

Case Report

JUVENILE OSSIFYING FIBROMA: A CASE REPORT AND LITERATURE REVIEW

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ABSTRACT

Juvenile ossifying fibroma are benign, potentially aggressive bone forming neoplasm. It can be distinguished from other fibro osseous lesion primarily by its age of onset, clinical presentation and aggressive behaviour. JOAF is often confused with malignant variations because of its rapidly progressive and osteolytic nature. A 11 year old male child reported to our centre with painful swelling on the left side of face since 3 months, clinical presentation and investigations were suggestive of juvenile ossifying fibroma of zygoma left side. Hemi maxillectomy was done.

Introduction

Fibro-osseous lesions are a diverse group of processes that are characterized by replacement of normal bone by fibrous tissue containing a newly formed mineralized product. The designation fibro-osseous lesion is not a specific diagnosis and describes only a process. Fibro-osseous lesions of the jaws include developmental (hamartomatous) lesions, reactive or dysplastic processes and neoplasms. The pathologic features may be very similar in lesions of diverse cause, behavior and prognosis. Clinical, radiographic and histopathologic correlation is usually most beneficial in establishing a specific diagnosis.¹ These lesions can be classified as fibrous dysplasia, ossifying fibroma (OF) and cement-osseous dysplasia. The OF

can be further divided into conventional and juvenile forms (JOF).²

The juvenile ossifying fibroma is a controversial fibro-osseous lesion that has been distinguished from the larger group of ossifying fibromas on the basis of the age of the patients, most common sites of involvement, and clinical behavior. The term juvenile (aggressive) ossifying fibroma was used in the second edition of the World Health Organization classification of odontogenic tumors, as a lesion affecting jaws of children under the age of 15 years³. It is a locally aggressive neoplasm with a predilection for paranasal sinuses and can extend into the orbit and anterior cranial base.^{4,5,2}

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Fig 1 FRONT PROFILE



Fig 2 INTRAORAL VIEW



Fig 3 PNS



FIG 4 OPG

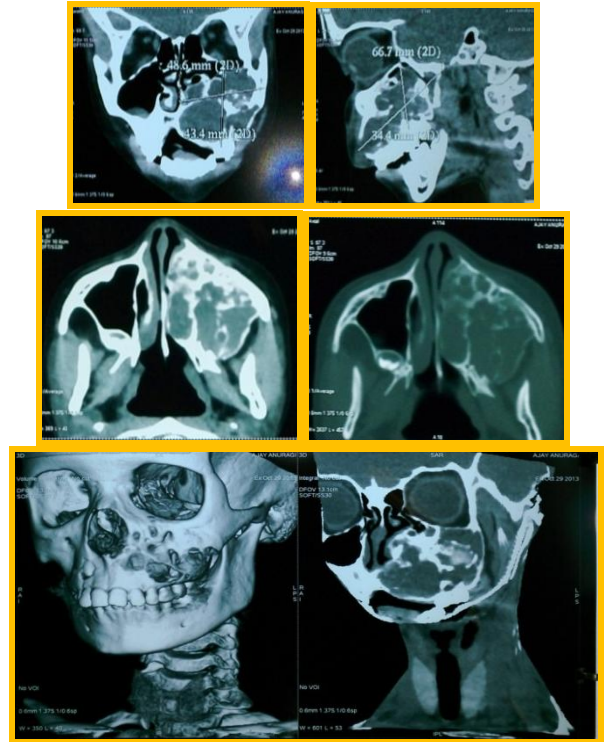


Fig 5 CT SCAN

Histologically, it consists of a cell-rich fibrous stroma, containing bands of cellular osteoid without osteoblastic rimming, together with trabeculae of more typical woven bone. Small foci of giant cells may be present. The lesion is non-encapsulated but well demarcated from surrounding bone.⁶ There recommended treatment is complete surgical excision to avoid recurrence. Reported is a case of massive juvenile ossifying fibroma of the maxilla in an 11 year old child along with a comprehensive review of literature on this entity.

CASE REPORT

An eleven year old systemically healthy male was referred to the Department of Oral and Maxillofacial Surgery with a chief complaint of a painful swelling

on the left side of the face since 3 months. Pain was dull, non-radiating and intermittent. The pain responded to various analgesics taken by the patient for symptomatic relief. The patient informed that a similar swelling in the same region had occurred 4 years ago, the diagnosis of which was histologically confirmed to be dentigerous cyst at that time. The cyst was removed under general anaesthesia and he had remained symptomless until last 3 months.

Detailed extra-oral examination revealed a single, diffuse, firm, tender swelling on the left side of the face extending from left infraorbital region to angle of mouth supero-inferiorly and from angle of mouth till the tragus antero-posteriorly. Overlying skin was normal in colour and surface was smooth with no secondary changes. There were no associated crepitations palpable.



Fig.6A. INCISION



Fig.6B.EXPOSURE



Fig.6C. ENUCLEATION



Fig.6D. SUBTOTAL MAXILLECTOMY



Fig. 7 Closure



Fig8 SPECIMEN

Radiographic evaluation with CT scan revealed an aggressive swelling with erosion of the antero-medial walls of the left maxillary sinus approaching the orbital floor and resorption of the anterior wall of the maxilla. Involvement of the left maxillary sinus as a radiolucent lesion was seen on the orthopantomogram and the paranasal sinus view as well.

FNAC and incisional biopsy were performed for histopathological evaluation. FNAC was inconclusive of the swelling as no fluid or tissue was aspirated. The incisional biopsy revealed characteristics of a fibro-

osseous lesion suggestive of juvenile ossifying fibroma.

