Juvenile Ossifying Fibroma of Mandible: A Case Report

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Abstract Fibro osseous lesions are benign mesenchymal skeletal tumours in which mineralized tissue, blood vessels and giant cells, in varied proportions replace normal bone. Included in this group are fibrous dysplasia (FD), cherubism, ossifying fibroma and osteoblastoma. Although fibro osseous lesions have similar histologic and radiographic features, they may exhibit a wide range of biological behaviours. Because the histologic appearance does not predict the rate of growth or prognosis, treatment is based on the clinical and biologic behaviour of the tumour. The term “Juvenile active ossifying fibroma”(JAOF) has been given to a less common, more aggressive variant of the central ossifying fibroma which occurs primarily in children and young adults. This lesion has been described in literature under variety of terms like “aggressive psammomatoid ossifying fibroma” or “juvenile ossifying fibroma”. We are presenting a case of Juvenile aggressive ossifying fibroma in a 17 years old girl who reported to our institute.

Keywords Ossifying fibroma · Mandible · Juvenile aggressive

Introduction

Ossifying fibroma of the jaw is a benign, fibro osseous lesion first described in 1927 by Montgomery [1]. Fibrous dysplasia (FD) and ossifying fibroma are similar and differentiation between the two may be difficult. They were considered as

one entity by most authors until 1948 [1]. In general, ossifying fibroma is a more circumscribed lesion than FD and is usually surrounded by a fibrous capsule [2]. Ossifying fibroma may exhibit a sclerotic margin on plain radiographs and CT [3]. Microscopically, Ossifying Fibroma is characterized by prominent osteoblasts, scant osteoclasts and relatively prominent stromal cellularity. On the contrary, osteoblasts and osteoclasts are scarce and the stroma presents low to moderate cellularity in fibrous dysplasia [4].

Ossifying fibroma is most commonly seen in the third and fourth decades of life. Juvenile ossifying fibroma, however is a variant that occurs in children usually younger than age 15. This article focuses on juvenile ossifying fibroma, also known as “aggressive”, “active” or “psammomatoid” ossifying fibroma [5].

A lesion with a characteristics of a juvenile ossifying fibroma was first reported by Benjamins in 1938. This lesion was located in the frontal sinus [1]. The term “Juvenile Ossifying fibroma” was first used by Johnson in 1952 to describe aggressive forms of ossifying fibroma that occurred in the craniofacial bones of children [1]. Kramer and Pindborg used the same term in the classification of odontogenic tumours in 1992 (world health organization) [16]. Juvenile ossifying fibroma is defined as a fibro osseous lesion that is characterized by cell-rich fibrous tissue, bands of cellular osteoid trabeculae and giant cells. It presents in children younger than 15 years of age, behaves aggressively, and tends to recur [6, 7].

In most of the cases, juvenile ossifying fibroma involves the paranasal sinuses, the orbit, the fronto-ethmoid bones and the maxilla [12]. Few cases of mandibular Juvenile Ossifying Fibroma have been reported [6, 9–12]. (See Table 1).

Juvenile Ossifying Fibroma sometimes develops in areas of congenitally missing teeth [12]. The reported

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Table 1 Reported cases of Juvenile active ossifying fibroma in the mandible

Authors	Year	No. of patients	Mean age	M/F
Test et al. [13]	1976	1	?	?
Reaume et al. [9]	1985	1	10	M
Scharffetter et al. [18]	1985	1	13	M
Slootweg and muller [10]	1990	1	9	F
Leimola-Virtanen et al. [11]	2001	1	10	M
	2001	1	8	F
Zama et al. [6]	2004	1	11	M
Rinaggro et al. [12]	2003	1	14	M
Dias et al. [13]	2008	1	12	F

recurrence rate for juvenile ossifying fibroma ranges from 30 to 58% [13, 14, 15]. Local recurrence probably is caused by incomplete removal of tumour, particularly in the maxillary sinus and orbital region.

Diagnosis

Clinically, Juvenile Ossifying Fibroma may present as an asymptomatic, painless expansion of the affected bone that leads to facial asymmetry. The tumour can grow to a considerable size and may behave as an aggressive lesion that destroys bone. Sometimes Juvenile Ossifying Fibroma exhibits rapid growth. Pain and paresthesia are rarely manifested. Nasal obstruction, epistaxis, exophthalmos and rarely, intracranial extension can be associated with lesions that arise in the paranasal sinuses, orbit and maxilla. Age range is from 2 to 15 years, and a slight male predominance is observed [7, 8].

Radiographically, juvenile ossifying fibroma appears as a unilocular or multilocular radiolucency with well defined borders and occasional central opacification. Aggressive lesions may show cortical thinning and perforation. A demarcating line between neoplasm and surrounding healthy bone tissue may be present [6]. CT findings of Juvenile Ossifying Fibroma may include well defined borders identified by thin sclerotic shell. Cortical disruption and involvement of adjacent anatomic structures also may be present. The lesion has predominantly soft tissue consistency with a variable amount of internal calcification or bone.

Radiolucent areas may also be noted. CT scan with intravenous contrast may show diffuse enhancement of the lesion. Juvenile ossifying fibroma appears to be more invasive and destructive on CT scans when compared to fibrous dysplasia and adult ossifying fibroma [7].

There is an intermediate to low signal intensity on MRI and lesion enhances after gadolinium contrast injection.

MRI offers greater specificity when there is neurovascular and ocular involvement [10]. Accurate diagnosis of juvenile ossifying fibroma is made by correlating the clinical, imaging and histopathologic findings. Clinical and biologic behaviour governs the treatment of this tumour.

