Juvenile Ossifying Fibroma in the Mandible

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Abstract

Juvenile ossifying fibroma is a rare benign fibroosseous tumor which involves maxilla more than mandible. It usually occurs in children below 15 years of age. This article reports a case occurring in the mandible with the surgical treatment followed by us.

Keywords: Fibroosseous tumor, juvenile ossifying fibroma, pediatric tumor, rare tumors

Introduction

Juvenile ossifying fibroma (JOF) is a rare benign fibro-osseous neoplasm which arises in craniofacial bones of children under 15 years of age. They occur more commonly in maxilla and very rarely in mandible. The tumor grows asymptomatically achieving a large size suggestive of its aggressive behavior giving suspicion of malignancy. The lesion has high recurrence rate. Although the tumor is uncapsulated, it is well demarcated from the surrounding bone. Histopathologically, two types have been identified: the psammomatoid and trabecular types.

Literature names it "JOF," "aggressive ossifying fibroma," or "active ossifying fibroma." It has a higher rate of occurrence in the maxilla involving the maxillary sinus, nasal cavity pushing the globe superiorly. Extension to cranial base is reported. Tumor rarely involves mandible, and only few cases have been reported. Surgery with wide excision is the suggested treatment. This paper reports JOF in a young boy in the mandible treated surgically with a long-term follow-up.

Case Report

An 8-year-old child reported to us with a complaint of a painless growth over the right side of lower jaw since 6 months. The growth was rapidly increasing in size. On examination, the swelling was well defined, extending from the right mandibular first molar to the left mandibular first molar with buccal and lingual cortical expansion and inferiorly the lesion involved the lower border of the mandible. Size of the lesion was around 7 cm. On palpation, the swelling was painless hard in consistency. The mucosa overlying was stretched and ulcerated due to trauma from maxillary teeth. There was no paresthesia over the lower lip or chin. The skin overlying the lesion was free. Lymph nodes were nonpalpable [Figure 1].

Radiograph revealed a mixed radiopaque radiolucent mass over the right body of mandible extending to the left side body. The teeth were malpositioned and pushed by the expansile lesion. Computed tomography represented a heterogeneous radiodense/radiolucent mass expanding the buccal cortical plate. Lingual cortex was unaffected. The inferior border was intact [Figure 2a-c].

The differential diagnosis of JOF or osteosarcoma was made. Biopsy of the lesion gave the diagnosis of JOF.

The patient was operated under general anesthesia with nasoendotracheal intubation. Intraoral incision was given from distal aspect of mandibular right first molar to left first molar. The tumor was exposed and resected with a wide margin. The specimen was sent for frozen section to rule out malignancy and to confirm the clarity of margins. Frozen section reported margins clear by 6–8 mm. A reconstruction plate was contoured and positioned with screws on either side [Figure 3]. The defect was closed in layers with 3/0 and 4/0 vicryl sutures.

Histopathological report revealed bundles of spindle-shaped cells showing large zones of sclerosis. Interspersed amidst are

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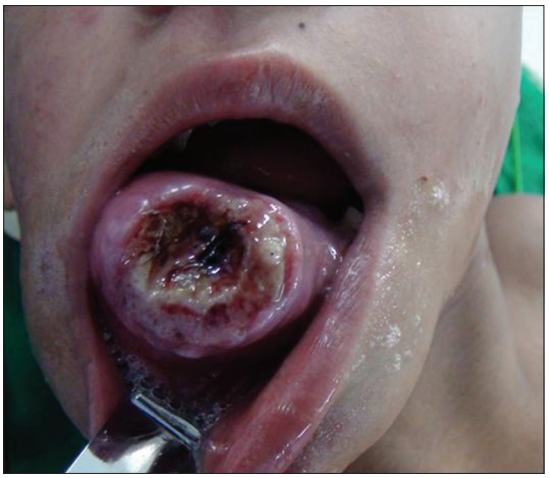

Figure 1: Clinical picture

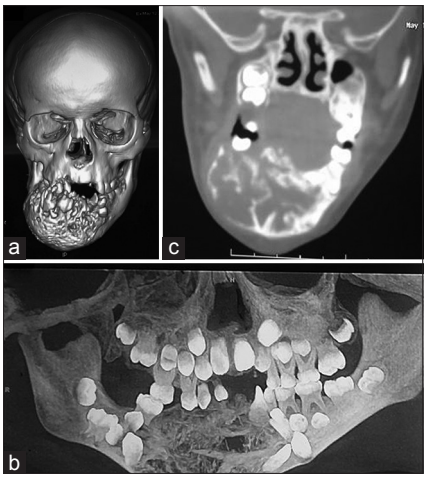

Figure 2: (a) Three dimensional computerized tomography scan of the patient, (b) orthopantomograph, (c) coronal computerized tomography scan

