Diagnostic aspects of juvenile ossificant fibroma: A case study

João Paulo Martins de Oliveira[1]; Maria Caroline Ferreira Cardoso[1]; Ana Paula Silva Lopes[1]; Aline Reis Stefanini[2]; Paulo Henrique Bortoluzo[3]; André Luís da Silva Fabris[4]; Tais Cristina Nascimento Marques[5]; Rogério Rodrigo Ramos[6,7]* Nilton Cesar Pezati Boer[8]; Luciana Estevam Simonato[9]

[1]School of Dentistry, Universidade Brasil, Fernandópolis, SP, Brazil

[2]Specialization in Medical Residency, Professor of the Department of Pathology, Universidade Brasil, Fernandópolis, SP, Brazil.

[3]Master's in Periodontics, Professor of the Postgraduate Program in Implantology, Universidade Brasil, Fernandópolis, SP, Brazil.

[4]PhD in Buccomaxillofacial Surgery and Traumatology, Professor of the Postgraduate Program in Implantology, Universidade Brasil, Fernandópolis, SP, Brazil.

[5]PhD in Dentistry, Professor at the School of Dentistry, Centro Universitário de Brasília-UDF, Brasília, DF, Brazil.

[6]PhD in Health Sciences, Professor at the School of Dentistry, Universidade Brasil, Fernandópolis, SP, Brazil.

[7]Bentham Science Ambassador, Fernandópolis, SP, Brazil.

[8]PhD in Biomedical Engineering, Professor at the Department of Integrated Dentistry Clinics and the Postgraduate Program in Bioengineering, Universidade Brasil, Fernandópolis, SP, Brazil.

[9]PhD in Biomedical Engineering, Professor of the Department of Clinical Propaedeutics and the Postgraduate Program in Bioengineering, Universidade Brasil, Fernandópolis, SP, Brazil.

*Corresponding author:
e-mail address: rogerio.enfer@gmail.com tel. +55 17 3465 4200

Abstract

Juvenile ossifying fibroma is an uncommon, fast-growing benign neoplasm classified within the group of fibro-osseous lesions that directly affect the face and the jaws. Although it is not followed by metastasis, it usually presents a highly aggressive nature. It is histopathologically divided into two patterns: trabecular and psammomatoid. It can be differentiated through the age of the patients, anatomic location and behavior of the lesion. In this context, this paper reports on the findings of a rare pathology case: juvenile ossifying fibroma, focusing primarily on its clinical, medical imaging and histopathological features characteristics, in order to familiarize the dentist, provide correct diagnosis and the most appropriate treatment.

Keywords: Ossifying fibroma; Oral pathology; Oral surgery; Oral diagnosis; Oral rehabilitation;

1. Introduction

There is a significant diversity of fibro-bone lesions that occur in the face and the jaws, it is characterized by the replacement of the normal bone by a fibrous tissue containing a newly formed mineralized product[1,2].

These lesions were first described in 1938 in a case of juvenile psammomatoid ossifying fibroma, known, at the time, as an atypical osteoid fibroma with ossification of the frontal sinus[3]. Eleven years later, there were two histopathologically similar reports on children's paranasal sinuses, after that, this pathology was renamed as a psammomatoid ossifying fibroma[4].

In 1952, the same lesion was named as active juvenile ossifying fibroma and it was described as a kind of aggressive ossifying fibroma, occurring more often in younger children[5]. Some years later, in 1965, two other authors described two cases of juvenile trabecular ossifying fibroma in two children, they were the first ones to report such a variant[6].

Juvenile ossifying fibroma is a fast-growing benign neoplasm. Although it is not followed by metastasis, it still presents a highly aggressive nature[7]. It is histopathologically divided into two patterns: trabecular and psammomatoid[8]. Despite being similar to other fibro-osseous lesions, it can be differentiated through the age of the patients, common anatomic locations and behavior of the lesion[9].

The age for its occurrence is variable, ranging from 6 months to 70 years[10]. However, the majority of reported cases are patients under 15[11]. It also affects more male than female patients and it is associated with the congenital absence of teeth, however, there are no records of malignant transformation[1,8]. It can occur in both gnathic bones, the maxilla being the most common local. Its recurrence rate varies between 30% to 58%, which could be associated with incomplete tumor removal[12,13].

Clinically, there may be painless bone expansion which, in association with faster lesion development, leads to facial deformity characterized by the presence of swellings or deformities[14]. There are no local phlogistic signs and, paresthesia or pain is rare[15]. They are able to reach the paranasal sinuses, orbit or maxilla, and it may potentially cause nasal obstruction, proptosis and intracranial involvement[9,16].

