Case Report

Psammomatoid Ossifying Fibroma- A case report with review of literature highlighting the change in classification

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ABSTRACT:

Psammomatoid ossifying fibroma is a rare entity that affects all age groups and is grouped as a variant of ossifying fibroma. PsOF is grouped under benign fibro-osseous lesion as a separate entity. The lesion is seen in all age groups and is not restricted to younger age only. It is highly aggressive and needs to be differentiated from other benign fibro-osseous lesions that are not aggressive. The lesion typically affects paranasal sinuses commonly and shows presence of ossicles known as a psammoma bodies. The classification of this lesion has seen a dramatic change over a period of 2 decades. We hereby report a case of Psammomatoid ossifying fibroma of mandible with review of literature.

KEYWORDS: Psammomatoid; ossifying fibroma; psammoma bodies; fibro-osseous lesions

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Submitted: 01.10.2023, **Accepted:** 04.12.2023, **Published:** 26.12.2023

INTRODUCTION:

Psammomatoid ossifying fibroma (PsOF) is a rare entity that affects all age groups and is grouped as a variant of ossifying fibroma (OF). It is seen to affect paranasal sinuses most commonly. This is followed by maxillary and mandibular bone involvement[1]. OF show 2 variations viz; juvenile trabecular and psammomatoid types. The psammomatoid variant has been reported in various age groups ranging from 3 months to 72 years. PsOF's radiographically depicts ground glass radiopaque appearance of the lesion as an outer thick mantle with central radiolucent areaor as a solid radiopacity[2]. PsOFs are grouped under locally aggressive lesions affecting orofacial regions that histopathologically are characterized by dense cellular fibrous stroma with spheroidal calcifications called psammoma bodies[3]. The typical diagnosis of this entity is done by careful clinical features along with radiological and histopathological examination. The commonly advocated treatment is complete surgical excision. We report a case of PsOF in the mandible with review of literature.

CASE REPORT:

A 17-year-old female patient reported to our institute complaining of swelling in lower jaw since 1 year. The swelling was small initially and increased in size over a period of time. The medical & family history were non-contributory. The swelling measured 4 cms X 5 cms and extended from left commissure of the lip to lower border of mandible. No discharge was evident. Radiographic examination revealed well

Access this article online

Quick Response Code:

Website: www.pjsr.org

DOI: doi.org/10.5281/zenodo.10432078

How to cite this article: Kavle P, Nagendra BS, Buva K, Deshpande SM. Psammomatoid Ossifying Fibroma- A case report with review of literature highlighting the change in classification. PJSR. 2023;16(2):31-33.

defined radiolucency in relation to mandibular incisor extending to mandibular second molar. There were radiopaque masses seen within the radiolucency. Incisional biopsy was carried out. A provisional diagnosis of fibroosseous lesion was made. The patient underwent complete surgical excision and the specimen was submitted for histopathology. The histopathology examination revealed dense cellular proliferation of spindle cells resembling fibroblasts. There were numerous ossicles resembling psammoma bodies. These bodies were acellular showing concentric pattern of lamination and were evenly distributed within the stroma [Figure 1& Figure 2]. There was no evidence of necrosis and cytological atypia. A final diagnosis of PsOF was made based on the above findings. A regular follow-up for 2 years revealed no recurrence.

