Case Report

Psammomatoid Type Juvenile Ossifying Fibroma of Mandible: A Rare Entity

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Juvenile ossifying fibroma (JOF) is a rare fibro-osseous neoplasm that arises within the craniofacial bones. Two distinct histopathological variants have been described; trabecular JOF and psammomatoid JOF (PsJOF). The later occurs predominantly in craniofacial skeleton and rarely in jaws and more so in mandible (50% of the times than that in maxilla). Here we report a rare case of PsJOF in mandible body ramus region with its unique radiographic characteristics to help the diagnostician approach the diagnosis very closely for this rare entity. The tumor has been present since 1 year with unilocular radiographic appearance; having homogeneous ground-glass appearance with higher imaging, without any focal radio-opacity and inferior alveolar canal was displaced but still intact. Thus these lesions can expand to large sizes even without maturing to its radiopaque stages and without destruction of adjacent vital structures although displacing them.

Keywords: Fibro-osseous lesions, Mandible, Psammomatoid type

INTRODUCTION

Juvenile ossifying fibroma (JOF) is a fibro-osseous lesion that occurs in the facial bones.[1] It is also called aggressive ossifying fibroma due to its aggressiveness and the high tendency to recur, unlike other fibro-osseous lesions, such as cemento-ossifying fibroma, which it may resemble radiographically. Due to its distinct histological features, it has been recognized as a separate histopathological entity among the fibro-osseous group of lesions.[1,2] El-Mofty (2002) identified two histopathological variants of JOF as trabecular JOF (TrJOF) and psammomatoid JOF (PsJOF).[3] JOF appears at an early age.[4] The age predilection for psJOF is less than 21.8 years.[5] JOF constitutes 2% of all oral tumors in children. It has an equal predilection for males and females. JOF commonly occurs in the facial bones (85%), calvarium (12%) and mandibular region (10%). Very rarely it has been reported extracranially (3%).[6,7]

PsJOF and TrJOF are considered as the variants of aggressive subclass of ossifying fibroma,[8-10] and share many similarities including their frequency of occurrence.[3] Radiographically both exhibit a mixture of radiolucent and radiodense areas with thin sclerotic rims that may be incomplete.[3] Both lesions typically lack fibrous capsules and tend to infiltrate adjacent bone. Thus tissue specimens are usually submitted fragmentedly.[3,9,11] In terms of location, TrJOF occurs more in the jaws, particularly the maxilla, while PsJOF is more commonly seen within the sino-naso-orbital regions.[3] PsJOF and TrJOF can both present with symptoms such as nasal obstruction; rhinorrhea and orbital displacement.[3,12] Most patients with PsJOF are older than those with TrJOF by about a decade, but younger than COF patients by a similar margin.[8] Here we report an additional case of PsJOF of mandible, an unusual site.

CASE REPORT

A 15-year-old girl visited the clinic with the swelling of left mandible body-angle-ramus region; present since last 1 year. A year back patient experienced pain in carious #36 and #37 which lasted for 7-8 days and noticed a swelling in left body-angle-ramus region which gradually increased in size and was asymptomatic. Patient reported to us with the swelling and sutures placed at the site of #36 and #37 being extracted along with surgical exploration of the swelling 7 days back with previous records mentioning profuse bleeding through the sockets during extraction.

Extraoral examination (Figure 1a) of the involved area revealed a 3 cm × 2.5 cm swelling in mandibular left body-angle region, ovoid in shape, bony hard and febrile on palpation. Intraorally (Figure 1b), buccal expansion and obliterated buccal vestibule seen. Anterioposteriorly, the swelling was extending from first premolar to retromolar area, involving part of the ramus as well.

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Conventional antero-posterior and lateral skull radiographic views and orthopantomogram (Figure 2a and b) showed large unilocular radiolucency involving the body angle ramus region and well-defined sclerotic border with an inferior border intact. Computed tomography (Figure 3a and b) revealed a 2.2 cm × 2.7 cm well-defined expansile lesion with homogeneous ground glass attenuation involving the left body-angle-ramus region. Expansion and thinning of adjoining cortical boundaries without any disruption of cortex noted.

