Surgical Treatment of Fibroosseous Lesion in Young Patient with Reduced Mouth Opening

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Abstract

Keywords

- fibrous dysplasia
- functional impairment
- surgical treatment
- case report

Fibrous dysplasia is a benign fibroosseous disorder that can affect the maxillary bones, causing aesthetic deformity and functional impairment. This article reports the case of a 13-year-old male patient at the time of diagnosis. The patient showed increased facial volume with relevant asymmetry, having reported the onset of the condition 12 months before. Upon examination, the patient presented an 8-mm mouth opening and an acute inflammatory process associated with tooth 37 pericoronal hood. Upon imaging exam, exuberant bone growth in the left mandibular ramus area of ground glass aspect was observed. After incisional biopsy, fibrous dysplasia was diagnosed and sequentially treated with osteoplasty and coronoid process removal. The patient evolved to a 43-mm mouth opening and favorable aesthetics without recurrence in a 3-year follow-up period. In this case, coronoidectomy and bone plasty proved to be effective, returning aesthetics and function. The patient is supposed to be followed up in the long term.

Fibroosseous lesions comprise a group of benign pathological entities associated with disordered collagen deposition and immature matrix ossification.^{1,2} These lesions can be classified into three types: fibrous dysplasia, bone dysplasia, and ossifying fibroma. They have similar histological aspects and are often confused; however, they also have different clinical behaviors and their development can be differentiated by radiographic or tomographic follow-up.^{1–4}

Fibrous dysplasia is a benign noninflammatory bone alteration which has been reported in literature to have a 1:30,000 individual incidence; all fibrous dysplasia lesions are thought to develop in childhood or puberty and evolve along the patient's life until they are clinically perceptible.^{4,5}

This condition can manifest in three different ways: monostotic, involving only one bone; polyostotic, when two or more

bones are affected; and McCune-Albright syndrome (MAS), when associated with *café-au-lait* spots and endocrine hyperfunction alterations.^{6–8}

Differential diagnosis of fibrous dysplasia lesions in the maxilla includes ossifying fibroma, low-grade central osteosarcoma, giant cell granuloma, as well as other fibroosseous lesions. One of the imaging keys for diagnosis is its frosted glass aspect and the observation of poorly defined boundaries as compared with healthy bone.^{4,9,10}

The disease results from a post zygote mutation and its severity is related to the stage of embryonic development in which a mutation of the GNAS gene (on chromosome 20q13), which encodes the G α subunit of the heterotrimeric G protein complex, occurs. If the mutation develops during internal cell mass formation, the three cell layers will be affected and the

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individual will develop MAS. Late mutations, such as monostotic fibrous dysplasia, have more limited phenotypes.^{6-8,10,11}

Treatment of fibrous dysplasia lesions depends on the severity and functional damage caused and includes several alternatives, ranging from drug therapy for lesion progression control with antiresorptive agents such as bisphosphonates (including pamidronate) and RANKL system inhibitors (e.g., denosumab)¹²⁻¹⁵ to surgery (e.g., osteoplasty)¹⁶ or direct rehabilitation with dental implants¹⁷ and, in some cases, resection of the lesion followed by autogenous grafting.^{18,19} However, the medications such bisphosphonates or others antiresorptive agents were associated, in the past 15 years, with the medication-related osteonecrosis of the jaws (MRONJ).²⁰ Although MRONJ has its main cause associated with dental surgery in patients submitted to a chronic use of the antiresorptive and antiangiogenic medications,²⁰ there is no evidence of MROJN in patients using these medications to treat fibrous dysplasia.²⁰

The aim of this study was to report the diagnosis and treatment of a mandibular monostotic fibrous dysplasia case which caused facial deformity associated with severe mouth opening limitation in a young patient.

Case Report

A 13-year-old male patient sought the Maxillofacial Surgery service at the University Hospital of the Federal University of Pelotas (CTBMF/HE-UFPel) complaining of an increased volume on the left side of the face, whose first signs had appeared

1 year before, and progressive mouth opening difficulty in the past 9 months. No other underlying pathologies, nor the use of continuous medication, were identified in his medical history.

Initial examination identified a diffuse growth of hard consistency in the preauricular and mandibular areas on the left which was painless on palpation. He showed an 8-mm mouth opening and presented poor oral hygiene due to this limitation, with suppurative pericoronitis associated with erupting tooth 37.

Upon the first consultation, systemic antibiotic therapy was prescribed for pericoronitis treatment (amoxicillin 500 mg daily divided into three doses) until the patient's return 7 days afterward to initiate treatment.

Upon the patient's return, the pericoronitis had receded and there were no phlogistic signs of infection at the site. However, there were no trismus or volume increase reduction improvements.

