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# Psammomatoid Juvenile Ossifying Fibroma of the Maxilla Misdiagnosed as Fibrous Dysplasia: A Clinicopathologic Case Report

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## Abstract

Juvenile ossifying fibroma (JOF) is a variant of the ossifying fibroma and includes two histopathological subtypes: trabecular and psammomatoid. Psammomatoid JOF (PJOF) in craniofacial structures should be distinguished from other fibro-osseous lesions, such as fibrous dysplasia (FD), considering the difference in the treatment protocols. Here, we present a rare case of PJOF that was initially misdiagnosed as a case of FD and emphasize the importance of considering JOF in the differential diagnosis of patients with craniofacial fibro-osseous lesions. A 4-year-old boy demonstrated progressive enlargement of the zygomaticomaxillary area on his left side for the last 6 months. The patient was diagnosed as a case of FD based on the clinical features and radiographic findings, and was operated considering the rapid progression. To achieve facial symmetry, contouring of the zygomatic bone and arch was performed. However, the patient demonstrated rapid enlargement at the 3-month postoperative follow-up. The decision was made to surgically remove the tumor due to visual field impairment. Intraoperatively, a rubbery mass, which was separated from the surrounding cortical bone, was identified and excised. The lesion was confirmed as PJOF by histopathological examination. The possibility of PJOF should not be ruled out in the differential diagnosis of patients with fibrous-osseous lesions. In the event of suspected PJOF, accurate diagnosis should be made through definitive biopsy.

**Keywords**

- psammomatoid juvenile ossifying fibroma
- fibrous dysplasia
- fibro-osseous lesions
- surgical treatment
- differential diagnosis

## Introduction

Juvenile ossifying fibroma (JOF) is a variant of ossifying fibroma and usually develops in the bones of the orbit and frontal and ethmoidal sinuses.[1] JOF includes two histopathological subtypes: trabecular and psammomatoid. Trabecular JOF is characterized by the presence of trabeculae of

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fibrillary osteoid, whereas psammomatoid JOF (PJOF) demonstrates small uniform spherical ossicles that resemble psammoma bodies.[2] Margo et al described PJOF as a characteristic fibro-osseous lesion in the orbit, demonstrating distinctive histologic features.[3] PJOF in craniofacial structures should be distinguished from other fibro-osseous lesions, such as fibrous dysplasia (FD), considering the difference in the treatment protocols.[4,5] We present a rare case of a 4-year-old boy who was initially misdiagnosed as a case of FD, who underwent a bone contouring procedure, and subsequently presented with rapid enlargement of the zygomatic bone. We report this case to emphasize the importance of considering JOF in the differential diagnosis of patients with craniofacial fibro-osseous lesions, to avoid unnecessary and harmful interventions.

## Case

A 4-year-old boy was referred to the Department of Plastic and Reconstructive Surgery at Seoul National University Hospital by an ophthalmologist at another hospital. The patient demonstrated progressive enlargement of the zygomaticomaxillary area on his left side for the last 6 months. Computed tomography (CT) images showed a bony expansile lesion with a predominantly homogeneous ground glass appearance and a focal radiolucent area in the left zygomatic bone. The patient was diagnosed as a case of FD based on the clinical features and radiographic findings, and was operated considering the rapid progression of the condition and for cosmetic reasons. Under general anesthesia, contouring of the zygomatic bone and arch was performed by bone shaving to achieve facial symmetry. There were no immediate postoperative complications. Histopathological diagnosis based on the specimen obtained from the shaving procedure was consistent with FD. (►**Fig. 1**) However, the patient demonstrated rapid enlargement of the zygomatic bone at the 3-month postoperative follow-up. CT scanning revealed rapid growth of the lesion, which was well-circumscribed and involved the left zygomaticomaxillary area (►**Fig. 2**). Due to compression of the eyeball and visual field impairment, the decision was made to surgically remove the tumor. The lesion was exposed through a subciliary incision. Intraoperatively, a rubbery mass, which was separated from the surrounding cortical bone, was identified and excised. (►**Figs. 3** and **4**) The excised specimen was sent for histopathological examination, and demonstrated a cellular stroma with small uniform spherical ossicles that resembled psammoma bodies (►**Fig. 5**). The lesion was confirmed as PJOF by definitive histopathological examination. The postoperative period was uneventful. Postoperative 6 months follow-up CT scan showed maxillary bony defect after tumor removal and no sign of recurrence (►**Fig. 6**). Skeletal reconstruction is planned after follow-up for recurrence.

