

Juvenile ossifying fibroma: Psammomatoid variant

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Abstract

Juvenile ossifying fibroma is a rare fibro-osseous lesion containing variable amount of calcified masses, which resembles bone or cementum within a fibrocellular connective tissue stroma. It has variable clinical behavior, highly aggressive in nature including invasion and destruction of adjacent anatomic structures with a strong tendency to recur. We reported a 28-year-old female patient with a growth in the upper left vestibule region extending from canine to molar region with clinical, histopathological, and radiological features are presented. Surgical management was done, and regular follow-up was advised.

Keywords: Benign neoplasm, juvenile ossifying fibroma, psammoma

Introduction

Juvenile ossifying fibroma (JOF) is an uncommon benign osteogenic neoplasm. It was first described by Benjamins in 1938 as “osteoid fibroma with atypical calcification.” (Khoury *et al.*, 2002). Later on in 1952, Johnson coined the term “juvenile active ossifying fibroma” (Neville *et al.* 2002).^[1]

As a term “juvenile” underlines, the tumor largely develops in children, 79% of whom under age of 15 years old. In reviews published by Slootweg *et al.* and Hamner *et al.*, the mean age of onset was 11.5 and 11.8 years, respectively, but it does occur in adults.^[1]

JOF is a controversial lesion, and it has been distinguished from the adult variant of ossifying fibroma on the basis of age of occurrence, most common site of involvement, and clinical behavior. Most of them affect extra-gnathic bones; some affect maxilla and craniofacial bones but rarely mandible.^[2]

Hereby, we present a case report of juvenile ossifying fibroma, psammomatoid Variant.

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Case Report

A 28-year-old female patient reported to an oral and maxillo-facial surgeon in a private dental clinic with a complaint of swelling on the left side of face over a 5-year period. The main concern of the patient was esthetic rehabilitation. Past history revealed a small painless swelling, which first appeared at the age of 12 years at the same site. Incisional biopsy was done; it was then diagnosed as cementifying fibroma. She underwent surgery for that and remained asymptomatic for 10 years. After that, swelling recurred at the same site, and it was excised. A diagnosis of fibrous dysplasia was given. After a period of 5 years, it recurred again at the same site, which gradually increased in size to attain the present size.

Extra-oral examination revealed gross facial asymmetry on the left side of the face with obliteration of the nasolabial fold. This was also visible in the left nasal cavity. On palpation, swelling was non-tender, hard in consistency with no bony crepitus present. [Figure 1]

Intra-oral examination revealed solitary well-defined swelling in the left upper vestibule in the canine-molar region, measuring approximately 5 × 3.5 cm in size. Slight mobility was present in relation to 23 and 25, and 24 was missing. There was gross expansion of the left maxillary process extending to the hard palate. [Figure 2] There was no history of pain, toothache, discharge of pus, and paresthesia. The overlying mucosa was smooth and intact and was of normal color.

CT scan was carried out. Under coronal view, mixed radioopaque-radiolucent lesion was seen on the antero-lateral and medial wall of the maxilla, involving the entire maxillary antrum on the left side and extending into the lateral wall of the nose.

Under axial view, destruction of buccal and palatal cortical plate was seen in the anterior maxilla. More extension of

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the lesion was seen on the buccal side, extending into the nasal cavity with complete obliteration of the antrum on the left side.

Under 3D view, nasolabial fold and vestibular obliteration was noticed. [Figure 3]

All routine blood examination was conducted before the surgical procedure. Excisional biopsy of the mass was performed under general anesthesia. Gross examination of tissue specimen revealed single tissue bit measuring 5 × 3.5 cm in size, brownish-white in color, gritty in consistency.

