Monostotic Fibrous Dysplasia of the Mandible

Cagri Delilbasi[1], Ediz Deniz[2], Isin Dogan Ekici[3]

[1]Department of Oral and Maxillofacial Surgery, İstanbul Medipol University, School of Dentistry, Ataturk Bulvari, Unkapani-Istanbul, Turkey. [2]Department of Oral and Maxillofacial Surgery, Yeditepe University, Faculty of Dentistry, Istanbul, Turkey. [3]Department of Pathology, Yeditepe University Faculty of Medicine, Istanbul, Turkey.

Abstract

Fibrous Dysplasia (FD) is a developmental disorder of bone that can affect one bone (monostotic type) or multiple bones (polyostotic type). The disease can be associated with hyperpigmentation and endocrinological disorders. It is usually observed in adolescents and young adults and comprises 7% of benign bone tumors. It has a predilection for long bones as well as the craniofacial skeleton. The etiology of FD is not clear but genetic predisposition is suspected. The diagnosis is based on radiological and histopathological examination. There are different treatment approaches including monitoring, medical treatment or surgery. In this article, we present a case of monostotic mandibular fibrous dysplasia of the mandible with clinical, radiographic and histopathological features.

Key Words: Fibrous dysplasia, Mandible, Fibro-osseous lesion

Introduction

Fibrous Dysplasia (FD) is a benign chronic fibro-osseous disease which is characterized by abnormal development of bone or fibrous tissue [1-6]. Mutation in the guanine nucleotide-binding protein-coding gene in early stages of life is responsible in the etiology of the disease [7]. A cell population with this genetic feature, which is not capable of producing normal tissue, instead generates a substitute of disorganized woven bone. The lesion has mainly two forms; monostotic form defines single bone involvement whereas polyostotic form defines multiple bone involvement [1,3,6]. The most affected bones are; femur, tibia, ribs and facial bones. It accounts for about 2.5% of all bone tumors and 7.5% of the benign bone neoplasms [8]. The involvement of facial and cranial bones in FD occurs in approximately 50% of patients with the polyostotic form and in 10-27% of patients with monostotic form. In many patients, the disease is recognized in early childhood, grows slowly and stabilizes in early adult life. Cases of FD showing rapid growth with extensive bone destruction during childhood are rare. A special form of FD is McCune-Albright syndrome, which is characterized by endocrine dysfunction including acromegaly, Cushing syndrome, hyperthyroidism and vitamin D resistant rickets. The most common features of this syndrome are; precocious puberty in girls and brownish pigmentations of the skin (café-au-lait spots) with irregular borders [3,7-9].

The aim of this article is to present clinical, radiological and microscopic findings of a case diagnosed as FD of the mandible.

Case Report

A 42-year-old female patient referred to our clinic with pain in the right lower teeth. Intraoral examination revealed missing upper right first premolar, lower right and left first and second molars and as well as carious lesions in upper right first molar and lower right second molar. Extra-oral examination findings were normal; the overlying skin was normal in color, texture and temperature and there was no motor or sensory deficit. Panoramic radiography revealed poorly defined ground glass radiopaque lesion around the roots of lower anterior and premolar teeth (*Figure 1*). Vitality test of the affected tooth was positive. Dental CT revealed a diffuse lesion that caused expansion and thinning of cortical bone, containing osseous and fibrous structures in the center (*Figure 2A*). Perforation of the buccal cortex was noticed on 3D reconstruction (*Figure 2B*). She had neither any systemic disease nor history of trauma. Incisional biopsy was performed and processed for histopathological examination. A texture like fibrous tissue mixed with soft bone was encountered at biopsy. Microscopic findings were consistent with fibrous dysplasia. The patient was referred to nuclear medicine section to investigate the possible involvement of other bones by scintigraphy. Whole body scintigraphy showed hyperperfusion and hyperemia associated with increased osteoblastic activity in the mandible. There was no sign of any other bone involvement in the skeleton (*Figures 3A and 3B*). Biochemical parameters including serum calcium, phosphorus and alkaline phosphatase levels were normal. Under local anesthesia the lesion was totally removed by curettage and processed for histopathogical examination. Mental foramen and the nerve were carefully exposed and protected during the surgery. Root canal treatment was

