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Case Report Open Access

Monostotic Fibrous Dysplasia: A Case Report

Canıtezer G*, Gunduz K, Ozden B and Kose HI
Department of Oral and Maxillofacial Radiology, Faculty of Dentistry, Ondokuz Mayıs University, Turkey

Abstract

Fibrous dysplasia (FD) is a benign fibro-osseous bone disease of unknown etiology and uncertain pathogenesis. When bone maturation is completed, indicating the occurence of stabilization is a strong evidence of mechanism. The lesion frequently affects the craniofacial skeleton. The maxilla is affected twice comparing mandible and occurs more frequently in the posterior area. In this case, a 16 year-old female patient is presented who was diagnosed as having maxillofacial fibrous dysplasia.

Keywords: Fibrous dysplasia; Maxilla; Jaw diseases; Monostotic form

Introduction

Fibrous dysplasia (FD) is a benign fibro-osseous pathologic condition characterized by the replacement of bone with fibrous tissue [1,2].

The lesion is firstly described by Lichtenstein [3] in 1938. In 1937, Albright et al. [4] described a syndrome characterized by polyostotic fibrous dysplasia that included: areas of pigmentation, skeletal changes and endocrine failure (the most striking example is precocious puberty in girls).

If fibrous dysplasia affects only one bone, it is called monostatic FD, but multipl bones may also been affected this form is called polyostatic FD [1]. In addition to these forms, Jones [5] described hereditary familial form of localized FD which is called cherubism.

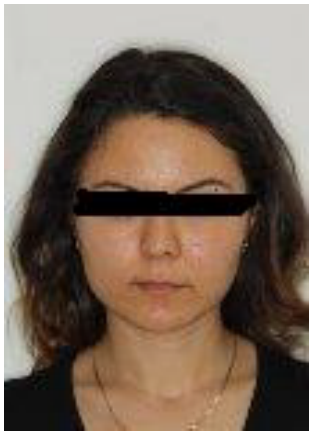

Figure 1: A 16 year-old female with facial asymmetry involvement of the left maxilla.

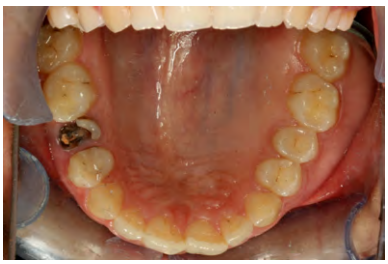

Figure 2: Intraoral sweelling of the buccal cortical plates.

The clinical findings are asymptomatic involved bone enlargement which causes facial asymmetry, loss teeth and facial deformity [6,7]. If the craniomaxillofacial bones are affected by FD, due to megacranium, the face of the patient is referred to 'lion face' [6].

The complications of the lesions involving sphenoid, orbital, frontal bones, are proptosis, visual disturbances, facial asymmetry and orbital dystopia [8,9]. The fifth nerve impairment, hearing loss and seizure disorders have been reported as neurological complications [10].

In our case, monostotic FD localized in the left side of maxilla that caused bone expansion and facial asymmetry is presented.

Case Report

A 16 year-old female referred to our clinic with the complain of sweeling on the left side of maxilla. In the history of the patient, the swelling was noticed by her friends 4 years ago but the patient didnt know when the swelling had occured. There was trauma history when she was a baby.

On extra oral examination diffused swelling of about 0.2 cm was present on left side of the face extending superio-inferiorly 0.5 cm below the cantho tragus line; to line joining corner of mouth to tragus and anteroposteriorly 0.2 cm away from the corner of the mouth to 1 cm in front of left tragus (Figure 1). On palpation the consistency was bony hard, nontender & no local rise of temperature. Buccal and palatal cortical plates were expanded (Figures 2 and 3). Overlying mucosa appeared normal, firm and was nontender.

Blood & Biochemical investigations showed alkaline phosphates 87 IU/l, Serum Calcium 9.8 mg% & Serum Phosphorus 3.1 mg% which were with-in normal range.

