J Istanbul Univ Fac Dent 2016;50(2):38-42.
http://dx.doi.org/10.17096/jiufd.99328 *CASE REPORT*

MONOSTOTIC FIBROUS DYSPLASIA PRESENTING IN MAXILLA: A CASE REPORT

Maksillada Monostatik Fibröz Displazi: Olgu Sunumu

Taha Emre KÖSE [1], Onur DİNÇER-KÖSE [2], Mehmet Ali ERDEM [2], Abdulkadir Burak ÇANKAYA [2], İlknur ÖZCAN-DUMAN [1]

Received: 27/10/2014
Accepted:25/02/2015

ABSTRACT

Fibrous dysplasia (FD) is a bone disorder in which fibrous tissue replaces normal bone. FD can be monostotic (10%–25%) or polyostotic (50–90%) and is typically diagnosed accidentally on radiographs. Craniofacial lesions are typically unilateral and are diagnosed by clinical assessment and radiographic evaluation. This report describes the case of a 50-year-old female patient who had presented with painless swelling of the right maxilla and was diagnosed with FD. Subsequent bone scintigraphy identified the lesion as monostotic. Based on the patient's age and the radiographic data, contour correction was performed. However, 1 year after surgery, the lesion regrew and the treatment was repeated.

Keywords: *Fibrous dysplasia; Monostotic; Maxilla*

ÖZ

Fibröz displazi(FD), normal kemik dokusunun yerini fibröz dokunun aldığı bir kemik hastalığıdır. Monostatik ve poliostotik formlarda gözlenebilir. Genellikle rastlantısal olarak tespit edilen fibröz displazi lezyonları, radyografilerde buzlu cam görüntüsüne neden olur. Kraniofasiyal bölgede genellikle tek taraflıdır. Kraniofasyal FD' lerin %10-25'i monostatik formda %50-90'ı polyostatik formdadır. Klinik ve radyografik değerlendirme ile ön tanısı konulabilir. Bu olguda sağ maksiller alanda ağrısız bir şişlik şikayeti bulunan ve fibröz displazi ön tanısı koyulan 50 yaşındaki hasta değerlendirildi. Hastanın kemik sintigrafisi neticesinde fibröz displazinin monostatik form olduğu tespit edildi. Hastanın yaşı ve radyografik değerlendirmesi sonucunda konservatif yaklaşımla cerrahi kontur düzeltmesi planlandı. Ancak 1 yıllık takip sonucunda lezyonun tekrar büyüdüğü tespit edildi ve tedavi prosedürleri tekrarlandı.

Anahtar kelimeler: *Fibröz displazi; Monostotik form; Maxilla*

[1] Department of Oral and Maxillofacial Radiology Faculty of Dentistry Istanbul University
[2] Department of Oral and Maxillofacial Surgery Faculty of Dentistry Istanbul University

Introduction

Fibrous dysplasia (FD), a chronic disease in which fibrous tissue replaces normal bone, was first observed by Lichtenstein in 1938 (1, 2, 3, 4, 5). Mutations in the α-stimulating activity polypeptide 1 gene (GNAS1), which encodes a guanine nucleotide-binding protein, lead to the development of FD (2, 6). Monostotic FD affects a single bone, whereas polyostotic FD affects multiple bones (7). McCune-Albright syndrome also presents with polyostotic FD, as do endocrine disorders and cases of abnormal skin pigmentation (6). Monostotic FD occurs more frequently than polyostotic FD and is typically asymptomatic (2, 8). In the maxillofacial region, the mandible and posterior maxilla are the most frequent locations of FD (9). In this article, we aim to report on the case of a patient with monostotic FD who was treated using a conservative contour correction approach.

Case Report

A 50-year-old female patient was referred to the Department of Oral and Maxillofacial Surgery, Faculty of Dentistry Istanbul University for evaluation of the right edentulous maxillary alveolar crest. Other than hypertension, the patient had no significant medical history, and no pathology was observed during the extraoral examination. Intraoral examination revealed expansion of the alveolar crest in the right edentulous premolar–molar area. Panoramic radiographs revealed the loss of trabecular structure and a ground-glass appearance between the right first incisor and second molar (Figure 1).

