

## Case Report

# A rare cause of maxillary mass: juvenile ossifying fibroma

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### ABSTRACT

Juvenile ossifying fibroma (JOF) is a unique fibro-osseous neoplasm. It has 2 histopathological variants. Psammomatoid juvenile ossifying fibroma (PsJOF) and Trabecular juvenile ossifying fibroma (TrJOF) affecting the jaws of children. Only 20% of the patients are over 15 years of age. JOF is more common in maxilla than mandible. It presents as an asymptomatic progressive, rapid expansion of jaws. It has a recurrence rate of 30-58%. Surgery is the only cure for this kind of lesion and total excision should be the goal of the treatment. We present a case report of 12 year-old female patient with clinical, radiographic and histopathological features of Psammomatoid JOF which clinically admitted to our clinic as maxillary sinus mass.

**Keywords:** Juvenile ossifying fibroma, Neoplasm, Sinüs

## INTRODUCTION

Juvenile ossifying fibroma (JOF) is usually diagnosed in the first or second decade of life. JOF, which arises from the periodontal ligament, constitutes 2% of oral tumours in children. It is seen equally in men and women. While JOF is usually diagnosed in the facial bones (75%), it is also seen in the calvarium (12%) and mandible (10%), with rare extracranial involvement in 3% of JOF cases.<sup>1,2</sup>

Two histopathological types of JOF have been described: psammomatoid JOF (PsJOF) and trabecular JOF (TJOF). While the trabecular variant of JOF is frequently seen in the maxilla, PsJOF is often observed in the sinonasal and orbital bones.<sup>2</sup> We present a case report of 12-year-old female patient with clinical, radiographic and histopathological features of psammomatoid JOF.

## CASE REPORT

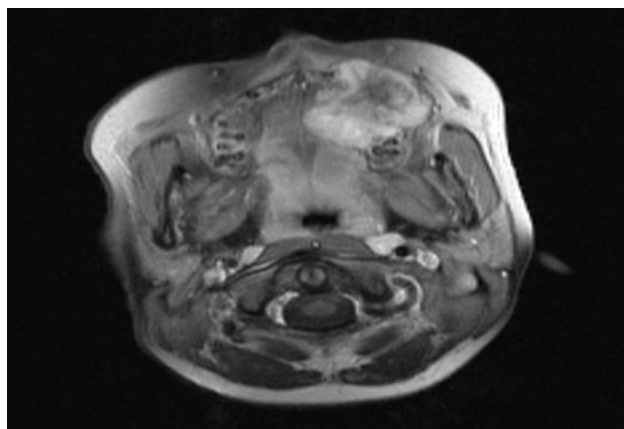
A 12-year-old female was admitted to our clinic for a growing swelling over her left cheek that had been present for approximately 1 year. On examination, a

mass, which was sensitive to palpation, covered the left side of the hard palate and invaded the maxillary sinus. The lesion extended toward the left second premolar (Figure 1). She had no symptoms of another systemic disease. A complete blood count and routine biochemical tests were normal. Computed tomography (CT) revealed a solid 37 x 41 x 33 mm hypodense mass at the level of the first and second premolars inferiorly, extending to the medial palatine suture medially, filling the left maxillary sinus completely, and eroding the left side of the maxilla. Facial magnetic resonance imaging showed a solid mass in the left maxilla causing marked bone destruction, extending between the first and second premolars inferiorly and the maxillary sinus superiorly, and the palatine suture medially (Figure 2). A diagnostic incisional biopsy was compatible with JOF. Subsequently, a left partial maxillectomy was performed. A left Weber Ferguson incision was made, exposing the lesion completely. A Le Fort I osteotomy was performed to excise the entire mass, which filled the maxillary sinus completely, extending to the zygoma laterally, piriform aperture medially, base of the orbit superiorly, canine teeth inferolaterally, and second premolar inferomedially

(Figure 3). Temporary soft palate prosthesis was inserted postoperatively at the Clinic of Prosthetic Dentistry in the Faculty of Dentistry (Figure 4). The pathology was reported as “psammomatoid type juvenile ossifying fibroma”. There were no complaints or evidence of relapse at the 1-year follow-up exam.



**Figure 1: Preoperative photo of the patient.**



**Figure 2: Preoperative MR imaging.**



**Figure 3: Postoperative photo of the patient.**



**Figure 4: Postoperative photo of the patient with soft palate prosthesis.**

## DISCUSSION

Ossifying fibroma, first described by Menzel in 1872, is a rare primary benign bone tumour; in 1927, Montgomery named the tumour juvenile aggressive ossifying fibroma when it is seen in children.<sup>3</sup> Psammomatoid juvenile ossifying fibroma (PsJOF) was first reported in 1938 and described as an atypical ossifying osteoid fibroma in the frontal sinus. In 1949, two more cases were reported and named PsJOF. Recently, El Mofty separated ossifying fibromas into TJOF and PsJOF based on histological criteria.<sup>4,5</sup> PsJOF commonly involves the paranasal sinus and orbital bones, whereas TJOF mainly affects the jawbones, although there is controversy as to whether the maxilla or mandible is affected more often. These lesions are more common in women.<sup>4,6</sup> In one series, 70% of PsJOF cases were observed in the paranasal sinus, 20% in the maxilla, and only 10% in mandible. This lesion recurs in 30-58% of cases.<sup>6</sup> Since patients are generally asymptomatic and the mass grows slowly, it may be large when initially diagnosed.<sup>4</sup>