Radiographically, the lesions are circumscribed radiolucent, unilocular or multilocular[2,15]. Margins may get thinner in more aggressive lesions[14,17]. In certain cases, it is possible to observe central radiopacities and a line between the tumor and healthy bone tissue. In some cases, it is also possible to observe ground-glass opacity. Still, when the tumor is inside the sinus, it can appear radiodense and fog-like aspect and, thus, be confused with sinusitis[10,13,18].

Both histological patterns are not encapsulated, presenting themselves as well-defined masses having more fibrous cellular connective tissue in some of its areas the in others. Myxomatous foci can be found and it is often associated with pseudocystic degeneration[19]. Mitotic figures can be found in fewer numbers[11]. Bleeding and small clusters of multinucleated giant cells may be observed[10]. The mineralized tissue in the two patterns is very different; the trabecular type has irregular clusters of highly cellular osteoid typically including bulky and irregular osteocytes. However, the psammomatoid type shows concentric lamellae and spherical ossicles which vary in shape, having basophilic centers with an eosinophilic peripheral osteoid margin[12,14].

The treatment for juvenile ossifying fibroma is defined in terms of tumor's behavior and stage. Many

reports state that conservative interventions such as curettage and enucleation of the lesion, presented better results, but only in less aggressive cases[16,19]. When the tumor is too big along with a fragile bone cortex, tooth displacement or resorbed roots, the common procedure is to have a block resection with clear tumor margins and bone graft reconstruction[17].

The present study reports a case of juvenile ossifying fibroma, a rare and rapidly developing pathology[7], which, if not diagnosed and treated in its early stages may present excessive local aggressiveness, directly affecting dental positioning and facial symmetry and in rarer cases, the noble anatomic structure, result in patient's death[18]. A primary focus is given to its clinical, visual and histopathological characteristics, which are fundamental for the identification, correct diagnosis and appropriate treatment for the lesion.

2. CLINICAL CASE REPORT

Female patient, 14 years old, parda/mulatta, was admitted in the Centro de Especialidade Odontologicas (CEO), Fernandópolis/SP, complaining of facial swelling which started approximately 2 months earlier. No risk habits or pre-existing systemic changes were reported during the anamnesis.

An extraoral cavity examination was performed (Figure 1) and a slight facial asymmetry with slight swelling was found on the left labial commissure of the mouth, there were no signs of inflammation, asymptomatic and fibrous on palpation. The local was normochromic, with no lymphadenopathy or any other alteration.

Figure 1. Physical appearance of the face. It is possible to observe a slight swelling on the left side of the maxillary-zygomatic region.

During the intraoral cavity examination (Figure 2), swelling was observed at the left upper alveolar ridge, expanding through both the buccal and palatal regions and covering the premolars and the entire

hard palate. There was crowding of the teeth, however, there were no signs of displacement. The lesion was covered by intact and normochromic mucosa and was asymptomatic and hard on palpation.

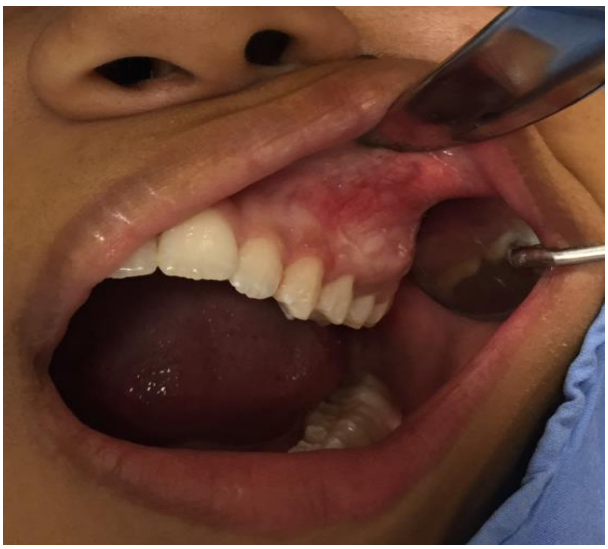

Figure 2. Early intraoral appearance. There is a significant increase in the premolar region on the left side.

The patient had a panoramic radiograph (Figure 3) which showed a circumscribed unilocular "ground-glass opacity" between teeth 24 and 25, with slight displacement.