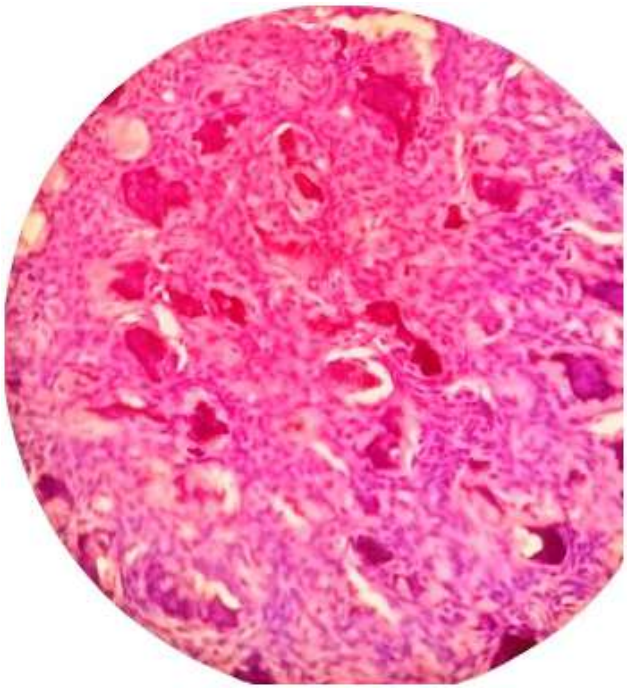

Figure 1: H&E stain shows psammoma bodies in a hypercellular area.

DISCUSSION:

PsOF is a rare aggressive lesion that was first classified as Juvenile form of OF and was known as Juvenile ossifying fibroma in the year 2005 WHO classification. Since earlier reported cases occurred mostly in 1st or 2nd decade of life it was grouped under juvenile variant of OF [4]. The pathogenesis is believed to be associatedwith *MDM2* and *RASAL1* gene amplifications [5]. The mineralized material consists of spherical or curved ossicles that are acellular or show sparsely distributed cells. These ossicles are characterized by thick irregular collagenous rim and sometimes they are deeply basophilic concentrically

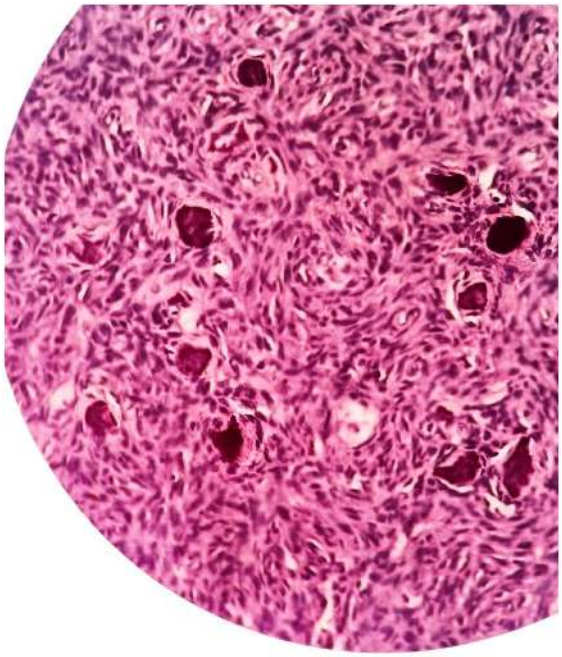

Figure 2: H &E stained section shows plump spindle shaped fibroblasts with psammoma bodies.

lamellated particles[4]. 2017 WHO classification stated that there are 3 variants of ossifying fibromas: cemento-ossifying fibroma, juvenile trabecular ossifying fibroma, and juvenile psammomatoid ossifying fibroma. PsOF was initially considered as a tumor of odontogenic origin (2005 WHO classification). In 2017 WHO classification, PsOF was considered as a distinctive type of ossifying fibroma, under benign fibro- and chondro-osseous lesions[6]. The psammoma bodies (ossicles) may fuse to form trabeculae, or may be grouped by fusion of their thick irregular collagenous rim. PsOFs show positivity for vimentin, SMA and CD10[7]. According to the new classification of WHO 2022, the word "***juvenile***" is removed from the psammomatoid variant and is retained only for the trabecular variant. Moreover, juvenile trabecular ossifying fibroma and PsOF are separated from the odontogenbic cemento-ossifying fibroma and are included as separate benign fibro-osseous lesions[8]. PsOFs share several common features with other fibrous neoplasms. Hence it becomes imperative to rule out other neoplasms as PsOFs are aggressive. The diagnosis is classically based on a triad including clinical symptoms, radiological features and histopathology[1].

CONCLUSION:

PsOF is an aggressive variant that may mimic other fibro-osseous lesions and demands attention. This lesion is no longer termed as juvenile as was seen in

early classifications. The treatment is essentially complete surgical excision and recurrence is unusual.

Financial Support and sponsorship
Nil.

Conflicts of interest
There are no conflicts of interest.

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