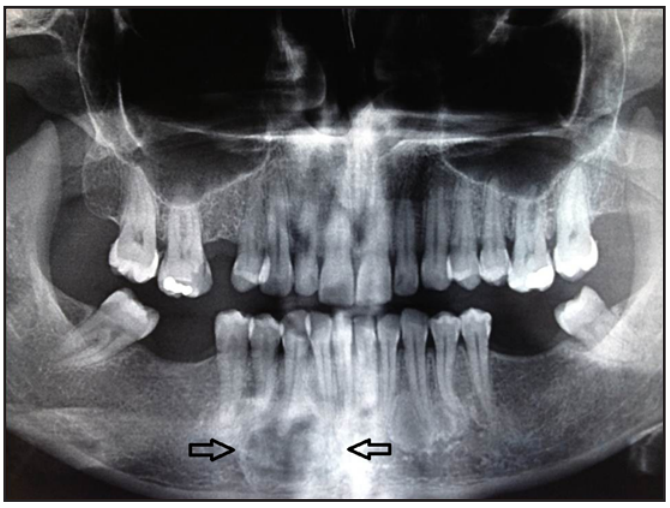

Figure 1. *Ground-glass appearance of the lesion on panoramic radiography.*

Corresponding author: Cagri Delilbasi, Department of Oral and Maxillofacial Surgery, İstanbul Medipol University, School of Dentistry, Ataturk Bulvari, Unkapani-Istanbul, Turkey, Tel: +90-212-453-49-50; Fax: +90-212-531-75-55; e-mail: cdelilbasi@yahoo.com

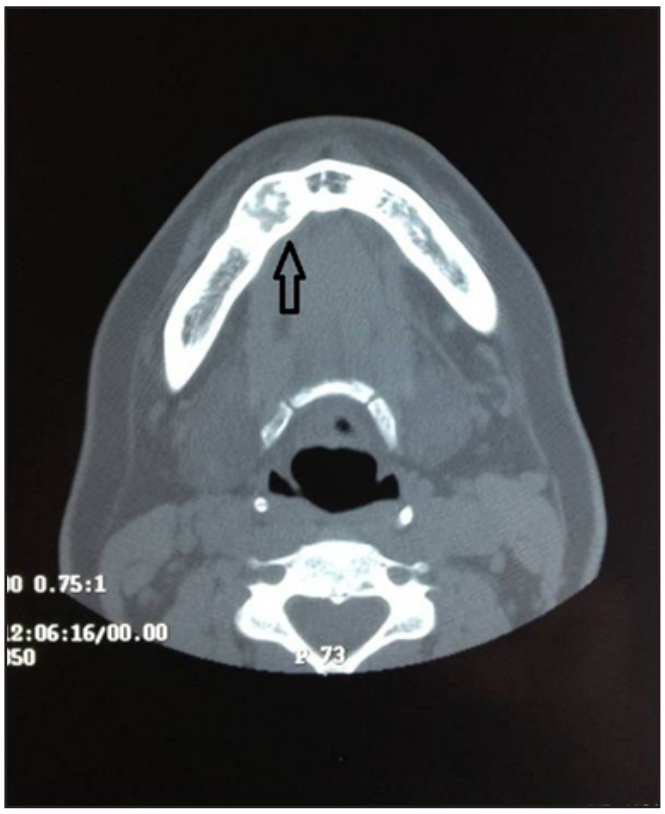

Figure 2A. *Fibro-osseous lesion causing expansion at the buccal plate.*

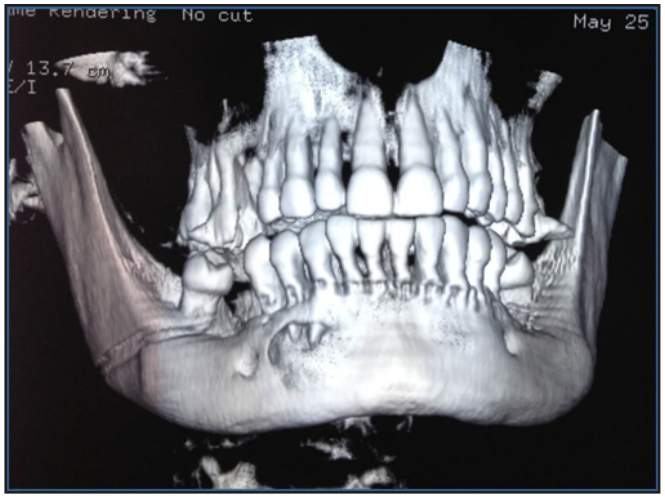

Figure 2B. *3D reconstruction of the mandible showing perforation.*

performed for lower right premolars and canine. Postoperative microscopic findings confirmed the diagnosis of FD (*Figures 4A and 4B*). The patient has been followed-up for 3 years without any signs of recurrence.

