

## Up-to Date Review And Case Report

# Juvenile ossifying fibroma: case report and literature review. Management and differential diagnosis

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**Abstract – Introduction:** Juvenile ossifying fibroma (JOF) is a rare neoplasm characterized by the replacement of the normal bone matrix with osteo-fibrous tissue. It has the tendency to be locally aggressive despite its benign character and to have a strong tendency for recurrence. **Observation:** In this case report, the patient is a young man, aged 16, with rapidly advancing maxillary swelling. We describe the diagnostic procedure, the surgical procedure and the differential diagnosis to be eliminated. **Discussion:** The clinical presentation of JOF, and its rapid growth, can cause fear of other pathologies such as osteosarcoma. The radiological characteristics should reassure the practitioner and a histological examination confirmed the diagnosis. **Conclusion:** JOF is a benign tumor. It should be operated on at an early stage because of its rapid growth. In its clinical and histological presentation, its trabecular form may mimic an osteosarcoma.

## Introduction

The juvenile ossifying fibroma (JOF) is a rare benign tumor [1], and it belongs to the group of fibro-osseous tumors according to the 2005 WHO classification [2]. The incidence of JOF is so difficult to estimate because no values have been reported in the current literature. JOF shows a significant locally aggressive nature along with a significant growth potential which can raise fears of malignant pathology. It predominantly affects children but may occur in adults [3].

## Observation

A 16-year-old patient presented to us for an oral checkup with a firm vestibular swelling in the right maxillary area and which was otherwise completely asymptomatic.

During questioning, the patient reported a rapid and progressive increase in size over a 2-year period, with a slight aesthetic issue but without pain. There was no other relevant medical history. The general state had not been altered nor was there any evidence of lymphadenopathy.

There was a visible facial asymmetry with a curvature of the upper right lip. Intraorally, there was vestibular swelling to the right, which extended from teeth 13–16 and measured approximately 2 cm on the major axis. On palpation, the mass was continuous with the maxilla. The mass was uniformly hard in consistency without fluctuation or tenderness. The mucosal lining was normal in appearance. The teeth were healthy and immobile (Fig. 1). Clinically, the relatively rapid growth and size of the lesion raised the suspicion of malignancy despite the lack of pain. The differential diagnoses were fibrous dysplasia, JOF, and malignant bone tumor.

As first step, a panoramic X-ray was obtained (Fig. 2). A discrete osteocondensing lesion adjacent to teeth 13–16 was observed, and its center was more dense. The lesion appeared unilocular, and no other lesions were visible.

Given the lack of information provided by the panoramic X-ray, a cone beam computed tomography (CT) was also performed. It showed a well-defined bone lesion with a heterogeneous content pressing on the cortex without traversing it. Areas of bone condensation were visible within the lesion. The lesion appeared to develop around the roots of tooth 14, its largest axis measuring 20 × 35 × 11 mm (Fig. 3).

The well-defined and homogeneous nature of the lesion on suggested its benign origin. Considering the patient's aesthetic issue and the benign radiological features, an excision without

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**Fig. 1.** Vestibular tumefaction. The recovering mucosa appears normal.

prior biopsy was decided on. Surgical intervention under general anesthesia was scheduled for lesion excision and a histological diagnosis.

The surgical intervention is illustrated (Fig. 4). The site was infiltrated with xylocaine–adrenaline to promote the detachment of the mucoperiosteal flap, and to decrease bleeding and postoperative pain. With the aid of an osteotome and a mallet, most of the lesion was removed, taking care not to injure the adjacent dental roots. The lesion was easily separated from the underlying bone. Curettage of the operative area was performed to remove the remaining tumor tissue on visual evaluation. The closure of the site was performed with simple, resorbable sutures. The lesion was then sent to the pathology department. There were no complications associated with the surgical intervention, and the patient had no complications during postoperative follow-up at 24 h.

Histological examination revealed a benign osseofibrous bone tumor corresponding to a trabecular JOF (Fig. 5). The excision was intralesional.

