Case Report:

Juvenile psammomatoid ossifying fibroma of the ethmoid sinus: A rare case report
Wan Draman WN1,2, Ramli R2, Mohd Noor R3, Baharudin Abdullah B4, Abdul Aziz RC5

Abstract
Juvenile psammomatoid ossifying fibroma (JPOF) is a rare, aggressive benign bony tumour that has been distinguished from a larger group of ossifying fibromas (OF) based on age of occurrence, most common site of involvement, and its clinical behaviour. We reported a case of asymptomatic 14 year old boy who had incidental finding of left ethmoidal bony mass on his CT brain imaging done for post traumatic cerebral concussion. The diagnosis of juvenile psammomatoid ossifying fibroma (JPOF) was made based on radiological and histological findings.

Keywords: Psammomatoid Ossifying Fibroma, Juvenile, Ethmoid sinus

Introduction
Ossifying fibroma (OF) is a rare, benign and aggressive fibro-osseous lesion which found mostly in the mandible, accounting for more than 70% of all cases, followed by the maxilla and rarely found in the orbit and paranasal sinuses, with only 55 reported cases in the literature from 1971 until February 20131-4,5. OF is highly cellular neoplasm and contains cementum-like deposits which have a smooth contour with a radiating fringe of collagen fibres.6

On the basis of histomorphological features, juvenile ossifying fibromas are further categorized into psammomatoid (JPOF) and trabecular (JTOF) variants.6,7 Psammomatoid ossifying fibroma (POF) characterized by numerous calcified "psammomatoid" ossicles that histologically resemble psammoma bodies. Distinctive features of JPOF include predilection for the sinonasal complex and orbit in young people, an aggressive infiltrative growth pattern and propensity for recurrence.8

Case Report
A 14 year old boy was referred to the our clinic for left ethmoidal mass. This was an incidental finding noted on follow up CT scan which was done as he sustained cerebral concussion due to motor vehicles accident. The patient was otherwise asymptomatic. He did not suffer any nasal or ocular symptoms such as hyposmia, epistaxis, nasal obstruction, any visual disturbances, epiphora and headache. Naso-endoscopic examination revealed no abnormal findings. Other head and neck examination was unremarkable. Laboratory results were all within the normal range.

Computed Tomography (CT) scan of the paranasal sinus revealed a mass in the left ethmoid sinus with extension into the left orbit. The mass was well-defined with a heterogeneous appearance on T1 and T2 weighted images. There was no evidence of bone destruction or invasion of adjacent structures.

Histopathologically, the lesion was composed of typical ossifying fibroma with areas of calcification. The trabecular pattern was also observed.

Conclusion
Juvenile psammomatoid ossifying fibroma is a rare entity which should be included in the differential diagnosis of lesion in the sinonasal region. Early diagnosis and appropriate treatment are important to prevent recurrence and complications.

1. Wan Nur Anis Wan Draman, Department of Otorhinolaryngology - Head and Neck Surgery, Hospital Raja Perempuan Zainab II, 15586 Kota Bharu, Kelantan, Malaysia, Department of Otorhinolaryngology - Head and Neck Surgery, School of Medical Sciences, Universiti Sains Malaysia Health Campus,16150, Kubang Kerian, Kelantan, Malaysia
2. Rosdi Ramli, Department of Otorhinolaryngology - Head and Neck Surgery, Hospital Raja Perempuan Zainab II, 15586 Kota Bharu, Kelantan, Malaysia
3. Rosli Mohd Noor, Department of Otorhinolaryngology - Head and Neck Surgery, Hospital Raja Perempuan Zainab II, 15586 Kota Bharu, Kelantan, Malaysia
4. Baharudin Abdullah, Department of Otorhinolaryngology - Head and Neck Surgery, School of Medical Sciences, Universiti Sains Malaysia Health Campus,16150, Kubang Kerian, Kelantan, Malaysia
5. Roziasyazni Che Abdul Aziz, Department of Pathology, Hospital Raja Perempuan Zainab II, Kota Bharu, Kelantan, Malaysia

Correspondence to: Wan Nur Anis Wan Draman (MD), Department of Otorhinolaryngology - Head and Neck Surgery, School of Medical Sciences, Universiti Sains Malaysia Health Campus,16150, Kubang Kerian, Kelantan, Malaysia, E-mail: nuranisarman@gmail.com
sinuses showed a hyperdense mass associated with calcification and thinning of lateral wall and roof of left posterior ethmoid sinus with possible areas of bony erosion (Figure 1a and Figure 1b). Functional endoscopic sinus surgery was performed and a near total resection of tumour was done. We received multiple grey white soft tissue bits along with bony bits (Figure 2). The operation was uneventful with no intra operation and post operation complications.

Histopathological examination revealed tumour is composed of benign fibro-osseous neoplasm with adjacent areas showing multiple small uniform psammomatoid bodies embedded in cellular stroma that contain spindle to oval cells (Figure 3). No nuclear atypia, necrosis and mitotic figures seen. This result in correlation with radiological findings supported a final diagnosis of Juvenile psammomatoid ossifying fibroma (JPOF). Endoscopic evaluation in subsequent follow up showed complete wound healing and the patient was clinically well. Patient has been scheduled for 6 months clinical and imaging follow up to prevent further recurrence of the tumour.