Radiological investigation includes panoramic radiography and cone beam CT (CBCT) scan. Panoramic radiography shows gross radio-opacity in the maxillary bone from first premolar region to tuberosity region. Which gives ground glass appearance (Figure 4). There was no root resorption or displacement of teeth on the effected

***Corresponding author:** Gözde Canıtezer, Department of Oral and Maxillofacial Radiology, Faculty of Dentistry, Ondokuz Mayıs University, 55139 Kurupelit, Samsun, Turkey, Tel: +90 362 3121919-3012, +90 505 8659063; Fax: +90 362 4576032; E-mail: gozde.canitezer@omu.edu.tr

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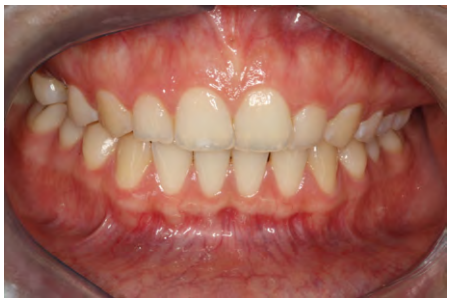

Figure 3: Intraoral sweelling of the palatal cortical plates.

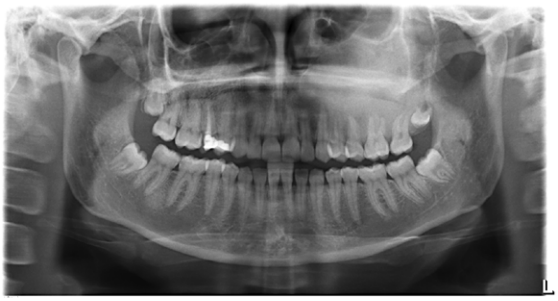

Figure 4: Panoramic radiograph shows a massive, radiopaque lesion which has ground glass appearance in the left side of maxilla that involves maxillary sinus and floor of the orbit.

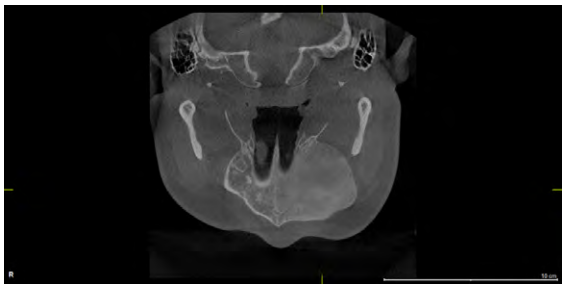

Figure 5: Axial and coronal scans of CBCT shows radiopaque lesion which extends from premolar area to the tuber maxilla, vertically the floor of orbit and comprises maxillary sinus on the left side.

side. CBCT scan coronal section revealed thick radio-opacity extending from premolar region to the tuberosity region on the left side. Complete obliteration of the lumen of the left maxillary antrum (Figure 5). The neurosensory examination didn't reveal any abnormalities.

After clinical and radiological findings the lesion was confirmed as fibrous dysplasia. The patient was referred to the Department of Oral and Maxillofacial Surgery for further treatment, but treatment was deferred till the age of 18 years. The patient was advised to visit her dentist regularly for ongoing re-examination and to observe and inform for any change in growth formation and appearance of pain. In the re-examine periodically is measured the level of alkaline phosphatase.

Discussion

FD is commonly benign lesion in which irregularly distributed spicules of bone lie in cellular fibrous stroma [11,12]. The lesion is believed to be hamartomatous develeopmental abnormality of bone of unknown etiology [13]. However, there is some evidence that the etiology of FD may be local infection or trauma [14]. In our case, we found a trauma history in the childhood of patient.

Studies of FD show no sexual predilection except for Mc Cune Albright syndrome which affects females almost exclusively. FD is seen maxilla more than mandible and occurs frequently in the posterior area [15]. This condition is being supported with our report. The polyostotic form usually is seen in children younger than 10 years, where as monostotic form typically is found in an slightly older age group [15]. Ozek et al. [13] reported in the series of 16 patients with FD of the craniomaxillofacial bones, one patient was in his first decade, 11 patients were in their second decade, 3 patients were in their third decade and one patient in the series was in his fourth decade when the symptoms occured. Hence our patient was in her second decade, our case supported to the case of Ozek et al. [13], Keijser et al. [16] reported the case with 13 patients after 20 years of age, the two of cases are polyostotic and the rest of cases are monostotic. In our case, the lesion is also monostotic form of FD.

