Juvenile Aggressive Ossifying Fibroma: A Rare Case Report

1S Manoj Kumar, 1S Elangovan, 2S Shanmugam, 3S Srividhya

1Professor, Department of Oral Medicine and Radiology, Ragas Dental College and Hospital, Chennai, Tamil Nadu, India
2Professor and Head, Department of Oral Medicine and Radiology, Ragas Dental College and Hospital, Chennai, Tamil Nadu, India
3Postgraduate Student, Department of Oral Medicine and Radiology, Ragas Dental College and Hospital, Chennai, Tamil Nadu, India

Correspondence: S Manoj Kumar, Professor, Department of Oral Medicine and Radiology, 2/102, East Coast Road, Uthandi Chennai-600119, Tamil Nadu, India, e-mail: smanojk@indiatimes.com

ABSTRACT

Juvenile ossifying fibroma considered as a variant of ossifying fibroma by some authors is rare. Its aggressive behavior necessitates early detection and proper surgical management. It may mimic different pathologic entities of jaw at different stages. A case report of juvenile aggressive ossifying fibroma in a 12-year-old girl which was diagnosed, investigated and treated successfully is presented here.

Keywords: Juvenile, Ossifying fibroma, Aggressive, Fibro-osseous.

INTRODUCTION

Fibro-osseous lesions are pathologic entities in which normal bone is replaced by fibrous tissue which in turn contains foci of mineralized substance. Fibro-osseous lesions of the jaws include developmental (hamartomatous) lesions, reactive or dysplastic processes and neoplasms.1

Ossifying fibroma is a well-demarcated benign fibro-osseous tumor with capsule, composed of metastatic bone, fibrous tissue and varying amounts of osteoid. The ossifying fibromas are subdivided into conventional and juvenile clinicopathologic subtypes.2

Juvenile ossifying fibroma is a rare fibro-osseous neoplasm that arises within the craniofacial bones in individuals under 15 years of age. It is usually asymptomatic achieving a large size and exhibiting aggressive behavior and is often diagnosed as juvenile ossifying fibroma, aggressive ossifying fibroma or active ossifying fibroma in the literature.3

A case report of juvenile aggressive ossifying fibroma is presented here because of its relative rarity.

CASE REPORT

A 12-year-old girl reported to the department of oral medicine and radiology, complaining about a swelling over the left side of the face for the past 3 months. She stated that the swelling was fresh, started suddenly and spontaneously adjacent to the left nostril about 3 months ago and kept increasing in size. The swelling was neither preceded nor followed by trauma, toothache, pain, discomfort or fever. There were no other swellings in the body. She had not attained menarche. She was moderately built, well nourished. There were no signs of anemia or jaundice. There was no significant lymphadenopathy. Review of the systems was not remarkable.

Examination of the face showed facial asymmetry because of the swelling over the left maxilla. The level of the left orbit was slightly at a higher level than the right orbit. On further enquiry, the patient did not report any visual disturbance. The swelling was single, diffuse, round and measured 5 cm in diameter extending from the ala of the nose obliterating the nasolabial fold to about 4 cm in front of the lobule of the left ear, from the lower border of the left orbit to the left corner of the mouth. The skin over the swelling appeared normal but stretched. Palpation confirmed the site, size and extent but the swelling was well circumscribed, nontender, hard in consistency, smooth surfaced and fixed to the underlying bone. There was no evidence of paresthesia.

Intraorally, a single, well-circumscribed swelling was seen in continuity with the extraoral swelling measuring 5 × 3 cm extending from the distal aspect of 21 to distal aspect of 25 (Fig. 1). It was situated above the alveolar mucosa obliterating the buccal sulcus. The surface was covered by normal mucosa with areas of blanching. None of the associated teeth showed evidence of caries.

The crown of the lateral incisor was tilted distally and labially and retained deciduous canine was seen. The marginal and attached gingiva appeared normal. The right and left maxillary second molars were yet to erupt. On palpation, the swelling was hard in consistency, nontender and had a nodular surface with mild blanching on pressure. There was no swelling on the palatal aspect. There was no mobility and tenderness on percussion or pocket formation in the associated teeth, which were also found to be vital on vitality tests.