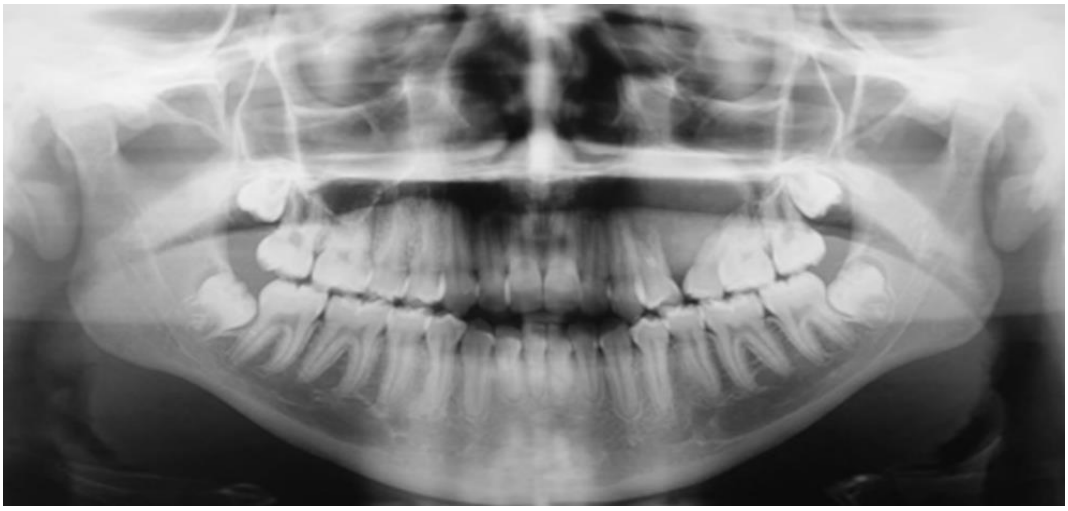

Figure 3. First panoramic radiograph. It indicated the presence of a circumscribed radiopaque unilocular lesion causing displacement of the 24 and 25 elements.

Then, a computed tomography (Figures 4, 5 and 6) was requested, which showed a hyperdense, homogeneous, well-defined image of osteolytic character and apparent disorganization of the medullary bone. The lesion was shown to be expansive, indicating bone cortices and adjacent structures displacement (maxillary sinus, nasal fossa floor and alveolar process). Teeth 24 and 25 were dislocated and there was external root resorption on the 23, 24, 25 and 26 elements.

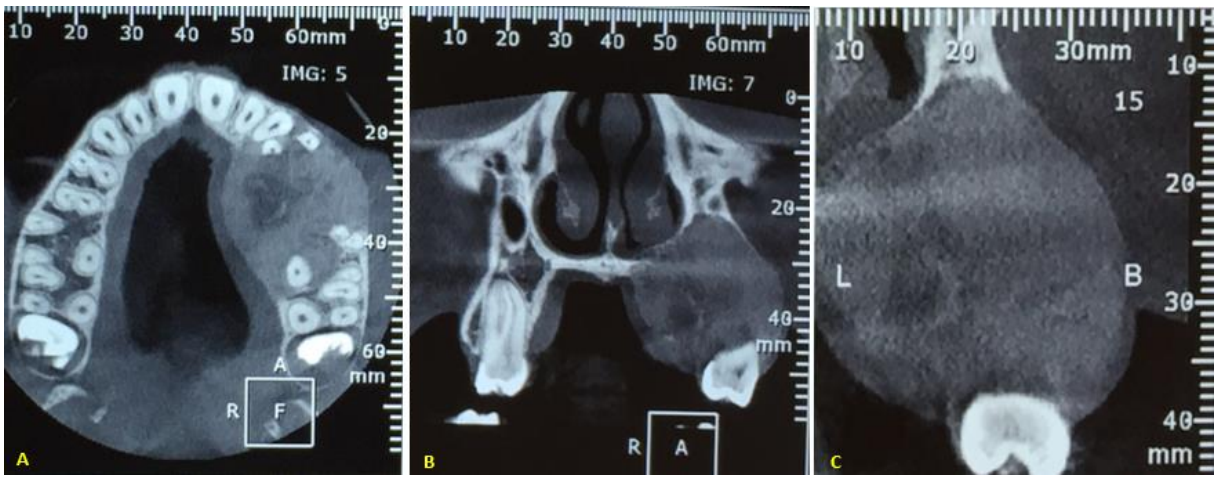

Figure 4. First CT scan. It is possible to observe some expansion of the vestibular and palatal bone margin, involving the 23, 24, 25 and 26 teeth, apical reabsorption, invasion of the maxillary sinus and nasal cavity floor. A: axial cut. B: coronal cut. C: oblique cut.