Differential Diagnosis and Discussion

FD is a benign bone lesion characterized by replacement of normal bone by fibro-osseous connective tissue exhibiting varying degrees of osseous metaplasia. Although it is not a common disease, it may cause fractures in long bones, deformities and severe bone pain [4,7,8]. Differential diagnosis of FD is based on clinical, radiographic, laboratory and histological findings. Pathological conditions mimicking FD can be classified as other fibro-osseous lesions (cemento-osseous dysplasia, ossifying fibroma), bone cysts, cementoma, Paget's disease, Cherubism, hyperparathyroidism, chronic sclerosing osteomyelitis and osteogenic sarcoma. Fibro-osseous lesion is a common heading of a group of diseases, but the treatment option varies from none to surgical recountering and complete removal. The lesion most frequently confused with FD is Ossifying Fibroma (OF). Diagnosis of these two lesions should be based on clinical, radiographic and microscopic findings. OF discloses a well-delineated border whereas FD is expansive and diffuse with ill-defined margin [6,10,11]. Paget's disease is characterized by abnormal resorption and deposition of bone. Radiographic findings may resemble to those of cemento-osseous lesions and in case of clinical jaw expansion, further evaluation is necessary for differential diagnosis [12]. Chronic sclerosing oteomylelitis presents increased radiodensity adjacent to the apex of a tooth with thickened periodontal ligament. Clinical expansion is not seen. The lesion does not show a radiolucent border as in cement-ooseous dysplasia [12].

There is not a definite etiology of FD, but it is believed to arise from abnormal activity in the bone forming mesenchymal tissue. Trauma was also believed as a causative factor but the disease has been reported at the time of birth. Most of the reported FD cases are monostotic type, and maxilla is more frequently affected than mandible. Monostotic type is seen more often in children and adults [1,8]. On the contrary in our case it affected a middle-aged woman and the lesion located in the mandible. Radiological assessment of FD can be made by plain radiography, scintigraphy, magnetic resonance imaging and computerized tomography scans. The typical appearance of FD on radiographs is a thin cortex, well-marginated borders and radiolucent ground-glass appearance. Total or partial deformation or enlargement of the bone can be noticed [7,8,13]. Laboratory findings including serum calcium and phosphorus levels are normal, but serum alkaline level may be elevated but not a specific marker for the lesion [9,10]. Histopathological features include benign

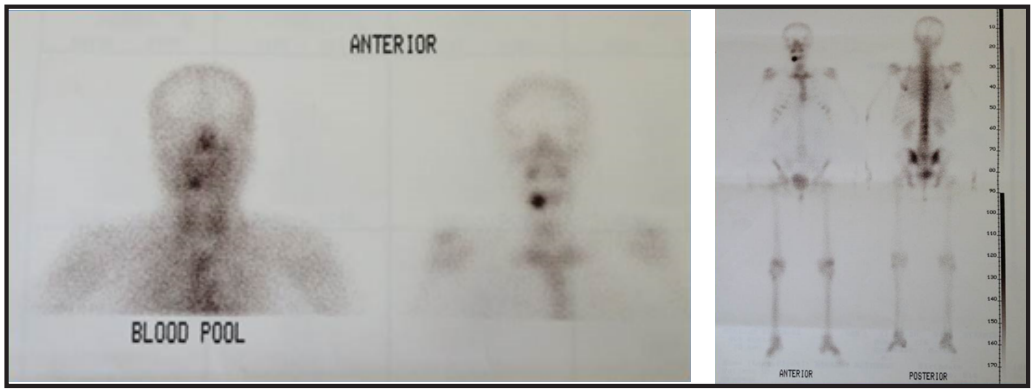

Figure 3A and 3B. *Whole body scintigraphy revealed only involvement of the mandible but not other parts of the skeleton.*

