CASE REPORT

A massive sinonasal psammomatoid variant of juvenile ossifying fibroma: Report of a rare entity

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ABSTRACT
Juvenile ossifying fibroma (JOF) is an uncommon, benign, bone-forming neoplasm with an aggressive local growth that is distinguished from other fibro-osseous lesions primarily by its age of onset, clinical presentation and aggressive behaviour. JOF is considered as a variant of the ossifying fibroma (OF) and the former includes psammomatoid JOF (PsJOF) and Trabecular JOF (TrJOF). Both variants involve the craniofacial bones with the trabecular variant being more common in the jaws and the psammomatoid variant being more common in the craniofacial skeleton. PsJOF is an unique variant of JOF that has a predilection for the sinonasal tract and the orbit particularly centered on the periorbital, frontal, and ethmoid bones. We report a rare case of massive PsJOF involving the maxillary sinus in a 20-year-old female.

Key words: Juvenile ossifying fibroma, maxillary sinus, psammomatoid, sinonasal tract

INTRODUCTION

Fibro-osseous lesions of the jaws represent a diverse group of entities which include developmental (hamartomatous) lesions, reactive or dysplastic processes and neoplasms.[1] Benign fibro-osseous lesions of the head and neck region are uncommon and represent a wide range of tumors sharing some histopathological features; which comprise fibrous dysplasia, ossifying fibroma (OF), and cement-osseous dysplasia. OF can be further divided into conventional and juvenile forms (juvenile ossifying fibroma (JOF)).[2]

JOF has to be distinguished from a larger group of OFs on the basis of the age of the patient, site of involvement and clinical behaviour.[3] According to the World Health Organization (WHO) classification of odontogenic tumors 2005, JOF is further subdivided into psammomatoid JOF (PsJOF) and trabecular JOF (TrJOF).[2]

Benjamins in 1938 first reported PsJOF as an "osteoid fibroma with atypical ossification of the frontal sinus",[4] Golgi in 1949 called it as "Psammomatoid Ossifying Fibroma",[2] and Johnson in 1952 coined the term "Juvenile Active Ossifying Fibroma".[2,4] According to WHO classification of odontogenic tumors 2005, it was named as "Juvenile Psammomatoid Ossifying Fibroma".[2] The term juvenile psammomatoid ossifying fibroma is now used to designate the neoplasm of the craniofacial skeleton of young age with well-defined clinicopathological features.[5]

The pathogenesis of these jaw lesions are related to the abnormal development of basal generative mechanism that is essential for root formation.[4] This paper highlights a rare entity of PsJOF involving the maxillary sinus and nasal cavity on the left side. Radiological and histological features along with the treatment have been discussed avidly.

Access this article online
Quick Response Code:
Website: www.jomfp.in
DOI: 10.4103/0973-029X.119751

CASE REPORT

A 20-year-old female patient reported with a painless progressive swelling in the left cheek region and difficulty in breathing since 1 year. Past medical history revealed that the patient underwent surgery in a private dental clinic 5 years back for the impacted tooth 23 which was associated with pathology. She had no noteworthy family history. Gross facial asymmetry was noted on left side of the face. The swelling was 6 × 4 cm in size approximately extending anteroposteriorly from ala of the nose to 5 cm from tragus of the ear on the left side. Nasal polyp was seen in the left nostril with obliteration of left ala of the nose [Figure 1].

Intraoral examination presented a swelling extending anteroposteriorly from the distal aspect of 21 to mesial aspect of 26; medially till the mid palatine raphe and laterally the buccal vestibule was obliterated and the swelling extended from 21 to 26. On palpation, the swelling was hard in consistency with no fluctuation.

Paranasal sinus (PNS) X-ray revealed haziness in the left maxillary sinus. Computed tomography (CT) scan confirmed well-defined mixed radiolucent and radiopaque areas with calcifications extending superoinferiorly from infraorbital rim to alveolus and anteroposteriorly from the nasal septum to post zygomatic buttress on left side [Figure 2].

Histopathological examination of incisional biopsy revealed connective tissue stroma with numerous spherical/irregular ossifications interspersed with cellular fibrous tissue [Figure 3]. The ossifications showed peripheral brush border surrounded by an eosinophilic rimming [Figure 4]. Haemorrhagic areas were also seen. The constellation of clinical, radiological, and histopathological features of this lesion supported an interpretation of psammomatoid variant of JOF.

