Juvenile psammomatoid ossifying fibroma of the paranasal sinuses: A report of two cases

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Introduction
Juvenile psammomatoid ossifying fibroma (JPOF) is characterized by a fibroblastic stroma containing small ossicles resembling psammoma bodies. The stroma varies from being loose and fibroblastic to intensely cellular with minimal intervening collagen. These ossicles should not be confused with the cementum-like deposits that are present in conventional OF. We report two cases in female, first case of fronto-ethmoid sinus and second one of maxillary sinus.

Case Report
Case 1
A 15 year old female presented with 5.5 x 5 cm in sized, hard forehead swelling extended to bridge of nose. MRI of the paranasal sinuses revealed a well-defined, spherical expansile lesion in the frontal bone.

Case 2
A 38 year old female presented with right sided facial swelling with a mucosa covered mass was obstructing the right maxillary cavity. On examination, the swelling was 5.5x5 cm in size, hard in consistency and extended till the bridge of the nose. MRI PNS revealed a well-defined expansile radiolucent lesion in the frontal bone involving both the frontal sinuses with cortical thinning however no cortical breach was present. (Figure 1a) The mass was surgically resected by giving Lynch Howarth incision and we received multiple grey white soft tissue bits. Histopathological examination revealed stroma comprised of bland fibroblastic spindle cells along with spherical and curved ossicles having concentric pattern of lamination and focal osteoblastic rimming.

Conclusion
PJOF is a very aggressive lesion which mimics malignancy and due to its benign nature, early diagnosis can prevent over management and helpful in long term.

Abstract
Introduction
Juvenile psammomatoid ossifying fibroma (JPOF) is an uncommon benign bone-forming neoplasm in the craniofacial skeleton of juvenile patients and defined as a variant of ossifying fibroma (OF). It has been distinguished from a larger group of ossifying fibromas on the basis of age of the patients, site of involvement and clinical behaviour.¹,²

OF is highly cellular neoplasm and contains cementum-like deposits which have a smooth contour with a radiating fringe of collagen fibres.² On the basis of histomorphological features, juvenile ossifying fibromas are further categorized into psammomatoid (PJOF) and trabecular (TJOF) variants.²,³ Distinctive features of PJOF include predilection for the sinonasal complex and orbit in young people, an aggressive infiltrative growth pattern and propensity for recurrence.⁷ We report two cases of PJOF involving the paranasal sinuses.

Case report
Case 1
A 15 year old female presented with a complaint of forehead swelling for three years. On examination, the swelling was 5.5x5 cm in size, hard in consistency and extended till the bridge of the nose. MRI PNS revealed a well-defined expansile radiolucent lesion in the frontal bone involving both the frontal sinuses with cortical thinning however no cortical breach was present. (Figure 1a) The mass was surgically resected by giving Lynch Howarth incision and we received multiple grey white soft tissue bits. Histopathological examination revealed stroma comprised of bland fibroblastic spindle cells along with spherical and curved ossicles (Figure 2a) having concentric pattern of lamination (Figure 2b) and focal osteoblastic rimming. No necrosis or atypia were identified however rare mitotic figures were seen. The histopathological features along with clinical-radiological finding supported a diagnosis of Juvenile psammomatoid ossifying fibroma (JPOF).

Case 2
A 38 year female presented with right sided facial swelling for six years and nasal obstruction for six months. On local examination, a mucosa covered mass was seen obstructing

Figure 1: a) MRI PNS (Fronto- ethmoid sinus) shows hypointense central soft tissue component seen in frontoethmoid sinus causing expansion with thinning of the overlying skull tables. No intracranial extension was observed. b) CT Maxilla revealed a partially ossified mass deviating the nasal septum and lateralizing the medial wall of the right maxillary sinus and effacing the right maxillary cavity.
the right nasal cavity. CT of paranasal sinus revealed a well-defined ovoid mass measuring 7x6.8 cm in the right nasal cavity. The partially ossified mass caused deviation of the nasal septum, lateralizing the medial wall of the right maxillary sinus and causing effacement of the right maxillary cavity. (Figure 1b) The mass was surgically excised by lateral rhinotomy and sent for histopathological examination. We received multiple soft tissue bits along with bony bits. The histopathology section revealed irregular ossicles exhibiting varying degrees of calcification which were interspersed among fibroblastic stroma (Figure 2c and Figure 2d). No atypia, necrosis and mitotic figures were observed in the section examined. The final diagnosis of psammomatoid ossifying fibroma was supported by histomorphological features in correlation with radiological and clinical findings.

Discussion
The classification of odontogenic tumours in the second edition of the WHO, used the term juvenile (aggressive) ossifying fibroma. It included two distinct histological types of maxillofacial ossifying fibroma which were PJOF and TJOF. In 1938, Benjamins designated PJOF as osteoid fibroma with atypical ossification of the frontal sinus and in 1949 Golg termed it as psammomatoid ossifying fibroma of the nose and paranasal sinuses. Finally in the year 1985, Margo et al. introduced the term psammomatoid juvenile ossifying fibroma.

Reports vary on gender predilection. The average age of occurrence for PJOF is 16 to 33 years. Both of our cases were females and ages were beyond the average range as reported in literature, although in some reports patients age ranged from 3 months to 72 years. PJOF develops predominantly in the orbit and paranasal sinuses mainly in the frontal and ethmoid sinuses. The pathogenesis for these maxillofacial lesions is related to the maldevelopment of basal generative mechanism that is essential for root formation.

Diagnosis of PJOF is based on clinical-radiological and microscopic findings. Clinically PJOF manifests as an asymptomatic bony-hard swelling, the duration and extent of which may vary depending on the site and aggressiveness of the lesion. The symptoms such as pain, paresthesia, sinusitis and proptosis can also occur. Radiologically PJOF appears as radiolucent, mixed or radiopaque unilocular, oval or spherical lesion, depending on the degree of calcification. A ‘ground-glass’ appearance on radiographs has also been reported in the literature.

Microscopically the pathognomonic feature is small spherules resembling psammoma bodies, which are referred to as psammoma-like bodies or psammomatoid ossicles. The intervening stroma is highly cellular consisting of spindle shaped fibroblasts with minimum intervening collagen. The other features that can be seen are pseudocystic stromal degeneration and haemorrhages with aneurysmal bone cyst like spaces.

Figure 2: a) Cellular fibroblastic stroma admixed with spherical and curved ossicles. (H&E 400x). b) Acellular psammoma body-like ossicles with concentric pattern of lamination (H&E 400x). c) Irregular ossicles exhibiting varying degrees of calcification which were interspersed among fibroblastic stroma (H&E 400x). d) Stratified squamous epithelium, subepithelium showed spindle shaped fibroblastic cells and irregular ossicles (H&E 100x).

Prognostically PJOF has an aggressive course with tendency to recur. The aggressive nature of this entity and its high recurrence rate, surgical resection is the preferred line of treatment.

Conclusion
PJOF is very aggressive lesion which mimics malignancy and common in childhood, it rarely occurs in maxilla. Diagnosis can be aided by awareness of the entity, characteristic histologic features and clinic-radiological finding. Due to its benign nature, early diagnosis can prevent over management and helpful in long term management.

References


Case report

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