Radiographic examination showed an enlargement of the left mandibular ramus, asymmetry of radiopaque aspect with poorly defined boundaries, and ground glass appearance suggestive of a fibroosseous lesion (►Fig. 1). Due to the clinical conditions and panoramic imaging, a face tomography was requested and an incisional biopsy of the lesion was scheduled.

The tomographic examination revealed a bone alteration in the mandible with abnormal growth of the entire mandibular ramus involving the coronoid process, which was markedly anomalous, showing increased volume that was the major mouth opening limitation cause; however, no impairment of the articular anatomy was found (►Fig. 2a).

Fig. 1 (a) Frontal view of patient in the first appointment. (b) Patient's maximum mouth opening in the first appointment. (c and d) Volumetric reconstruction of patient's tomography, showing the abnormal growth in left mandibular ramus and coronoid process. (e) Sagittal TC slice of mandibular left ramus evidencing the irregular bone density. (f) Coronal TC slice provides a view to compare the asymmetric thickness of the left mandibular ramus with the right mandibular ramus.

Fig. 2 (a) Preoperative panoramic radiography presenting the classic ground-glass aspect of fibrous dysplasia. (b) Hematoxylin and eosin (H&E) section of bone biopsy (40 $\times$) showing the *Chinese letters* aspect of immature bone deposit. (c) H&E section of bone biopsy (400 $\times$) with spindle cells in an immature osteoid matrix.

Because of the great difficulty of access, incisional biopsy was performed under general anesthesia, with intraoral approach both for the ramus and mandibular angle. After the access, it was found that the periosteum was also involved in the lesion, making tissue detachment and division difficult. With the help of a tapered waist drill (H33L 702; Brasil Komet, Brazil), an osteofibrous fragment was removed from the lesion.

Histopathological examination revealed fibrous dysplasia ($\rightarrow$ Fig. 2b, c). A new surgery was then scheduled for coronoidectomy and osteoplasty to improve mouth opening, as well as marginal ostectomy to correct the facial deformity.

Coronoidectomy and osteoplasty ($\rightarrow$ Fig. 3a, b) were conducted under general anesthesia through intraoral access for the ramus and mandibular angle, this access being at the same site as the anterior extended in its proximal aspect until mandibular temporal crest exposition. After detachment and division of tissues until there was exposure of the entire lateral aspect of the mandibular ramus, the coronoid process was held by means of Rochester Peane forceps; ostectomy was performed by a truncated conical drill, followed by the coronoid process removal. Afterward, osteoplasty of the ramus lateral side was performed with surgical steel cutters (H251E Maxicut; Brasil Komet), until the desired contour and shape were reached; tooth 37 and the germ of tooth 38 were removed due to their lesion involvement. The patient progressed well in the immediate post-

operative period, without complaints, being medicated with ketoprofen (1.5 g per day divided into three doses) and Dipirone (4 g per day in four doses) and was discharged the next day.

On the 5th day after surgery, the patient presented an edema which was compatible with the procedure, and complained of mild pain and discomfort. Mouth opening was still 8 mm. Oral opening physiotherapy was to be performed with bidigital action and the use of a progressive number of tongue depressors four to five times a day, accompanied by heat packs in the bilateral masseteric area. After 20 days, the patient showed a 13-mm mouth opening, edema remission, and had no complaints; therefore, physical therapy continuation was recommended and a panoramic postoperative radiograph was requested ($\rightarrow$ Fig. 3c).

Five months after the surgery, the patient had a 35-mm mouth opening and adequate facial contour. He also showed aesthetic satisfaction, good masticatory function, and no complaints. At 8-month follow-up, the patient had 43-mm mouth opening, was satisfied with the aesthetic results, and showed new bone formation in the ostectomy area on a new imaging examination, compatible with the anatomy of a new coronoid eminence ($\rightarrow$ Fig. 4a–c). After a 3-year postoperative follow-up, the patient currently shows no recurrence of mouth opening limitation and shows satisfactory bone contour and symmetry, and the coronoid eminence fully recovered its shape and size ($\rightarrow$ Fig. 4d–f).

Fig. 3 (a) Surgical image after the coronoid process removal. (b) Surgical image of bone shape. (c) Immediate postoperative radiography.