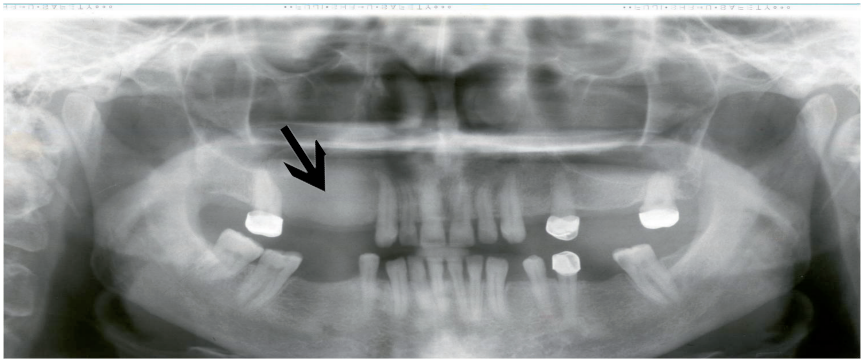

Figure 1. *Panoramic radiograph showing the lesion in the right maxillary region.*

Computerized tomography (CT) confirmed the ground-glass appearance, and revealed the horizontal expansion of the crest in the premolar -molar area (Figure 2). A biopsy sample was obtained from the lesion, and histopathologic analysis revealed irregular immature bone trabeculae in the fibrous stroma (Figure 3). Bone scintigraphy revealed that the lesion was monostotic and is limited to the right maxillary region (Figure 4). Surgery was performed using 2 ml articaine hydrochloride/epinephrine (Maxicaine®; VEM, Turkey) as the local anesthetic. A crestal incision originating at the right maxillary canine and extending to the right maxillary second molar was made, and a full-thickness mucoperiosteal flap was elevated. Contour correction of the alveolar crest was performed using a stainless-steel drill, and soft-tissue reduction was also performed. Amoxicillin/clavulanic acid (Augmentin-BID®, 1,000 mg; GlaxoSmithKline, England) and naproxen sodium (Apranax® Fort, 550 mg; Syntex Corporation, USA) were prescribed for twice-daily consumption. Sutures were removed 7 days after surgery. Following surgery, the patient attended routine follow-up assessments. After 1 year, buccal and palatal expansion of the lesion was observed (Figure 5). Panoramic radiographs obtained at that time were found to be similar to those obtained initially (Figure 6). A second operation was performed (Figure 7), after which the patient continued to attend routine follow-up assessments.

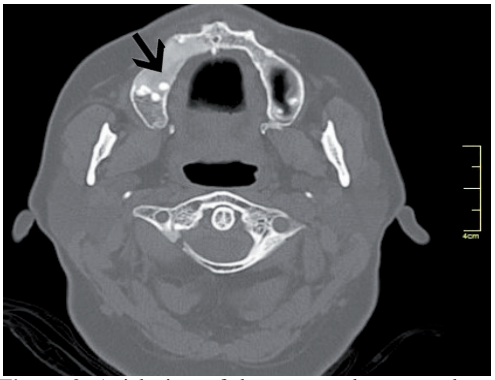

Figure 2. Axial view of the computed tomography showing the lesion's ground-glass appearance and position between the right central incisor and second molar.

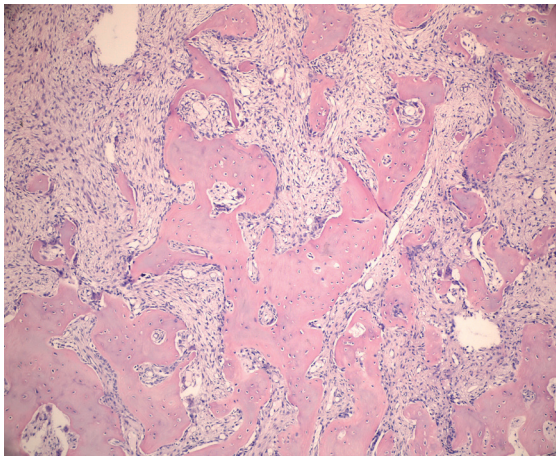

Figure 3. *Anastomosis of new bone in the stroma consisting of fibrocytes, fibroblasts, and new mesenchymal cells. H&E stain, 200× magnification.*

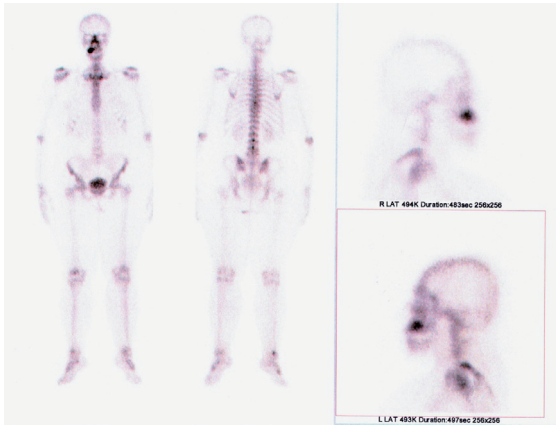

Figure 4. *Bone scintigraphy showing theincreased uptake at right maxillary region.*