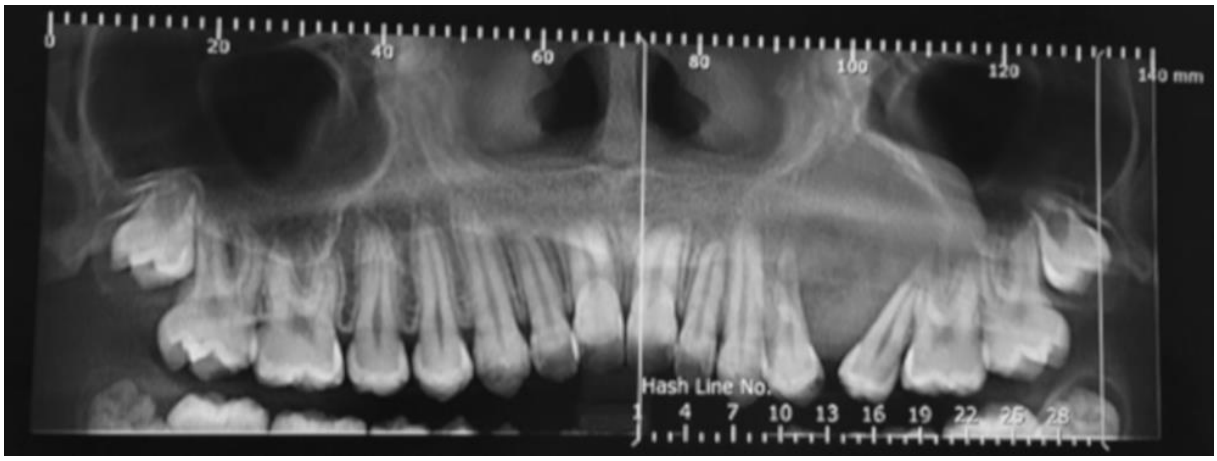

Figure 5. Panoramic reconstruction of the first CT showing the extent of the lesion and involvement of the teeth 23, 24, 25 and 26; teeth displacement and the anterior wall and floor of the maxillary sinus.

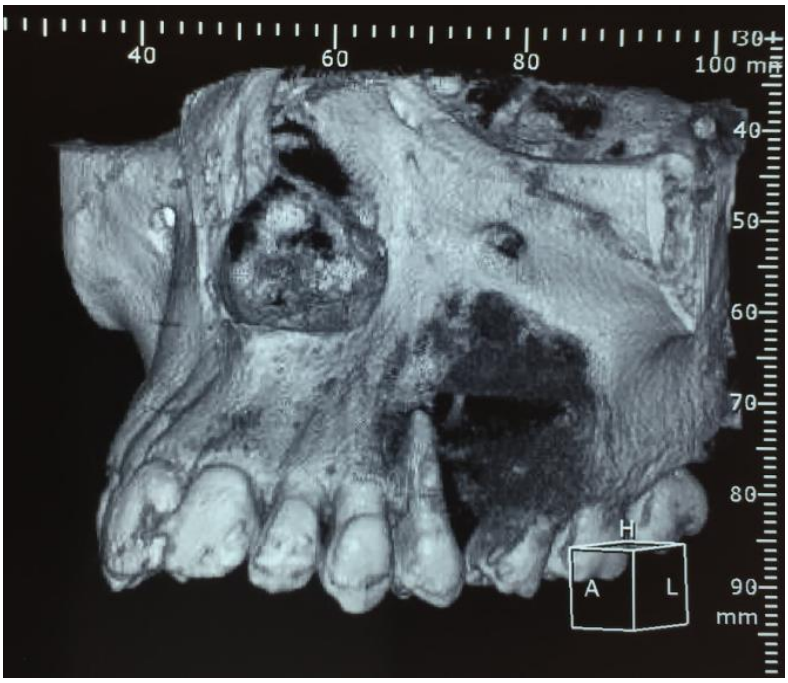

Figure 6. 3D reconstruction of the first CT scan indicating bone tissue loss at the injury location and its extension.

The clinical and imaging results strongly suggested fibrous dysplasia or ossifying fibroma as a differential diagnosis. Therefore, an incisional biopsy was performed (Figure 7) through intraoral cavity, and the collected material was conditioned and sent to further analysis.

