Juvenile trabecular ossifying fibroma involving mandibular angle and ramus region

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Abstract

Juvenile ossifying fibromas (JOFs) are the fibro-osseous lesions known to occur in children. Although benign, they are locally aggressive and are known to reoccur. They have two microscopic patterns; trabecular and psammomatoid. Here, we report a case of JOF in a 4-year old male patient who reported to our department with a chief complaint of swelling over the right side of the lower third of his face.

Keywords: Fibro-osseous lesions, juvenile ossifying fibroma, ossifying fibroma

Introduction

Juvenile ossifying fibroma (JOF) is a fibro-osseous lesion arising within the craniofacial bones of children. It is a rarity and is distinguished from the adult type on the basis of age, location, clinical features, and microscopic picture.[1] Within the confusing ray of fibro-osseous lesions, it is the most enigmatic. Whenever pathologists consider such a diagnosis, they are faced with too many criteria and find little consolidation in the literature. Two distinct microscopic patterns have been described; trabecular and psammomatoid.[2] JOF is considered as a unique lesion because of its tendency to occur in children and adolescents, its locally aggressive growth pattern and the complex histological features. Hence, early detection and complete surgical excision are essential.[3]

Case Report

A 4-year-old male patient reported to our department with his father complaining of a swelling over the right side of his son's face since 25 days. The swelling was insidious in onset, slowly progressing in size to the present size, and associated with mild intermittent dull pain. Patient’s father also gave a history of trauma over the right side of his son’s face 25 days back, wherein he got hit by a slide in the school. No history of restricted mouth opening, difficulty in jaw movements, and difficulty in swallowing or speaking was revealed. No history of any topical application was given. Patient’s father has consulted a pediatrician before for his son, wherein he was prescribed antibiotics and analgesics and was asked to consult a dentist.

On general physical examination, the patient was moderately built, well oriented to time, place, and person, and showed no signs of pallor, icterus, cyanosis, clubbing, and lymphadenopathy.

On intraoral examination, there was a solitary swelling over the buccal aspect of 85 and posterior to it [Figure 3]. It was roughly oval in shape, measuring approximately 3 cm in size. It extended superoinferiorly from approximately 2 cm below the ala-tragal line till the lower border of the mandible, and mesiodistally from the corner of the mouth till angle of the mandible. The borders of the swelling were diffuse. The skin over it appeared normal in color as the surrounding skin. On palpation, all the inspector findings were confirmed. There was no local increase in surface temperature of the swelling. It was soft at the periphery and firm in the center and was non-tender.

On intraoral examination, there was a solitary swelling over the buccal aspect of 85 and posterior to it [Figure 3]. It was roughly oval in shape, measuring approximately 3 cm in size. It extended superoinferiorly from the level of occlusal plane till the pterygomandibular raphe. The mucosa over it appeared pink. The borders were...
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diffuse. On palpation, all the inspectory findings were confirmed. It was firm in consistency, non-tender, and non-mobile. Full complement of primary dentition was present. The teeth 84 and 85 showed no odontogenic pathology, no displacement, and no mobility.

On the basis of history and clinical examination, a provisional diagnosis of cystic lesion involving the right posterior mandible and ramus was given. The differentials were listed as dentigerous cyst in unerupted 46, adenomatoid odontogenic tumor, ameloblastic fibroma, and unicystic ameloblastoma.

Orthopantomograph [Figure 4] was then taken for the patient, which revealed a full complement of primary dentition and tooth buds of permanent teeth present. Solitary oval-shaped multilocular radiolucency measuring approximately 5 cm in diameter was seen involving the angle and ramus of the mandible on the right side. Anteriorly, the radiolucency was attached to the cementoenamel junction of 46 on the distal aspect and covered its occlusal surface. It extended posteriorly till the sigmoid notch. Superiorly and inferiorly, it showed thinning of the cortices. The internal structure appeared homogeneously radiolucent with few pinpoint-sized radiopaque flecks seen near the occlusal surface of 46. The crown of 46 showed slight distal tilt.

Cone beam computed tomography (CT) [Figure 5] of the right mandible revealed solitary oval-shaped hypodense area in the posterior aspect of mandible on the right side extending from 46 to posterior border of ramus of mandible and from subcondylar region to the lower border of mandible, measuring 5 cm in its greatest diameter.

The axial section revealed the expansion of buccal and lingual cortical plates in the region of right mandibular angle and ramus with thinning of the cortices. Thick linear septa were seen extending internally from the anterior wall toward the posterior wall.

