Mandibular Fibrous Dysplasia in Cotton Wool Appearance: A Rare Case Report

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INTRODUCTION

In 1891, Fibrous dysplasia was described for the first time by Von Recklinghausen. Later, in 1938, Lichtenstein and Jaffe gave the name of Fibrous dysplasia.¹ Fibrous dysplasia is pathology of bone development in which the normal bone is replaced by a disorganized fibrous bone.² Fibrous dysplasia may affect single or multiple bones.³

The Polyostotic form of the disease has nearly 100% involvement of the craniofacial bones.⁴ The diagnosis of fibrous dysplasia is based on the correlation between the clinical, radiological and histological aspect, which constitutes a challenge in clinical practice.² Through the following case, the purpose in this article is to show a rare case of fibrous dysplasia with a less common radiographic appearance.

CASE REPORT

A 50-year-old woman in good general health and with no medical antecedents consulted for implant placement advice in the mandible. At the X-ray examination, the attending physician discovers a suspicious image in the mandible.

The clinical examination shows a slight bilateral swelling. The patient indicates that it lasts for several years. No lymphadenopathies were palpable. The oral examination showed normal mucosa. We note the absence of 36 and 46. The vitality test on all the mandibular teeth was positive except on the 47. (Fig.1)

Palpation next to the mandibular molar sites shows a filling of the vestibule floor. The filling is hard, non-compressive, and covered with a healthy mucous membrane.

The panoramic radiograph shows a mixed image in cotton wool. The image is present on both sides’ right and left. It extends from the premolar region to the molar region. The 47 presents a canal radioopacity synonymous with a root filling. (Fig.2)

The dentascan shows a sparse radiolucent image of rounded opacities. The image respects the internal and external cortices of the mandible. (Fig.3,4)

In view of the clinical and radiological information, the diagnosis referred to are: fibrous dysplasia, ossifying fibrous, osseous dysplasia. A biopsy was performed to determine the nature of the lesion. (Fig.5)

Pathological examination specified the diagnosis of fibrous dysplasia.

ABSTRACT

Fibrous dysplasia is pathology of bone development in which the normal bone is replaced by a disorganized fibrous bone. This condition is considered rare in the maxilla. Usually, its evolution is progressively along the years, leading to an asymmetry whose aesthetic prejudice may be important. Other manifestations are also found, such as pain, paraesthesia, or malocclusion. There are several radiological aspects of fibrous dysplasia. The most common form is the “ground glass” appearance. The appearance in “cotton wool” being less found. We present a rare case of fibrous dysplasia in a 50-year-old woman. The diagnosis was made after a complete clinical examination including examination of the patient, radiological examination, and incisional biopsy. The patient is regularly monitored in our department.

KEYWORDS: Fibrous dysplasia, cotton wool

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Maxillofacial fibro-osseous lesions comprise a group of face and jaw disorders characterized by the replacement of bone by a benign connective-tissue matrix with varying amounts of mineralized substances. Fibrous dysplasia (FD) belongs to the category of a benign fibro-osseous in which the normal medullary bone and cortices are replaced by a disorganized fibrous woven bone due to disturbance in the maturation and remodeling of the bone.

It is accepted that mutation, or suppression of an intracytoplasmic transducer protein would be responsible for this disturbance. The resultant fibro-osseous bone is more elastic and structurally weaker than the original bone.

Fibrous dysplasia is considered a rare bone disease that represents 2.5% of all bone diseases and 7% of all benign bone tumors. In general, FD presents in three forms: monostotic, polyostotic, and polyostotic with endocrinopathies, which can be associated with hyperpigmentation and endocrinological disorders and is called McCune-Albright syndrome. The most common form is monostotic. The polyostotic type is less found.

The average age of the affected patients is 20-30 years, whereas in the polyostotic form, the patient is much younger (less than 10 years on average).

The monostotic form usually involves the femur and the bones of the craniofacial skeleton. When it has this location, the maxilla is most frequently affected than the mandible. It involves the maxilla 50% more often than the mandible, usually in the posterior region.

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Mandibular localization is considered extremely rare. In fact, chin et al. has performed a “zone” classification of fibrous dysplasia. The fourth zone, which is the least frequent, DF involves the maxilla and mandible separately (monostotic form) or together (polyostotic form) with the alveolar process.

The signs and symptoms are correlated to the type and location of the FD and include facial deformity and asymmetry, visual alteration, auditory disability, nasal congestion and/or obstruction, pain, paraesthesia, or malocclusion. In our case, the patient mentioned a slight asymmetry that did not disturb her. Several radiological aspects are possible in the case of fibrous dysplasia.

The most frequent X-ray characteristic of craniofacial FD is “The ground glass” appearance. It takes on a granular appearance resembling the small fragments of a shattered windshield with a fine cortex and no defined borders. Another radiological manifestation is orange peel and cotton wool. The first is a pattern resembling the surface of an orange. The appearance in cotton wool is in the form of a wispy arrangement. Early lesions may be more radiolucent than mature ones.

In our case, the lesions are clearly in the aspect of cotton wool. The radiographic aspect is more radiolucent on the right side than on the left side, suggesting that the DF was earlier on the left side than on the right side.

The differential diagnosis of FD includes ossifying fibroma, osseous dysplasia, chronic sclerosing osteomyelitis, osteosarcoma, and Paget’s disease. Among these assumptions, one is particularly difficult to differentiate. Indeed, ossifying fibroma and fibrous dysplasia represent a dilemma for the oral surgeon. The similarity in the clinical and radiological aspects and microscopic features is so close that the difference between these two entities are very difficult. To make the difference between ossifying fibroma and fibrous dysplasia, an exploration of the margins of the latter and the normal adjacent bone using a microscope is necessary.
The characteristics noted on the microscope and which can be attributed specifically to fibrous dysplasia are “Chinese character” or “fish bone”.  

Nevertheless, differentiation between them is crucial since these lesions require different clinical approaches in their management. In fact, because of its risk for recurrence, ossifying fibroma needs to be completely enucleated from the surrounding bone.

The treatment of bony lesions of fibrous dysplasia includes surgical and nonsurgical therapies. Prevention of pathological fractures and reduction of bone deformities is the goal of surgical treatment.

Conservative management seems to be the standard of care, which involves removing the diseased bone via an intraoral approach. Complete excision with graft reconstruction could be considered in severe cases. Clinical observation is also suggested for fibrous dysplasia lesions that have no risk of pathologic fracture or deformity.

In our case, the patient wants to place implants at sites with dysplasia. The possibility of conservative treatment with removal of dysplastic sequestration, bone grafting, and implant placement was discussed.

In fact, the complete reossification of the defected bone and the osteointegration of the implants have been reported in the literature. Samman, in 1991, in his studies demonstrated that the use of titanium materials would result in good integration and histologic adaptation in the dysplastic bone.

This eventuality was declined by the patient. She prefers to wear a removable prosthesis. In this case, DF has been evolving very slowly for years, with no noticeable changes or functional genes. Note that craniofacial fibrous dysplasia tends to stabilize when the patient reaches skeletal maturity. Likewise, the progression of the lesion stops with its maturation.

To the sum of these findings, it was agreed to adopt regular monitoring of the patient for any changes or growth of the lesion.

**CONCLUSION**

Fibrous dysplasia is a rare benign bone tumor that progresses silently over the years. The diagnosis is based on clinical, radiological and histological examination. The appearance in "cotton wool" is very rarely found, it looks like a wispy arrangement what differentiates it from the orange peel aspect. Fibrous dysplasia requires abstention, however, surgical management can be considered in case of significant facial deformation.

**REFERENCES**


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