Microscopic examination revealed a highly cellular fibrous stroma exhibiting a whorled pattern. The connective tissue stroma consists of proliferating plump cells having hyperchromatic nuclei intermingled with psammoma-like bodies or psammomatoid ossicles without any osteoblastic rimming. The intervening fibrous connective tissue stroma consists mainly of spindle-shaped fibroblasts and areas of chronic inflammatory cells. [Figures 4]

Immunohistochemical analysis was conducted to check out the aggressiveness and origin of tumor. Ki 67 [Figure 5] and vimentin [Figure 6] positivity was noticed.

Thus, the final diagnosis of juvenile ossifying fibroma was given.

The patient was subjected to clinical and radiological follow-up after excision of the lesion to discard any possible relapses or recurrences.

Discussion

Benign fibro-osseous lesions consist of varied group of pathological conditions that include developmental lesions, reactive or dysplastic disease, and neoplasms.^[3] They encompass a class of histopathology characterized by the presence of a fibrous stroma with varying amounts of mineralized material, which resembles bone or cementum. One such rarely-occurring fibro-osseous lesion is JOF.^[4]

The etiology of JOF has not been fully understood. Genetic differences in fibro-osseous lesion of craniofacial bone are not well-established, but there are reports that these were related to non-random break points at Xq26 and 2q33. A few cases of facial trauma have been suggested as a possible etiologic factor in the JOF development.^[3]

Clinically, the symptoms are variable and include facial swelling, enlarging hard mass, sinusitis, nasal obstruction, teeth displacement, eye proptosis, and pain. The present patient was 28-years-old and demonstrated a swelling on the left side of face, which was non-tender and was hard in consistency.

Our case is in accordance to the findings of Slootweg *et al.*^[5] who described maxilla as the most frequent site while Johnson *et al.*^[6] even reported that 90% of facial bone fibromas occur in the paranasal sinuses and only 10% in the mandible.

Radiographically, they present as well-circumscribed radiolucencies, and in some cases, contain central radiopacities. The term “mature” has been used to denote the transformation of the lesion from the radiolucent to radiopaque stage. A period of at least 6 years is required for the lesion to pass from the radiolucent to the radiopaque stage.^[7] However, the case presented here showed mixed radiolucency and radio opacity, which is indicating towards more mature lesion.

The histological criteria for accepting any case as JOF was defined by the WHO and Slootweg *et al.* According to them, well-demarcated lesions showing a fibrocellular stroma containing trabeculae of osteoid and woven bone with or without osteoblastic rimming with paucity of cementicles and containing giant cells can be considered as JOF. The lesion is non-encapsulated but well-demarcated from surrounding bone, which is in contrast to fibrous dysplasia (FD), which has poorly-defined borders and blends with surrounding normal bone.^[8]

Histologically, the psammomatous (Ps) and trabecular (Tr) subtypes of JOF have been well-documented. 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.^[9]

The psammomatous variants have a basophilic center and a peripheral rim that sometimes showed radiating fibers, thus assuming the appearance of a fringe. Osteoblastic rimming may or may not be present. The trabecular subtype possesses irregularly-shaped trabeculae of immature bone with or without osteoblastic rimming instead of spherical or ovoid bone.

The case presented here shows psammoma-like bodies or psammomatoid ossicles without any osteoblastic rimming within fibrocellular connective tissue stroma. In our case, it is the pre-dominant calcified tissue produced, and such lesions may be designated as psammomatoid ossifying fibromas- from the Greek word ‘psammos,’ i.e. sand.^[4]

Immunohistochemical markers were applied to explore the behavior and origin of tumor. Ki 67 positivity indicates the aggressiveness, which were in accordance with the findings of Bohn OL *et al.*^[12] and vimentin positivity indicates the connective tissue origin of tumor, which were in accordance with the findings of Granados R *et al.*^[10]

On CT scan, JOF is characterized as an expansive lesion, having well-defined sclerotic borders, locally aggressive and

destructive at cortex.^[1] In our case, The CT showed expansion and destruction of buccal and palatal cortical plates with obliteration of nasolabial fold, which are consistent with the above-mentioned findings.

The diagnosis of fibro-osseous lesions is often arbitrary and subjective since both droplet cementum and bone formation are found in many of these tumors. Thus, clinical, radiological, and histological correlation is required.



