Psammomatoid Juvenile Ossifying Fibroma: A Case Report with Literature Review

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ABSTRACT
Psammomatoid Juvenile Ossifying Fibroma (PJOF) is an uncommon neoplasm that is distinguished from the adult variant of ossifying fibromas on the basis of age, site, clinical behavior and microscopic appearance. It is seen in children younger than 15 years of age, and the maxilla is more commonly involved than the mandible. PJOF may exhibit erosion and invasion of the surrounding bone accompanied by rapid enlargement. PJOF can be distinguished from other maxillofacial fibro-osseous lesions by its tendency to recur and its clinical mimicry of malignant bone tumors, but some clinical and histological features of PJOF overlap with the other fibro-osseous lesions as well. We report a case of a 15-year-old female patient with a painless apparently slow growing tumor in the left malar region, which occupied almost the whole of the left maxillary sinus, eroding the orbital border of the sinus.

Keywords: Psammomatoid, Juvenile ossifying fibroma, Psammoma bodies.

INTRODUCTION
Maxillofacial fibro-osseous lesions are a generic designation of a group of jaw disorders characterized by the replacement of bone by a connective tissue matrix, which displays varying degrees of mineralization in the form of woven bone or of cementum-like structures indistinguishable from “cementicles”.

Psammomatoid juvenile ossifying fibroma is an uncommon neoplasm characterized by the replacement of bone by a connective tissue matrix, which displays varying degrees of mineralization in the form of woven bone or of cementum-like structures indistinguishable from “cementicles”. The lesion is considered as a variant of juvenile ossifying fibroma. It is characterized by the presence of psammoma bodies, which are calcified, ossified or ossicle-like structures resembling cementicles. These lesions are of particular interest to the maxillofacial radiologist because they are frequently encountered in the diagnostic process and can be distinguished from other maxillofacial fibro-osseous lesions by their tendency to recur and their clinical mimicry of malignant bone tumors. Psammomatoid juvenile ossifying fibroma (PJOF) has its peak incidence in the first and second decades of life with a slight male predilection. The preferred sites are paranasal sinuses, maxilla, ethmoid and frontal bones.

CASE HISTORY
A 15-year-old female patient reported to the Department of Oral Medicine and Radiology with a painless swelling on left side of the face since one year. Patient noticed a small swelling of the face which gradually increased in size over a period of one year and the swelling was asymptomatic. On consulting the local dentist, she was advised antibiotics and analgesics. Patient did not report any reduction in the swelling. Later on, her left upper first molar was removed suspecting it to be a causative factor. Past medical and family history was noncontributory.

On general physical examination, patient was conscious, cooperative, healthy, moderately built and well nourished. All vital signs were in the normal range. She had attained menarche at the age of 12 years. Extraoral examination showed facial asymmetry due to swelling over the left malar area. The left eye appeared smaller in size. Local examination of the area revealed a single diffuse swelling present on left malar area measuring approximately 2 × 3 cm size. Anteriorly, the swelling extended 1 cm from the ala of the nose to posteriorly 2 cm in front of the tragus of left ear. Superioinferiorly, it extended from the left orbit to the midbrow.

The following radiographic findings were noted:

- **Cone beam CT**
  - The swelling was well demarcated from the surrounding bone.
  - There was no evidence of bone erosion or permeation.

- **Plain X-ray**
  - The swelling was well demarcated from the surrounding bone.
  - No evidence of bone erosion or permeation.

- **MR imaging**
  - The swelling was well demarcated from the surrounding bone.
  - No evidence of bone erosion or permeation.

The radiographic findings were consistent with a diagnosis of psammomatoid juvenile ossifying fibroma. The lesion was excised and the histological examination confirmed the diagnosis. The patient was advised to undergo follow-up to monitor for any recurrence.

**DISCUSSION**
Psammomatoid juvenile ossifying fibroma is a variant of ossifying fibroma that is characterized by the presence of psammoma bodies. The lesion is distinguished from the adult variant of ossifying fibromas by its tendency to recur and its clinical mimicry of malignant bone tumors. Psammomatoid juvenile ossifying fibroma has a peak incidence in the first and second decades of life with a slight male predilection. The preferred sites are paranasal sinuses, maxilla, ethmoid and frontal bones.

