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SM Journal of Case Reports

Case Report

Monostotic Fibrous Dysplasia of Maxilla in a Postmenopausal Female- A Rare Case Report with Review of Literature

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Article Information

Received date: Mar 24, 2016
Accepted date: Apr 29, 2016
Published date: May 02, 2016

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Keywords Monostotic fibrous dysplasia; Postmenopausal female; Ground glass appearance

Abstract

Fibrous dysplasia is a condition characterized by excessive proliferation of bone-forming mesenchymal cells. It can affect one bone (monostotic type) or multiple bones (polyostotic type). It is usually observed in adolescents and young adults and comprises 7% of benign bone tumors. The etiology is not clear but genetic predisposition is suspected. It has a predilection for long bones as well as the craniofacial skeleton. The maxilla is the most commonly affected facial bone, with facial asymmetry being the usual complaint. The diagnosis is based on radiological and histopathological examination. There are different treatment approaches including monitoring, medical treatment or surgery. A 45-year-old female reported with a complaint of painless swelling on the left side of maxilla since 1 year. A diffuse intraoral bony hard, non tender swelling was seen in the left maxilla involving the premolar-molar region. Plain film radiographs and Computed tomography revealed ground glass appearance of the left maxilla. The lesion was excised and on histopathology showed features of fibrous dysplasia. Very few cases of Fibrous dysplasia manifesting in the older and postmenopausal age group are reported in the literature. Once diagnosed, routine follow-up should be done on a yearly basis with x-ray examination.

Introduction

Fibro osseous lesion is a commonly used term that includes bone dysplasia's, as well as neoplasms and other lesions of bone [1]. In 1891, Fibrous Dysplasia (FD) bone was first described by Von Recklinghausen. In 1938, Lichtenstein and Jaffe first introduced the term fibrous dysplasia [2]. In 1942, Lichtenstein and Jaffe reported two primary categories of the disease: monostotic fibrous dysplasia that involves only one bone and polyostotic fibrous dysplasia, which involves several bones. A monostotic form does not progress into a polyostotic form of the disease [3]. Craniomaxillofacial FD usually begins in childhood and slowly progresses through puberty, resulting in devastating facial deformities and functional deficits. Maxilla is more commonly affected than mandible and usually involves adjacent bones such as sphenoid, zygoma and frontal bone [4]. Very few cases of fibrous dysplasia manifesting in post menopausal patients are reported in literature. Hereby we are presenting a case of fibrous dysplasia of maxilla in an elderly female patient.

Case Report

A 45 year-old female reported to the department of Oral Medicine and Radiology with the complaint of painless swelling on the left side of maxilla since 1 year. The patient noticed the swelling three years ago which was gradual in onset, initially small in size and later progressed to attain the present size. Patient gave no relevant history of pain and fever. Her medical history was insignificant. No evidence of pigmentations was present on the body. She attained menstruation at the age of 12 yrs and has now reached menopause.

On extra oral examination facial asymmetry was evident due to slight fullness on the left malar region. On intra-oral examination a solitary, diffuse, roughly elliptical swelling measuring 4X2cms was seen extending antero-posteriorly from the distal surface of 23 to mesial surface of 27 and laterally to obliterate the buccal vestibule in relation to 24, 25, 26 and 27. Swelling was bony hard, non tender swelling with an undulated surface. Overlying mucosa appeared normal with no palpable pulsation (Figure 1).

OPEN ACCESS
ISSN: 2473-0688

How to cite this article Deepa JP, Samriddhi S, Puri G, Aravinda K, Dixit A, Gupta R, et al. Monostotic Fibrous Dysplasia of Maxilla in a Postmenopausal Female- A Rare Case Report with Review of Literature. SM J Case Rep. 2016; 2(2): 1025.

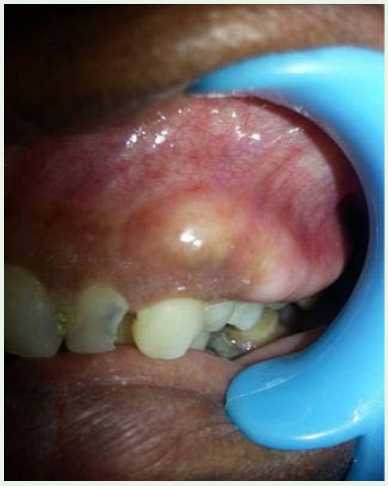

Figure 1: Intra-oral swelling extending from 23to 27 with buccal cortical plate expansion.