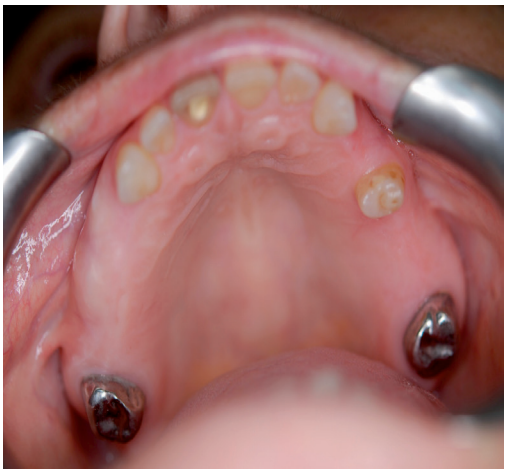

Figure 5. *Intraoral image showing the expanded alveolar crest.*

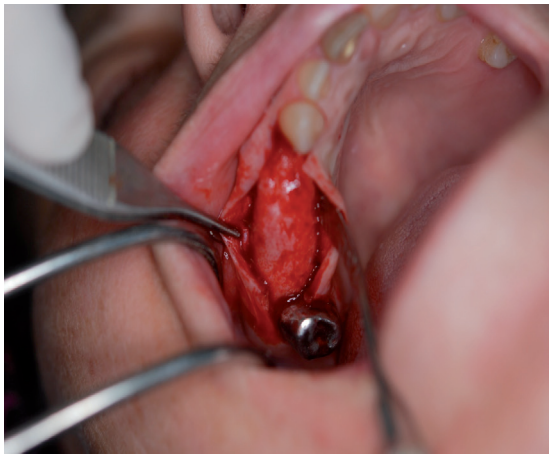

Figure 7. *View of the lesion, prior to the second surgery.*

Discussion

FD accounts for 7% of all benign bone disorders (2, 7, 9). As FD involves the replacement of cancellous bone by fibrous tissue, it is classified as a disorder and not a tumor (8). Some investigators have suggested that FD is more prevalent among females, whereas others have shown similar prevalence among males and females (2, 9). The symptoms of FD vary according to the bones that are affected; however, the most common symptom is the presence of a slow-growing painless swelling (5, 10, 11). Facial deformities, pain, headache, and visual or olfactory impairment are other symptoms of FD affecting the craniofacial bones (11). Typically, monostotic FD appears during the second or third decade of life and is passive (12). Most FD lesions in the craniofacial region are unilateral (2), as was observed in this study. Radiographs revealed that the FD in our patient had a ground-glass appearance. The appearance of FD depends largely on the stage of the lesion, with early-stage lesions appearing radiolucent or mottled and late-stage lesions appearing sclerotic (10, 13). CT or cone-beam CT images are generally used to determine the extent of FD in the craniofacial region (6). FD lesions typically mature and stop progressing after adolescence; however, for aesthetic reasons, surgery remains necessary (6, 7). Scintigraphy, which measures the radioisotope uptake of lesions, is a practical approach for the determination of whether FD is monostotic or polyostotic (14). Mutations in the GNAS1 gene result in FD. When mutations occur in undifferentiated stem cells during early embryogenesis, FD causes altered skin pigmentation or endocrine disorders. When mutations occur in skeletal progenitor cells,

FD lesions develop in multiple bones. The treatment options for FD include conservative shaving, surgical removal, and bisphosphonate treatment. Lesion regrowth after surgery occurs in 25–50% of patients, and although FD lesions may stop growing after skeletal maturation, some lesions continue to grow slowly (8, 10), necessitating follow-up assessment.

