Fibrous dysplasia of the mandible: a case report

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Summary

Fibrous dysplasia is a benign fibro-osseous disease of bone of unknown etiology. Its occurrence in the maxillo-facial region is frequent. Because of the benign nature of the condition, the surgery itself should be relatively conservative. Recently, bisphosphonates have shown progress in the treatment of patients with fibrous dysplasia. In the current case report, combination of enucleation and performance of alendronate to the fibrous dysplasia is reported.

Key words: fibrous dysplasia, alendronate, mandible.

Introduction

Fibrous dysplasia (FD) of bone is a skeletal disorder characterized by extensive proliferation of fibrous tissue in bone marrow, leading to osteolytic lesions, fractures and deformations [1]. Initial symptoms are most often present during childhood or adolescence, as bone pain and repeated fractures. The other usual clinical findings are bone deformity and neurological compression, especially when the facial bones or the skull are involved [2]. It may be divided into three categories: monostotic (74%), polyostotic (13%) and craniofacial (13%) [3]. Monostotic forms are often asymptomatic [4].

Radiological signs of FD consist mainly of lytic and cystic lesions, with reduction of cortical thickness, and sometimes widening of the diaphysis. Computed tomography and magnetic resonance imaging can be used for differential diagnosis with malignancies [5]. It is generally held that the radiographic presentation of FD varies according to the degree of maturation which determines the degree of opacity [6]. Although the most frequent description is „ground glass“ [6], other patterns were reported by Waldron and Giansanti as „smoky and cloudy“ [7] and By Obisesan and coworkers [8] as „peau d'orange“, „whorled“ or „diffuse sclerosis“. The initial radiolucent stage of FD may suggest central giant cell granuloma, traumatic bone cyst, aneurysmal bone cyst [6], and cemento-osseous fibroma. The generally younger age of onset and its unilateral distribution allows FD to be readily differentiated from Paget's disease, which affects older patients and is frequently bilateral [9].

Fibrous dysplasia as an entity has been described for about 50 years but it's management remains controversial. Trials of therapy in craniofacial diseases are difficult to perform due to the great variation between presentations [10]. In our case report, we aimed to reveal the effects of locally applied alendronate to the lesion side.

Case report

A 45-year-old female presented to our clinic with a complaint of swelling in the anterior mandibular region *(Figures 1, 2)*. Radiological examination showed a cyst-like lesion in the mandible, located in the mental region *(Figures 3, 4)*. The incisional biopsy report revealed FD. The serum levels of creatinine, calcium, phosphate and alkaline phosphatase were all under normal levels *(Table 1)*. Enucleation and curettage of the lesion was planned and they were done under general anaesthesia. During the operation, the teeth from left canine to the right second premolar were extracted. After all the curet-

tage procedure, corticocancellous bone grafts (Tutodent®) soaked with alendronate were placed in the operation area and covered with a membrane *(Figure 5)*. The specimen is investigated under x100 magnification *(Figures 6, 7)* and pathology report named the lesion as "Fibrous Dysplasia". The suture removal of the wound lesion was performed 10 days later and the postoperative period was uneventful. After 4 years of follow-up, no recurrence was seen.

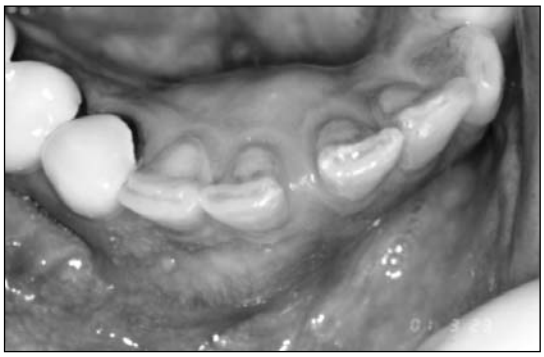

Figure 1. Clinical presentation of the patient revealing swelling in the anterior region

