FD in both of the patients. The first patient was followed up for 2 years after the diagnosis without performing any operation. The area with FD causing malocclusion and asymmetry was remodeled and followed for 2 years in the other patient. After 2 years, no change was observed (Figure 6). The patients are planned to be rehabilated with im-

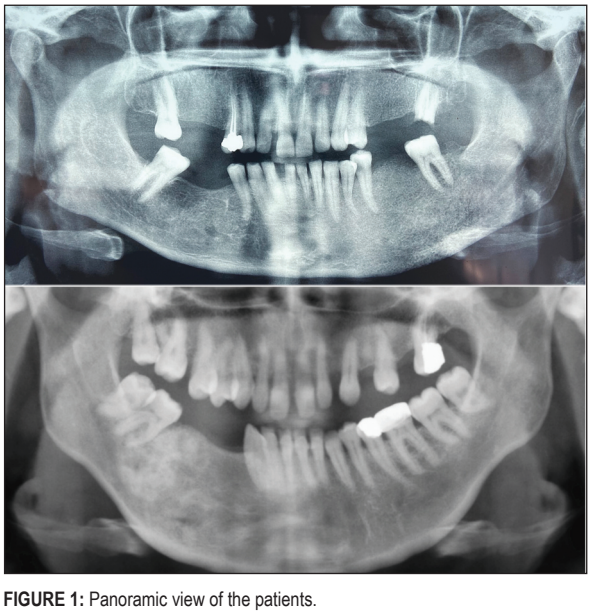

FIGURE 1: Panoramic view of the patients.

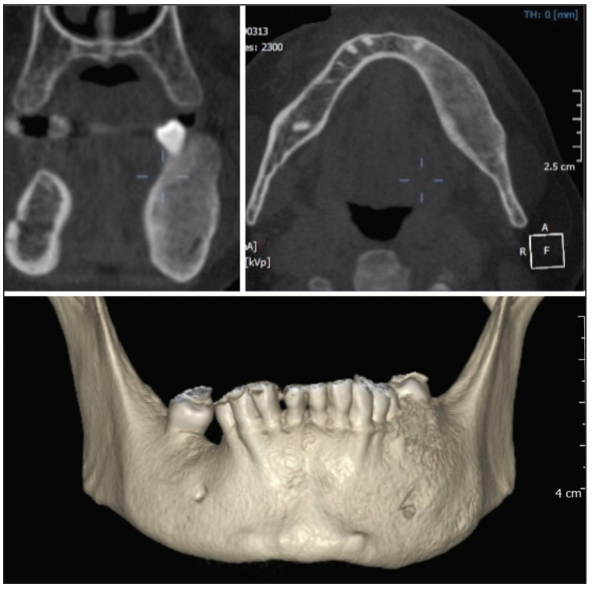

FIGURE 2: The cone beam computed tomography of the lesion.

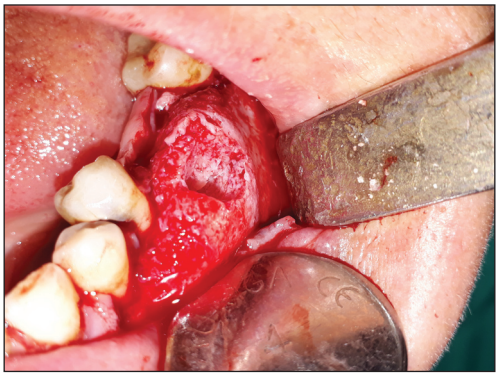

FIGURE 3: Intra-operative view of the fibrous dysplasia.

plant supported prothesis. Informed consent was obtained from the patients for the use of their information and images in the article.

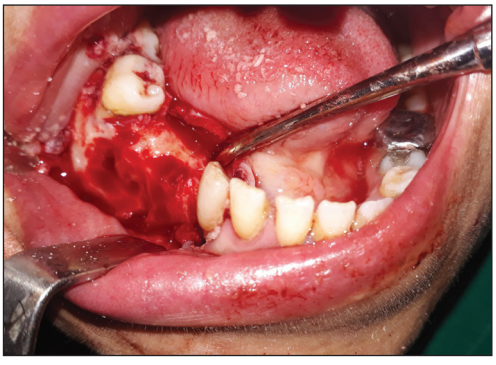

FIGURE 4: Intra-operative view of the fibrous dysplasia.