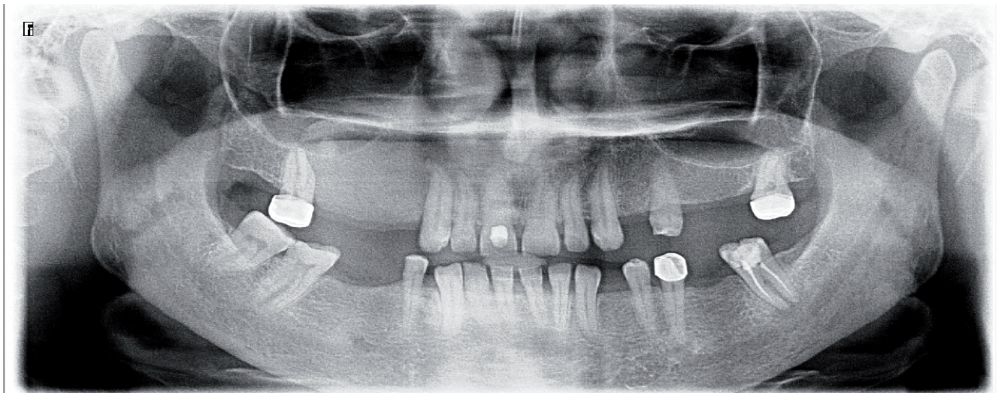

Figure 6. *Panoramic radiograph obtained 1 year after surgery.*

Conclusion

Lesions with unilateral involvement and a ground-glass appearance should signify FD to clinicians. Additionally, because 50% to 90% of craniofacial FDs are polyostotic, patients with these lesions must be assessed to identify the involvement of other bones or for the presence of endocrine disorders.

Source of funding
None declared

Conflict of interest
None declared

References

1. Verma A, Jindal N, Singh V, Sethi P. Unusual presentation of monostatic fibrous dysplasia in zygoma. J Craniofac Surg 2013;24(6):e592-594.
2. Wu H, Yang L, Li S, Jin X, Xu J, Lu J, Zhang C, Teng L. Clinical characteristics of craniomaxillofacial fibrous dysplasia. J Craniomaxillofac Surg 2014;42(7):1450-1455.
3. Anitha N, Sankari SL, Malathi L, Karthick R. Fibrous dysplasia - Recent concepts. J Pharm Bioalliedn Sci 2015;7(Suppl 1):S171-172.
4. Li J, Li H, Liu X, Han Z. Surgical treatment of polyostotic craniomaxillofacial fibrous dysplasia in adults: a case report and review of the literature. Int J Clin Exp Med 2015;15;8(9):16756-16764.
5. Wei WB, Chen MJ, Yang C, Zhang W, Wang Y. Decompression of the inferior alveolar nerve to treat the pain of the mandible caused by fibrous dysplasia - Case report. Int J Clin Exp Med 2015;8(10):19535-19539.
6. Nityasri V, Haris PS, Bose T, Balan A. Fibrous dysplasia - A 13-year retrospective radiographic analysis in a south Indian population. Dentomaxillofac Radiol 2011;40(5):282-289.
7. Hakim DN, Pelly T, Kulendran M, Caris JA. Benign tumours of the bone: A review. J Bone Oncol 2015;2;4(2):37-41.
8. Lietman SA, Levine MA. Fibrous dysplasia. Pediatr Endocrinol Rev 2013;10(Suppl 2):389-396.
9. De Melo WM, Sonoda CK, Hochuli-Vieira E. Monostotic fibrous dysplasia of the mandible. J Craniofac Surg 2012;23(5):e452-454.
10. Neville BW DD, Allen CM, Bouquot JE. Oral and maxillofacial pathology. St. Louis, Missouri: Saunders Elsevier, 2009, p.635-640.
11. Tsakiropoulou E, Konstantinidis I, Chatziavramidis A, Constantinidis J. Olfactory dysfunction as the first presenting symptome of cranial fibrous dysplasia. BMJ Case Rep 2013. doi: 10.1136/bcr-2013-200432
12. Chandar VV, Priya, Amita. Bilateral fibrous dysplasia of the mandible in a 7-year-old male patient - A rare case. J Indian Soc Pedod Prev

Dent 2010;28(2):126-129.
13. Nair PP, Bhargava D, Thomas S, Shreenivas K. Immature fibrous dysplasia: A mixed radio-opaque radiolucent lesion. BMJ Case Rep 2013;3:2013. doi: 10.1136/bcr-2012-007934.
14. Park HJ, Cho MS, Lee SS. Fibrous dysplasia of the inferior turbinate. Int J Clin Exp Pathol 2013;6(3):531-535.

Corresponding Author:
Onur DİNÇER KÖSE
Department of Oral and Maxillofacial Surgery
Faculty of Dentistry Istanbul University
34093 - Çapa Fatih, Istanbul / Turkey
Phone: +90 212 414 20 20/30289
e-mail: dt.onurdincer@hotmail.com