Craniofacial Fibrous Dysplasia of the Mandible: A Case Report and Literature Review

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Abstract

Objective: Report a case of craniofacial fibrous dysplasia of the mandible with a literature review about the management of this disease.

Clinical case: A 47-years-old female patient was referred to our unit due to an incidental radiographic finding of a mixed radiolucent-radiopaque lesion in the right mandible ramus. Complementary blood tests, an updated panoramic radiography and a cone beam computed tomography were performed. The incisional biopsy’s histopathological report described a craniofacial monostotic fibrous dysplasia of the mandible.

Discussion: The fibrous dysplasia is a low prevalence fibro-osseous lesion that can affect one or more bones, causing different degree of alterations. Treatment must be assessed according to several factors like age, location, size, aggressiveness, among others. Conservative management with long-term follow-up is the treatment of choice in certain cases, avoiding surgical morbidity.

Conclusion: The craniofacial fibrous dysplasia is a rare disease of which diagnosis is difficult due to multiple differential diagnosis. The conservative or surgical management must be assessed depending on factors such as aggressiveness, synanthomatology and presence of aesthetics and/or functional alterations.

Keywords

Monostotic fibrous dysplasia, Craniofacial fibrous dysplasia, Bone disease

Introduction

Fibro-osseous lesions of the jaws are a group of benign disorders that can affect bone. Within this group is Fibrous Dysplasia (FD), whose name was mentioned for the first time by Lichtenstein in 1938 [1] and then by Lichtenstein and Jaffe in 1942 [2].

In this condition normal bone tissue is replaced by fibrous tissue and immature bone characterised by abnormal proliferation of fibrous tissue between normal and immature bone, caused by poorly differentiated osteoblasts. Its etiology remains unknown [3]. It represents 3% of all bone tumors and between 5-7% of all benign bone tumors [4,5].

In decreasing order, its most frequent locations are the maxilla, mandible and frontal bone [6,7]. It commonly involves adjacent structures such as dental arch, maxillary sinus, airway, orbit, and skull base [8,9].

Regarding the alterations associated with its presence in the mandibular region, several signs and symptoms have been described such as neurosensitive disturbances due to compression of the inferior alveolar nerve canal [10-13], malocclusion, facial deformity, among others [14].
In the orthopantomography (Figure 2), a heterogeneous, radiolucent/mixed-aspect lesion is evidenced that involves part of the body and mandibular branch on the right side.

A Cone Beam Computed Tomography (CBCT) was taken, which revealed an extensive and irregular hypodense area, with clear limits, which involves the middle and lower third of the ramus and mandibular body from the right side to the distal side of tooth 46, compromising the entire alveolar process alveolar from the crest to the mandibular canal. It displaces the mandibular canal towards the basilar edge, generating a solution of continuity in the canal roof. It expands buccal and lingual bone tables, without drilling or thinning the bone cortex, does not displace teeth or generate rhizalysis (Figure 3).

Our diagnostic hypotheses were calcifying epithelial odontogenic tumor, ossifying fibroma, and fibrous dysplasia. Through an incisional biopsy (Figure 4), a histopathological study was performed where the definitive diagnosis of Monostotic Fibrous Dysplasia of the Mandibular bone was established.
Discussion

The latest WHO head and neck tumor classification, published in 2017, located FD in the group of “osteochondromatous and fibroosseus lesions [15].

It can occur at any age, although it is more frequent in children and adults between the first and second decade [5], without predilection for sex [5,11]. There are also no clear conclusions regarding its progress and arrest, although most studies imply a decrease in the rate of progression or remission after puberty [5,16].

Regarding the location, its incidence in the maxilla is almost double that in the mandible. In the maxilla, it usually presents unilaterally in its posterior region [7,17]. In this case, the lesion affected the mandibular branch and body on the right side.

It usually has a slow and progressive growth with associated complications of pain, facial deformity, fractures, functional impotence, dental and neurological...
Genetically it is expressed in two ways, monostotic (56-70%) or polyostotic (30-47%) [5, 7, 21-23]. The most frequent location for FD in descending order is the maxilla, mandible, and frontal bones [7, 18]. They commonly involve adjacent structures, such as the dental arch, maxillary sinus, airway, orbit, and skull base.

Malignancy is rare, being approximately 0.5% in the monostotic variable and 4% for SMA. This situation has been seen with radiation therapy and growth hormone (GH) alterations. [4, 5, 24].

