

Case Report

Giant juvenile ossifying fibroma from diagnosis to management: A rare case

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ABSTRACT

Juvenile aggressive ossifying fibroma is a rare, benign tumor of the maxillofacial region that is aggressive and recurrent leading to multiple surgeries causing maxillofacial defect and deformities. The management of patients with ossifying fibroma is controversial. The aim was to report a case of huge ossifying fibroma involving maxilla, zygoma, and orbit in a young female with a duration of 15 years. The patient was operated multiple times but the lesion recurred. Patient-specific implant was fabricated to replace the defect area which would be created after surgery but with meticulous intraoperative decision the high morbid surgery was changed to conservative surgery. No recurrence was seen after two year follow up. The patient aesthetic has also improved. In conclusion it can be said in huge benign ossifying fibroma morbid surgery should be avoided.

Key words: Juvenile aggressive ossifying fibroma, maxillofacial, conservative surgery

INTRODUCTION

Ossifying fibroma is a benign fibro-osseous lesion that is divided into subtypes depending on the age of onset as conventional ossifying fibroma and juvenile ossifying fibroma (JOF). Juvenile ossifying fibroma is further subdivided into Juvenile psammomatoid ossifying fibroma (JPOF) and juvenile trabecular ossifying fibroma (JTOF) according to histological characteristics (El-mofty, 2002). Age of onset of juvenile ossifying fibroma is generally younger than 15 years. Unlike conventional ossifying fibroma JOF is characterized by aggressive growth and causing damage to cortical bone, eye nose, and even involving cerebrum. The imaging characteristics of JOF show expansile, well defined, spherical outline with radiolucent to radio-opaque image separated from surrounding normal bone. The management of JOF remains controversial. According to literature recurrence rate after surgery is around 30-58% (Olentino *et al.*, 2013). Treatment can be controversial or radical (Abuzinada and Alyamani, 2010). Radical surgery is advocated for aggressive lesion having high recurrence rate. (El-mofty, 2002; Smith *et al.*, 2009; Han *et al.*, 2016) while others advocate conservative treatment (Leimola-Virtanen *et al.*, 2001; Abuzinada and Alyamani, 2010).

Case presentation: A 23-year-old female patient reported to our department with a chief complaint of facial deformity secondary to painless swelling on the right side of face since last 15 years and epiphora and stuffy nose. No diplopia was present. She was previously operated multiple times in the years 2005, 2006, and 2008 but met with recurrence every time. The swelling was around 10x15 cm in size occupying right side maxilla, zygoma, and even caused orbital dystopia.

Intra orally the swelling resulted in obliteration of buccal vestibule and expansion of buccal and palatal vestibule. (Figures 1 and 2) Computed tomography was performed which shows spherical shaped lesion occupying right side maxilla, zygoma, and invading right side of lateral orbital wall. The lesion was crossing the midline and encroaching the nasal region. The lesion was mixed radiolucent and radio-opaque. (Figure 3) Incisional biopsy of the lesion was performed which was consistent with previous biopsy report of ossifying fibroma. No malignant transformation was present. Due to the huge size of the lesion and multiple recurrences resection of lesion and reconstruction with patient-specific implant was planned. 3 D model of the patient along with customized patient-specific implant was made (Figure 4). The Patient was taken under general anesthesia and intraoral vestibular incision was. The entire mass was meticulously enucleated in multiple fragments. The mass was present in multiple small compartments in the zygoma and orbit region (Figures 5 and 6). Removing lesion in the palatal region created palatal fistula is accidentally due to loss of palatal bone. No reconstruction was required. After 2 years of follow up, no recurrence was noted. Facial deformity, ocular dystopia, and epiphora to a great extent is resolved.

DISCUSSION

JAOF is benign odontogenic pediatric tumor which is characterized by aggressive behavior and high recurrence rate. It has two histologic variants JTOF and JPOF. One is characterized by small uniform spherical ossicles resembling psammoma bodies (psammomatoid juvenile ossifying fibroma). The other is distinguished by trabeculae of fibrillary



Figure 1: Swelling around 10x15 cm in size occupying right side maxilla, zygoma, and orbit

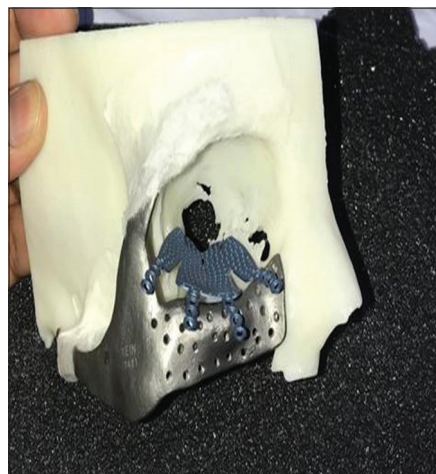


Figure 4: Stereolithographic model and customised patient specific implant



Figure 2: Intra oral digram showing swelling involving palate and buccal vestibule