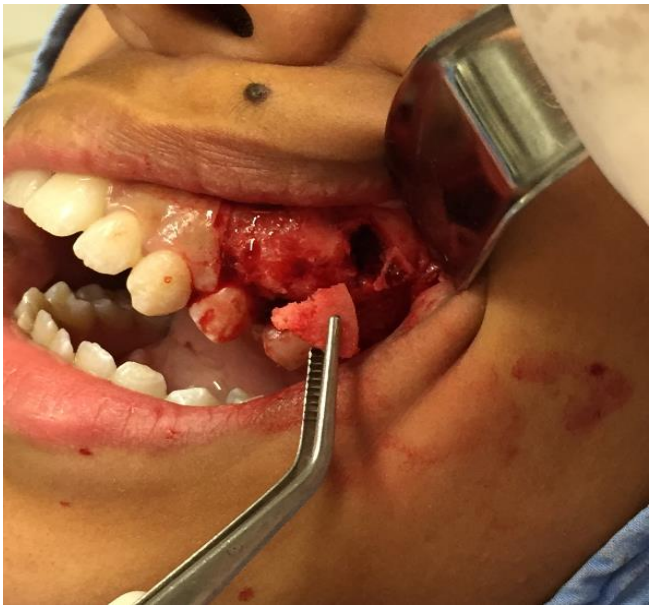

Figure 7. Material collected from the incisional biopsy, through intraoral access, and taken to anatomopathological analysis.

The anatomopathological result showed a non-encapsulated lesion with irregularly shaped immature bone trabeculae in a loosely arranged fibrous cell stroma, suggests fibrous dysplasia to a greater extend (Figure 8). However, certain areas were more disorganized, presenting a mixture of immature and lamellar

bone, which is compatible with ossifying fibroma. Thus, the histopathological diagnosis detected a benign mesenchymal neoplasia, indicating complete excision of the tumor for a definitive diagnosis.

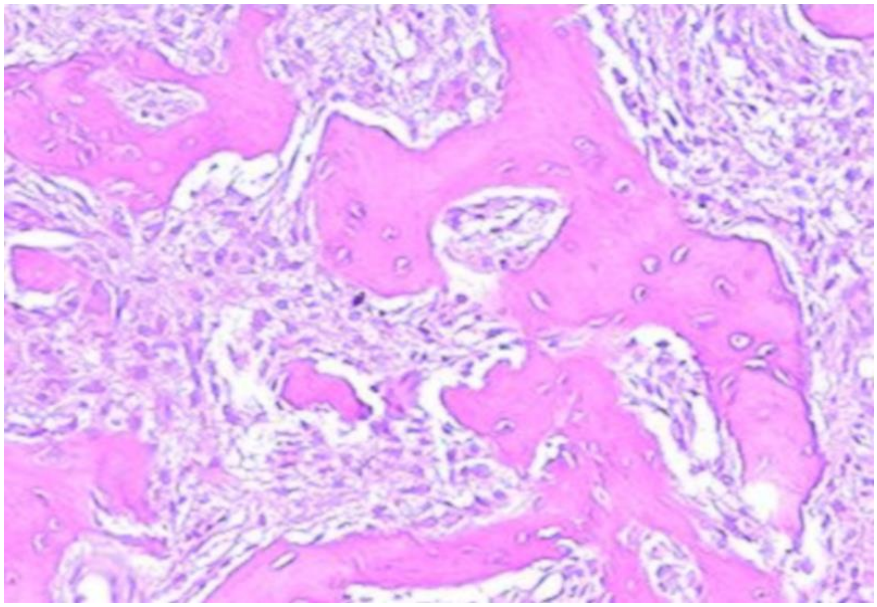

Figure 8. Histopathological aspect showing immature bone trabeculae and lamellar bone in a loosely arranged fibrous cell stroma - H.E. (200x).

The patient was referred to the Hospital de Base of São José do Rio Preto / SP and two months after the first surgical procedure (incisional biopsy), the enucleation of the lesion was performed through intraoral access in a hospital environment under general anesthesia. The removed part was properly packed and sent for further histopathological analysis, which confirmed the diagnosis of juvenile ossifying fibroma.

After six months of follow-up, a new panoramic radiograph (Figure 9) and computed tomography (Figure 10) were taken. Both, the imaging tests and the clinical aspect (Figure 11), indicated no signs of recurrence. The patient remained asymptomatic, without paresthesia, with no mobility in the adjacent teeth and normal facial appearance.