Under general anesthesia, a Weber-Ferguson incision was given to expose the complete lesion. Subtotal maxillectomy was performed and the tumor mass along with nasal and ethmoidal polyps were removed with the help of a chisel and mallet. After complete removal of the mass, borders were carefully osteotomized to avoid the chances of recurrence [Figure 5]. The excised specimen was sent for histopathological examination [Figure 6] and the diagnosis of PsJOF was confirmed. Postoperative facial appearance and oral function were satisfactory. Follow-up examination of the patient showed no signs of recurrence.

DISCUSSION

JOF are benign, potentially aggressive fibro-osseous lesions of the craniofacial bones.[3] The word "Psammos" is descended from Greek word "psammos" which means sand.[2,3] PsJOF is a unique variant of JOF that has a predilection for orbit and PNSs accounting for about 72% followed by calvarium 11%,

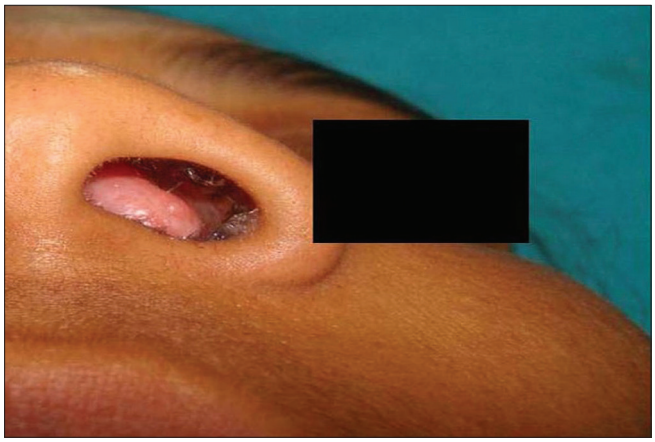

Figure 1: Nasal polyp seen in left nostril with obliteration of left ala of nose

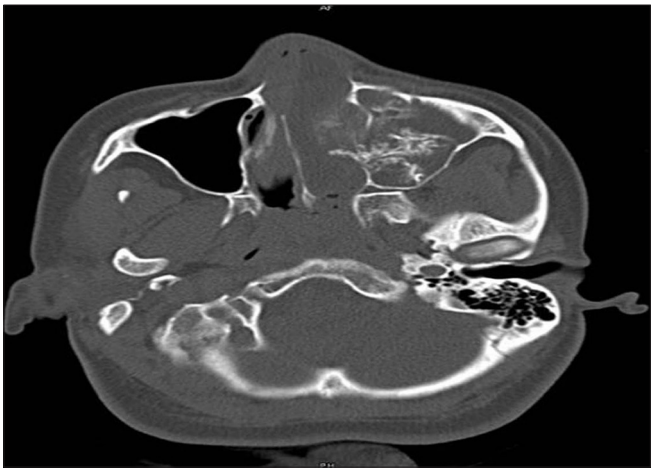

Figure 2: Axial computed tomography revealing well-defined radiolucent and radiopaque areas with calcifications

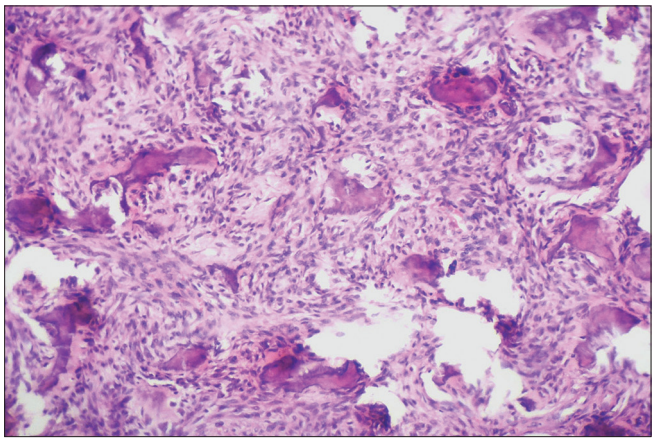

Figure 3: Photomicrograph of hematoxylin and eosin (H&E) stained section (×100) showing numerous spherical and irregular calcifications interspersed with fibrous tissue