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Based on the history and findings of the clinical examination, a provisional diagnosis of a benign odontogenic neoplasm was made. The possibility of a dentigerous cyst, a fibro-osseous
lesion and a benign tumor of the bone was considered in the differential diagnosis.

An attempted aspiration was not successful because of the bony hard nature of the swelling. The IOPA of incisor canine region showed the retained 63, distally inclined 22, the impacted 23 above the incisors. A mixed radiolucent, radiopacity was seen adjoining the root of 22 which extended beyond the radiograph.

Maxillary topographic occlusal view showed a large mixed radiolucent, radiopaque lesion which was extending from the root of 22 to distal aspect 26. It was well demarcated without a hyperostotic border. The impacted canine was situated on the periphery of the lesion, pushing the floor of the nasal fossa inwards. It also showed the buccal cortical expansion from 22 to 25 region (Fig. 2).

The panoramic radiograph showed similar findings. The opacification of the inferior portion of the left maxillary sinus was also noted.

An incision biopsy under local anesthesia was performed. The histopathology showed cellular proliferation of spindle-shaped cells arranged in streamy fascicle with minimum amount of collagen fibers, peripheral arrangement of fibers along with cortical bone, suggesting it to be a juvenile aggressive ossifying fibrom.

A well-encapsulated lesion was removed in toto under general anesthesia. The histopathological examination of the resected lesion confirmed the diagnosis. The postoperative period was uneventful (Fig. 3).

DISCUSSION

JOF considered to be a variant of ossifying fibromas distinguished because of its tendency to occur in younger patients, its predilection for maxilla and aggressive behavior. Juvenile ossifying fibroma was first described by Benjamin in 1938 as 'osteoid fibroma with atypical calcification'. Johnson in 1952 coined the term 'juvenile active ossifying fibroma'. According to the new edition of the classification of the World Health Organization, ossifying fibromas which appear as fast growing mass between 5 and 15 years of age, radiologically well bordered and consistent with ossifying fibroma histologically, are referred as juvenile (aggressive) ossifying fibroma. Facial trauma has been suggested as a possible etiologic factor in the JOF development.

JOF has no gender predilection. However, Johnson et al stated that mandibular tumors are more frequently common in girls between the age of 5 to 11 years or during the second to fourth decades of life. JOF has been reported to occur as a hybrid lesion along with aneurysmal bone cyst. 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 lesions. It can expand the involved bones, causing facial asymmetry. Depending on the site and symptoms, such as pain, paresthesia, malocclusion, sinusitis, proptosis, can also occur due to the swelling. Very recently, Pace, Crosher, Holt estimated the rate of growth in a case of juvenile aggressive ossifying fibroma using panoramic radiographs and 3D CT. The result showed that the lesion had doubled in length and the cross-sectional area had increased more than 3.5 times in the span of 35 months. This emphasizes the aggressive nature of JOF.

Radiographically, JOF appears as a well-demarcated lesion involving the alveolar and basal bone of either jaw. The internal structure can be radiolucent, mixed, or radiopaque, depending on the degree of calcification. Root displacement is common and resorption can occur. Though it causes cortical expansion in both the directions, it expands the buccal cortex to greater extent. A periapical, occlusal, panoramic radiographs and CT may be necessary to evaluate the lesion for its periphery, internal structure extent and its relationship with the adjacent teeth and bone. On CT, JOF is characterized as expansive, having defined sclerotic borders, locally aggressive and destructive at cortex. On MRI, JOF is isointense in T1-weighted images and hypointense or isointense in T2-weighted images.
Histopathologically, JOF appears in two distinct forms, the psammomatoid and trabecular types.

Our case showed increased areas of cellularity with areas of minimal mineralization with no mitotic figures. Intraoperatively, the lesion was shelled out as a whole. These features were consistent with a diagnosis of juvenile aggressive ossifying fibroma. Though aggressive, JOF is a benign lesion and can be completely removed by local excision or thorough curettage when the lesion is relatively small. When allowed to attain a larger size, more extensive surgical procedure may be needed. So, an early diagnosis will pave way for a simple surgical procedure and preclude greater tissue destruction, complicated surgery and the need for reconstruction.

REFERENCES