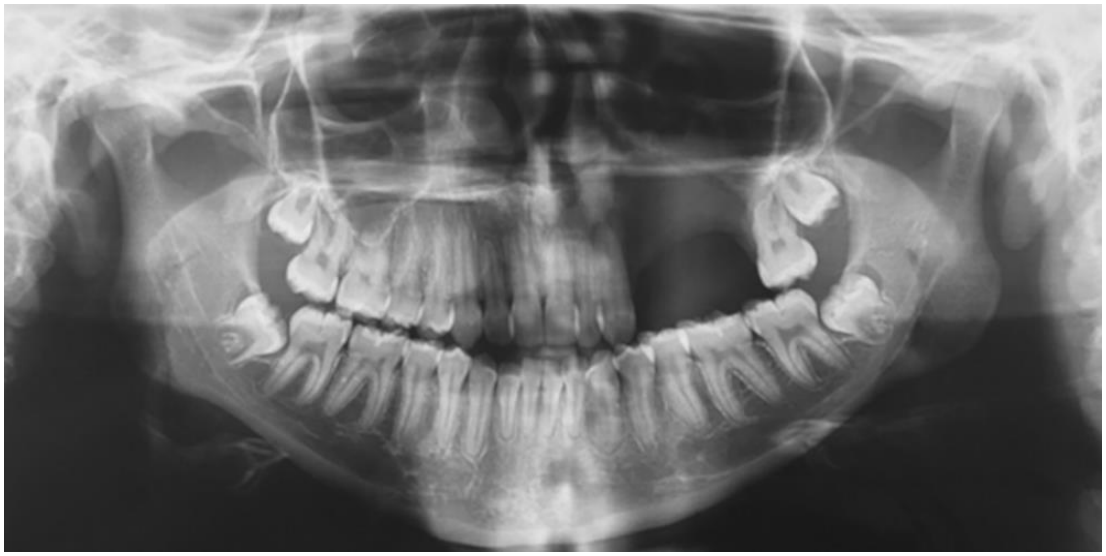

Figure 9. Panoramic radiograph after a 6 six months postoperative period showing the absence of teeth 24, 25 and 26, which were removed along with the lesion, it also indicates the presence of cortical bone line on the anterior wall and nasal sinus floor with no signs of oroantral communication.

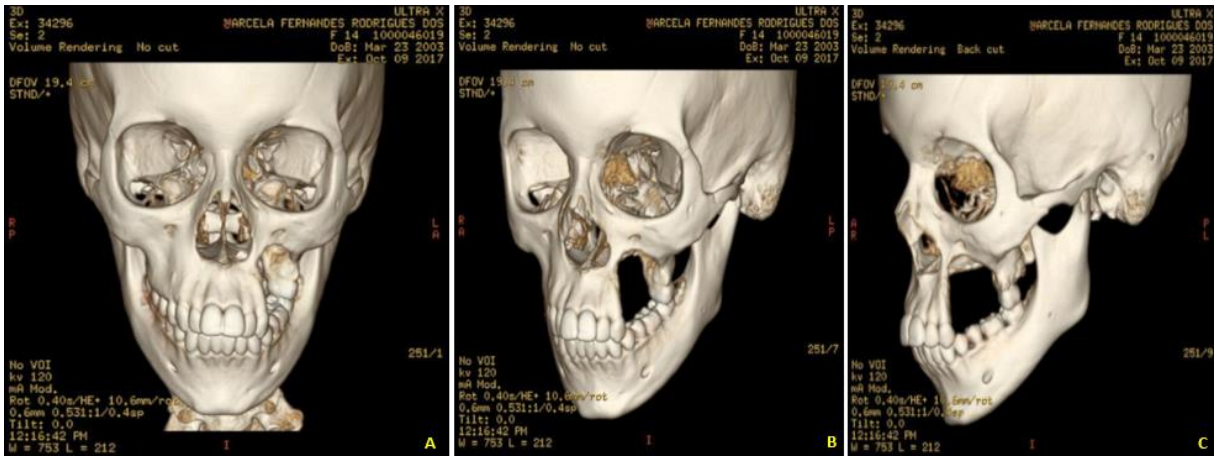

Figure 10. 3D reconstruction of CT scan after 6 months postoperative period showing where the enucleation was performed.

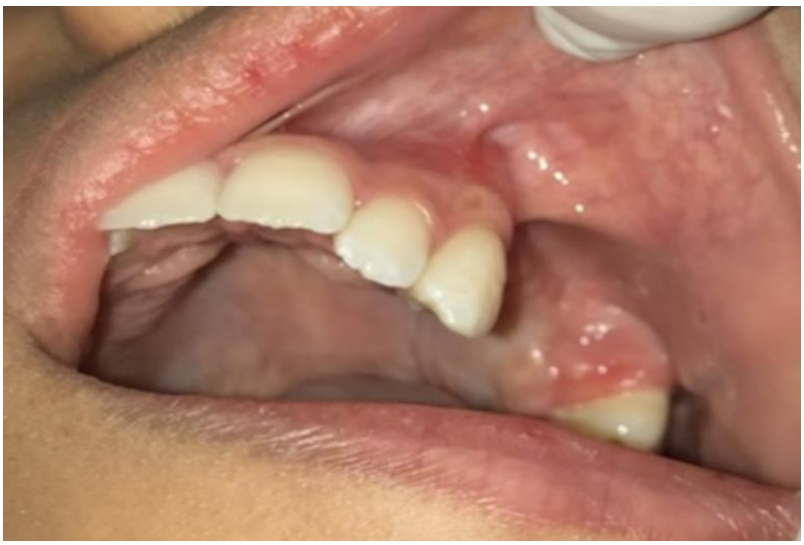

Figure 11. Intraoral appearance 6 months after the operation. It is observed absence of the 24, 25 and 26 teeth, healthy mucosa, without periodontal disease, without oroantral communication.