**REFERENCES**


**KEYWORDS**
Psammomatoid, Juvenile ossifying fibroma, Psammoma bodies.
to 3 cm short of inferior border of mandible. Skin over the swelling and the surrounding area appeared normal. On palpation temperature of the swelling was not raised, but swelling was tender and all inspectory findings were confirmed. Swelling was hard in consistency.

Intraoral examination revealed obliteration of the vestibule in relation to the maxillary left canine and premolars. On inspection a single diffuse swelling was present in the vestibule sulcus. Anteriorly, the swelling extended from the maxillary left canine to the second premolar posteriorly. Superioinferior borders of the swelling could not be evaluated (Fig. 1). On palpation swelling was tender and inspectory findings were confirmed. Swelling was hard in consistency and expansion of the buccal cortical plate was present. No expansion was noticed on the palatal aspect.

A provisional diagnosis of monostotic fibrous dysplasia involving the left maxilla was made. Differential diagnosis included ossifying fibroma and benign odontogenic tumor.

Radiographic investigations included, maxillary lateral topographic occlusal view, which showed expansion of the buccal cortical plate and diffuse radiopacity in the molar region. PNS view showed circumscribed radiopacity occupying almost the whole of the maxillary sinus with well-defined borders (Fig. 2). Serum alkaline phosphatase was found to be within normal limits. CT examination revealed sclerosis, bone expansion, softening and bone ossification noted extensively in the left maxilla and maxillary sinus, which measures 3.1 × 2.8 cm (Fig. 3). Nasal septum was mildly deviated to the right. Axial section CT showed the lesion destroying orbital margin of the sinus. CT findings were suggestive of either an ossifying fibroma or fibrous dysplasia involving the left maxilla. A more probable diagnosis of juvenile ossifying fibroma was considered because of the age of the patient.

An incisional biopsy was done from the left maxilla. Microscopic examination of specimen revealed highly fibrocellular mesenchymal tissue containing abundant spherical and globular calcified masses or ossicles. The mesenchymal cells are mostly plump and spindle shaped. Upon correlating with clinical and radiographic findings, a final diagnosis of psammomatoid juvenile ossifying fibroma was established (Fig. 4).
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The lesion was surgically excised and microscopic examination of the specimen revealed psammomatoid bodies with osteoid rims, embedded in a cellular stroma composed of uniform stellate and spindle shaped cells with little collagen. Immature osteoids with some showing irregular mineralization in the center were also seen. Excisional biopsy report confirmed the diagnosis of psammomatoid juvenile ossifying fibroma.

Postoperative healing was uneventful and patient was reviewed after one month and has been advised to report every six months for review, keeping in mind the high recurrence rates of this particular neoplasm.

REVIEW OF LITERATURE

Psammomatoid juvenile ossifying fibroma is a very uncommon lesion. In an extensive review by El Mofy in 2002, 230 cases were reported till date. The term psammomatoid ossifying fibroma was originally used by Gogl in 1949 to describe two cases, one affecting the frontal sinus in a 5-year-old boy and the other, the ethmoid sinus in a 9-year-old girl. Before this Benjamins in 1938 reported a lesion of the frontal sinus, which he termed “osteoid fibroma with atypical calcification” that was histologically similar to Gogl’s cases. He cited earlier references by Moser in 1899, Fetissof in 1929, and Ringertz in 1938, who had described similar to Gogl’s cases. Although, Benjamins identified these lesions as the same entity, his nomenclature is no longer applied. In the interim, however, other synonymous nomenclature had been used, including “least differentiated form of fibro-osteoma,” young ossifying fibroma, cementifying fibroma, psammo-osteoid-fibroma, and psammomatoid ossifying fibroma. Johnson et al in 1952 was the first to use the term juvenile active ossifying fibroma. Makeke in 1983 reviewed 86 cases and considered the lesion to be a variant of osteoblastoma and termed it psammous desmo-osteoblastoma and he called the trabecular variant as trabecular desmo-osteoblastoma. In a later review of 112 cases, Johnson et al in 1991 described PJOF as a cellular mass, which generates innumerable small uniform sized osteoid bodies.

It has been proposed that the mesenchymal cells of the periodontal membrane serve as multipotential precursor cells capable of differentiation into cementum, osteoid, or fibrous tissue and give rise to a spectrum of fibro-osseous lesions. According to some investigators, they perhaps originate from maldevelopment of the tooth. Johnson et al hypothesized that the lesion arose from the overproduction of myxofibrous cellular stroma normally involved in the development of septa in the paranasal sinuses as they enlarge and pneumatize. Recent studies also demonstrate the presence of nonrandom chromosome break points at Xq26 and 2q33 resulting in (X; 2) translocation.

