Case Report

A rare case of bone lesion: Mandible's fibrous dysplasia

ABSTRACT

Fibrous dysplasia is a rare genetic syndrome that affects bone tissue. This pathology replaces the mineralized matrix of the bone affected with connective and fibrous tissue. This article describes a mandibular fibrous osseous dysplasia case and its surgical treatment. A 45-year-old woman complained about a slow development of swelling of the left mandibular bone. The orthopantomography (OPT) and the cone beam computed tomography (CBCT) revealed a well-circumscribed sclerotic lesion with a ground-glass appearance apical to the 3.5 element. The surgery was performed to excise the lesion. Anatomopathological examination of tissue confirmed the suspects among the diagnosis of fibrous dysplasia. The patient underwent to follow-up of 4 years, and no recurrences were found. In the absence of a univocal consensus on therapy, surgery remains the treatment of choice for unifocal forms.

Keywords: Fibrous dysplasia, fibro-osseous lesions, oral surgery

INTRODUCTION

Fibro-osseous lesions are a group of pathologies characterized by the resorption of bone and replacement with fibrous connective tissue. Benign fibro-osseous lesions are divided into three categories: ossifying fibroma, fibrous dysplasia (FD), and cement-bone dysplasia.

FD was defined as "a benign lesion, presumably developmental in nature, characterized by the presence of fibrous connective tissue with a characteristic whorled pattern and containing trabeculae of immature non-lamellar bone."[1] This pathology represents a rare condition that develops at an early age: the diagnosis of 90% of FDs occurs within 4 years of life.[2]

Symptoms related to FD depend on the anatomical area involved.[3] One-third of all patients are asymptomatic, and the diagnosis is done through radiological findings that are completely random.[4]

The most common symptoms linked to FD that affect the cranial bone usually are characterized by unilateral bone swelling associated with slow growth and, in some cases, with pain.[2] In rare cases, this pathological condition could lead to malocclusion, impaired vision, and hearing problems.[5]

Access this article online	
Website: www.njms.in	Quick Response Code
DOI: 10.4103/njms.njms_35_23	

The radiological aspect could be various, but, in most cases, the FD appears as a ground-glass radiopaque lesion with faded edges. Furthermore, the FD can lead to thinning of the lingual or vestibular cortical bone, dislocation of the teeth, the inferior alveolar nerve, the maxillary sinus, or the floor of the orbit.[6,7]

If symptomatic, the standard treatment for this condition is surgical excision. Although bisphosphonates have been proposed as a medical therapy against the progression and the pain symptoms, in the literature there is not a univocal opinion on the real effectiveness of the use of these drugs.[5]

CASE REPORT

A 45-year-old woman visited the Santi Paolo and Carlo Hospital complaining about a slow development of swelling

Pietro Fusari, Federico Guerri, Matteo Arcari, Andrea Sardella

Department of Health Sciences - Santi Paolo and Carlo Hospital, University of Milan, Italy

Address for correspondence: Dr. Federico Guerri, Beldiletto Street 1, Milan, 20134, Italy.
E-mail: fede.guerri@gmail.com

Received: 04 March 2023, **Revised:** 10 January 2024, **Accepted:** 12 January 2024, **Published:** 16 November 2024

How to cite this article: Fusari P, Guerri F, Arcari M, Sardella A. A rare case of bone lesion: Mandible's fibrous dysplasia. Natl J Maxillofac Surg 2024;15:518-20.

of the left mandibular bone was sent by a colleague after the casual discovery of a bone lesion after taking an OPT.

Oral examination confirmed the swelling of the vestibular cortical mandibular bone in the area from 3.4 to 3.6 with an extraction of circa 3.5 cm. After seeing the OPT, a CBCT was prescribed and revealed a well-circumscribed sclerotic lesion with a ground-glass appearance apical to the 3.5 element [Figure 1]. The lesion was associated with focal thinning of the vestibular cortex.

The excision of the lesion was planned under general anesthesia.