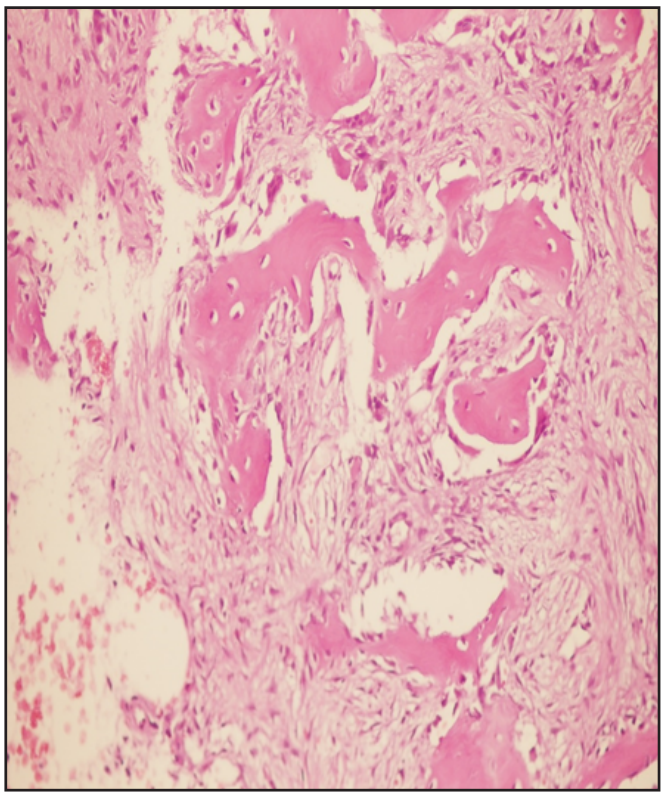

Figure 4A. *Stromal fibroblasts producing bony matrix without morphologic evidence of osteoblastic cells at the periphery of the bony spicules could be seen (H&E, x200).*

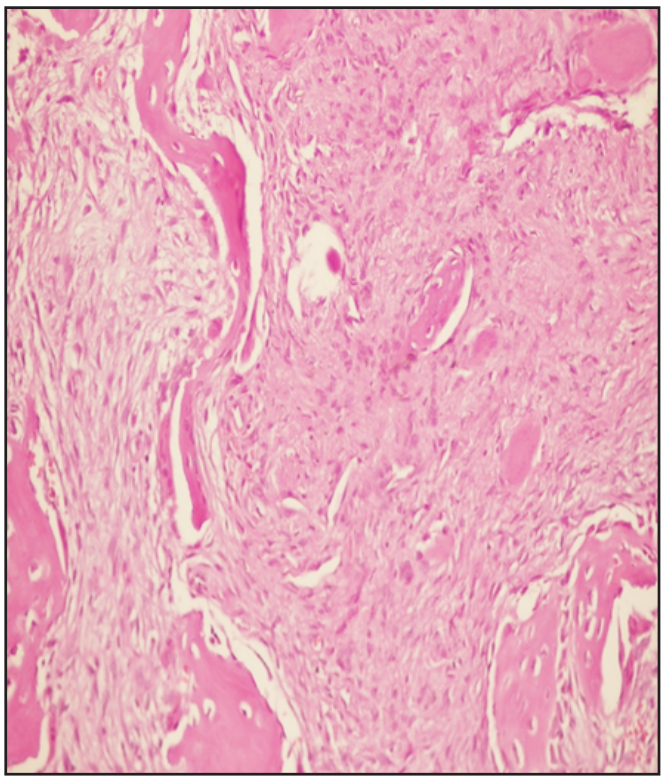

Figure 4B. *Thin irregular-shaped woven bone that resembles membranous ossification (Chinese characters) could be seen (H&E, x200).*

fibroblastic tissue arranged in a loose, whorled pattern and irregular spicules of woven bone with typical osteoblastic rimming embedded in fibrous tissue. The bony trabeculae vary in shape, which resemble Chinese characters, but this appearance is not prerequisite for diagnosis of FD [5,10]. Malignant transformation is rare but it may transform into sarcomas. Radiation therapy is important factor for the malign transformation of FD, therefore it is not included in the treatment options for FD [7,8]. Recurrence of FD is rare in adults but more common during the growth period. The risk of recurrence increases in cases where conservative surgery is used and the lesion cannot be totally removed. The surgical treatment of FD ranges from biopsy specimens to modeling osteotomies, bi-maxillary osteotomies and calvarial remodeling. The aim of the surgical therapy for FD is to prevent pathological fractures, control the pain and to reduce bone deformities [1,2,11,14]. Only curative surgical treatment was performed in our case. No recurrence was seen in the 3-year-follow-up period.