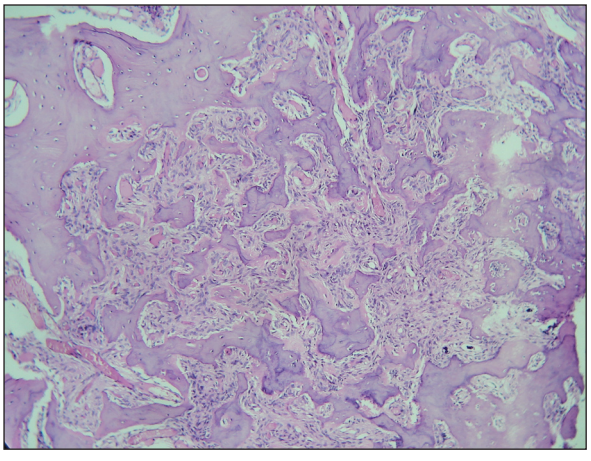

FIGURE 5: Histological view of the specimen.

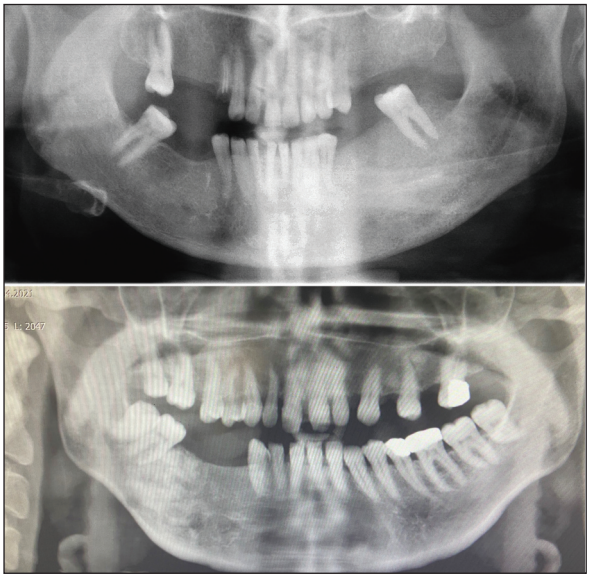

FIGURE 6: After 2 years, panoramic view of the patients.

DISCUSSION

Benign fibro-osseous lesions (BFOLs) are difficult to diagnose by the pathologist. The three major forms of BFOLs are FD, ossifying fibroma and cemento-osseous dysplasia. They differ from each other by their anatomical locations, symptoms and histological appearance.[11] FD is characterized by the differentiation of the bone with abnormal proliferation into fibrous connective tissue.[3]

It usually occurs in the first or second decade of life; is asymptomatic, progresses slowly, and affects women twice as often as men. FD causes facial asymmetry and deformity. Patients' complaints are generally aesthetic, hearing problem in severe cases, visual change, nasal obstruction, pain, paresthesia and malocclusion. Although the maxillary bone is more affected than the mandible, mandible was more affected in this report. And may involve one (monostotic) or less commonly two or more bones (polyostotic).[3] In this report, both of the patients were women as literature mentioned and they had malocclusion.

The definition of craniofacial FD is used when more than one bone is affected in the craniofacial region.[11] Polyostatic FD is often associated with McCune-Albright Syndrome. These changes in the bone can be seen from mild expansion to large expansions that cause cosmetic problems and disrupt its structure.[12] Our patients did not have any accompanying syndrome.

In large lesions, bisphosphonates have been used to reduce bone pain and bone fragility, but it increased the risk of osteonecrosis in the jaws.[13] In mild lesions, contouring the bone that causes asymmetry and malocclusion is performed. Increasing the life comfort of patients should be aimed and they must be followed up regularly. In the first case, after the enlarged bone region causing malocclusion and asymmetry was remodeled, bone overgrowth was not observed during the 2 years follow-up period and it was decided to be rehabilitated with implants. The other patient was only followed up for 2 years without the need for surgical procedure. At the end of 2 years follow-up, no change was observed and it was decided to be rehabilited with implant supported prothesis.

As a conclusion, the treatment method for FD should be considered according to the degree of pathologic and clinical changes and the patients should be followed regularly.

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Conflict of Interest

No conflicts of interest between the authors and / or family members of the scientific and medical committee members or members of the potential conflicts of interest, counseling, expertise, working conditions, share holding and similar situations in any firm.

Authorship Contributions

Idea/Concept: *Mert Özlü, Serpil Altundoğan;* ***Design:*** *Mert Özlü;* ***Control/Supervision:*** *Serpil Altundoğan;* ***Data Collection and/or Processing:*** *Mert Özlü, Serpil Altundoğan;* ***Analysis and/or Interpretation:*** *Serpil Altundoğan;* ***Literature Review:*** *Mert Özlü;* ***Writing the Article:*** *Mert Özlü;* ***Critical Review:*** *Serpil Altundoğan.*

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