Figure 1: Facial asymmetry along with obliteration of nasolabial fold



Figure 2: Well-defined swelling in the upper left vestibule region



Figure 3: 3D reconstruction showing nasolabial fold as well as vestibule obliteration

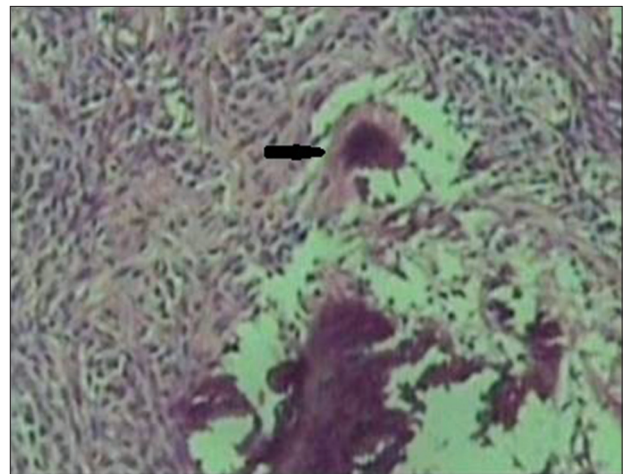


Figure 4: Fibrocellular connective tissue stroma containing psammoma-like body (H and E, x10)

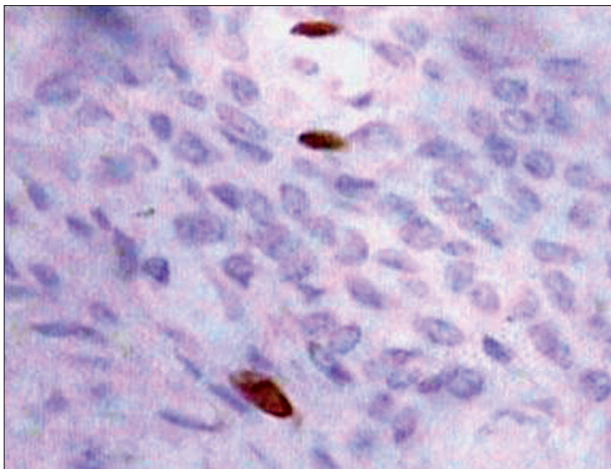


Figure 5: Ki-67 Positivity

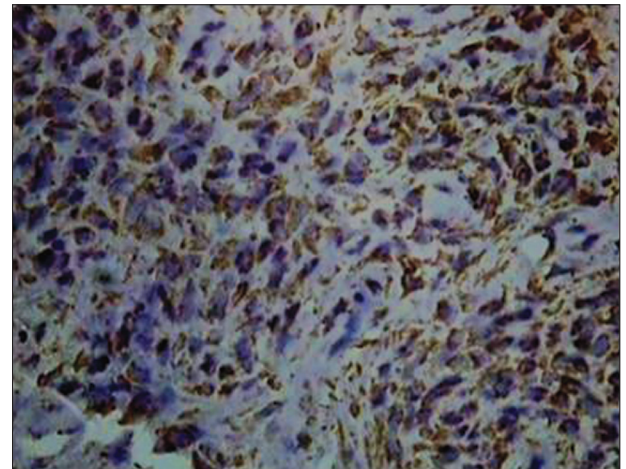


Figure 6: Vimentin positivity

Owing to its aggressive nature and high recurrence rate, early detection and complete surgical excision is required. Thus, it is important to perform a clinical and radiological follow-up for as many years as possible because of the possibility of recurrence in this type of neo-formation.

Conclusion

A case of juvenile ossifying fibroma is presented. Clinical, histopathological, and radiographic features were correlated to reach the final diagnosis of JOF and to differentiate from the other fibro-osseous lesion. Although the lesion was excised completely, still a regular follow-up was maintained as the tumor has proved to be aggressive.

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