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

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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%,
maxilla 10%, and mandible 7%. The ethmoidal sinuses are most commonly involved, followed by the frontal sinuses, the maxillary sinuses, and the sphenoid sinus. Both the variants of JOF show a predilection for males. 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 exophthalmoses, bulbar displacement, and nasal obstruction. 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. Sclerotic changes are evident in the lesion which may show a ground-glass appearance. 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. These ossicles are round to oval in shape which have a basophilic center and peripheral pink rim showing some radiating fibers which corroborates with the microscopic features provided in the present case.

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. 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. Prognosis is good with a recurrence rate of about 30-58%. No malignant transformation has been documented. 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.

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

| Reference | No. of cases | Age (years) | Male: Female | Sinonasal region | Maxilla |
|-----------|--------------|-------------|--------------|------------------|---------|
| Dehner [9] (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-Mofdy [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 |
excision of the lesion followed by long-term follow-up is necessary for proper clinical management.

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