Discussion

Although maxillary fibroosseous lesions are very similar by histological analysis, their behaviors follow different characteristics and the radiologic aspect is almost always necessary to reach a final diagnosis—without it, the histopathological report will often be described as a benign fibroosseous lesion—and is the only way to avoid inappropriate treatment.^{4,5,21}

While monostotic fibrous dysplasia (as in the present case) represents approximately 60% of all fibrous dysplasias, the other 40% consist of polyostotic variants, including 5 to 7% of cases associated with MAS.¹²

In spite of the fact that the diagnosis of polyostotic variants is usually clinical and by imaging, a biopsy should be performed for monostotic fibrous dysplasia, except when there is a risk of fracture of the bone involved.^{4,22}

According to the literature, approximately 25% of fibroosseous lesions in the craniofacial complex are fibrous dysplasias, with a higher incidence in the second decade of life and a discrete male preference, without race predilection. In most cases, the posterior maxillary area is the most affected, followed by the mandibular body.^{4,9,10}

Maxillary fibroosseous lesions are usually diagnosed during routine exams. With the development of the CT scan, these lesions have been better diagnosed in both extent and

volume. Lesions of significant sizes are usually diagnosed at the hospital by conventional tomography,⁴ which was the case of the patient under discussion.

In cases where the dysplasia variant is monostotic, other injuries which have similar behavior and radiographic imaging, such as giant central cell lesion, ameloblastic fibroma, ameloblastic fibro-odontoma, and ossifying fibroma, should be included in the differential diagnosis.^{4,10,22}

Fibrous dysplasia, despite being mostly asymptomatic, may present clinical manifestations such as bone deformities, pathological fractures, pain, or function limitation. In these cases, there are therapeutic management alternatives, such as the use of bisphosphonates.^{13–15} However, these lesions are best treated when diagnosed early.²³

In case of fully developed conditions, bone surgery is a safe and effective way of managing lesions that may cause deformities or function impairment.^{3,12,24,25}

Despite the apparent bone fragility that supposedly results from fibrous dysplasia, which could contraindicate or make surgery more difficult, whether lesion resection or plasty, a low rate of surgical complications can be observed.^{2,3} There is no consensus in the literature as for lesion growth recurrence, where some studies have shown recurrence in up to 50% of the cases, with a mean time of 6.6 years. Other studies do not show recurrence in a mean 9-year follow-up period.²⁶

Fig. 4 (a and b) Clinical aspect of patient 250 days after bone shaping and coronoidectomy, with a 33-mm mouth opening. (c) Volumetric reconstruction of patient tomography at 250th postoperative day, showing the growth of a new coronoid process. (d and e) Clinical view after 3 years postoperatively, patient with good facial symmetry and 38-mm mouth opening. (f) Volumetric reconstruction of 3-year postoperative tomography, with a completely newly formed coronoid process. (g) Sagittal TC slice at 250th postoperative day evidencing the bone irregular mineralization. (h) Sagittal TC slice at 3-year follow-up, with a complete newly formed coronoid process.

Even when resection is radical, making use of bone grafts as an alternative for defect reconstruction has been successfully reported, in cases when the lesion does not affect vital structures and the remodeling of the lesion with long-term follow-up is able to restore aesthetics and function;^{24,25} therefore, the decision to treat these lesions should be extremely individualized.

The case discussed presented epidemiological characteristics that fitted the patient profile as described in the literature: young, and in his second decade of life; however, the lesion growth occurred rapidly, according to the patient's report, and the difficulty in maintaining oral hygiene contributed to the development of pericoronitis associated with tooth 37, concomitantly with the development of trismus at the time of examination. The patient in this case showed an accelerated growth of the lesion, which could be associated with the inflammatory response in the injured area caused by the pericoronitis. However, there are no data in the literature that suggest of an association of the two events.

Surgical treatment of fibroosseous lesions is usually recommended after the end of the patient's growth. Nevertheless, in the present case, aesthetic and functional impairments, as well as interference in the patient's social life, recommended surgical intervention. It is known that the treatment in these cases should be followed up until facial growth has stopped, and may imply future interventions. The patient in this study will

continue his follow-up until the end of his growth phase, with the possibility of future corrective interventions.

Final Considerations

Fibroosseous lesions are a group of benign bone changes that, due to specific characteristics, can cause comorbidities in affected patients. Therefore, a precise diagnosis and the correct treatment, whether surgical or nonsurgical, are necessary to ensure better quality of life to the patient, who should be closely monitored so that the physician can intervene early in case of alterations.

Conflict of Interest
None.

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References

- 1 Waldrón CA. Fibro-osseous lesions of the jaws. *J Oral Maxillofac Surg* 1993;51(08):828–835