In most cases, the radiographic and clinical findings are sufficient to allow the practitioner to diagnose without a biopsy [15]. The differential diagnosis with similar radiographic appearance such as ameloblastoma, ameloblastic fibroma, ameloblastic odontoma, ameloblastic fibro-odontoma, cental giant cell granuloma, odontogenic cyst, ossifying fibroma, osseous dysplasia, chronic sclerosing osteomyelitis and osteosarcoma should be considered [17]. In our case, there was no compelling indication to seek a biopsy, any sudden change in the clinical presentation or behavior of the lesion might warrant further investigation.

The density and trabecular pattern of FD lesions is variable. Early lesions may be more radiolucent than mature lesions and in rare cases may appear to have granular internal septa, giving the internal aspect a multilocular appearance. The abnormal trabeculae usually shorter, thinner, irregularly shaped and more numerous than normal trabeculae. This creates a variable radyopaque pattern, it may have a granular appearance ('ground-glass' appearance, resembling the small fragments of a shattered windshield), a pattern resembling the surface of an orange (peau d'orange), a wispy arrangement (cotton wool), or an amorphous, dense pattern. A distinctive characteristic is the organization of the abnormal trabeculae into a swirling pattern similar to a fingerprint [15]. Prapayasatok et al. [18] reported a case which was seen a rare radiographic 'sunray' appearance in 19-year-old woman. In this presented case, the panoramic radiography revealed a 'ground-glass' appearance of the affected area.

FD is a rare but severe bone disease which may cause fractures in long bones, deformities and bone pain. Although most lesions appear to stabilize when approaching bone maturity, some cases can reach severe asymmetry, visual impairment, diplopia, pain, paresthesia, proptosis, hearing loss, anosmia, nasal obstruction, epistaxis and epiphora. The patients generally complain of swelling (%94) and pain (%15) [19]. In our case, the patient referred to our clinic with complain of swelling.

When the lesion involves frontal bone, nasal bones, orbit, ethmoid, zygoma and upper maxilla, radical surgery is suggested but this approach is difficult in treatment of recurrences. Because radical surgery would possibly increase morbidity by removal of teeth. Hence, conservative treatment has been treatment of choice. Shaving and debridement of lesion are parts of conservative treatment [20]. Previous radiation and spontaneous degeneration may be the reason of malignant transformation. The frequency of sarcoma which occurs after radiotherapy, is high [21].

If FD is asymptomatic, it can be noticed incidentally in CBCT, CT scans and radiographs. If there is no symptom or evidence of progression during follow-up, surgical treatment isn't considered [13].

Recurrence of FD is rare when the lesion has occured in adults but it is seen more commonly in growth period [22]. Because of the conservatice surgery and unsuccessful removal of the lesion cause the increased risk of recurrence. Patients with craniofacial fibrous dysplasia have the risk of recurrence ranged from 15 to 20% [23,24]. Concentration of serum alkaline phosphatase (ALP) may be important marker for detection of the recurrence of the lesion. The patients who had FD, have higher ALP, this may be a reliable marker for estimating tumor progress and a sudden rise in ALP was correlated with the regrowth of FD by Park et al. [25].

The maxillary area and also other areas of cranio-maxillo-facial skeleton which includes the structures such as the orbital region, mandibular or tha zygomatic bone, the cranial base, may cause some problems to the surgeon because of their anatomical relationship to important structures [25].