In order to be able to have an adequate life condition and to keep her aesthetic and masticatory functions. The patient was rehabilitated using a maxillofacial prosthesis (Figure 12) due to the significant tissue loss during the procedure.

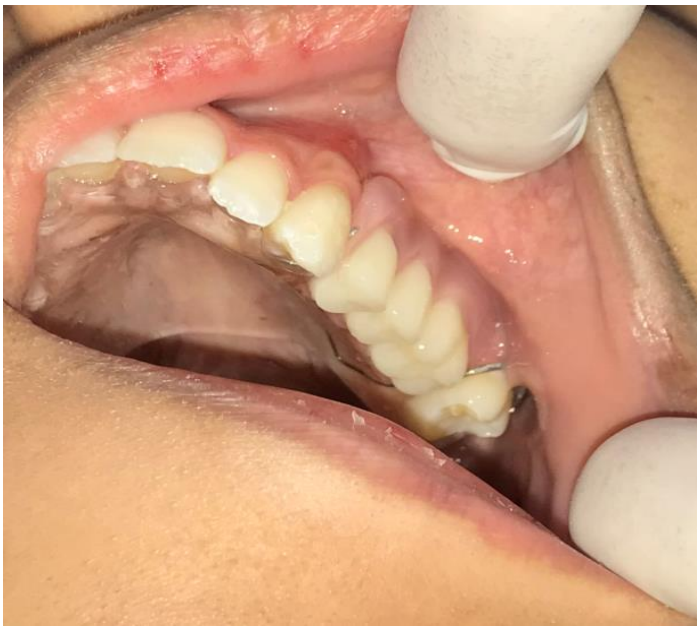

Figure 12. Oral rehabilitation using maxillofacial prosthesis, enabling aesthetics and masticatory function.

3. Discussion

Juvenile ossifying fibroma is a fibro-osseous lesion with rapid development and great local aggressiveness[7,9,14], the case study in this paper presented a patient with an expansive tumor, teeth and bone cortical displacement, which led to facial asymmetry within two months' time.

Its occurrence is more common in patients under 15 and male patients are slightly more affected than female, it can reach both gnathic bones, but the most common location is maxilla[8,18]. In this case report, the age group and location matched the existing literature, there were no significant relevance regarding gender for the differential diagnosis of the lesion.

The radiographic examination indicated the lesions to be well-defined radiolucent, unilocular or multilocular, with central radiopacities and ground-glass opacity[2,11] in cases like this. CT scans and MRIs can also provide a better visualization of the affected bone and structures, the latter being important when there is ocular and neurovascular involvement[1,12,13].

Two histopathological patterns could be observed: psammomatoid and trabecular. Both are shown as well-defined, non-encapsulated masses, differing each other due to the mineralized component[8,10,11]. In this case report was not possible to define the exact pattern, even after a second analysis with a larger amount of material, because characteristics of the two variants were present. Thus, the pathology was referred only as juvenile ossifying fibroma.

The differential diagnosis is usually made with fibrous dysplasia[15,19], it typically requires a complete anatomopathological analysis, as described in this case. Thus, the final diagnosis is reached by combining clinical, imaging and histopathological exams.

There are multiple options for treatment it and must be chosen according to the behavior and stage of the tumor. Many reports suggest that, in less aggressive situations, better results are obtained from conservative interventions, such as surgical enucleation, successfully performed on this patient. In more aggressive cases, less conservative treatments may be chosen, such as block resection with a safety

margin[10,17]. This stage is of paramount importance since it is assumed that high rates of recurrence are associated with deficient removal of the lesion, which can range from 30 to 58% of cases[13,16].

4. Conclusion

After the presenting this case study, it can be concluded that knowing the clinical, imagological and histopathological characteristics of juvenile ossifying fibroma favors its correct diagnosis and has profound influence on choosing the most appropriate treatment in each situation.

Conflicts of interest

The authors declare no conflict of interest.