The treatment planned for the patient was hemimaxillectomy owing to the aggressive behaviour of the lesion. The modified Weber Ferguson incision including the infraorbital approach for complete exposure of the maxilla was given. Along with hemimaxillectomy involving the affected bone, a wide and adequate margin of healthy bone was also excised. The excision included complete maxillary bone in both vertical and horizontal dimensions excluding the floor of orbit. Finally, layered closure with 3-0 vicryl and 5-0 ethicon was done.

The patient has been on regular follow-up for the past 1.5 years. There have been no signs of recurrence in terms of any post surgical swelling, pain or any other discomfort associated with the region where the excision of the lesion was performed. The intraoral defect has an adequate soft tissue cover. The extraoral incision also healed adequately and with esthetically acceptable scars.

DISCUSSION

Benign fibro-osseous lesions are uncommon, poorly defined and relatively controversial group of lesions affecting the craniofacial skeleton wherein pathological processes replace normal bone by fibroblasts and collagen fibres containing variable amounts of mineralized material. These lesions present histological similarities and therefore a definitive diagnosis requires a precise correlation of clinical, histopathological and imaging findings.

Ossifying fibroma is a type of fibro-osseous lesion of the jaws arising from the undifferentiated cells of the periodontal ligament affecting the tooth-bearing areas especially premolars and molars.⁷ It occurs mostly in adults. Its relatively more aggressive variant

predominant in children below 15 years of age has been termed as juvenile ossifying fibroma (JOF).

Initial clinical manifestation of JOF is in the form of a swelling of the maxilla which is usually asymptomatic. When the orbital bone and the paranasal sinuses are involved, signs such as pain along with exophthalmos, bulbar displacement and nasal obstruction appear.⁶ On radiographs, the lesions appear as circumscribed radiolucencies that in some cases contain central radiopacities giving a “ground-glass” or “honey-comb” appearance.⁷

Histologically, the lesions are nonencapsulated but well demarcated from the surrounding bone. The mass consists of cellular fibrous connective tissue that exhibits areas that are loose and other zones that are so cellular that the cytoplasm of individual cells is hard to discern because of nuclear crowding. Myxomatous foci are not rare and often are associated with pseudocystic degeneration. Mitotic figures can be found but are not numerous. Areas of hemorrhage and small clusters of multi nucleated giant cells are usually seen.⁶

According to the WHO classification of odontogenic tumors 2005, JOF is further subdivided into juvenile psammomatoid ossifying fibroma (JPOF) and juvenile trabecular ossifying fibroma (JTOF). The trabecular type is seen more commonly in children whereas the psammomatoid type has been reported in relatively adult age groups as well. The basic difference between the two types lies in the pattern of calcification of the masses of osteoid dispersed in fibrous stroma. JPOF presents with concentric calcifications imparting a psammoma body-like appearance to the spherical masses. However, JTOF shows calcification progressing into the osteoid strands, producing woven

bone trabeculae that are rimmed with osteoblasts and incorporated osteocytes.⁴

Differential diagnosis can be established with cemento-ossifying fibroma (clinically predominant in adults and predilection for mandible, histological variations), osteo-fibrous dysplasia and fibrous dysplasia (radiographically diffuse margins as compared to clearly defined cortical margin in JOF).⁸

The treatment of such lesions is controversial and includes conservative surgery such as enucleation and curettage (risk of recurrence); aggressive management (preferred due to low recurrence rate) with enblock resection and wide excision of the lesion. This also includes reconstruction of the lost soft and hard tissues following excision of the primary lesion.

Important factors that determine the type of excision planned for the lesion are age of the patient, the rate of growth, vital structures involved or at risk, aggressiveness in terms of resorption of the adjacent bone.

Since JOF in patients below 15 years the need for reconstruction is also greatly increased. The child face keeps growing until the age of 16 – 18 years on an average. Hence, many authors prefer the late reconstruction of the defect in growing age group irrespective of the lesion being recurrent or non recurrent in behavior.

In cases where the lesion is small in dimension along with no involvement of the adjacent bone, curettage and enucleation of the lesion can be chosen as a better treatment option. There are also case reports of adequate healing and regeneration of the bone even in cases of juvenile ossifying fibroma. The theory postulated for such regenerate formed is the preservation of the periosteum. However, the recurrence rate of such treatment ranges between 30%

to 58 % owing to which some authors prefer the treatment of this lesion with extensive surgery that is mainly en-bloc or wide excision with adequate margins especially in cases where adjacent structures are also involved. G Sun et al⁶ have discussed the occurrence of the lesion to be more aggressive in cases that involve the maxilla and hence en block resection is the preferred treatment option.

Due to the aggressive nature of the lesion as well as its predilection in growing children, Colmenero et al¹⁰ suggested a two stage treatment wherein, the first stage involves wide and complete excision of the lesion. Following this, regular monitoring for recurrence is done and reconstruction is planned as the second stage procedure after the growth of the child is complete.

CONCLUSION

Juvenile ossifying fibroma is a locally aggressive lesion with high tendency to resorb adjacent bony structures and recur even after following a conservative treatment of the pathology. The pathology hence is treated more aggressively and preferably reconstructed at a later stage. The present case was treated aggressively with wide margins taken for excision and is still on follow up for past 1.5 years. The patient is awaited to reach the age of 16 years when the growth will almost be complete following which reconstruction will be carried out as the second stage procedure.

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