The patient did not come in for his 3-month postoperative appointment, but eventually came for follow-up 8 months afterward. There was no facial asymmetry or intraoral swelling, and mucosal healing was satisfactory. A repeat cone-beam CT was performed (Fig. 6) revealed an osseodense lesion between teeth 13 and 14. As the patient was no longer experiencing any discomfort, another surgical procedure was deemed unnecessary and was advised regular monitoring. The patient did not attend the scheduled 1-year follow-up.

## Discussion

Ossifying fibromas (OF) are rare fibro-osseous tumors. They are made up of osteogenic calcified matrix, and they are not of odontogenic origin as previously thought [4]. The terms

cementifying and cemento-ossifying are no longer used because they designate the same entity [5]. Their epidemiology is poorly known because they have long been confused with cemento-osseous dysplasia [6]. Their location is mainly limited to the craniofacial bones [4].

OFs can be present in two forms: conventional and juvenile [7]. The first usually occurs in the mandibles of 30–40-year-old women [8]. On the other hand, the active juvenile form is very rare, grows rapidly, affects children or young adults, and has is more common in the maxilla [9]. It occurs before age 15 years in 80% cases [10]. In literature, there has been some debate regarding the sex predilection. According to El Mofty, [10] males tend to be more frequently affected whereas others argue that females are more frequently affected [11]. Clinically, JOF presents a painless swelling with rapid growth. It may cause dental displacements [12] and may be accompanied by proptosis if it occurs in the maxilla.

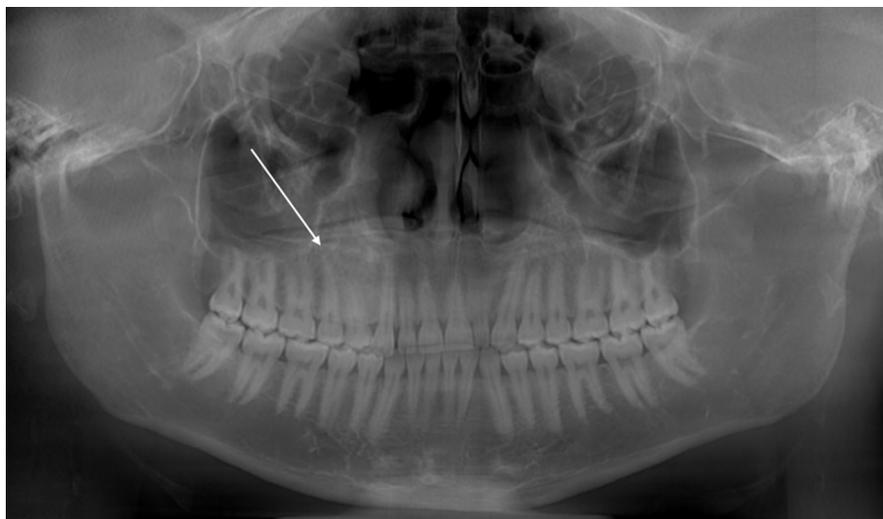
Two distinct histoclinical entities have been described: the psammomatoid and the trabecular. The former can be found in the sinus, paranasal, and orbital regions, whereas the latter is generally located in the maxillary region in men [9,13]. The psammomatoid occurs more frequently and is also more aggressive with a higher recurrence rate than the trabecular form [14]. In its trabecular form, JOF may be similar to an osteosarcoma in its aggressiveness [15]. In the case reported here, despite its relatively rapid growth, the painless nature and healthy appearance of the mucosa were reassuring.

Radiologically, JOF presents itself as a single, expansive, and well-defined lesion of variable and increased density with mineralization. Therefore, it may be completely radiopaque in the initial stages [16]. In most cases, the cortex is swollen but not traversed, which indicates a benign character. However, a cortical intrusion has been reported in cases previously [1,10].