Chapurlat et al. [26] reported that they provide evidence that in FD treated with intravenous pamidronate, bone turnover could be reduced, bone pain could be alleviated and radiological lesions could be improved. Few studies have reported the nonsurgical treatment of FD. At least in aggressive forms of FD, increased remodeling activity and bone resorption encouraged some open therapeutic trials with calcitonin in order to inhibit osteoclastic resorption [26]. Bell et al. [27] reported a decrease in elevated urinary excretion of hydroxyproline in a patient who had been treated with calcitonin for 16 days. There was no report about the effects of using calcitonin on clinical symptoms or X-ray abnormalities. The effects of treatment using disodium etidronate in an 18-year-old male patient diagnosing polyostotic fibrous dysplasia who had used calcitonin for 3 months as an unsuccessful treatment, were reported in one study [28]. Pamidronate is a potent inhibitör of bone resorption like other biophosphonates, a lasting effect on bone turnover [29]. Pamidronate has been successful treatment in Paget disease [30,31], malignant hypercalcemia [32], lytic bone metastases [33,34], multiple myeloma [35] and osteoporosis [36,37].

In the study of Chapurlat et al. pamidronate has led to decrease in pain severity and in the number of painful areas per patient and partially an open study permits to result [26].

Treatment of mandibular defects is complex and includes free vascularized flaps. Donor areas involve the iliac crest, radius, scapula and fibula [38-40]. Taylor et al. [41] firstly reported the transfer of the free fibula flap. In 1989, the method was used for the segmental mandibulectomy defects by Hidalgo [42]. The fibular flap is considered as one of the ideal flaps for long mandibular defects and is superior to the iliac crest for this purpose, especially in older patients [43]. Munoz Guerra et al. [44] reported 26 cases with using vascularized free fibular flap for mandibular reconstruction. Six patients weren't operated because of development of a vascular crisis and 5 flaps were successfully salvaged (83.3%). In 2 cases, a false positive exploration occured, in 2 vascular compression occured and in 1 case each of venous thrombosis and cervical hematoma occured. Except 1, the rest of fibular flaps survived (loss rate of flap 3.84%). In 2 patients, the donor area was closed primarily without a skin graft. In the rest of patients, skin grafting was performed to the donor site [44]. In addition, fibula provides sufficient amount of bone width and height to support osseointegrated implants which serve fort he support of overdentures and for functional reconstruction [45-47]. Barber et al. [48] evaluated the osseointegration of implants which were placed into fibular flap used in cancer patients following radiation therapy and subsequent hyperbaric oxygen therapy. 20 implants that were placed in the flap, has osseointegration clinicallyat the time the implants were uncovered and during the 6 months, the cases were followed up.

In our case report, we document the clinical and radiological features in a case of FD which is thought that trauma history may be a reason. The lesion exhibited extension into the extraosseous tissues. Our case with its clinical and radiographic features represents an addition to the literatüre of monostotic FD.