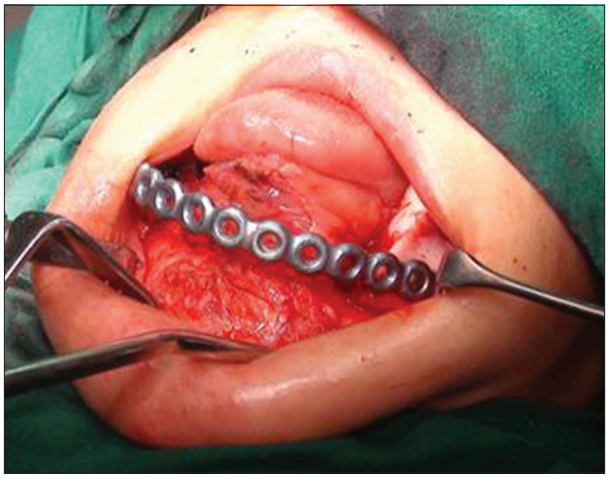

Figure 3: Intraoperative view of tumor resection and reconstruction plate placement

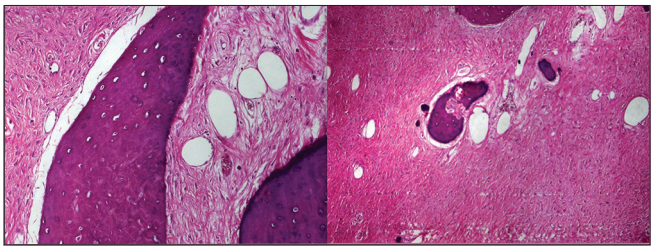

Figure 4: Histopathological slide

few curved bony islands. A large number of these consisted mainly of woven bone devoid of osteoblastic rimming. Only a rare island showed few osteoblasts around the bone. The report confirmed the diagnosis of trabecular variant of JOF [Figure 4].

The patient has been kept under regular follow-up for 3 years. There are no signs of recurrence [Figures 5-7]. The patient will be taken for reconstruction with free fibula flap followed by placement of dental implants.

Discussion

JOF is a fibro-osseous neoplasm described as an actively growing lesion consisting of a cell-rich fibrous stroma, containing bands of cellular osteoid without osteoblastic lining, together with trabeculae of more typical woven bone. It has been thought that JOF arises as a result of differentiation of multipotential precursor cells or mesenchymal cells of the periodontal ligament to form cementum, osteoid or fibrous tissue combination.

Fibro-osseous lesions involving craniofacial skeleton display a variety of clinical behavior. Fibrous dysplasia is diffuse, non-neoplastic proliferation of fibro-osteoid tissue which slows down and gets arrested with age. It shows no respect to anatomical landmarks and demonstrates no clear demarcation. It occurs in two forms; monostotic and polyostotic forms. In monostotic form, single facial bone is involved. It affects the maxilla more commonly than the mandible. It is slow growing, painless swelling causing facial asymmetry. In polyostotic form, multiple craniofacial bones are affected resulting in increased pressure symptoms intracranially, displacing the orbital contents. As a rule, monostotic variety is treated conservatively primarily for cosmetic reason, and polyostotic type is treated by shaving and contouring of facial bones for relieving pressure to the vital organs.

In contrast, ossifying fibroma is a neoplasm seen in young patients having aggressive expansile growth causing severe facial asymmetry, having high rate of recurrence and thus demanding wide resection and long-term follow-up.

Ossifying fibroma was first described by Montgomery in 1927, as a benign fibro-osseous lesion.[1] The term "JOF" was first used by Johnson in 1952 when he was describing aggressive forms of ossifying fibroma as it occurred in the craniofacial bones of children.[2]

JOF is an aggressive expansile variant of ossifying fibroma seen in patients below the age of 15 years. According to Hamner *et al*.[3] and Slootweg *et al*.,[4] the mean age of onset was 11.5 and 11.8 years, respectively. There is no gender

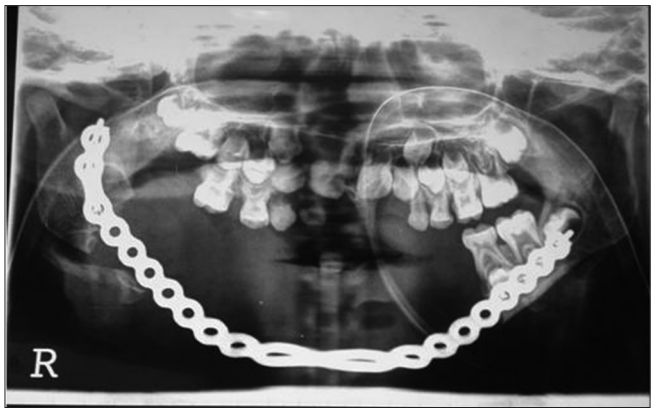

Figure 5: Follow-up orthopantomograph showing plate in position and no signs of recurrence