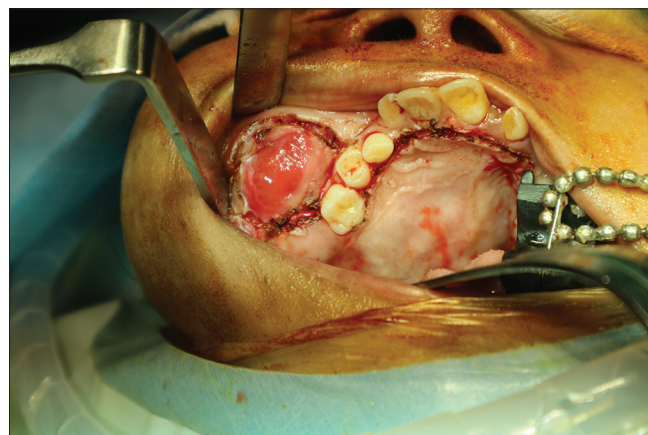


Figure 5: Intra op digram showing tumor mass

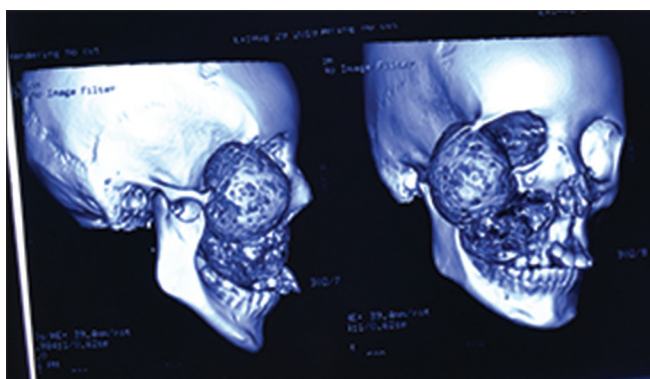


Figure 3: D CT scan showing spherical tumor mass involving maxilla, zygoma and orbit



Figure 6: Intra op digram showing tumor mass

osteoid and woven bone (trabecular juvenile ossifying fibroma).

It differs in age of onset and radiographic characteristics. The age of onset of JPOF is 10 to 25 years (Margo *et al.*, 1985) whereas JTOF patients are relatively younger, at 8.5–12 years (El-mofty, 2002). The age of onset in our case was around 10

years. JOF is seen to be more common in females with a ratio of 2:1. JPOF is seen to be more common in Paranasal sinus (Linhares *et al.*, 2011) while JTOF is more commonly seen in jaws (El-mofty, 2002). In our case also as the tumor was occupying paranasal sinus so the radiographic presentation resembles with JPOF. The radiographic presentation was consistent with the histologic report.

Tumor located in maxilla extensively expands in size involving maxilla, zygoma, it can involve eye resulting in Exophthalmos or can involve nasal cavity resulting in nasal obstruction. JPOF radiographically ground glass in appearance while JTOF is mixed radiopaque radiolucent in appearance. Pathologies that need to be distinguished from JOF using imaging studies include osseous dysplasia, odontoma, fibrous dysplasia, and ameloblastoma. Fibrous dysplasia can resemble ground glass; and has unclear boundaries and generally involve multiple bones. Osseous dysplasia usually presents as calcification in the space between the roots or as a cotton-like calcification involving the bilateral upper and lower jaws. Odontoma and ameloblastoma are also common in younger patients, although odontoma usually presents as a mass with mixed densities around the roots, with clear boundaries and visible non-erupted teeth or shadows of teeth of varying sizes. Ameloblastoma usually presents as multiple radiolucent regions with discrete, corticated borders.

Conventional ossifying fibroma can be treated with surgical excision as it has clear margins. It has rare recurrence and has a very good prognosis. However there is great controversy for the treatment of JOF. Treatment can be conservative or radical and histological subtypes have no significance. As the tumor has high recurrence rate and aggressive behavior so radical surgery removing as much tumor and surrounding involved tissue while preserving as much as important vital structures are advocated (El-mofty, 2002; Smith *et al.*, 2009). Conservative treatment include curettage and Enucleation. The aim is to limit facial deformity, preserve normal appearance, save dentition and preserve growth and development (Leimola-Virtanen *et al.*, 2001; Abuzinada and Alyamani, 2010). As there are different schools of thought for the treatment of aggressive treatment but according to our experience the reason for recurrence is incomplete removal of tumor. If surgery is performed meticulously removing the entire tumor there is no chance of recurrence. The tumor is present in small pockets separated by septae so the surgeon

should be highly vigilant in removing tumor and in case of very large tumor an operative strategy should be made to start from a location and move in a mannered direction. So the key to limit radical surgery and limit resection is meticulous surgery and moving in a mannered way. Although we have planned for resection and reconstruction with patient-specific zygoma implant but this ablative surgery is prevented.

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