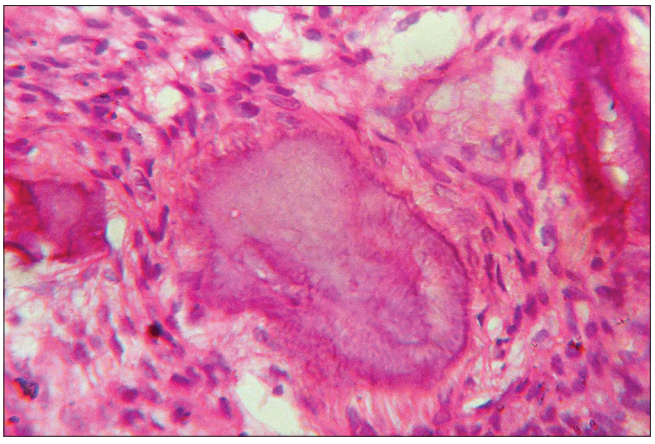

Figure 4: Photomicrograph of H&E stained section (×400) showing peripheral brush border surrounded by eosinophilic rimming

maxilla 10%, and mandible 7%.[6] The ethmoidal sinuses are most commonly involved, followed by the frontal sinuses, the maxillary sinuses, and the sphenoid sinus.[7] Both the variants of JOF show a predilection for males.[3] The swelling in this case is associated with maxilla involving maxillary sinus and nasal cavity in a 20-year-old female patient. Table 1 summarizes the clinical presentation of case series of PsJOF reviewed in the literature.

PsJOF initially manifests as an asymptomatic swelling. The patients may develop exopthalmoses, bulbar displacement, and nasal obstruction.[19] In the present case because of the extension of the tumor into the nasal cavity, nasal obstruction was present on the left side.

Radiographically, PsJOF can be radiolucent, radiopaque, or mixed depending upon cystic changes and the degree of calcification.[1,2] Sclerotic changes are evident in the lesion which may show a ground-glass appearance.[4] The present case revealed mixed radiolucent and radiopaque areas associated with the lesion.

Histologically, PsJOF shows highly cellular fibrous stroma often with whorled pattern containing closely packed spherical ossicles resembling psammoma bodies.[6] These ossicles are round to oval in shape which have a basophilic center and peripheral pink rim showing some radiating fibers[2] which corroborates with the microscopic features provided in the present case.