Its differential diagnoses include Paget’s disease, type I neurofibromatosis, osteofibrous dysplasia, ossifying fibroma, cherubism, osteosarcoma among others [22]. These lesions present a wide clinical spectrum of expression, from being practically asymptomatic to a disabling alteration for the senses such as hearing and vision.

Considering the high rate of casual radiographic findings of the lesion [25], we did not find reports describing the appearance of monostotic or polyostotic craniofacial lesions in conjunction with lesions in other parts of the body.

The diagnosis of FD represents a challenge in clinical practice, it requires clinical, radiological, histopathological correlation and surgical findings [26]. Although the clinical characteristics can provide a diagnostic approach for this type of lesion, they are not specific to each other, so histopathology is key for a definitive diagnosis and proper management of the lesions. In our case, prior to performing the biopsy, the first differential diagnoses pointed to an odontogenic tumor or cyst due to the higher prevalence of these lesions over the fibro-osseous ones, in addition to its unusual location.

Although histopathology is important to establishing a definitive diagnosis, sometimes it is inconclusive. This is due to the great similarity between the histopathological characteristics with other fibro-osseous lesions such as ossifying fibroma. It is important to consider the stage of maturation of the lesion and the variations in its histological patterns for the establishment of a definitive diagnosis. The gold standard for the diagnosis of FD is the histopathological confirmation of a fibro-osseous lesion without a capsule, which must be confirmed with radiographic findings [11]. Therefore, close collaboration between the surgeon and the pathologist is essential for the establishment of a definitive diagnosis.

The treatment of FDs must be highly individualized and depends on their clinical presentation [27]. Treatment protocols include observation, medical treatment, and/or surgery. The age, affected site, size and the biological behavior of the lesion are influencing factors in the selection of therapeutic management [28]. In relation to the biological behavior, lesions of the facial skeleton can be described as quiescent, non-aggressive or aggressive.

The first line of treatment is surgical, which can be conservative or invasive, through resection and subsequent reconstruction [5, 21, 29, 30]. It has also been determined that observation and constant monitoring is feasible in incidentally found lesions, without signs or symptoms associated. Recurrence of FD is common, reaching up to 50% in children until the end of growth, and is the most common cause for reoperation [6, 21].

On the other hand, it has been shown that therapy with intravenous bisphosphonates (Zoledronic acid or Pamidronate) and Denosumab (RANKL monoclonal antibody) have been effective in relieving pain and normalizing bone turnover. There are some studies that report the complete or partial disappearance of lesions after prolonged therapy with bisphosphonates [31, 32], however, its use is recommended in the symptomatic polyostotic variable [21, 31, 33].

In the presence of deformity, functional impotence, risk of pathological fracture or symptoms, conservative surgical management is recommended. In more severe cases, with a greater compromise of adjacent structures, remodeling, shaving or bone grafting is also a viable alternative [5, 6]. Treatment goals seek to correct or prevent functional problems caused by injuries and achieve aesthetic improvement. In our case, the patient did not manifest pain, deformity, functional or neurological alterations.

In the case of quiescent fibrous dysplasia in which facial deformation or relative risk of pathological fracture is not investigated, observation and follow-up is an acceptable treatment modality [5, 29].

In the present case, the characteristics of her pathology, prognosis and treatment options were explained to the patient. It was decided to maintain clinical, photographic and radiographic controls. Considering that the malignancy of the FD varies between 0.4%-4%, continuous and close monitoring, clinical and radiological, every 6 months is suggested to control the evolution of the pathology [5]. The patient should be informed that if signs or symptoms appear, they should promptly inform their caregiver.

The finding of mandibular FD in the area of the body and angle in a woman in her 4th decade is uncommon. However, the general dentist may be the first to detect it in routine practice with a panoramic radiograph. Therefore, the knowledge of the pathology is essential in the conformation of an appropriate diagnosis and in reducing potential complications.

**Conclusion**

The diagnosis of FD is challenging for the clinician and
requires a comprehensive clinical history and physical examination. When selecting the appropriate treatment for FD, the clinical characteristics should be considered individually for the case, considering factors such as age, the presence of comorbidities, aggressiveness, the stage of development, the associated symptoms and the presence of functional or aesthetic alterations. Close monitoring is essential in cases where the selected treatment is observation.

**Ethic Aspects**

The authors declare that the study was done in accordance with the code of ethics of the World Medical Association (Declaration of Helsinki) for human studies.

**Conflict of Interest**

No conflicts of interest are declared.

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