References

1. Jundt G (2005) Fibrous dysplasia. World Health Organization classification of tumours, pathology and genetics of head and neck tumours. Lyon: IARC Press.
2. Neville B, Damm D, Allen C, Bouquot J (2002) Oral and maxillofacial pathology. Philadelphia (PA): Saunders.
3. Lichtenstein L (1938) Polyostotic fibrous dysplasia. Arch Surg 36: 874-898.
4. Albright F, Butler AM, Hampton AO (1937) Syndrome characterized by osteitis fibroda disseminata, areas of pigmentation and endocrine dysfunction with precocious puberty in females, report of five cases. N Engl J Med 216: 727-746.
5. Jones WA (1965) Cherubism. Oral Surg 20: 648-653.
6. Chen YR, Noordhoff MS (1990) Treatment of cranio-maxillo-facial fibrous dysplasia: how early and how extensive. Plast Reconstr Surg 86: 835-842.
7. Karja J, Rasanen O (1972) Fibrous dysplasia of the jawbones: analysis of five cases. Acta Otolaryngol 74: 130-138.
8. Sassin JF, Rosenberg RN (1968) Neurological complications of fibrous dysplasia of the skull. Arch Neurol 18: 363-369.
9. Chen YR, Fairholm D (1985) Fronto-orbito-sphenoidal fibrous dysplasia. Ann Plast Surg 15: 190-203.
10. Schonder A (1977) Fibrous dysplasia of bone with proptosis. Am J Dis Child 131: 678-679.
11. Siegal GP, Bianco P, Dal Cin P (2013) Fibrous dysplasia. Pathology and genetics of tumours of soft tissue and bones. IARC, Lyon.
12. Dicaprio MR, Enneking WF (2005) Fibrous dysplasia. Pathophysiology, evaluation, and treatment. J Bone Joint Surg (Am) 87: 1848-1864.
13. Ozek C, Gundogan H, Bilkay U, Tokat C, Gurler T, et al. (2002) Craniomaxillofacial fibrous dysplasia. J Craniofac Surg 13: 382-389.
14. Arshad N, Kapala JT (1995) Monostotic fibrous dysplasia in an eight-year-old male: report of a case. ASDC J Dent Child 62: 145-147.
15. White SC, Pharoah MJ (2009) Oral Radiology: Principles and Interpretation. (6thedn), Elsevier.
16. Keijser LC, Van Tienen TG, Schreuder HW, Lemmens JA, Pruszczynski M, et al. (2001) Fibrous dysplasia of bone: management and outcome of 20 cases. J Surg Oncol 76: 157-166.
17. O'Connell KJ (1981) Bony enlargement of the left maxilla. J Am Dent Assoc 102: 340-342.
18. Prapayasatok S, Iamaroon A, Miles DA, Kumchai T (2000) A rare, radiographic 'sunray' appearance in fibrous dysplasia. Dento-Maxillo-Facial Radiology 29: 245-248.
19. MacDonald-Jankowski D (1999) Fibrous dysplasia in the jaws of a Hong-Kong population: radiographic presentation and systematic review. Dentomaxillofac Radiol 28: 195-202.
20. Yeow VK, Chen YR (1999) Orthognathic surgery in craniomaxillofacial fibrous dysplasia. J Craniofac Surg 10: 155-159.
21. Tanaka Y, Tajima S, Maejima S (1993) Craniofacial fibrous dysplasia showing marked involution postoperatively. Ann Plast Surg 30: 71-76.
22. Alvares LC, Capelozza AL, Cardoso CL, Lima MC, Fleury RN, et al. (2009) Monostotic fibrous dysplasia: a 23-year follow-up of a patient with spontaneous bone remodelling. Oral Surg Oral Med Oral Pathol Oral Radiol Endod 107: 229-234.

23. Rahman AMA, Madge SN, Billing K, Anderson PJ, Leibovitch I, et al. (2009) Craniofacial fibrous dysplasia: clinical characteristics and long-term outcomes. Eye 23: 2175-2181.

24. Valentini V, Cassoni A, Marianetti TM, Terenzi V, Fadda MT, et al. (2009) Craniomaxillofacial fibrous dysplasia: conservative treatment or radical surgery? A retrospective study on 68 patients. Plast Reconstr Surg 123: 653-660.

25. Park BY, Cheon YW, Kim YO, Pae NS, Lee WJ (2010) Prognosis for craniofacial fibrous dysplasia after incomplete resection: age and serum alkaline phosphatise. Int J Oral Maxillofac Surg 39: 221-226.

26. Chapurlat RD, Delmas PD, Liens D, Meunier PJ (1997) Long-Term Effects of Intravenous Pamidronate in Fibrous Dysplasia of Bone. J Bone Miner Res 12: 1746-1752.

27. Bell NH, Avery S, Johnston CC Jr (1970) Effects of calcitonin in Paget's disease and polyostotic fibrous dysplasia. J Clin Endocrinol Metab 31: 283-290.

28. Long A, Longhlin T, Towers RP, Mc Kenna TJ (1988) Polyostotic fibrous dysplasia with contrasting response to calcitonin and mythramycin: Aetiological and therapeutic implications. Int J Med Sci 157: 229-234.

29. Fleisch H (1989) Bisphosphonates: A new class of drugs in diseases of bone and calcium metabolism. Recent Results Cancer Res 116: 1-28.

30. Bijvoet OLM (1991) Disodium pamidronate therapy of Paget's disease. Elsevier Science Publishing Co., New York, NY, U.S.A.