In reviews published by Hamner et al and Slootweg et al the mean age of onset was 11.5 and 11.8 years old respectively. Bruce M Wenig et al in 1995 in a published report mentioned the youngest age in their study as five years, but also had patients in the fourth and sixth decade. The onset of psammomatoid ossifying fibroma is often in the mid-teenage years, and the average duration of symptoms before presentation is 4 to 5 years. Johnson et al suggested that these lesions may be present for extended periods of time, beginning in adolescence, but only manifest in adulthood once the lesion has gained an appreciable size, thus still qualifying as a juvenile lesion. In general, PJOF commonly occurs in patients between the ages of 5 and 15 years and is more often seen in males than females.

Johnson et al reviewed 3000 fibro-osseous lesions and found that 112 met the criteria for the juvenile active ossifying fibroma. Among the facial tumors, 90% arise from the paranasal sinuses and the remaining 10% involve the mandible. Cases involving the temporal bone have also been reported. Faizan et al reported that the maxillary tumors often fill and obliterate the maxillary sinus whereas mandibular tumors usually involve the ramus and angle. In 84% of the patients with PJO, the tumors are located in the facial bones, but calvaria (12%) and extracranial sites (4%) can be involved. Slootweg et al reviewed cases from the university hospitals at Groningen and Utrecht and noted that psammomatoid type, termed “juvenile ossifying fibroma—WHO [World Health Organization] type” in the study, had a predilection for the maxilla. Lehrer HZ in 1969 and Johnson LC et al in 1991 reported a predilection towards supraorbital locations (frontal and ethmoid bones), the ethmoid sinuses and the maxillary sinuses. Margo et al reviewed 21 cases involving the orbit and found 10 cases involving the orbital plate of the frontal bone (supraorbital) and nine cases centered in the ethmoid sinus. Lawton et al reports on ethmoid sinuses are the most commonly involved, followed by the frontal sinuses, maxillary sinuses, and sphenoid sinus.

Johnson et al in 1952 described PJOF as a cellular and aggressive form ossifying fibroma usually affecting children as the lesion proliferates, grows, invades and destroys tissue until the eyes are extruded and cerebrospinal fluid is reached. The first clinical manifestation is swelling of the maxilla. When the orbital bone and paranasal sinuses are involved, the patients may develop exophthalmos, bulbar displacement and nasal obstruction.
Clinically, PJOF presents with proptosis, visual disturbances, blindness, ptosis, disturbances in ocular mobility, papilledema, recurrent headaches, and nasal obstruction. Aggressive growth occurs in some but not all cases. Such behavior may be related to younger patient age and the concurrent development of aneurysmal bone cysts, which is seen more frequently in PJOF. Lawton et al report that tumor erosions bone partitions and encroaches on adjacent orbital, nasal, and cranial compartments, disturbing the face, displacing orbital contents, and blocking normal sinus drainage to form mucoceles. Visual disturbances or blindness can occur from compression of the optic nerves and orbit. Intracranial extension develops slowly with few neurological signs or symptoms, even though the lesions may be large enough to fill the anterior cranial fossa. The dura maintains an effective barrier to invasion of brain. Occasionally, patients will present with meningitis caused by communication between paranasal sinuses and subarachnoid space.

The radiographic appearance manifests as well-demarcated, unilocular or multilocular radiolucencies with a variable amount of radiopacity, usually manifesting as fine specks or as ground-glass opacification. It reveals an expansile well-circumscribed radiolucent or mixed radiolucent/radiopaque lesion surrounded by a thick bony wall. PJOFS are classically described as circumscribed unilocular lesions that are often surrounded by an eggshell-thin rim of bone. Aggressive forms tend to show more of an expansile growth pattern than the typical variety. However, they still tend to exhibit sharp demarcation from the normal adjacent bone and this occasionally reflected by an abrupt loss of the overlying cortex.