5. References

[1] L.M.G. Figueiredo, R.O.H. Valente, V.A. Sarmento, S.C. Trindade, T.F.L. Oliveira and W.R.M. Costa, Aspectos atuais no diagnóstico e tratamento do fibroma ossificante juvenil. Rev Bras Cir Cabeça Pescoço. 2012;41(2):99-102.

[2] S.M. Gondivkar, A.R. Gadbail, R. Chole, R.V. Parikh and S. Balsaraf, Ossifying fibroma of the jaws: report of two cases and literature review. Oral Oncology. 2011;47(9):804-809.

[3] C.E. Benjamins, Das osteoid-fibrom mit atypischer verkalkung im sinus frontalis. Acta Otolaryngol. 1938;26:26-43.

[4] H. Gögl, Das psammo-osteoid fibroma der nase und ihrer Nebenhöhlen. Monatsschr Ohrenheilkd Laryngorhinol. 1949;83:1-10.

[5] L.C. Johnson, M. Yousefi, T.N. Vinh, D.K. Heffner, V.J. Hyams and K.S. Hartman, Juvenile active ossifying fibroma: its nature, dynamics and origin. Acta Otolaryngol. Suppl 1991;448:1-40.

[6] R.J. Reed and D.M. Hagy, Benign non-odontogenic fibroosseous lesions of the skull: report of two cases. Oral Surg Oral Med Oral Pathol. 1965;19:214-27.

[7] A. Suarez-Soto, M.C.B.R. Hermosa, I. Minguez-Martínez, L.M. Floría-García, J. Barea-Gámiz, J. Delhom-Valero and P. Risueño-Mata, 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(3):479-485.

[8] S. El-Mofty, Psammomatoid and trabecular juvenile ossifying fibroma of the craniofacial skeleton: Two distinct clinicopathologic entities. Oral Surg, Oral Med, Oral Pathol, Oral Radiol Endod. 2002;93(3):296-304.

[9] J.G. Paiva, F. Böing, M.B. Benaglia and A. Nascimento, Fibroma ossificante: relato de 2 casos. Rev. Cir Traumatol Buco-Maxilo-Fac. 2009;9(1):33-40.

[10] A.C. Chi, Patologia óssea. In: B.W. Neville, D.D. Damm, C.M. Allen and J.E. Bouquot, editores. Patologia oral e maxilofacial. 4 ed. Rio de Janeiro: Elsevier; 2016. p. 615-678.

[11] L. Barnes, J.W. Eveson, P. Reichart and D. Sidransky, editors. World Health Organization Classification of Tumours. Pathology and genetics of head and neck tumours. Lyon: IARC Press; 2005.

[12] B. Keles, M. Duran, Y. Uyar, A. Azimov, A. Demirkan and H.H. Esen, Juvenile Ossifying Fibroma of the Mandible: a Case Report. J Oral Maxillofac Res. 2010;1(2):1-7.

[13] B.R. Peterson and B.L. Nelson, Juvenile Active Ossifying Fibroma. Head Neck Pathol. 2015;9(3):384-386.

[14] S.F. Smith, L. Newman and D.M. Walker, Juvenile aggressive psammomatoid ossifying fibroma: an interesting, challenging, and unusual case report and review of the literature. J Oral Maxillofac Surg. 2009;67:200-206.

[15] A.S. Espinosa, J. Villanueva, H. Hampel and D. Reyes, Spontaneous regeneration after juvenile ossifying fibroma resection: a case report. Oral Surg, Oral Med, Oral Pathol, Oral Radiol Endod. 2006;102(5):32-35.

[16] A.J. Saiz-Pardo-Pinos, M.O.V. Olmedo-Gaya, E. Prados-Sánchez and M. Vallecillo-Capilla, Fibroma óseo juvenil: a propósito de un caso clínico. Med oral Patol Oral Cir Bucal. 2004;9:454-458.

[17] M.E. Papadaki, M.J. Troulis and L.B. Kaban, Advances in diagnosis and management of fibro-osseous lesions. Oral Maxillofacial Surg Clin N Am. 2005;17:415-434.

[18] S. Ram, A. Kumar, I. Ali, P. Wadhwani, A. Saxena and M.I. Ali, Juvenile ossifying fibroma of mandible. Dentistry and Medical Reserch. 2014;2(1):14-16.

[19] R.L.M. Nogueira, C.F.W. Nonaka, R.B. Cavalcante, A.C. Carvalho and L.B. Souza, Fibroma ossificante juvenil localizado em mandíbula: relato de caso e breve revisão da literatura. Rev Cir Traumatol Buco-Maxilo-fac. 2009;9(1):25-32.