Report of a Case

A 17-year-old girl came to our hospital with a chief complaint of a bony hard painless swelling on the lower left side of the face since 2–3 months. On clinical examination, there was a bony hard expansion of the lingual and buccal cortex in the lower left premolar–molar region. The swelling was well defined measuring 4 cm × 2 cm from left canine to first molar region. The overlying mucosa was normal in colour and consistency and it was non tender to palpation and no pulsations were felt. No draining sinuses or fistulae were observed and all the teeth associated were vital, non tender and in good condition.

There was no complaint of paresthesia in mental region and no lymphadenopathy was detected.

The Orthopantomograph revealed a large, single, well corticated, oval shaped, unilocular radiolucency in left mandible extending from central incisor to mesial root of the second molar measuring 4 cm × 4 cm. There was ballooning of the buccal and lingual cortex and a bowing out of lower border. The roots of canine and premolars seemed to be hanging in the radiolucency, but no resorption of roots or displacement of teeth was observed (Fig. 1).

A CT scan of 2 mm sections in sagittal and coronal plane was taken which confirmed the above findings and also showed the radiopaque specks lying in the lesion. (Figs. 2, 3).

Hematological examination showed nothing unusual. Aspiration biopsy of the lesion was negative therefore hemangioma and cyst were ruled out. A differential diagnosis of central giant cell reparative granuloma, giant cell tumour, osteoma, osteoblastoma, unicystic ameloblastoma,

Fig. 1 OPG showing the radiolucency from 31 to 37

Fig. 2 Sagittal section

Fig. 3 Coronal section

monostotic fibrous dysplasia was made. Brown's tumour was ruled out as no hypocalcemia was detected. The patient underwent an incisional biopsy (Fig. 4) under local anesthesia from the buccal vestibule which was reported as Juvenile Ossifying Fibroma.

Microscopically, the specimen consisted of non capsulated psammomatoid patterns of concentric, lamellated and spherical ossicles that vary in shape, lying in cellular connective tissue that exhibits areas that are loose and other zones that are so cellular that the cytoplasm of individual cells is hard to see because of nuclear crowding.

Surgical Technique

Complete enucleation and curettage of tumour was planned after intentional root canal treatment of central incisors to

Fig. 4 Pre-op intraoral photograph

first molar. The enucleation was done through an intraoral crevicular incision extending from central incisor to second molar under naso-endotracheal intubation. The inferior alveolar nerve was found displaced inferiorly and was preserved. The surrounding bone was thoroughly curetted and primary closure was done without any bone grafting. Patient was put on iv injection Taxim 1 g 12 hourly, iv injection Metronidazole 100 cc 8 hourly and iv injection Dynapar aq 8 hourly. The healing was uneventful and sutures were removed on seventh post op day.

The patient has been followed up for last 6 months and there are no symptoms and a gradual remodelling of the mandible has taken place and the facial asymmetry has reduced. Follow up OPG has shown that the radiolucency has reduced in size and a good regeneration of bone has taken place (Fig. 5).

Discussion

Non aggressive forms of Juvenile Ossifying Fibroma are treated by a conservative operation that includes local

Fig. 5 Sixth month Post-op OPG

excision or curettage [17]. Recurrence of Juvenile Ossifying Fibroma also may be managed by local surgical excision. However, aggressive juvenile ossifying fibroma with rapid growth rate, cortical thinning or perforation, tooth displacement and root resorption may exhibit early recurrence unless treated by enbloc resection. The skeletal defect is reconstructed with bone graft once tumour margins are verified [10]. Troulis and colleagues [18] suggested a staged protocol for jaw tumours in children. Stage 1 includes the resection and placement of a rigid internal reconstruction plate. Stage 2 includes skeletal reconstruction with an autogenous bone graft (4–9 months after stage 1). Stage 3 consists of osseointegrated implant placement (5–12 months after stage 2) when possible. Stage 4 consists of prosthetic dental reconstruction (5–7 months after stage 3).

But in our case, the patient's parents were too poor to afford such a long drawn out treatment plan and so we had to manage the lesion with enucleation and curettage as we had adequate access and visibility to the tumour site. Our patient was older than the reported age group, but was malnourished. She reported aggressive growth in the last 3 months. The diagnosis of Juvenile Ossifying Fibroma was arrived at because of the histological report and the clinical behavior of the lesion. We had good access to the lesion and could ensure thorough curettage of the surrounding bone so that the lesion could be removed in toto. We have a follow up of 6 months during which bone regeneration is clearly visible on the radiograph. On studying the literature, we have realized that higher recurrence rates are seen in lesions of maxilla or orbit, which have less access and more vital structures to be taken care of, because of which it is possible that inadequate removal of the tumour takes place.

In fact, for children with aggressive Juvenile Ossifying Fibroma in maxilla, orbit or paranasal sinuses, Kaban and colleagues [18] have suggested thorough curettage or enucleation in combination with adjuvant interferon alpha therapy for 1 year. This protocol is based on the rapid growth, vascularity and bone fibroblast growth factor (a vascular proliferative marker) production of these tumours in children.

Conclusion

The approach to the management of Aggressive Ossifying Fibroma is continually evolving. Complete surgical excision should be the goal, taking into consideration the size, location and extent of the tumour. Small, accessible mandibular tumours may be amenable to surgical excision with

enucleation or with peripheral osteotomy without continuity defects. Larger or recurrent lesions especially in the maxilla need enbloc resection. Our patient shows no signs of recurrence and adequate bone regeneration radiographically in the follow up period of 6 months. We would like to conclude that the benefits of conservative surgical treatment are normal growth, mastication, esthetics and neural function in the mandible are preserved.

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