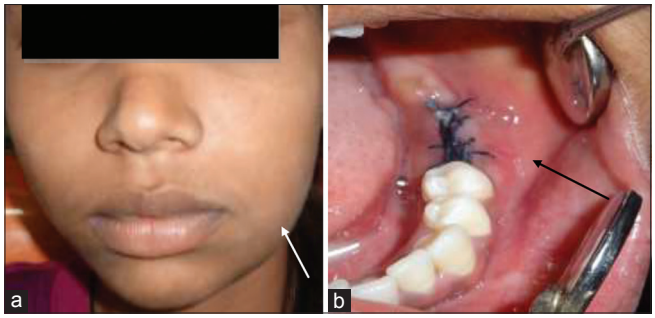

Figure 1: Clinical photographs revealing swelling of left mandibular body angle ramus region (a) swelling (arrow) causing facial asymmetry (b) obliterated vestibule (arrow) and missing first and second molar

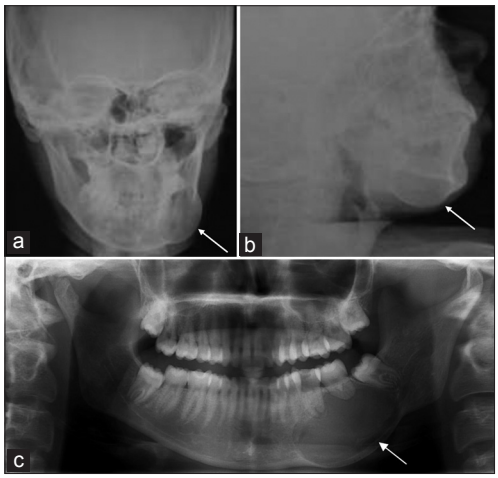

Figure 2: Findings on conventional imaging revealing unilocular osteolytic lesion with cortical expansion and thinning (arrow) in left mandibular body-angle-ramus region. (a) PA view skull, (b) Lateral skull view, (c) orthopantomogram

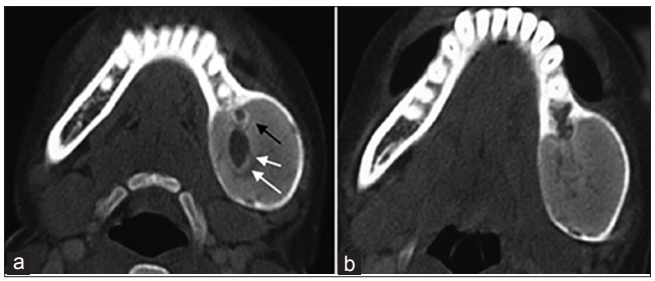

Figure 3: Computed tomography axial views show cortical plate expansion of left body-angle region, (a) hyperdense ground glass internal appearance and focal hypodense oval area (white arrow) within due to previous surgical exploration and anteromedially displaced inferior alveolar canal (black arrow), (b) ground glass internal appearance with central hypodence area

Blood chemistry was within normal limits. Biopsy through the unhealed extraction sockets was performed which revealed gritty firm mass with intervening areas having bony hard consistency with gross bleeding encountered. Histopathology examination confirmed psammomatoid type of JOF as psammoma body like structures seen in a highly cellular background of spindle-shaped fibroblasts (Figure 4a and b).