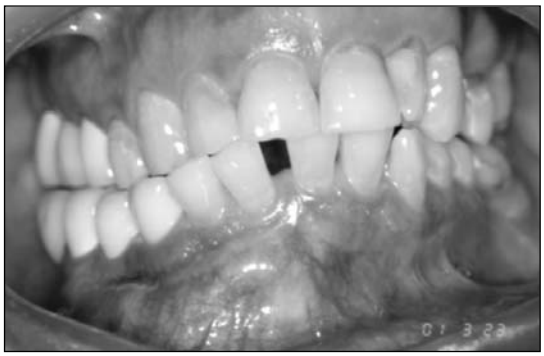

Figure 2. Clinical presentation of the patient revealing swelling in the anterior region

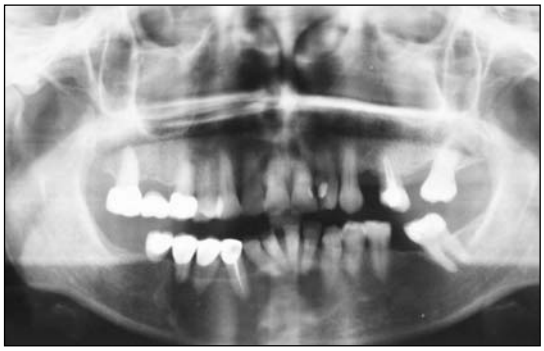

Figure 3. Radiographic view of the lesion

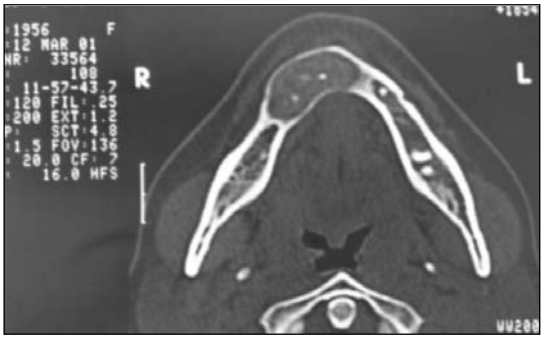

Figure 4. Radiographic view of the lesion

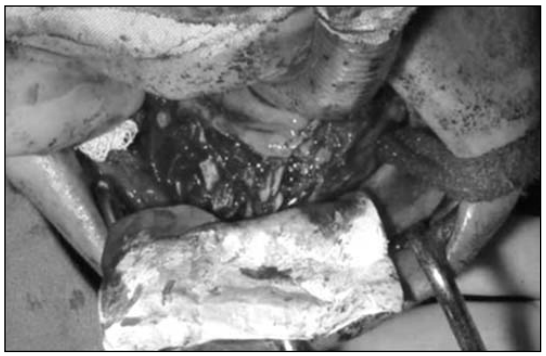

Figure 5. Reconstruction of the cavity with bone grafts soaked with alendronate

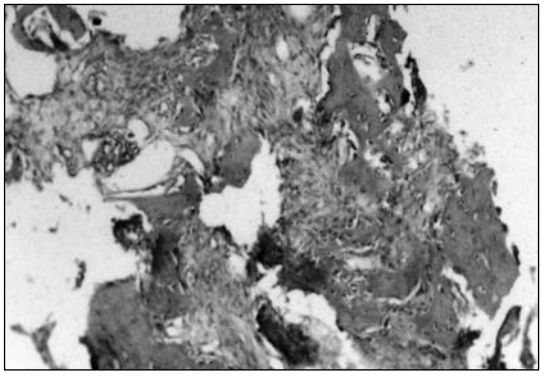

Figure 6. New bone trabeculaes in the fibrous tissue (H&E x100)

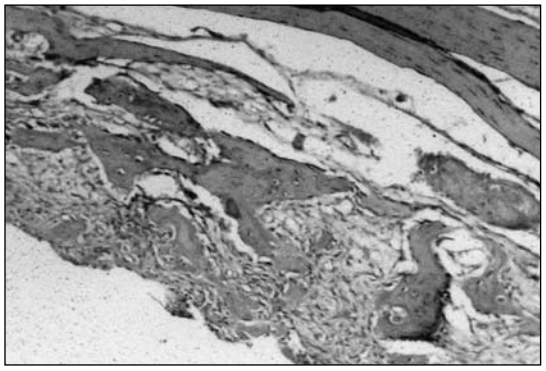

Figure 7. Fibrous areas are observed inner side of the cortex (H&E x 100)