The sagittal section revealed a discontinuity in the distal aspect of the follicular space at the cervical level of 46. A unilocular

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**Figure 1:** Profile picture showing solitary swelling over right side lower third of face

**Figure 2:** Worm's eye view showing diffuse solitary swelling involving the right side of lower third of face

**Figure 3:** Intraoral picture showing swelling in the right lower posterior vestibule region

**Figure 4:** Orthopantomograph showing radiolucency involving right mandibular angle and ramus region
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Ossifying fibroma, a rare tumor entity, is a well-demarcated benign fibro-osseous tumor, wherein the capsule is made of metaplastic bone, varying amounts of osteoid and fibrous tissue. They are subdivided into conventional and juvenile clinicopathologic subtypes. Conventional ones are usually slow growing and seen in the third and fourth decades of life, whereas the juvenile type is benign, potentially aggressive lesions of the craniofacial bones.

The JOF was first described by Sir Benjamin, in 1938, as an “osteoid fibroma with atypical calcification” and later on in 1952, Sir Johnson et al., for the first time, coined the term “juvenile active ossifying fibroma.” In literature, it is known by a variety of terms: Juvenile active ossifying fibroma, juvenile aggressive ossifying fibroma, trabecular osteo desmoplasia, and active fibrous dysplasia. It is a rare lesion and comprises 2% of oral tumors in children. Although it mostly occurs in children and young adults, Johnson et al. have reported cases of JOFs occurring between 3 months and 72 years.

The etiology of an OF is unknown, but developmental, odontogenic, and traumatic origins have been suggested, and it is also thought to be of a periodontal ligament origin because of its capacity to produce cementum and osteoid material. Piementa et al. have reported the association with tumor suppressor gene (HPRT2), suggesting that the lesion could arise as a result of haploinsufficiency of the particular gene.

JOF is located mainly in facial bones (85%), calvarium (12%), and very rarely (3%) extracranially. Ninety percent of the facial lesions involve the sinuses, mainly the maxillary antra. Mandibular lesions are seen in 10% of the cases. In the mandible, the angle and ramus are the most common sites of involvement. The lesion is usually asymptomatic. In general, the first sign of the tumor is swelling of the jawbone. Some lesions can enlarge rapidly causing extensive destruction of bone. In these cases, the tumor may cause facial asymmetry, local pain, teeth loss, root displacement, dental occlusion alterations, cortical thinning, and perforation. When JOF involves the maxillary bone, it can be associated with nasal obstruction, sinusitis, and proptosis. Radiographically, it is characterized by three stages. Stage I or the initial stage appears as a well-defined radiolucency, with no evidence of internal calcification. Stage II or mixed stage is characterized by flecks of radiopacities in the radiolucent area. Stage III or the mature stage is a completely radiopaque mass. Hence, on a plain radiograph, it can appear radiolucent, radiopaque, or of mixed opacity depending on the degree of calcification of the lesion. The radiograph may also show certain non-specific features that consist of a unilocular or multilocular radiolucent area having ill-defined borders and occasional central opacification. Erosion and invasion of the surrounding bone may be noted in more aggressive cases. On CT scans, JOFs appear as an expansile mass, surrounded by thick or thin hyperdense rimming. There may be islands of bone formation within the lesion. CT attenuation levels have been reported to range from

**Discussion**

Fibro-osseous lesions are a group of heterogeneous lesions including developmental, reactive, and neoplastic types, wherein the normal bone is replaced by fibroblasts which consequently form collagen fibers and also by different types of mineralized tissues, similar to bone or cement.

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**Figure 5:** Cone beam computed tomography of right posterior mandible and ramus region
34 to 513 Hounsfield depending on the fibrous tissue and bone content. On contrast imaging, it shows diffuse appearance enhancement. Magnetic resonance imaging is important to see the lesion extent but is inadequate for bony components. It appears isointense on T1-weighted and hypointense on T2-weighted images. On gadolinium injection, there is homogeneous tumor enhancement.[14] Large mandibular lesions may lead to downward bowing of the inferior border of the mandible, most apparently visible on radiographs and CT scans.[17]

Differentials can be listed as ameloblastic fibroma, central osteoma, odontogenic cyst, vascular lesion, ameloblastoma, and giant cell lesions.

Histologically, it shows the presence of a cellular fibrous stroma, bony strands, and cement particles. El-Mofty identified two histopathological variants, trabecular JOF and psammomatoid JOF (PsJOF). Trabecular variant shows the presence of trabeculae of fibrillar osteoid and woven bone and psammomatoid variant shows the presence of small uniform spherical ossicles that resemble psammoma bodies. Trabecular variant is a gnathic lesion, mostly involving maxilla, PsJOF occurs in the sinonasal and orbital bones.[18]

Although malignant transformation has not been reported in literature so far, JOF is a locally aggressive lesion with high recurrence rate if not adequately treated. It is known to have recurrence rate from 30% to 58%, and these are generally seen at early stage and these are more aggressive than the primary lesions.[19] It is also commonly found to be associated with secondary changes such as aneurysmal bone cysts and hemorrhage.

**Conclusion**

Although JOF is a rare lesion, its aggressiveness and high recurrence make it important to formulate an early diagnosis so as to provide appropriate treatment and follow-up the patient over the long-term.

**References**