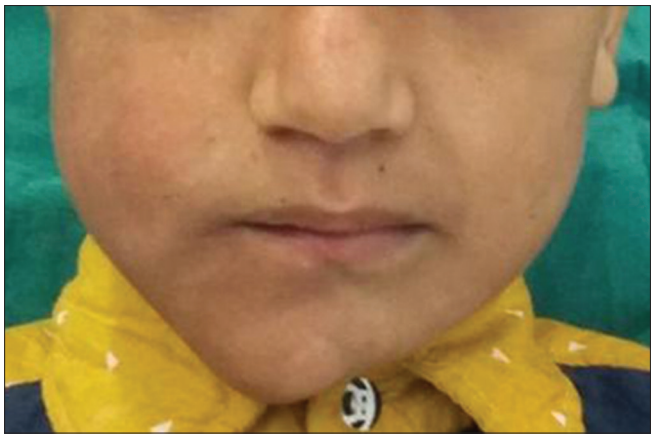

Figure 6: Follow-up clinical photograph

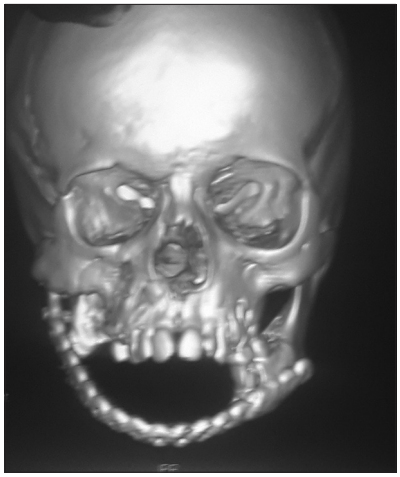

Figure 7: Follow-up three dimensional computerized tomography scan

predilection. Most cases affect sinonasal area and jaws (90%), out of which 10% cases involve mandible.[5]

JOF is rarely encountered, and when seen, it is more common in maxilla involving nasal cavity and orbit disturbing the globe contents causing dystopia. Mandibular involvement is rare as compared to maxilla. Intracranial involvement is reported.

JOF is characterized by its occurrence in the age of 5–15 years, painless but aggressive behavior seen as a rapid growth deforming the face and mimicking malignancy. Paresthesia is not commonly seen. Expansile growth displaces the teeth. The lesion assumes a large size in short time giving a suspicion of malignancy. It can grow to a considerable size and show an aggressive behavior of rapid growth with cortical thinning and perforation.[6]

Radiograph show well-demarcated lesion with area of radiolucency or radiopacity. Amount of calcified tissue produced will show varying degree of radiolucency and radiopacity.[7] Radiographs can show root displacement and resorption though rarely.[8]

Histologically, JOF shows heterogeneous morphology. Areas of dense cellularity may alternate with myxomatous regions. Distribution of bone trabeculae and ossicles are uneven and giant cells are often present. The most recent classification is by El Mofty who identified two categories, trabecular JOF and psammomatoid JOF; based on histologic criteria.[8] The psammomatoid JOF mainly involves the bones of the orbit and paranasal sinuses, whereas the trabecular type commonly involves the jaws.[9] The pathognomonic feature of the psammomatoid type of this fibro-osseous lesion is the presence of eosinophilic spherical structures dispersed in a fibrous stroma consisting of plump spindle-shaped cells that are arranged as strands or whorls; this unique spherical structure is termed as psammoma like bodies. They vary in appearance but usually have a central basophilic area and a peripheral eosinophilic fringe. The variant histopathology does not influence the clinical behavior of the tumor.

Aggressive behavior and high rate of recurrence demands more radical approach in treating these lesions. Curettage has high rate of recurrence; thus, resection of the lesion should be carried out with preservation of vital structures. Reconstruction with bone grafting should be delayed in children to ensure cure and make the donor area available.

The recurrence rate for JOF is very high and ranges from 30% to 58%.[5,10] Consequently, regular follow-up assessments are essential. There is no standardized follow-up protocol in literature due to the rarity of the lesion. Due to the fairly high recurrence rate, immediate reconstruction is not advised. The suggested treatment is wide resection with clear margins on frozen sections, and long-term follow-up of 5 years. The long-term follow-up excludes recurrence and creates adequate donor area for reconstruction and rehabilitation.

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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