## Discussion

Fibro-osseous conditions are characterized by the replacement of normal bone by a fibrous connective tissue matrix, of which ossifying fibroma and FD are the most common.[6] JOF usually affects children or adolescents younger than 15 years of age and has an aggressive growth pattern, which differentiates it from conventional ossifying fibroma and other fibro-osseous conditions.[1] JOF should be distinguished from other craniofacial fibro-osseous conditions such as FD considering the difference in treatment and progression.[4] The progression of FD is thought to stabilize after childhood in most cases, and surgical intervention is usually delayed as long as possible.[7–9] Although surgical treatment is required for cosmetic reasons, conservative procedures, such as bone shaving or contouring, are usually performed.[8,10]

PJOF is a variant of JOF, which is characterized by the presence of small psammoma-like bodies and has a high potential for recurrence.[11] The majority of PJOF lesions originate in the paranasal sinuses, particularly in the frontal

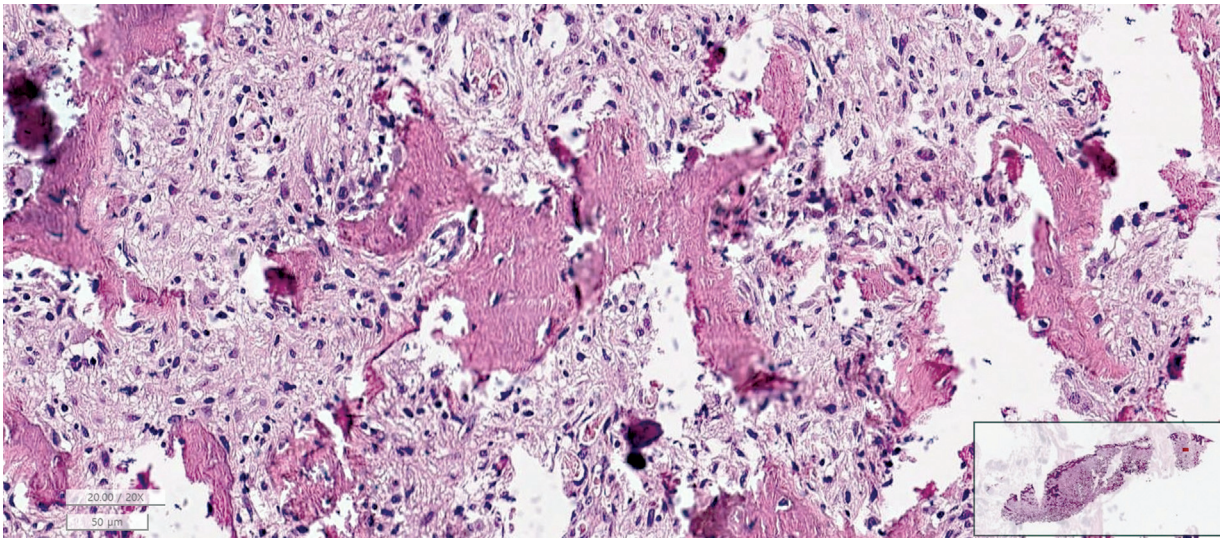


**Fig. 1** Histologic findings (hematoxylin and eosin staining, x20) of the specimen obtained from the shaving procedure show irregular trabeculae of woven bone.

 