31. Meunier PJ, Vignot E (1995) Therapeutic strategy in Paget's disease of bone. Bone 17: 489-492.

32. Ralston SH, Gallagher SJ, Patel U (1989) Comparison of three intravenous biphosphonates in cancer associated hypercalcemia. Lancet 1: 1180-1182.

33. Van Breukelen FIM, Bijvoet OLM, Van Oosternon AT (1979) Inhibition of osteolytic bone lesions by (3 amino 1 hydroxypropylilidene) 1.1 bisphosphonate (APD). Lancet 1: 803-805.

34. Burckhardt P, Thie´baud D, Pery L, Von Fliedner V (1989) Treatment of tumor induced osteolysis by APD. Recent Results Cancer Res 116: 54-66.

35. Berenson JR, Lichtenstein A, Porter L, Dimopoulos MA, Bordoni R, et al. (1996) for the Myeloma Study Group. Efficacy of pamidronate in reducing skeletal events in patients with advanced multiple myeloma. N Engl J Med 334: 488-493.

36. Reid IR, Wattie DJ, Evans MC, Gamble GD, Stapleton JP, et al. (1994) Continuous therapy with pamidronate, a potent bisphosphonate, in postmenopausal osteoporosis. J Clin Endocrinol Metab 79: 1595-1599.

37. Valkema R, Vismans FJFE, Papapoulos SE, Pauwels EKJ, Bijvoet OLM (1989) Maintained improvement in calcium balance and bone mineral content in patients with osteoporosis treated with the biphosphonate APD. Bone Miner 5: 183-192.

38. Sieg P, Zieron JO, Bierwolf S, Hakim SG (2002) Defect-related variations in mandibular reconstruction using fibula grafts. A review of 96 cases. Br J Oral Maxillofac Surg 40: 322.

39. Moscoso JF, Keller J, Genden E, Weinberg H, Biller HF, et al. (1994) Vascularized bone flaps in oromandibular reconstruction. A comparative anatomic study of bone stock from various donor sites to assess suitability for endosseous dental implants. Arch Otolaryngol Head Neck Surg 120: 36.

40. Beckers A, Schenck C, Klesper B, Koebke J (1998) Comparative densitometric study of iliac crest and scapula bone in relation to osseous integrated dental implants in microvascular mandibular reconstruction. J Craniomaxillofac Surg 26: 75-83.

41. Taylor GI, Miller GDH, Hamm FJ (1975) The free vascularized bone graft. Plast Reconstr Surg 55: 533-544.

42. Hidalgo DA (1989) Fibula free flap: A new method of mandible reconstruction. Plast Reconstr Surg 84: 71-79.

43. Peled M, El-Naaj IA, Lipin Y, Ardekian L (2005) The Use of Free Fibular Flap for Functional Mandibular Reconstruction. J Oral and Maxillofac Surg 63: 220-224.

44. Munoz Guerra MF, Gias LN, Rodriguez Campo FJ, Díaz González FJ (2001) Vascularized free fibular flap for mandibular reconstruction: A report of 26 cases. J Oral Maxillofac Surg 59: 140-144.

45. Dalkiz M, Beydemir B, Gunaydin Y (2001) Treatment of a microvascular reconstructed mandible using an implant-supported fixed partial denture: Case report. Implant Dent 10: 121-125.

46. Nagy K, Borbely L, Kovacs A, Fazekas A, Vajdovich I, et al. (1999) Implant-prosthetic rehabilitation after segmental mandibulectomy and bone grafting. J Long Term Eff Med Implants 9: 185-191.

47. Pogrel MA, Podlesh S, Anthony JP, Alexander J (1997) A comparison of vascularized and nonvascularized bone grafts for reconstruction of mandibular continuity defects. J Oral Maxillofac Surg 55: 1200-1206.

48. Barber HD, Seckinger RJ, Hayden RE, Weinstein GS (1995) Evaluation of osseointegration of endosseous implants in radiated, vascularized fibula flaps to the mandible: A pilot study. J Oral Maxillofac Surg 53: 640-645.