Histologically JOFs correspond to very limited proliferation of fibrocellular tissue and mineralized substance. The rapid growth of JOF, reflected in the presence of mitoses upon microscopic examination, may give rise to fears of a malignant tumor. The absence of cytonuclear atypia and abnormal mitoses confirms its benign character. The trabecular form of JOF has a characteristic distinct border of turgescent osteoblasts [15].

The treatment is surgical and consists of performing an enucleation as completely as possible by first adapting the approach to the location [5].

Monitoring must be long-term because 36%–50% cases experience recurrence. There have been no malignant transformations reported in the literature [17]. In the present case, the patient discontinued their follow-up at 8 months after procedure. Recurrence was therefore highly probable given the intralesional excision. However, it must be noted that scheduling a complete excision of the lesion with a partial maxillectomy would have been too invasive.



**Fig. 2.** Panoramic X-ray showing a slightly osteodense lesion in relation to teeth 13–16. Root divergence is noted on 13 and 14.



**Fig. 3.** Cone-beam CT highlighting a well-defined right maxillary bone lesion consisting of homogeneous content compressing the cortex without traversing it. There are bone condensations within the lesion.

The differential diagnosis to consider in osseous tumefaction are osteomas and osteochondromas. These are benign, asymptomatic tumors that are mostly slow-growing. Radiologically, they present as a dense, homogenous, and well-defined masses. The occurrence of such tumors in multiple locations must raise suspicion of Gardner's syndrome [18].

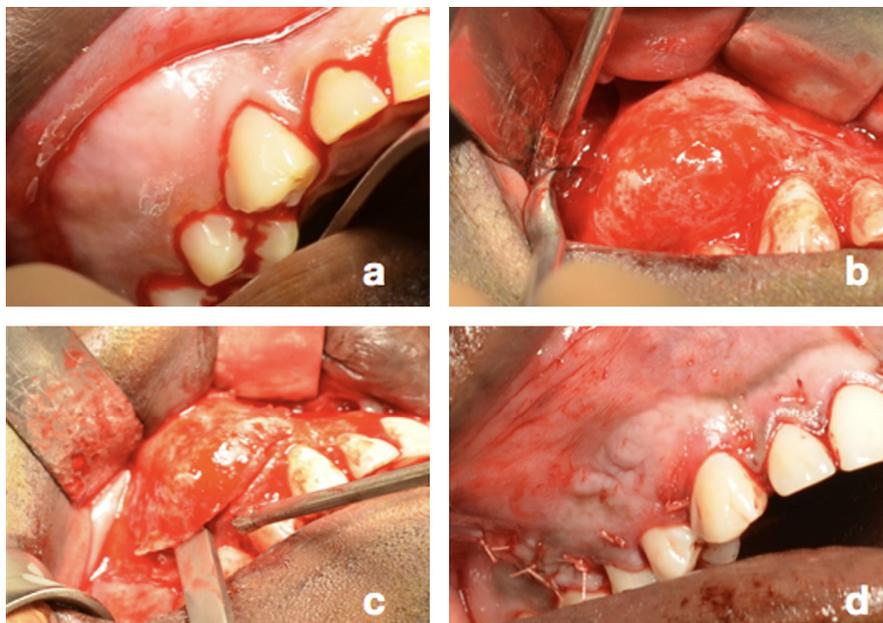
Benign bone growths, such as tori or exostoses, should also be considered [19]. They tend to be asymptomatic and bilateral, and excision is not always justified. They form a dense, very corticalized image on the vestibular edge of the maxillary alveolar bone.