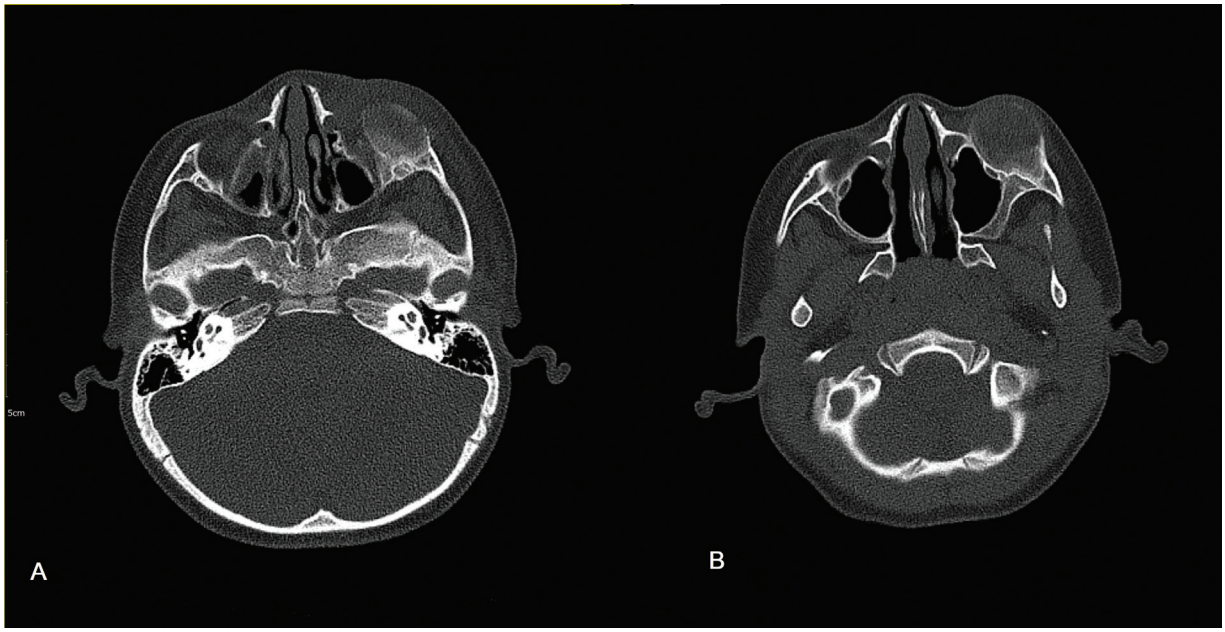


**Fig. 2** (A) Computed tomography (CT) scanning shows a 2.4 × 1.8 × 1.5cm-sized bony expansile lesion with predominantly homogeneous ground glass appearance and focal radiolucent area in the left zygoma. (B) CT scanning revealed rapid growth of the lesion, which was well-circumscribed and involved the left zygomaticomaxillary area.

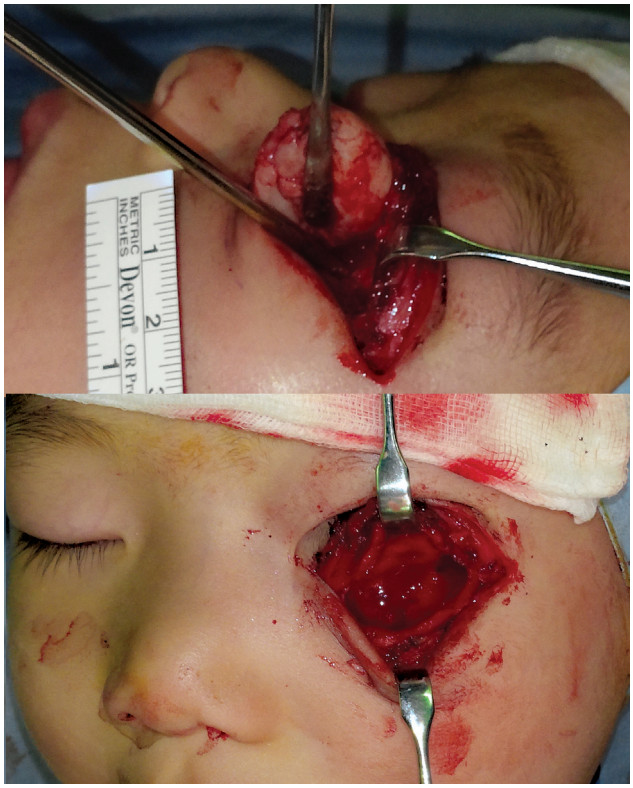

**Fig. 3** Intraoperative finding. A rubbery mass, which was separated from the surrounding cortical bone, was identified and excised.