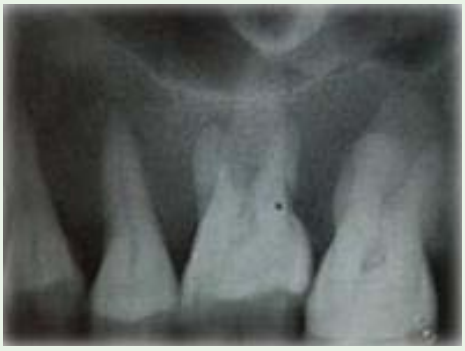

Figure 2: Intra-oral periapical radiograph demonstrating ground glass appearance and loss of lamina dura.

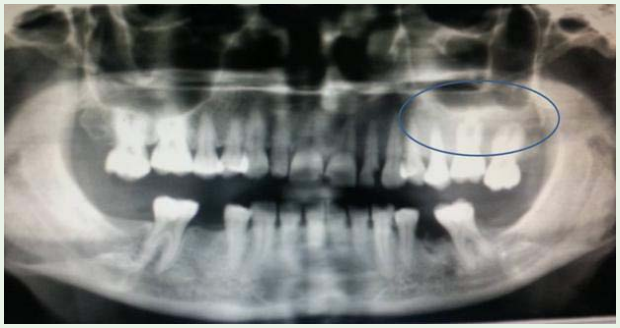

Figure 3: Panoramic radiograph depicting the ground glass appearance.

Based on the clinical findings a provisional diagnosis of exostoses was given. Ossifying fibroma, Paget's disease and fibrous dysplasia were considered under differential diagnosis. Loss of lamina dura was seen with 24,25,26,27 (Figure 2). Panoramic radiography shows gross radio-opacity in the maxillary bone from canine region to second molar region, which gives ground glass appearance (Figure 3). Skeletal survey was performed with plain films of pelvic bone, lower limbs and chest to rule out systemic involvement. No bony changes were evident.

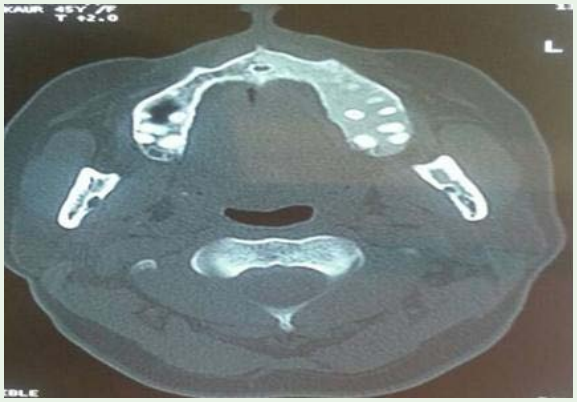

Figure 4: Axial section of maxilla depicting the expansion, thinning of the cortex with ground glass appearance of left maxilla.

CT scan of the lesion revealed ill defined expansible lesion causing medial-lateral widening of alveolar process of maxilla on left side (Figure 4). The lesion caused increased density of bone with ground glass appearance with thinning of the overlying cortex. There was no evidence of extension of the lesion into the left maxillary sinus and its shape and size was maintained.

Blood & Biochemical investigations showed alkaline phosphates 46.2 IU/l, Serum Calcium 9.8 mg% & Serum Phosphorus 3.1 mg% which were with-in normal range. A bone biopsy was performed from the left premolar region and the specimen sent for histopathological examination. The reports showed presence of trabeculae of demineralised bone arranged in a Chinese letter pattern. The bone trabeculae were lined by osteoblasts and showed the presence of resting lines. The marrow spaces in between were hypocellular with few chronic inflammatory cells in a fibrous connective tissue stroma (Figure 5).

Based on the clinical, radiological and histo-pathological findings a final diagnosis of Monostotic fibrous dysplasia was given. Surgical recontouring was done under local anesthesia in the department of Oral and Maxillofacial Surgery. The patient is under follow-up since six months with no recurrence.

