CASE REPORT

Psammomatoid variant of juvenile ossifying fibroma

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ABSTRACT

Juvenile ossifying fibroma (JOF) is a rare benign tumor of the craniofacial bones differing from other fibro-osseous lesions in terms of early age of onset, aggressive clinical behavior, and high recurrence rate. Histopathologically, it is divided into two as trabecular JOF (TrJOF) and psammomatoid JOF (PsJOF). In PsJOF, psammoma-like spherical ossicles constitute pathognomonic histopathological images, whereas TrJOF has trabeculae of fibrillary osteoid and woven bone. Despite the histopathologic separation, both lesions have similar clinical behavior, thus the treatment procedure is also the same. Complete surgical resection is preferred for the treatment. We report a rare case of PsJOF involving the maxillary sinus and resultant facial symmetry in a 13-year-old female child.

KEY WORDS: Fibro-osseous lesion, juvenile ossifying fibroma, psammomatoid

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INTRODUCTION

Juvenile ossifying fibroma (JOF) is a benign tumor that occurs in the craniofacial bones and differs from other fibro-osseous lesions in terms of age of onset and clinical behavior. Although it has high recurrence potential and aggressive nature, malign transformation and metastasis have not been reported.[1] Histopathologically, it is divided into trabecular JOF (TrJOF) and psammomatoid JOF (PsJOF).[2] In PsJOF, psammoma-like spherical ossicles constitute pathognomonic histopathological image, while TrJOF has trabeculae of fibrillary osteoid and woven bone.[3]

CASE REPORT

A 13-year-old girl admitted to our clinic as a result of 3-month progressive growth of her painless swelling on the left side of the face that began 1 year ago and complaints of ongoing nasal obstruction. Clinical examination showed a hard swelling with smooth surface starting from the left superior alveolar arch, causing diffuse expansion throughout the anterior wall of the maxillary sinus, and creating asymmetry [Figure 1a]. On the radiological examination, a 42 mm × 42 mm × 39 mm solid mass that caused expansion of the bones by surrounding the multiple number of tooth roots was found in the left superior alveolar arcuate. The maxillary sinus volume was reduced, but the osteometal unit was open. The left nasal cavity was obliterated [Figure 1b].

In the light of these findings, the patient underwent medial maxillectomy by combining left endoscopic sinus and Caldwell–Luc surgery. The excised specimen was sent for histopathological examination. During microscopic examination of very fragmented tumor tissues, the largest one of which was 2.5 cm × 2 cm × 1.5 cm in size, numerous psammoma-like spheroidal ossicles and having peripheral eosinophilic, basophilic centers were seen in the cellular fibrous stroma that did not show atypia [Figure 1c]. Mitosa was rarely seen as 1/50 high-power field. Necrosis was not seen. The histologic features were suggestive of psammomatoid variant of JOF. On radiological control at the postoperative 9th month, a curvilinear style thickening which extended to superior alveolar arcuate hard palate and the left maxillary sinus bed was observed. It was isointense in T1A and hyperintense in T2A compared to adjacent muscle tissues. It also reached 9 mm in some places and showed contrast retention in postcontrast sections [Figure 1d]. On endoscopic examination, there was a mucosal hypertrophy appearance in the cavity.

DISCUSSION

JOF is a rare, locally aggressive tumor with a high potential for recurrence. JOF is thought to originate from mesenchymal cell differentiation of the periodontal

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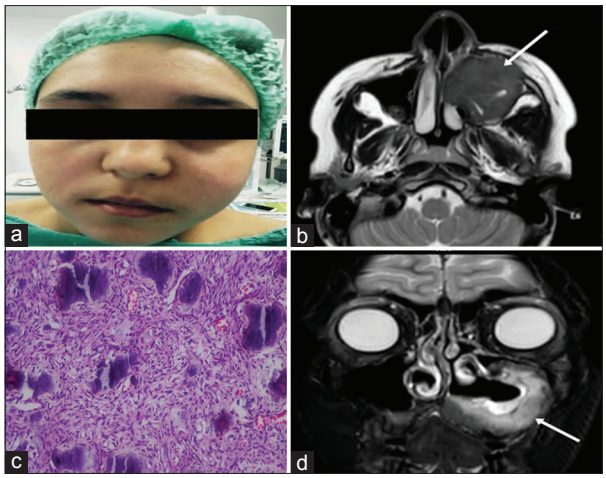