DISCUSSION

In a study by El-Mofty (2002) two categories of JOF, TrJOF and PsJOF, had been identified based on the histologic criteria.[3] However, the two categories also have a distinct predilection for specific age-groups: The average age of occurrence of TrJOF is 8½-12 years, whereas that of PsJOF is 16-33 years.[3] The psammomatous type mainly involves the bones of the orbit and paranasal sinuses, whereas the trabecular type commonly involves the jaws, although there is controversy as to which jaw has greater predilection, maxilla or mandible. These lesions exhibit a slight male predilection.[9] In addition to its aggressive behavior, PsJOF also has a very strong tendency to recur, and recurrence rate as high as 30-56% have been reported.[13] In rare cases both TrJOF and PsJOF features have been seen together; in a recent study PsJOF has been reported to be occurring as a recurrent lesion after resection of TrJOF.[9]

Although the term "juvenile" is a misnomer because both PsJOF and TrJOF can occur in a wide age range including the elderly.[9] JOF usually manifests as an asymptomatic bony-hard swelling, the duration and extent of which may vary depending on the site and aggressiveness of the lesion; however, it does not demonstrate the chronic, long-standing evolution of some of the other fibro-osseous

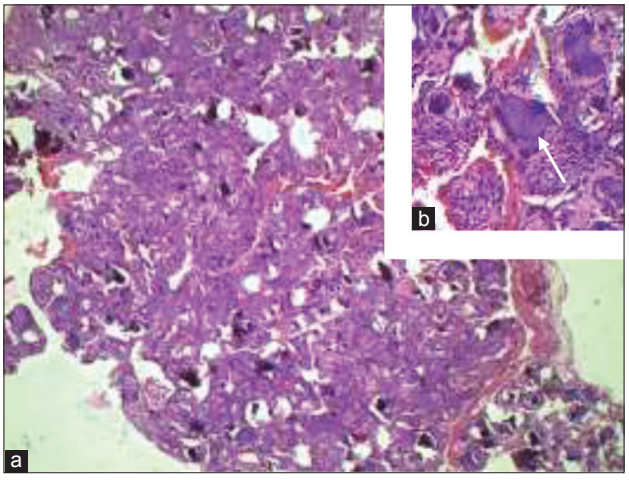

Figure 4: (a) Highly cellular stroma composed of plump fibroblasts and numerous psammoma body like mineralized structures, background stroma is fibrovascular with moderate vascularity (H and E, ×10); (b) psammoma bodies (arrow) with basophilic oval to spherical target like ossicles and peripheral eosinophilic rim (H and E, ×40)

lesions. It can expand the involved bones, causing facial asymmetry. Depending on the site, symptoms such as pain, paresthesia,[14] malocclusion, sinusitis, proptosis, etc. can also occur due to the swelling.[15] Radiographically the internal structure can be radiolucent, mixed, or radiopaque, depending on the degree of calcification.[3] A period of at least 6 years is required for the lesion to pass from radiolucent to radiopaque stage, so usually only radiolucent and mixed density lesions are reported as in this case but in rare cases rapid maturation to complete radiopacity may occur in as less time as 1 year.[4] Radiolucency along with waterline can be seen suggestive of cystic changes with fluid accumulation.[3] In earlier stages it appears as a unilocular radiolucency in extraoral radiographs and solid hypodense mass in CT images[6,14,16,17] as seen in our case. Root displacement is common and resorption, though rare, can occur.[14,15,18] The lesion can cause expansion as well as perforation.[3,14] However in our case the lesion has expanded to large size even without maturing to its radiopaque stage and without destruction of adjacent vital structures and without cortical plate destruction, although displacing adjacent structures.