The etiology of the disease is unclear. Genetic factors and facial trauma are possible etiological factors in JOF.<sup>1</sup> The clinical symptoms are variable and include facial swelling, a palpable hard mass, sinus complaints, nasal obstruction, tooth displacement, proptosis, and eye pain. Our patient complained of a tender swelling involving the left half of the face and hard palate. While Johnson, et al found paranasal sinus and mandible involvement from the facial bones in 90% and 10% of cases, respectively, Slootweg claimed that the maxilla was the bone most often involved.<sup>7,8</sup> In our case, the mass filled almost the entire left maxillary sinus.

The JOF lesion is not encapsulated, but is clearly separated from bone, in contrast with fibrous dysplasia, which has unidentifiable borders and is mistaken for normal bone tissue.<sup>9,10</sup> The psammomatoid variant has a

basophilic center, while the peripheral portion sometimes produces optical emissions, giving it a fringed appearance. The trabecular variant reflects the trabecular structure of immature bone, and may or may not be surrounded by osteoblastic activity, unlike spheric or ovoid bone.<sup>2,11</sup> Histopathologically, our case had psammoma corpuscle-like structures, in addition to cement-like islands and bone trabeculae in cellular fibrous tissue. These spherule structures have brush borders.

On radiographic examination, an aspheric, rarely cystic, cortical osteolytic lesion is observed. On CT, the lesion is hypodense compared to normal bone. Lesions may range in size from 2 to 8 cm. On imaging, PsJOF is multilobular. Low-density areas seen on imaging reflect cystic changes.<sup>4,5</sup> In our patient, maxillofacial CT showed that the lesion was hypodense compared with normal bone tissue; it was solid, measuring 37 x 41 x 33 mm, at the level of the first and second premolars inferiorly, extending to the palatine suture medially, filling the left maxillary sinus completely, and eroding the left side of the maxilla.

As an ossifying fibroma is separated from the surrounding bone by a characteristic border and has a capsule, the treatment is enucleation. Small tumours can be treated with curettage, and relapse is rare. However, ossifying fibromas of the jawbones have relapse rates of 30-58% in children, due to strong osteoblastic and cementoblastic activities.<sup>5,9,11</sup> Active, aggressive cellular production of cement tissue rather than osteoid material results in clinical changes and affects the surgical outcome, often necessitating radical procedures.<sup>6,9,10</sup> We performed a left partial maxillectomy, and we have just reached the 1 year follow-up.

## CONCLUSION

JOF is a rare fibro-osseous bone tumour of the sinonasal tract. It has a unique histomorphological structure. It is invasive and can destroy surrounding anatomical structures. When a JOF is large and expanding into the surrounding tissues, it should be completely excised. For small lesions, more conservative procedures such as curettage and enucleation can be performed. Due to the high relapse rate, patients should be educated and followed for an extended period.

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## REFERENCES

1. Shaik M, Babu KT, PrabhuSekhar G, Murthy VRV. Juvenile ossifying fibroma of the maxilla: a case report. J Maxillofac Oral Surg. 2015;14(1):69-72.
2. Peterson BR, Nelson BL. Juvenile Active Ossifying Fibroma. Head Neck Pathol. 2015;9(3):384-6.
3. Mohsenifar Z, Nouhi S, Abbas FM, Farhadi S, Abedin B. Ossifying fibroma of the ethmoid sinus: Report of a rare case and review of literature. J Res Med Sci. 2011;16(6):841-7.
4. El-Mofty S. 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.
5. Ranganath K, Kamath SM, Munoyath SK, Nandini HV. Juvenile psammomatoid ossifying fibroma of maxillary sinus: case report with review of literature. J Maxillofac Oral Surg. 2014;13(2):109-14.
6. Pandit N, Saini N, Kler S, Jindal S. Juvenile trabecular ossifying fibroma. J Indian Soc Periodontol. 2014;18(2):232-5.
7. Johnson LC, Yousefi M, Vinh TN, Heffner DK, Hyams VJ, Hartman KS. Juvenile active ossifying fibroma. Its nature, dynamics and origin. Acta Otolaryngol Suppl. 1991;488:1-40.
8. Slootweg PJ. Juvenile trabecular ossifying fibroma: an update. Virchows Arch. 2012;461(6):699-703.
9. Goyal S, Sharma S, Arora V. Juvenile ossifying fibroma diagnosed on fine needle aspiration cytology: a diagnostic challenge. Diagn Cytopathol. 2015;43(1):75-9.
10. Yadav N, Gupta P, Naik SR, Aggarwal A. Juvenile psammomatoid ossifying fibroma: An unusual case report. Contemp Clin Dent. 2013;4(4):566-8.
11. Reddy AV, Reddy KR, Prakash AR, Rajinikanth, Vidhyadhari P. Juvenile ossifying fibroma with aneurysmal bone cyst: a case report. J Clin Diagn Res. 2014;8(10):ZD01-2.

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