Figure 1: (a) Preoperative photograph of the patient showing facial asymmetry due to unilateral swelling on the left side of the face. (b) Axial T2-weighted magnetic resonance image shows the solid mass (arrow) which are mostly hypointense (it sometimes includes hyperintense foci), fills left maxillary sinus, and cause expansions. (c) Cellular fibrous tissue with numerous spheroidal ossicles (H and E, ×200). (d) Coronal T2-weighted magnetic resonance image shows postoperative absenteeism in the medial wall of the left maxillary sinus, air and fluid signals in the sinus lumen, and hyperintense thickening (arrow) in the lower, upper, and lateral sections of the sinus

ligament, a precursor for osteoid, fibrous tissue, and cementum.[4] In these lesions, the normal bone structure is replaced with collagen fibers and fibroblasts containing mineralized material in various quantities.[5] PsJOF was first reported by Benjamins in 1938 as osteoid fibroma with atypical ossification of the frontal sinus. In 1949, Gögl renamed these lesions as psammomatoid ossifying fibroma of the nose and paranasal sinuses. In 1952, the same lesion was named by Johnson *et al.* as juvenile active ossifying fibroma.[6] Slootweg *et al.* divided JOFs into two as JOF-WHO type and JOF-PO (psammoma-like ossicles) based on the difference in the age of formation.[7] In the most recent classification by El-Mofty, it was divided into two as trabecular JOF (TrJOF) and psammomatoid JOF (PsJOF) according to the histopathological criteria of JOFs.[2]

Although JOF is mostly seen in children and young adults, it can be seen in a wide range from 3 months to 72 years (1, 7). Hammer *et al.* and Slootweg *et al.* reported that the mean age of onset was 11.5 and 11.8, respectively.[8]

Clinical symptom of JOF is usually swelling in asymptomatic bone stiffness. The size and duration of swelling vary depending on the aggressiveness and the location of the tumor. PsJOF mostly occurs in paranasal sinuses, whereas TrJOF usually occurs in the maxilla.[7] Contrary to this, our patient showed involvement in maxilla even though she had PsJOF.

It might radiologically be radiolucent, mixed, and radiopaque according to the degree of calcification or cystic changes.[6,7] In addition, the radiological image varies greatly depending on the stage of development of the lesion.[9]

JOF diagnosis is made through correlation with clinical, imaginogical, and histopathological findings.[5] JOF is classified as TrJOF and PsJOF according to histopathologic criteria. However, it has also some features such as the age of onset and locality.[2] However, their clinical behavior is similar, thus the treatment is also the same. While 75% of the PsJOF is seen in orbit, paranasal sinuses, and calvaria, TrJOF is mostly seen in the maxilla.[10] The age range of TrJOF (8.5-12) is smaller than that of PsJOF (16-33.[2]). In our 13-year-old patient, the placement of PsJOF was in the form of expanding the maxillary sinus to superior by originating from tooth roots in the superior alveolar arch.

Histologically, both variants of JOF have similar stroma. However, numerous spherical/lamellated ossicles resembling psammoma bodies of meningioma's in the cell-rich fibrous stroma are seen in PsJOF, while trabeculae of woven bone with coarse lacunae, swollen osteocytes, and a lining of plump osteoblasts are observed in TrJOF.[10] Whether PsJOF is a variant of cemento-ossifying fibroma that has long been discussed due to the similarity between cementicles in cemento-ossifying fibroma and psammoma-like ossicles in PsJOF. However, the significant cellularity of JOF is generally contradictory to the stroma rich appearance of cemento-ossifying fibroma.[7]

JOF should be treated like local aggressive neoplasia because of its aggressive behavior and due to its high recurrence rate (30%–58%). Complete surgical resection should be preferred because partial or incomplete resections will cause recurrences.[10] In our patient, at the end of the 3rd year, thickening on the cavity wall with mucosal hypertrophy occurred instead of aggressive and invasive recurrence. It did not cause any pressure or asymmetry and the controls of the patient have been still continuing.

CONCLUSION

JOF is an uncommon benign but potentially locally aggressive fibro-osseous lesion. In the management of this lesion, due to its aggressive nature and high recurrence rate with infiltrative growth pattern, early diagnosis together with a multidisciplinary approach of clinic, radiology, and pathology, and long-term follow-up with nasal endoscopy and serial imaging are very important.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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