Table 1. Bone markers of the patient

	Patient Values	Normal Values
Creatinine	0.6 mg/dL	0.6-1.2 mg/dL
Alkaline Phosphatase	92 U/L	30-120 U/L
Calcium	8.9 mg/dL	8.5-10.5 mg/dL
Phosphate	3.1 mg/dL	2.7-4.5 mg/dL

Discussion

FD is essentially a disease of the young population although often going unnoticed until middle age. The bone disease may be monostotic (affecting a single bone) or polyostotic (affecting many bones) and is usually asymmetrical and often unilateral. The rib, femur, tibia and maxilla are most commonly involved [10]. Mandibular involvement may threaten the functions of mastication, speech, respiration and facial and jaw cosmetics. Therefore, the disease progress is of utmost importance. Our case was about her 45's and was presenting a cyst-like lesion in the anterior mandibula.

As no one investigation is pathognomonic, the diagnosis only becomes apparent after collation of history, examination, histology and radiology [11]. However, Camileri [10] remarks that the biopsy is not always mandatory for the management of the patient.

It is clear that CT was invaluable for the assessment of extensive lesions, especially those affecting the anatomically complex maxilla [12, 13]. Furthermore, sclerosis and thickening can create overlapping images, which may impede adequate assessment by plain film radiography [12]. Fries [14], has described three radiological patterns in craniofacial fibrous dysplasia. The first is pagetoid with bone expansion and alternate areas of radiodensity and radiolucency [10]. It occurs in more than half of the patients, most of whom are older than 30 years of age and have had symptoms for an average of 15 years. The second pattern is sclerotic, with bone expansion and a homogenous radiodensity (a ground glass appearance). The third type is cyst-like, usually a round or oval lesion with a sclerotic border. The sclerotic and cyst-like patterns occurred in younger individuals (average age 20 years). The ground-glass appearance with an expanded cortex is the most common. The most effective method of monitoring growth and estimating extent of disease seems to be computerized tomography [10]. Our case matches with the third type of Fries' descriptions and unilateral nature of FD is apparent. Meanwhile, in spite of the normal level of the bone markers, our case was identified as FD.

MacDonald Jankowski [9] reported that tooth displacement might also occur. He suggested also lamina dura could be absent. Petrikowski et al. [15] suggested that the loss of lamina dura could be used as an ancillary diagnostic feature for FD. In the present case, the absence of lamina dura was observed.

Donald-Jankowski [16] mentioned about cystic degeneration in fibrous dysplasia of the jaws.

According its treatment, chemotherapy is proved ineffective in retarding the progression of disease. Oral aluminum acetate can be used to reduce the danger of hyperphosphataemia in severe forms. Steroids have been used with partial success in treating painful lesions. Radiotherapy is contraindicated and has been associated with sarcomatous change.

Formerly, orthopedic surgery was the only therapy for FD; it consisted of preventive means such as bone grafts, fixation, curettage and treatment of fractures. Calcitonin has failed in the treatment [17]. It appears wise, however, to use antiresorptive drugs, such as potent bisphosphonates, in a condition of bone diseases such as FD in which there is frequently an increase in bone turnover.

Surgical treatment for cranio-maxillo-facial lesions is controversial. Excision of all the affected bone is usually fruitless since it is impossible to be sure of the limits of the disease. However, it is only indicated if an important function is threatened, deformity becomes substantial or complications develop. Lane et al. [18] treated FD with oral or i.v. biphosphonates and his therapy diminished pain, prevented fractures, lowered N-telopeptide values, and led to partial resolution of fibrous dysplasia lesions.

Chapurlat et al. [19] proposed treatment with the bisphosphonate pamidronate without surgical intervention. They noticed that severe bone pain and the number of painful sites

appeared to be significantly reduced and all biochemical markers of bone remodeling were substantially lowered. They also noticed a radiographic response in nine patients with refilling of osteolytic lesions.

Pfeilschifter and Ziegler [20] remarked that pamidronate appears to be an effective and well tolerable treatment option for patients with fibrous dysplasia.

Weinstein [21] stated intravenous pamidronate increased total proximal bone mineral density in FD and prevented early fractures and deformities while making the patient pain free.

Unlike the other authors, we applied alendronate directly in FD with bone grafts and we observed that local alendronate administration showed high rate of bone healing in a very short time.

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