JOF must be distinguished from other OF lesions that are characterized by the replacement of normal bone tissue with more or less cellular fibrous tissue including ossified structures. These include the following:

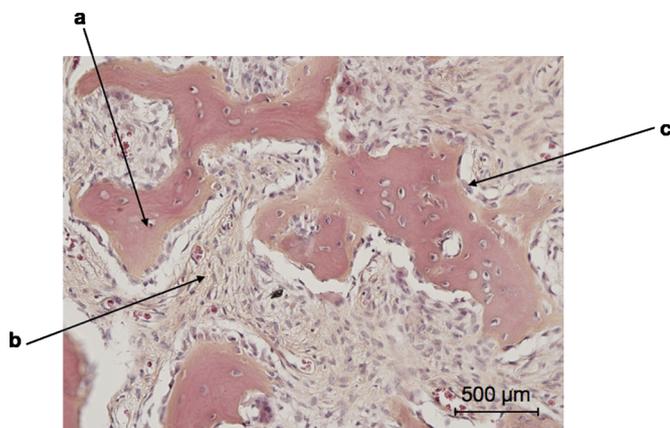
- fibrous dysplasia, which develops slowly, can cause bone pain or deformities. Radiologically, it has a somewhat homogeneous, frosted glass appearance and a continuity with the normal bone unlike JOF. A sarcomatous transformation is reported in 0.5% of cases;
- cemento-osseous dysplasia, a reactionary or dysplastic lesion surrounding the alveolodental ligament, may occur close to dental apices. It is asymptomatic and generally does not exceed 2 cm. These lesions must not be biopsied because of the risk of a secondary healing disorder [20];
- osteosarcoma, especially in its trabecular form that can mimic JOF. Clinically, the symptomatology is generally painful, unlike JOF [15]. Radiologically, the lesion is both osteolytic and osteosclerotic and causes cortical destruction. There may be a periosteal reaction and soft tissue invasion. Histologically, cytonuclear atypia favors this pathology.

## Conclusion

The diagnosis of JOF made by a combination of clinical, radiological, and histological evidence. JOF is a rare, locally aggressive lesion with a high potential for recurrence. This is why excision must be as complete as possible while ensuring the safety of the neighboring anatomical structures, and treatment should include regular patient follow-ups.



**Fig. 4.** Operating time during the excision of the lesion (a) vestibular incision in relation to teeth 11–16 with distal discharge; (b) and (c) with the aid of an osteotome and a mallet, most of the swelling was removed; (d) closure of the operative site.

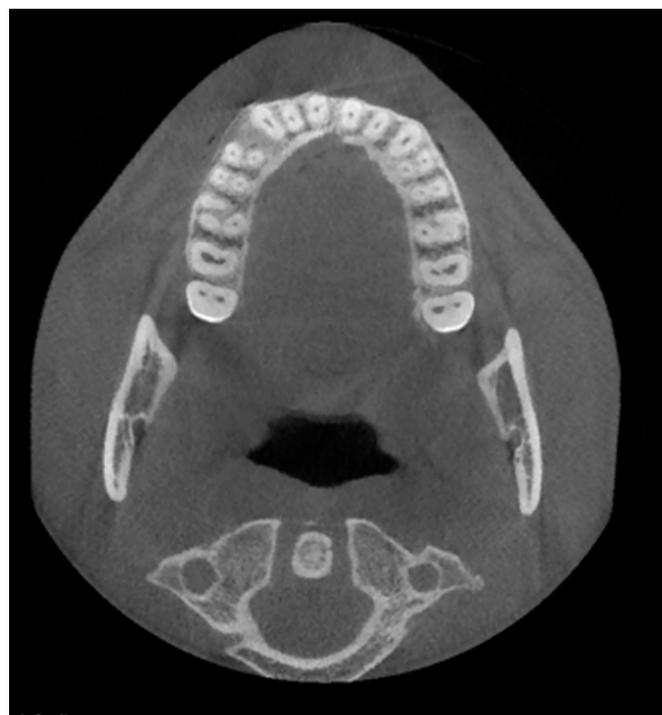


**Fig. 5.** Histological section of the largest fragment. H&E staining (a) bone spans of irregular contours; (b) very cellular collagenic background; (c) osteoblastic border testifying to tumor aggressiveness, specific to the trabecular form of JOF.

**Conflicts of interests:** The authors declare that they have no conflicts of interest in relation to this article.

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**Fig. 6.** CBCT 8 months after the surgical intervention. Axial section shows no recurrence of the initial lesion.

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