Local anesthesia was followed by an intrasulcular incision and the elevation of a mucoperiosteal flap from 3.3 to 3.6. The lesion was exposed and removed after the osteotomy, allowing the creation of a small operculum. The margins of the neo cavity were scraped to remove any residues and to prevent any relapses. The flap was sutured with 4/0 Vicryl in its original position [Figure 2].

A histopathological examination of the lesion removed was requested.

The following post-operative therapies were provided to the patient:

- antibiotic therapy with amoxicillin + clavulanic acid for 6 days with a dosage of 1 g every 12 h;
- rinses with chlorhexidine mouthwash 0.12% for 10 days, to start 24 h after the surgery;
- local applications of 1% chlorhexidine gel, 2 times a day for 10 days;
- non-steroidal anti-inflammatory drugs as needed.

Post-operative instruction was given to the patient.

RESULTS

Immediately after the surgery, the patient reports a lower left lip paresthesia. A therapy with alpha-lipoic acid (1 pill every day for the first 3 months) and l-acetyl-carnitine (2 pills per day for 3 months) was prescribed. The patient underwent clinical and neurosensory evaluation with touch, nociceptive, and thermal tests to map the affected area. The paresthesia has been recovered spontaneously within 3 months.

The radiological signs associated with the execution of the anatomical pathological examination led to the diagnosis of FD.

The patient underwent periodical clinical and radiological follow-up that lasted for 4 years. At the last clinical examination, there were not any pathological signs and the last orthopantomography shouted on this occasion did not show any relapse [Figure 3].

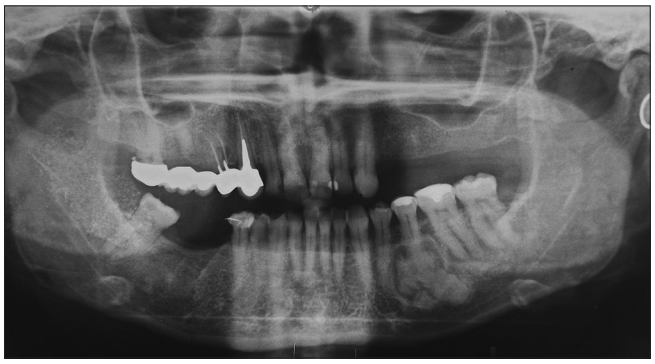

Figure 1: Orthopanoramic radiography that shows the presence of ground-glass appearance lesion apical to the 3.5 element

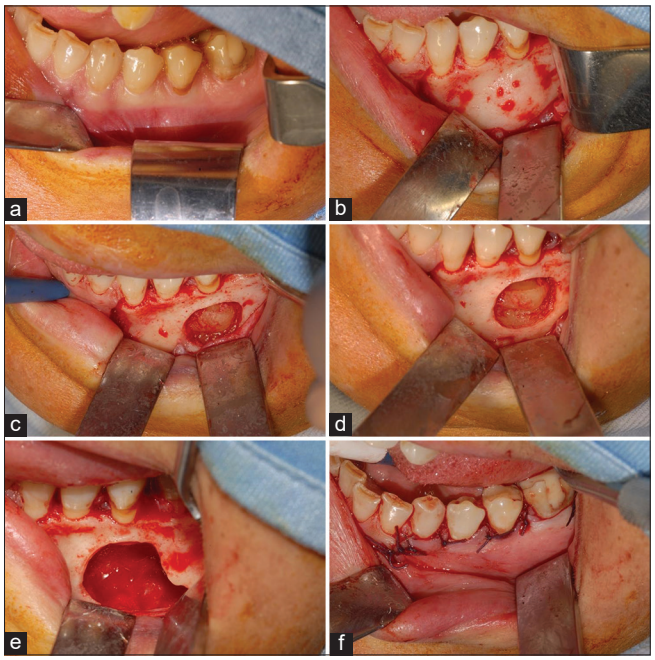

Figure 2: The sequence of the surgical procedure for the excision of the lesion: keratinized gingiva and alveolar mucosa area 33-36 (a), intrasulcar incision and subperiosteal flap elevation (b), ostectomy (c), root section (d), lesion excision (e), and stitching (f)