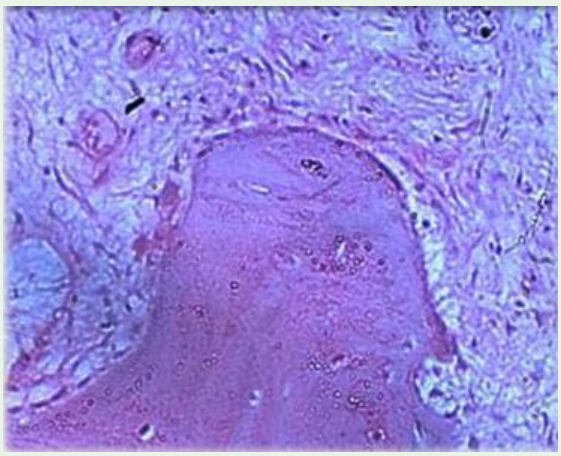

Figure 5: Hematoxylin & Eosin staining showing trabeculae of demineralised bone arranged in a Chinese letter pattern.

Table 1: Cases of FD manifesting in the older and postmenopausal age group.

S. no	Authors	Age at presentation	Sex	Site	Radiographic Features
1	Ogunsal[6] et al	47	F	Post Mandible	Radio-opaque
2	Ogunsalu[6] et al	47	F	Anterior & Posterior Mandible	Evidence of excess bone production
3	Lustig[7] et al	45	F	Craniofacial	Ground glass appearance
4	Gupta[8] et al	36	F	Left Maxilla	Ground glass appearance
5	Bhadada[9] et al	44	F	Polyostotic	Ground glass appearance
6	Gambhir[10] et al	63	M	Left Maxilla	Ground glass appearance
7	Nadaf[11] et al	40	F	Mandible Bilaterally	Ground glass appearance
8	Sasankoti[12] et al	44	F	Craniofacial areas & Mandible	Ill-defined Cotton wool appearance
9	Bijai[13] et al	64	M	Left Maxilla	Cotton wool to Ground glass appearance

Discussion

The present case focuses on fibrous dysplasia manifesting in the postmenopausal age group which is a rarity. According to Eisenburg and Eisenbud [5], majority of the cases burn out in early adulthood after skeletal maturity. In our case, the lesion might have been a slow growing lesion initially, then entered into a quiescent stage and reactivated in later stages of life. As it caused a deformity the patient reported to us. It is important to underscore here, the occurrence of these lesions in the later years of life. The possible mechanism behind this could be traced to trauma, hormonal changes or aberrant activity in the bone forming mesenchymal tissues, in addition to a delay in presentation. Search of literature revealed very few cases of FD reported in the older age group. Table 1 [6-13] summarizes the cases of FD reported in the older age group. The age ranged from 36 to 63 years with a female predilection (7:2). A systematic review by Mac- Donald-Jankowski14 and Slootweg and Muller 15 showed a female predominance and has been demonstrated in our case. The most common site involved was the mandible, maxilla and craniofacial bones in the descending order. One case of polyostotic FD was seen in a 44 year old female patient. The most common radiographic feature was ground glass appearance. In the study by Mac- Donald-Jankowski [14] and Li [15] and in the systematic review by MacDonald-Jankowski [16], fibrous dysplasia most commonly presented radiographically as a poorly defined ovoid (fusiform) area of dysplastic bone exhibiting a ground-glass appearance.

Fibrous dysplasia is a benign and rare bone disorder that often affects the craniofacial skeleton. It usually causes significant morphological and functional disturbance, such as malocclusion and disfigurement. Fibrous dysplasia represents about 5% of all bone tumors and over 7% of all benign tumours [17]. Cranial or facial bones are affected approximately in 30% of the patients. The average age of the patients with FD is 25.8 years (from 5 to 67) usually manifests before the 3rd decade of life, which is a striking difference in our case [18].

The monostotic form of fibrous dysplasia is the most common, comprising 70% of cases, most likely to quiesce at puberty. A typical monostotic lesion, usually present unilaterally, will involve the femur, tibia or ribs, with 25% occurring in the bones of the skull. Affection of the craniofacial bone is observed with 10% of the patients suffering from monostotic FD [18]. Schlumberger [19] first reported single bone involvement by the disease process and described it as "monostotic fibrous dysplasia". Monostotic fibrous dysplasia occurs with equal predilection for males and females, with a mild predominance for females. It is more common in children and young adults than in older persons. It occurs at the age of 20-30 years; with the mean age of occurrence is 27-34 years. Ribs and craniofacial bones are most commonly affected. Other bones affected include clavicle, tibia, femur etc. Maxillary involvement is more common than mandibular. Both maxillary and mandibular lesions occur as bony hard swellings that expand the jaws and are not tender on palpation. The overlying mucosa is almost invariably intact over the lesion. There may be some mal-alignment, tipping or displacement of the teeth due to the progressive expansible nature of the lesion [18].