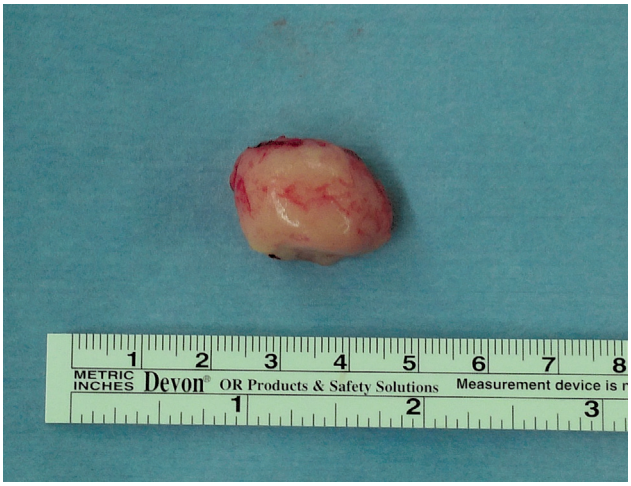


**Fig. 4** Macroscopic specimen.

or ethmoidal sinuses.[12,13] PJOF has the potential to invade surrounding tissues and such lesions usually require complete extirpation.[14] In addition, complete extirpation of PJOF lesions from the surrounding tissues is important considering the high risk of recurrence.[15] Despite the completely different treatment protocols for PJOF and FD, a diagnostic dilemma is often present due to the uncertainty of radiographic and histological features.[5] A typical radiographic finding of JOF is a round, well-defined lesion that occasionally demonstrates a corticated osteolytic lesion with a cystic appearance. However, some JOF lesions can demonstrate sclerotic changes showing a ground-glass pattern, thereby complicating the differential diagnosis with FD.[16] A key for the differential diagnosis is that JOF shows a circumscribed and well-defined radiopaque margin that is absent in FD.[17] However, some studies have reported that the differential diagnosis based on clinical manifestations and radiographic findings is controversial.[4,5] These differences are summarized in ►**Table 1**. There have been several reports that discussed on the differential diagnosis of FD and PJOF.[18,19] In our case, a typical radiopaque halo was not clearly observed due to diminished resolution of the CT images,

 

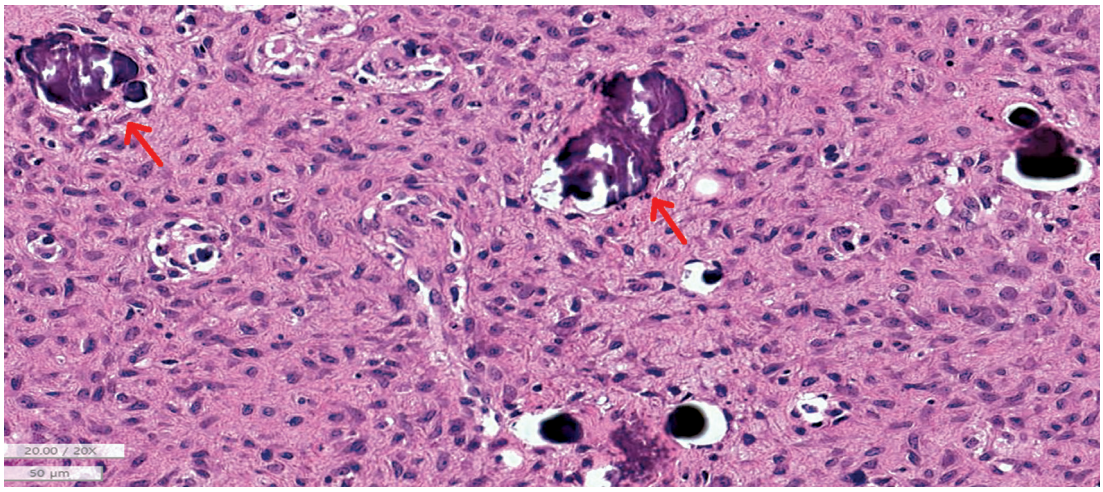


**Fig. 5** Histologic findings (hematoxylin and eosin staining, x40) show cellular stroma with small psammoma-like bodies, indicated by the red arrow.