The radiographic features of JOF can resemble that of other lesions, such as fibrous dysplasia and cemento-ossifying fibroma, aneurysmal bone cyst (ABC), Osteogenic sarcoma and Osteoblastoma.[14,18,19] JOF is not capsulated but is separated from surrounding bone by a radiopaque border[17,18,20,21] and this finding can help in differentiating it from fibrous dysplasia,[18] also root resorption is not a common feature of fibrous dysplasia in contrast to JOF. A "ground-glass" appearance on radiographs has been reported[3,7,17] as in this case. PsJOF usually presents as round well defined, sometimes corticated osteolytic lesion with a cystic appearance, with sclerotic changes being evident as ground glass appearance, while ground glass as well as multilocular honeycomb appearance has been described with TrJOF.[1,21] The bony shell is evidence of old stable lesion. Some lesions penetrate their shell, grow to a new size and produce an outer new shell.[21] It usually has a concentric or centrifugal growth pattern, which can lead to an erroneous clinical diagnosis of cemento-ossifying fibroma.[16,18,19] JOF has also been reported to be associated with other bony lesions such as ABC associated with both PsJOF and TrJOF.[3,16,22] Aggressiveness of the lesion with marked destruction of adjacent structures[15] and presence of osteogenic elements make it sometimes difficult to differentiate from osteogenic sarcoma radiographically; the lack of periosteal reaction in JOF may help in differentiation, however malignant tumors of craniofacial bones often do not exhibit periosteal reaction.[14] Despite the rapid increase in size of JOF there is relative lack of destruction[14] as seen in our case. Anesthesia of Inferior alveolar nerve has been reported rarely but no perineural spread and neural invasion reported,[9] however in our case there was no anesthesia or paresthesia reported along the distribution of inferior alveolar nerve while it was being displaced anteromedially. Craniofacial skeleton is a very uncommon site for osteoblastoma (another osteogenic tumor).[14] Malignant transformation has never been reported.[14,18]

The microscopic features of the lesion are distinctive and include a cell-rich fibrous stroma containing bands of cellular osteoid without osteoblastic lining, osteoid strands, and trabeculae of woven bone.[3,19] PsJOF is slightly more cellular than TrJOF. Due to the resemblance of the psammoma-like ossicles seen in PsJOF to the cementicles in cemento-ossifying fibroma, it has been argued that PsJOF is a type of cemento-ossifying fibroma.[2,3] However, the marked cellularity of JOF is in sharp contrast to the usually stroma-rich appearance of the latter group of lesions. In our case was seen highly cellular stroma composed of plump fibroblasts interspersed with numerous psammoma bodies showing basal oval to spherical ossicles with peripheral eosinophilic rim suggestive of PsJOF. Background stroma is fibrovascular with moderate vascularity with some extravasated red blood cells. The pathogenesis of PsJOF jaw lesions are related to the maldevelopment of basal degenerative mechanism that is essential for root formation.[17]

JOF is most frequent in sinuses that lengthen septa therefore it is presumed that focal proliferation for splicing is excessive producing the cellular stroma, ossicles and gelatinous myxoma and occasional septa. When ossicles predominate they may suggest a JOF. As mandibular lesions are related to ramus and are most common in females aged 6-12 years i.e., the place and time associated with molar development. Between 6th and 12th years erupted first molar completes root development and second molar initiates its root growth for eruption. Basal myxoid generative mechanism for the socket may occasionally produce a JOF with proliferation expanding into the ramus; the tooth may persist, be displaced, or be missing,[1,15,23] although in present case in a 15-year-old girl the swelling has been present since 1 year, but the above theory may suggest that swelling could have been present since the time period of molar tooth development not being noticed by the patient until it acquired a large size, although in this case molar teeth were missing due to previous extraction. However cases of PsJOF, which have been reported to arise in older ages[12] may conflict the theory of tumor origin from basal myxoid tissue; also in Wenig's study of the seven cases of PsJOF, in all the five cases occurring in adults there was no dormant or early onset lesion identified,[12] also more aggressive nature of the tumor in younger ages[6] may suggest some different origin for the two age groups and a

need to further classify the histologically similar appearing lesions based on a possibly different origin still to be found out, which may further help a clinician to diagnose and manage the lesions accordingly based on the predicted aggressiveness and other possible criteria if found to be having a different origin.

There is no standardized follow-up protocol in the literature; because of the fairly high recurrence rate, immediate reconstruction is not advised. The smaller tumors may be treated by enucleation and curettage successfully.[1,5] Resection should also be considered in cases where there is a recurrence,[24] invasion of adjacent cavities, or where preserving the inferior border is not feasible.[6,9]

CONCLUSION

Thus, it can be concluded that PsJOF can expand to large sizes even without maturing to its radiopaque stages and without destruction of the adjacent vital structures although displacing them.

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