Clinicians should be familiar with differential diagnostic criteria and features of FD in order to plan accurate treatment and to predict risk of recurrence.

References

1. De Melo WM, Sonoda CK, Hochuli-Vieira E. Monostotic fibrous dysplasia of the mandible. *Journal of Craniofacial Surgery*. 2012; **23**: e452-e454.
2. Mendonca Caridad JJ, Platas F Jr. Fibrous dysplasia of the mandible: Surgical treatment with platelet-rich plasma and a corticocancellous iliac crest graft-report of a case. *Oral Surgery, Oral Medicine*, *Oral Pathology, Oral Radiology, and Endodontics*. 2008; **105**: e12-e18.
3. Menon S, Venkatswamy S, Ramu V, Banu K, Ehtaih S, et al. Craniofacial fibrous dysplasia: Surgery and literature review. *Annals of Maxillofacial Surgery*. 2013; **3**: 66-71.
4. Kruse A, Pieles U, Riener MO, Zunker Ch, Bredell MG, et al. Craniomaxillofacial fibrous dysplasia: a 10-year database 1996-2006. *British Journal of Oral and Maxillofacial Surgery*. 2009; **47**: 302-305.
5. Ozek C,Gundogan H,Bilkay U, Tokat C, Gurler T, et al. Craniomaxillofacial fibrous dysplasia. *Journal of Craniofacial Surgery*. 2002; **13**: 382-389.
6. Prabhu S, Sharanya S, Naik PM, Reddy A, Patil V, et al. Fibro-osseous lesions of the oral and maxillo-facial region: Retrospective analysis for 20 years. *Journal of Oral And Maxillofacial Pathology*. 2013; **17**: 36-40.
7. Assaf AT, Benecke AW, Riecke B, Zustin J, Fuhrmann AW, et al. Craniofacial fibrous dysplasia (CFD) of the maxilla in an 11-year old boy: a case report. *Journal of Cranio-Maxillo-Facial Surgery*. 2012; **40**: 788-792.
8. Olasoji HO, Ugboko VI, Nggada HA. Aggressive form of fibrous dysplasia of the mandible in childhood: Case report. *International Journal of Pediatric Otorhinolaryngology*. 2006; **1**: 89-92.
9. McCarthy EF. Fibro-osseous lesions of the maxillofacial bones. *Head and Neck Pathology.* 2013; **7**: 5-10.
10. El Deeb M, Waite DE, Jaspers MT. Fibrous dysplasia of the jaws: Report of five cases. *Oral Surgery, Oral Medicine, Oral Pathology, Oral Radiology, and Endodontics*. 1979; **47**: 312-318.
11. Suarez-Soto A, Baquero-Ruiz de la Hermosa MC, Minguez-Martínez I, Floría-García LM, Barea-Gámiz J, et al. Management of fibro-osseous lesions of the craniofacial area. Presentation of 19 cases and review of the literature. *Medicina Oral, Patología Oral y Cirugía Bucal*. 2013; **18**: e479-e485.
12. Neville BW, Dougles DD, Allen CM, Bouquot JE. Oral and

Maxillofacial Pathology (2nd Edn) Philadelphia, Pennsylvania: WB Saunders Company; 2002.

13. Sontakke SA, Karjodkar FR, Umarji HR. Computed tomographic features of fibrous dysplasia of maxillofacial region. *Imaging Science in Dentistry*. 2011; **41**: 23-28.

14. Yetiser S, Gonul E, Tosun F, Tasar M, Hidir Y. Monostotic craniofacial fibrous dysplasia: the Turkish experience. *Journal of Craniofacial Surgery*. 2006; **17**: 62-67.