Discussion

Ossifying fibroma (OF) is a benign fibro-osseous lesion mostly found in craniofacial bones. It is a rare, locally aggressive and slow growing tumor. The mandible is considered as the most common location of this neoplasm accounting for more than 70%. However it could also be seen in the maxilla and paranasal sinuses in some occasions. The first case of ossifying fibroma in literature was described in 1872 by Menzel. It was later coined with this terminology by Montgomery in 1927. The ossifying fibromas (OF) are subdivided into conventional and juvenile clinicopathologic subtypes. On the basis of morphologic features, juvenile ossifying fibromas are further separated into trabecular (JTOF) and psammomatoid (JPOF) variants. The juvenile variants are characterized by distinctive trabecular or psammomatoid matrix production, occurrence in younger patients relative to conventional OF and a predilection for the bones of the paranasal sinuses, the periorbital region and the maxilla.

Juvenile ossifying fibroma is usually seen in the first and second decade of life but it does occur in adult. The average age of occurrence for JPOF is usually found in craniofacial bones.
is 16 to 33 years \(^6\). As a term “juvenile” underlines, the tumour largely develops in children, 79% of whom under age of 15 years old \(^{12}\). This was observed in our case. The mean age of onset was 11.5 and 11.8 years, respectively \(^{12,14}\). Reports vary on gender predilection \(^6,15\). Manes et al. reviewed 55 cases of ossifying fibroma in the paranasal sinuses \(^{15}\) and revealed that the male to female ratio with this type of tumor tends to be 1:1.04 \(^3\). JPOF develops predominantly in the orbit and paranasal sinuses mainly in the frontal and ethmoid sinuses. \(^6\) Manes et al. reviewed 15 psammomatoid ossifying fibroma cases with this type of tumor tends to be 1:1.04 \(^3\).

JPOF develops predominantly in the orbit and paranasal sinuses mainly in the frontal and ethmoid sinuses. \(^15\) No specific etiology has been attributed to the development of ossifying fibroma. However, it is suggested that odontogenic, developmental abnormality and traumatic origins could be some underlying causes \(^{1,5}\). There is no evidence of hereditary predominance \(^{16}\). Presenting symptoms of this tumour is variable, depending on the location and rate of growth \(^6\). Patients with ossifying fibroma tumors are often symptomless. They are mostly discovered incidentally on imaging taken for another reason as in our case. When they present with mass effect symptoms, facial swelling tends to be the most common sign followed by nasal obstruction symptoms, anosmia, epistaxis and headache. Ocular symptoms include visual loss, diplopia, proptosis and epiphora \(^{1,3}\). Larger tumours may also lead to a painless swelling of the involved bone \(^1\). Meningitis and pneumocephalus are two rare intracranial complications of ossifying fibroma. \(^6\) Our patient was fortunate that even though asymptomatic, he had early diagnosis of JPOF detected by the CT imaging following his post traumatic cerebral concussion.

Diagnosis is mostly done by radiographic images showing a well-circumscribed demarcated lesion. Initially, the lesion is radiolucent but progresses to radiopaque and gets involved by a radiolucent or sclerotic periphery. Paranasal CT scan presents the central area consist of a non-homogeneous matrix with a “ground-glass” opacification representing diffuse calcifications and low attenuation areas containing fibrous tissue or retained mucus with possible contrast enhancement. \(^1\) The walls of the involved sinuses may undergo further remodelling and thickening, sometimes along with erosions. \(^{5,10}\) Other diagnostic tools such as histopathological examination and MRI might be needed to confirm the diagnosis or to rule out intraorbital or intracranial extension \(^{1,5}\).

Microscopically the pathognomonic feature is small spherules resembling psammoma bodies, which are referred to as psammomma-like bodies or psammomatoid ossicles \(^{17}\). 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. \(^8\) The mainstay treatment of a paranasal sinus ossifying fibroma is total excision of lesion by different modalities. Endonasal endoscopic resection of the tumour is the recommended therapeutic approach. It holds high advantages including direct visualization, enhanced visibility and magnification of the lesion resulting in decreased intraoperative trauma and postoperative morbidity. The advent 3D navigation devices further enhances this advantages. Aesthetic outcome is excellent in the absence of facial scars. Total resection must be performed to avoid recurrence. \(^{10}\) With a mean follow up time of 26 months, the recurrence rate of paranasal sinus ossifying fibroma was 7% with total resection compared to 25% with sub-total resection \(^3\). Despite the aggressive nature of the lesion and high rate of recurrence, malignant transformation of this benign tumour to sarcoma has not been reported \(^{18,19}\).

Although radiation therapy has been advocated in the treatment of bony tumour, it has not been advocated in the treatment course of ossifying fibroma \(^3\). Radiotherapy is contraindicated for OF because it may increase the possibility of malignant transformation. Reported rates of malignant transformation range from 4% to 40% \(^{10,20}\). Long term follow-up with repeated nasal endoscopy and serial paranasal sinus CT is important to detect any tumour recurrence and prevents any possible complications and morbidity. \(^1,3\)

A 6 months follow up of our patient revealed no signs or symptoms of recurrence by physical examination and nasal endoscopy. A paranasal CT scan is planned in one year from the time of surgery.

**Conclusions**

Juvenile psammomatoid ossifying fibroma is a rare, locally aggressive lesion of cranio facial bone of juvenile patients which mimics malignancy. It is unusual to be found in the paranasal sinuses. Crucial attention to the clinical, radiographical and histopathological examination should be taken for more accurate diagnosis and thus early and appropriate management can be commenced.

**Funding:** No funding.

**Conflict of interest:** The authors declare that they have no conflict of interest.