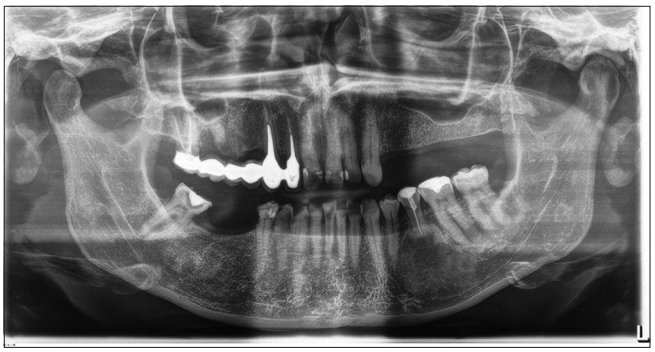

Figure 3: Orthopanoramic radiography after 4 years of follow-up

DISCUSSION

The diagnosis of FD is difficult to make with the radiological examinations alone, and it could be confused with cement-bone fibrosis, ossifying fibroma, osteonecrosis, fibrocystic osteitis (or Recklinghausen's disease), and giant cell granulomas.[8]

The only way to confirm the clinical suspects and make a definitive diagnosis is to perform a biopsy and request a histologic examination of the sample.[9]

The biopsy should be an excisional biopsy, and it is important for complete surgical removal of the mass. In literature, indeed, it is estimated that the regrowth rate affects 25-50% of the patients that undergo the surgery treatment. The regrowth risk is higher among younger patients rather than in older ones, for this reason, is it advisable to plan the surgery at the end of the development of the patient. In extremely rare cases, less than 1%, the FB can undergo a malignant transformation, usually osteosarcoma. Changes in radiographic appearance, sudden appearance of pain, or neurosensory changes should alert the clinician.[9]

CONCLUSION

FD is rare and genetic but a non-hereditary condition that replaces the mineralized matrix of the bone with fibro-connective tissue. A certain diagnosis of FD with radiology alone is inaccurate, and the association with an anatomopathological examination after the surgery is strongly recommended. Medical therapy has been studied in the literature, but there is no real consensus on its effectiveness. The surgery remains the treatment of choice for unifocal forms. A long clinical and radiological follow-up for these lesions is mandatory to diagnose any relapse.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understands that her name and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Kamboj M, Baoz K, Mahajan S. Maxillofacial fibrous dysplasia. Indian J Dent Res 2005;16:151-2.
2. Burke A, Collins M, Boyce A. Fibrous dysplasia of bone: Craniofacial and dental implications. Oral Dis 2017;23:697-708.
3. Javaid MK, Boyce A, Appelman-Dijkstra N, Ong J, Defabianis P, Offiah A, *et al*. Best practice management guidelines for fibrous dysplasia/McCune-Albright syndrome: A consensus statement from the FD/MAS international consortium. Orphanet J Rare Dis 2019;14:139.
4. Yang L, Wu H, Lu J, Teng L. Prevalence of different forms and involved bones of craniofacial fibrous dysplasia. J Craniofac Surg 2017;28:21–5.
5. Metwally T, Burke A, Tsai JY, Collins MT, Boyce AM. Fibrous dysplasia and medication-related osteonecrosis of the jaw. J Oral Maxillofac Surg 2016;74:1983–99.
6. Sontakke SA, Karjodkar FR, Umarji HR. Computed tomographic features of fibrous dysplasia of maxillofacial region. Imaging Sci Dent 2011;41:23-8.
7. Akintoye SO, Lee JS, Feimster T, Booher S, Brahim J, Kingman A, *et al*. Dental characteristics of fibrous dysplasia and McCune-Albright syndrome. Oral Surgery, Oral Med Oral Pathol Oral Radiol Endodontology 2003;96:275–82.
8. Koenig LJ, Tamimi D, Petrikowski CG, Perschbacher SC. Diagnostic Imaging: Oral and Maxillofacial. 2nd ed. Elsevier; 2017.
9. Neville BW, Damm DD, Allen CM, Chi AC. Oral and Maxillofacial Pathology. Elsevier; 2016.