- 2 Kumar KA, Kishore PK, Mohan AP, Venkatesh V, Kumar BP, Gandla D. Management and treatment outcomes of maxillofacial fibro-osseous lesions: a retrospective study. *J Maxillofac Oral Surg* 2015;14(03):728–734
- 3 Suarez-Soto A, Baquero-Ruiz de la Hermosa MC, Minguez-Martínez I, et al. Management of fibro-osseous lesions of the craniofacial area. Presentation of 19 cases and review of the literature. *Med Oral Patol Oral Cir Bucal* 2013;18(03):479–485
- 4 MacDonald DS. Maxillofacial fibro-osseous lesions. *Clin Radiol* 2015;70(01):25–36
- 5 MacDonald-Jankowski DS. Fibro-osseous lesions of the face and jaws. *Clin Radiol* 2004;59(01):11–25
- 6 McCarthy EF. Fibro-osseous lesions of the maxillofacial bones. *Head Neck Pathol* 2013;7(01):5–10
- 7 Tabareau-Delalande F, Collin C, Gomez-Bouchet A, et al. Diagnostic value of investigating GNAS mutations in fibro-osseous lesions: a retrospective study of 91 cases of fibrous dysplasia and 40 other fibro-osseous lesions. *Mod Pathol* 2013;26(07):911–921
- 8 Akintoye SO, Boyce AM, Collins MT. Dental perspectives in fibrous dysplasia and McCune-Albright syndrome. *Oral Surg Oral Med Oral Pathol Oral Radiol* 2013;116(03):e149–e155
- 9 Prabhu S, Sharanya S, Naik PM, et al. Fibro-osseous lesions of the oral and maxillo-facial region: Retrospective analysis for 20 years. *J Oral Maxillofac Pathol* 2013;17(01):36–40
- 10 Eversole R, Su L, ElMofty S. Benign fibro-osseous lesions of the craniofacial complex. A review. *Head Neck Pathol* 2008;2(03):177–202
- 11 Chapurlat RD. Fibrous dysplasia of bone and McCune-Albright syndrome. *IBMS BoneKEy* 2010;7(01):18–26
- 12 Nardone V, D'Asta F, Brandi ML. Pharmacological management of osteogenesis. *Clinics (Sao Paulo)* 2014;69(06):438–446
- 13 Chapurlat RD, Gensburger D, Jimenez-Andrade JM, Ghilardi JR, Kelly M, Mantyh P. Pathophysiology and medical treatment of pain in fibrous dysplasia of bone. *Orphanet J Rare Dis* 2012;7(01, Suppl 1):S3
- 14 Lane JM, Khan SN, O'Connor WJ, et al. Bisphosphonate therapy in fibrous dysplasia. *Clin Orthop Relat Res* 2001;382(382):6–12
- 15 Lala R, Matarazzo P, Bertelloni S, Buzi F, Rigon F, de Sanctis C. Pamidronate treatment of bone fibrous dysplasia in nine children with McCune-Albright syndrome. *Acta Paediatr* 2000;89(02):188–193
- 16 Petrocelli M, Kretschmer W. Conservative treatment and implant rehabilitation of the mandible in a case of craniofacial fibrous dysplasia: a case report. *J Oral Maxillofac Surg* 2014;72(05):902.e1–902.e6
- 17 Bajwa MS, Ethunandan M, Flood TR. Oral rehabilitation with endosseous implants in a patient with fibrous dysplasia (McCune-Albright syndrome): a case report. *J Oral Maxillofac Surg* 2008;66(12):2605–2608
- 18 Villar-Puchades R, Ramos-Medina B. Virtual surgical planning for extensive fibrous dysplasia in the mandible. *Aesthetic Plast Surg* 2014;38(05):941–945
- 19 Mendonça Caridad JJ, Platas F Jr. Fibrous dysplasia of the mandible: Surgical treatment with platelet-rich plasma and a cortico-cancellous iliac crest graft-report of a case. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod* 2008;105(04):e12–e18
- 20 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(10):1983–1999
- 21 Eisenberg E, Eisenbud L. Benign fibro-osseous disease: current concepts in historical perspective. *Oral Maxillofac Surg Clin North Am* 1997;9:551–562
- 22 Abdelkarim A, Green R, Startzell J, Preece J. Craniofacial polyostotic fibrous dysplasia: a case report and review of the literature. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod* 2008;106(01):e49–e55
- 23 Majoor BC, Appelman-Dijkstra NM, Fiocco M, van de Sande MA, Dijkstra PS, Hamdy NA. Outcome of long-term bisphosphonate therapy in McCune-Albright syndrome and polyostotic fibrous dysplasia. *J Bone Miner Res* 2017;32(02):264–276
- 24 De Melo WM, Sonoda CK, Hochuli-Vieira E. Monostotic fibrous dysplasia of the mandible. *J Craniofac Surg* 2012;23(05):e452–e454
- 25 Zeng HF, Lu JJ, Teng L, et al. Surgical treatment of craniomaxillofacial fibrous dysplasia: functionally or aesthetically? *J Craniofac Surg* 2013;24(03):758–762
- 26 Macdonald-Jankowski DS, Li TK. Fibrous dysplasia in a Hong Kong community: the clinical and radiological features and outcomes of treatment. *Dentomaxillofac Radiol* 2009;38(02):63–72