Weil in 1922 recognized the case of polyostotic FD associated with skin lesions and endocrine disturbance. There are two apparently separate types of Polyostotic FD (PFD) which include Jaffe's type and Albright's syndrome [4].

The etiology of FD is not certain and is probably a genetic predisposition. It is assumed that the mutation is sporadic, postzygotic and located on the GNAS1 gene. This gene is found on chromosome 20q13 and is responsible for the formation of the alpha subunit of stimulating G-proteins. This mutation activates adenylate cyclase and consequently increases intracellular concentrations of cAMP resulting in abnormal osteoblast differentiation and production of dysplastic bone. On the other hand it stimulates release of several cytokines (mainly interleukin-6) which cause normal osteoclasts to congregate and increase bone resorption [20].

The radiographic appearance of fibrous dysplasia is a function of its histologic structure. If there is a predominance of osseous elements, the lesion is more opaque. A mixture of fibrous and bony elements produces a ground glass appearance, while predominance of fibrous elements produces a radiolucent cyst like picture. The radiographic appearance of the lesion may be seen in three basic patterns. It may appear as a small unilocular or large multilocular radiolucency with a well defied border. The lesion may be sharply defined when a sclerotic rim is present or may be ill defined if it lacks perilesional sclerosis and may fade into the adjacent normal bone [21]. In the second type, the pattern is similar except increased trabeculation renders the lesion more opaque and typically mottled in appearance. The third type is quite opaque with many delicate trabeculae giving a ground glass or orange peel appearance as seen in our case (Shafer et al 1983) [14,22].

Both CT and MRI are excellent imaging modalities in defining craniofacial fibrous dysplasia. Expansion of involved bone with a heterogeneous pattern of CT densities, along with intact thin cortex, is a characteristic of fibrous dysplasia. Fibrous dysplasia has an intermediate signal on T1- weighted and heterogeneous hypointense signal on T2- weighted MR images [23].

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. As per the literature, they all are usually within normal ranges in patients with Monostotic fibrous dysplasia as an evident in the present case [23]. Persons with extensive polyostotic FD may have hypophosphatemia, hyperphosphaturia and osteomalacia. ALP is a marker for detecting recurrence in FD [24].

Diagnosis of fibrous dysplasia is based on clinical examination, radiographic and histopathological findings. The microscopic appearances are those of a hypercellular and cytologically uniform fibrous stroma within which delicate and irregularly shaped trabeculae of woven bone are deposited. The configurations of these bony trabeculae are often referred to as resembling Chinese characters [25].

Therapeutic options for Craniomaxillofacial fibrous dysplasia include observation, medical therapy, conservative surgery and radical surgery. Though there are no uniformly accepted protocols for treatment of CFD, surgical therapy remains the mainstay of therapy for this disease and is directed at correcting or preventing functional deficits and achieving normal facial aesthetics. This can however result in recurrence, especially during the growth period and can range from 15-20%. Medical therapy has not occupied a prominent role in the management of fibrous dysplasia to date. Recognition of pathogenesis of this disease led to treatment with biphosphonates like Pamidronate [26]. They control bone erosion by the inhibition of osteoclastic action. It has been shown to reduce intensity of bony pain, bony resorption, and filling of lytic lesions.

Fibrous dysplasia is generally considered a benign, pediatric disease, which usually becomes dormant by adulthood. However, patients with FD should be made aware of the real potential for malignant transformation (1%) and should be on periodic follow up. The most common malignancy includes osteosarcoma (~70%), with fibrosarcoma (20%), chondrosarcoma (10%), and malignant fibrous histiocytoma (~4%) occurring less frequently [23].

Conclusion

This case report emphasizes on the occurrence of fibrous dysplasia in post menopausal patients. The possible mechanism behind this, need to be elucidated. Very few cases of FD manifesting in the older and postmenopausal age group are reported in the literature. Ours is one such case. Once FD is diagnosed, routine follow-up should be done on a yearly basis with x-ray examination. In addition, the patient should bring any changes in symptomatology such as (increased pain, weakness, deformity) to the attention of their physician. Plain radiographs, CT scans supplemented by histopathology plays an important role in the diagnosis of these lesions. Fibrous dysplasia should be considered in the differential diagnosis of bony enlargements in the elderly patients and a skeletal survey should be performed. Follow up of these patients is mandatory to recognize recurrence. Future trends should focus on genetic manipulation in the management of disease.

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