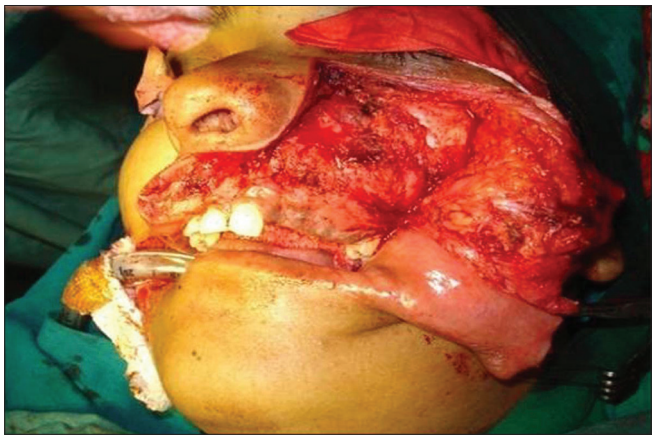

Figure 5: Weber-Ferguson incision was given to expose the lesion

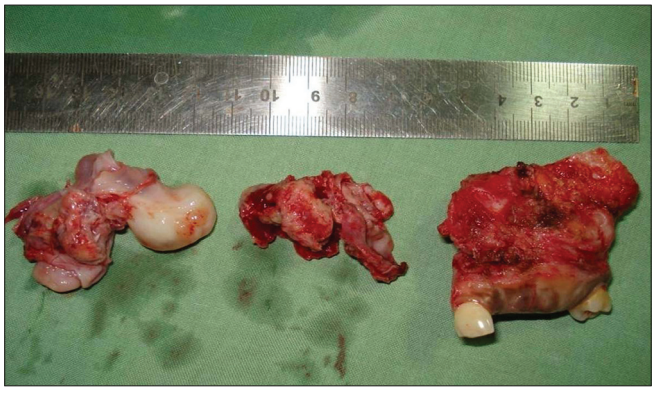

Figure 6: Excised specimen after subtotal maxillectomy

PsJOF should be differentiated from extracranial meningioma with psammoma bodies, which demonstrates epithelial membrane antigen (EMA) positivity and even the psammomatoid ossicles in PsJOF are clearly different from spherical true psammoma bodies.[3] Other differential diagnosis include fibrous dysplasia, osteoma, cementoblastoma, well-differentiated osteosarcoma, psammomatous extracranial meningioma [Table 2].

The clinical management of smaller lesions is simple excision with surrounding marginal bone, whereas larger lesions warrant more aggressive surgical management.[3] Prognosis is good with a recurrence rate of about 30-58%.[7] No malignant transformation has been documented.[3] Even though these lesions tend to locally invade, there were no cases of metastasis being reported. Occasionally meningitis, secondary invasion into the cranial cavity has been reported.[2]

CONCLUSION

PsJOF are unique rare entities occurring in the maxilla. They are unique because of their aggressive behaviour mimicking malignancy. Therefore, it is very important to correlate the clinical, radiographical, and histopathological findings for proper treatment. Early detection and complete surgical

Table 1: Review of previously reported PsJOF case series

Reference	No. of cases	Age (years)	Male: female	Sinonasal region	Maxilla
Dehner[8] (1973)	4	6-11	1:3	3	0
Makek[9] (1983)	86	3-49	1.2:1	53	17
Johnson *et al.*,[10] (1991)	112	3 months-72	1.1:1	71	20
Han *et al.*,[11] (1991)	5	12-56	1.5:1	5	0
Marvel *et al.*,[12] (1991)	3	7-32	2:1	2	0
Slootweg *et al.*,[13] (1993)	3	18-36	3:0	2	1
Wenig *et al.*,[14] (1995)	7	5-54	1.3:1	7	0
Lawton *et al.*,[15] (1997)	4	21-36	3:1	4	0
Hartstein *et al.*,[16] (1998)	3	10-24	2:1	3	0
El-Mofty[17] (2002)	3	15-53	2:1	1	0
Granados *et al.*,[5] (2006)	10	9-40	2:2	4	0
Gopinath *et al.*,[18] (2013)	8	9-20	3:5	0	1

Table 2: Differential diagnosis

Type	Clinical features	Radiological features	Histopathology
Fibrous dysplasia	1st-2nd decade Progressive and self-limited growth	Pagetoid or ground glass, sclerotic and cystic Homogenous dense sclerosis on CT	Bony trabeculae in fibrous background present in curvilinear forms Immature and woven type Rich in osteoid and lack osteoblastic rimming
Osteoma	2nd-4th decade Slow growth	Densely opaque, sharply defined mass that is usually broad based	Dense compact lamellar cortical bone with vascular canals Prominent cement lines with densely stained parallel accretion lines at peripheral margin
Cementoblastoma	Younger than 25 Mandibular 1st premolar region	Radiopaque mass fused to one or more tooth roots surrounded by a thin radiolucent rim	Presence of cementum droplets or cementicles which are characterized by the presence of numerous round to oval mineralized spherules
Well-differentiated osteosarcoma	Rapid growth with pain Male predominance	Calcifications, lytic and sclerotic changes on CT Symmetrical widening of periodontal ligament space with loss of lamina dura	Similar to fibrous dysplasia but exhibits nuclear pleomorphism
Psammomatous extracranial menigioma	Slow growth 6-7th decade Female prediliction	Hyperostosis and lysis, soft tissue tumor enhancement	Wide range psammoma bodies Psammoma bodies haphazardly distributed

excision of the lesion followed by long-term follow-up is necessary for proper clinical management.

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How to cite this article: Guttikonda V, Taneeru S, Gaddipati R, Madala J. A massive sinonasal psammomatoid variant of juvenile ossifying fibroma: Report of a rare entity. J Oral Maxillofac Pathol 2013;17:302-5.

Source of Support: Nil. **Conflict of Interest:** None declared.