Psammomatoid Juvenile Ossifying Fibroma of Maxilla: An Uncommon Neoplasm

Abstract

Juvenile ossifying fibroma (JOF) is a rare variant of ossifying fibroma. JOF is classified into two distinct clinicopathological variants: Trabecular and psammomatoid JOF. Based on the age group involved, most common site of occurrence and clinical behavior, JOF differs from the larger group of ossifying fibromas. We report a case of psammomatoid JOF of maxilla in a 10-year-old female who presented with a swelling over the right side of the face.

Keywords: *Fibroma, juvenile, maxilla*

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Introduction

Juvenile ossifying fibroma (JOF) is an uncommon benign bone-forming neoplasm involving the craniofacial skeleton of juvenile patients.[1] Its incidence is still underestimated because of its rarity.[1] JOF is a variant of ossifying fibroma. JOF has been distinguished from the larger group of ossifying fibromas on the basis of age, most common site of involvement and clinical behavior.[2] As a term "juvenile" underlines, the tumor usually develops in children, majority of whom under age of 15 years.[2] The lesion most commonly involves the paranasal sinuses, periorbital bones. Prognostically JOF behaves in a very aggressive fashion than does the ossifying fibroma.[3] JOF is further classified into two distinct clinicopathological variants: Trabecular JOF (TrJOF) and psammomatoid JOF (PsJOF).[1,3] We report a case of PsJOF of the maxilla in a 10-year-old female.

Case Report

A 10-year-old female presented with complaints of swelling over the right side of the face for past 8 months. It initially started as pustule and progressed slowly. On examination, the swelling was 7 cm × 7 cm in size extending superiorly till infraorbital margin, medially till ala of the nose and inferiorly above the ramus of the mandible. On palpation, the swelling was bony hard in consistency. Intraorally, mucosa over swelling appeared normal. Computed tomography of paranasal sinuses showed a partially ossified mass causing effacement of the right maxillary cavity. A fine needle aspiration was attempted, however, it did not yield any material. Partial maxillectomy was done under general anesthesia and the specimen was sent for histopathological examination. Grossly the lesion was globular, gray-white in color and hard in consistency [Figure 1a]. Microscopically, the lesion consisted of a cellular stroma comprised of spindle-shaped cells having elongated nucleus, opened up nuclear chromatin, moderate amount of eosinophilic cytoplasm with indistinct cellular margins. Small ossicles resembling psammoma bodies were interspersed in the stroma [Figure 1b]. Many ossified trabeculae of woven bone revealing focal areas of osteoblastic rimming were seen [Figure 1c]. No necrosis, atypia, or increased mitotic figures were identified. Immunohistochemical for Ki67 was 3% [Figure 1d]. A final diagnosis of PsJOF was made.

Discussion

Although described as early as in 1938 by Benjamins and in 1949 by Golg, it was in the year 1985 Margo *et al*. introduced the term "psammomatoid juvenile ossifying fibroma."[1,3,4] The PsJOF has an aggressive, infiltrative growth pattern, and propensity for recurrence.[1] The sites most commonly involved are the paranasal sinuses followed

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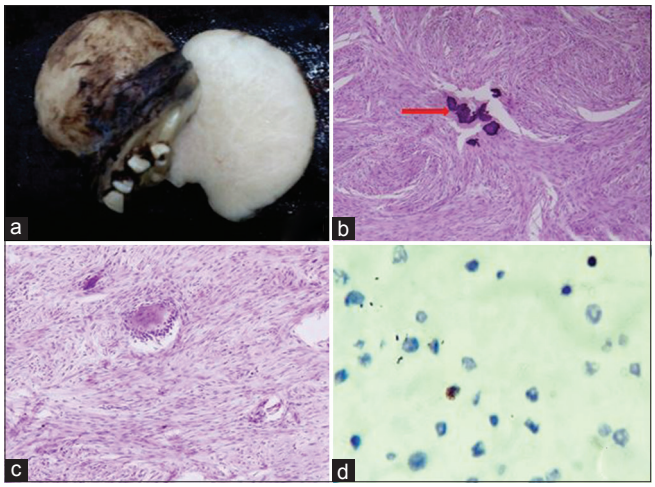

Figure 1: (a) Grossly the tumor was globular, gray-white in color and hard in consistency. (b) Microscopically, small ossicles resembling psammoma bodies seen interspersed in the stroma (H and E, ×40). (c) Ossified trabeculae of woven bone with focal areas of osteoblastic rimming (H and E, ×100). (d) Immunohistochemistry for Ki67 (×100)

by maxilla and mandible.[1] Maxilla is involved in 20% of cases only.[1]

The age of PsJOF patients ranges from 16 to 33 years.[1] It usually manifests as an asymptomatic bony-hard swelling. Sometimes it may be associated with pain, paresthesia, sinusitis and orbital proptosis.[3] Depending on the degree of calcification, it can be radiolucent, mixed or radiopaque on radiological imaging.[3] Microscopically, PsJOF is characterized by small spherules resembling psammoma bodies, which are referred to as psammoma-like bodies.[3,4] The intervening stroma is highly cellular consisting of spindle-shaped fibroblasts.[3] Immunohistochemical marker for Ki67 was performed to assess the aggressive behavior of the neoplasm. The Ki67 was 3%, which was in accordance with other case studies.[2]

The pathogenesis for these jaw lesions is related to the maldevelopment of basal generative mechanism that is essential for root formation.[3] The presence of nonrandom chromosomal breakpoints at Xq26 and 2q33 resulting in (*x*, 2) translocation have been reported.[5]

Treatment options range from simple curettage with peripheral osteoectomy to block resection and segmental resection.[1] Bone grafting has been used in severe cases. Surgical resection is the preferred choice of management because of the aggressive nature and high recurrence rate of PsJOF.[3]

Conclusion

JOF is an uncommon benign bone-forming neoplasm involving the craniofacial skeleton of juvenile patients. It has been distinguished from the larger group of ossifying fibromas on the basis of age, the most common site of involvement and clinical behavior. PsJOF has an aggressive course with tendency to recur.

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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