On gross inspection, PJOF appears as a yellow-white, homogeneous, and firm lobulated mass with gritty consistency. Cystic spaces, although rare, can be present. The most characteristic histologic feature of psammomatoid ossifying fibroma is the presence of numerous small, round ossicles or “psammomatoid” bodies that are embedded in a cellular fibrous stroma. The ossicles are mineralized collagenous foci that vary from small, smoothly contoured round-to-oval patterns to larger, irregularly shaped ossicle-like patterns with concentric layering similar to that of psammoma bodies. The ossicles are identified within the bony trabeculae as well as within the adjacent cellular stroma. A prominent marginal osteoid rim surrounds the ossicles. The number of ossicles varies from only a few to a dense population of innumerable spherical bodies.

**DISCUSSION**

Juvenile ossifying fibroma has been considered as a distinct disease entity from conventional ossifying fibroma and the other fibro-osseous lesions because of its tendency to occur at a young age and its locally aggressive behavior. Moreover, JOF may clinically manifest with rapid painless expansion of the affected bone as an aggressive lesion mimicking malignancy such as osteosarcoma. So, it is important to accurately recognize JOF for making the diagnosis and managing this disease.

The age and site of PJOF in our case are consistent with other reports, which state it as being common in the second decade of life and common in the paranasal sinuses. The literature reports a slight male predominance, ours being a female patient. The clinical features in our case were slightly different from the literature as the patient did not give a history of nasal obstruction or a feeling of heaviness in the sinus region. Radiographic and histological features are also similar to the reported cases.

PJOF needs to be distinguished from malignant bone tumors; in that there is a similarity of clinical manifestations, but it can be easily excluded from malignant bone tumors on histological examination. The differential diagnosis in conjunction with radiographic findings in this patient initially included aneurysmal bone cyst, central giant cell granuloma, osteogenic sarcoma, and progressive monostotic fibrous dysplasia and nonodontogenic primary tumors of bone, such as osteoblastoma. In addition, less likely consideration included locally aggressive odontogenic lesions, such as ameloblastic fibro-odontoma, as well as other entities consisting of calcifying odontogenic cyst (Gorlin cyst), adenomatoid odontogenic tumor, and primordial cysts (keratocyst). Vascular tumors can also have rapid growth and should be considered in the differential diagnosis. For example, central hemangioma grows rapidly and commonly presents as a radiolucent mandibular mass in children and young adults. Arteriovenous malformations also exhibit rapid growth, but usually display thrills or bruits on examination.

It is important to distinguish PJOF from central cementifying fibroma (ossifying fibroma, cementifying fibroma, cemento-ossifying fibroma), which is a benign jaw lesion of odontogenic origin arising in the molar and premolar regions of the maxilla and, most commonly, the mandible. Patients with central cementifying fibroma are in the third or fourth decade of life with a distinctly female predilection. The main presenting symptom is painless swelling of the involved bone. Radiographically, central cementifying fibroma is a well-marginated, unilocular, radiolucent or variably opacified lesion. Microscopically, central cementifying fibromas consist of fibrous stroma with dense cellularity and small, spherical calcifications—“cementicles” (osseous tissue related to dental cementum). Unlike PJOF, central cementifying fibromas rarely recur after removal.

Treatment of PJOF has been controversial. Complete removal by enucleation alone or with curettage as well as en block resections are common therapeutic options. Mass, as much as possible, must be removed while protecting adjacent vital structures to prevent recurrences. Our case required curettage of adjacent normal appearing surrounding tissue to ensure total excision of the mass. The reported recurrence rate ranges between 30 and 58%. Radiotherapy has been proven ineffective and contraindicated due to an increased incidence of malignant transformation ranging from 0.4 to 40%. Despite the aggressive behavior, no metastasis has been reported. Therefore, conservative treatment of the jaw lesions by enucleation and curettage has been reported to be successful.

**CONCLUSION**

PJOF is a rapidly growing benign but aggressive neoplasm that is encountered most commonly in children and young adults. Also, it poses diagnostic challenges since its clinical features overlap with many commonly occurring bone neoplasms. Histologic criteria for accurate classification remain complicated and are often controversial, secondary to confusing terminology as well as
overlapping morphologic features shared by several closely related entities. Consequently, accurate and reliable diagnosis of JPOF requires a multidisciplinary approach with close attention to pertinent clinical history, radiographic impression, and correlation with histomorphologic findings. The management of these lesions is directed at primarily surgical intervention and must consists of a complete resection, since partial removal is associated with the hazard of recurrence.

REFERENCES