Case Report: Juvenile psammomatoid ossifying fibroma of the ethmoid sinus: A rare case report

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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 2013. OF is highly cellular neoplasm and contains cementum-like deposits which have a smooth contour with a radiating fringe of collagen fibres.

On the basis of histomorphological features, juvenile ossifying fibromas are further categorized into psammomatoid (JPOF) and trabecular (JTOF) variants. 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.

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 homogeneous soft tissue dense mass in the left ethmoid sinuses.

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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 16 to 33 years. As a term “juvenile” underlines, the tumour largely develops in children, 79% of whom under age of 15 years old. This was observed in our case. The mean age of onset was 11.5 and 11.8 years, respectively. Reports vary on gender predilection. Manes et al. reviewed 55 cases of ossifying fibroma in the paranasal sinuses and revealed that the male to female ratio with this type of tumor tends to be 1:1.04. JPOF develops predominantly in the orbit and paranasal sinuses mainly in the frontal and ethmoid sinuses. It is 16 to 33 years. As a term “juvenile” underlines, the tumour largely develops in children, 79% of whom under age of 15 years old. This was observed in our case. The mean age of onset was 11.5 and 11.8 years, respectively. Reports vary on gender predilection. Manes et al. reviewed 55 cases of ossifying fibroma in the paranasal sinuses and revealed that the male to female ratio with this type of tumor tends to be 1:1.04.

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