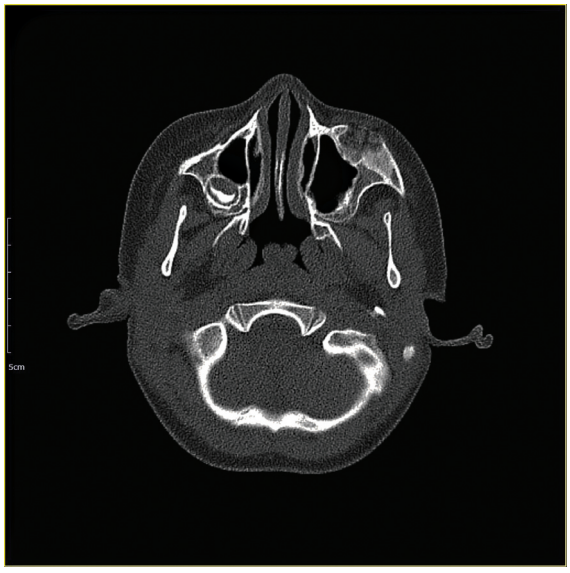


**Fig. 6** Postoperative follow-up computed tomography scan showed bony defect after removal of tumor and no sign of recurrence.

considering the low radiation dose recommended in children. Regarding the histopathologic findings, the specimen obtained from the shaving procedure was initially diagnosed as FD. It is difficult to accurately differentiate between these two entities based on histopathological examination of the shaved specimens. If an appropriate specimen is not obtained, there could be errors in the diagnosis considering their histologic similarity.[20] Therefore, the possibility of pathological misdiagnosis should be considered, and the findings must be correlated with the clinical features. FD is not clearly demarcated from normal bone, so its surgical removal is performed by radical resection or reductive surgery. On the other hand, in case of PJOF, "en bloc" surgical excision is often possible as in our case, which enables complete removal of the lesion with minimal morbidity. However, even in the case of radical resection, the recurrence rate of PJOF is high, so long-term follow-up will be required to confirm the safety and effectiveness of this method.

In conclusion, the possibility of PJOF should not be ruled out in the differential diagnosis of patients with fibrous-osseous lesions, because the treatment protocol for the

**Table 1** Comparison of craniofacial PJOF and FD

| | PJOF | FD |
|---|---|---|
| Age at presentation (y) | All ages can be affected, ranging from less than 1 to 72 years old[2] | Younger child to adolescent (< 15 y)[10] |
| Incidence | Rare | More common |
| Most common location | Paranasal sinuses (70%), maxilla (20%), and mandible (10%)[15] | Maxilla and its contiguous bone (sphenoid, zygomatic, frontonasal, and skull base)[14] |
| Radiological findings[15] | Relatively well-defined lesion with variable density Common cystic change (>50%) | Less-defined radiopaque lesion with gradual transition (may be radiolucent and well-defined lesion initially) |
| Histopathological findings | Spherical masses of osteoid dispersed in fibroblastic stroma with concentric calcification, producing a psammoma body-like appearance[1] | Irregular trabeculae of woven bone, blending into the surrounding normal bone[14] |
| Progression | More rapid, aggressive[11,15] | Slow growing, rarely grow after age 16 (may expand rapidly in growth period)[8,9] |
| Treatment | Earlier radical resection[11,15] | Conservative surgical approach (debulking, contouring, shaving) and total resection[8,9] |
| Prognosis | High recurrence rate (30–56%)[15] | Lower recurrence rate (15–20% in growth period)[9] |

Abbreviations: FD, fibrous dysplasia; PJOF, psammomatoid juvenile ossifying fibroma.

 

condition differs completely from that for other lesions. In the event of suspected PJOF, accurate diagnosis should be made through definitive biopsy and a correlation should be established based on clinical, radiographic, and histopathological findings for accurate diagnosis. In addition, if possible, "en bloc" excision can be considered as the treatment of PJOF for minimal morbidity.

### Authors' Contributions

JH Kim and BJ Kim conceptualized the study. JH Kim, J Kang, and S Kim were involved in data curation. JH Kim, J Kang, and S Kim did formal analysis. JH Kim was involved in methodology. BJ Kim did project administration. JH Kim and J Kang contributed to visualization. JH Kim was involved in writing—original draft. JH Kim, J Kang, and BJ Kim reviewed and edited the manuscript.

### Ethical Approval

Ethical approval for this study was obtained from the Institutional Review Board in Seoul National University Hospital (IRB no.: H-2206-047-1330)

### Patient Consent

Patients provided written informed consent for the publication and the use of their images.

### Conflict of Interest

None declared.

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