Case Report – Cyst & Tumors

Aggressive Ossifying Fibroma of the Maxilla

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Abstract

The aggressive ossifying fibroma is an uncommon benign fibro-osseous lesion which has been described in the literature under a variety of terms. This tumor is distinguished from standard ossifying fibroma based on its more clinically aggressive biological behavior, occurrence in children and young adults, and tendency to occur in different anatomic sites. We report a case of a 45-year-old female who presented with a unilateral swelling of the right middle face for 5 months. Clinical examination showed a mass extending over the right maxilla. Orthopantomogram and computed tomography scan were performed. Biopsy suggests a fibro-osseous lesion. The complete surgical excision of tumor was performed under local anesthesia. The histopathological examination revealed the diagnosis of an aggressive ossifying fibroma-trabeculae type. No recurrence was noted. Because of its aggressive and compressive nature, aggressive ossifying fibroma requires an early complete surgical excision. A long-term clinical and radiological surveillance is necessary to prevent recurrence.

Keywords: Aggressive ossifying fibroma, fibro-osseous tumor, maxilla, trabeculae variant

INTRODUCTION

The 4th Edition of the World Health Organization (WHO) Classification of Odontogenic and Maxillofacial Bone Tumours 2017 included ossifying fibroma in the classification of odontogenic tumors. It was however discussed under “bone-related lesions” in 2005. The ossifying fibroma occurs in the jaws, perhaps exclusively in the jaws, which is neoplastic and histologically distinct from juvenile trabecular or juvenile psammomatoid ossifying fibromas (JPOFs). The tumor is of periodontal ligament origin and therefore odontogenic. The ossifying fibromas, including the cemento-osseous fibroma, juvenile trabecular ossifying fibroma (JTOF), and JPOF, have similar and overlapping fibro-osseous appearance histologically.[1]

Ossifying fibroma is characterized by fibrous connective tissue matrix with varying amount of osteoid, immature, and mature bone. The ossifying fibroma is subdivided into conventional and juvenile or aggressive clinicopathologic subtypes.

Aggressive or juvenile ossifying fibroma (JOF), a rare tumor entity occurs predominantly but not exclusively in children, with no gender predilection. It is usually benign and locally aggressive. The lesion is located mainly (85%) in facial bones, in some cases (12%) in calvarium and very seldom (3%) extracranially.[2] Ninety percent of the lesions located in the face region, involve the sinuses, mainly the maxillary antrum.[2] Mandibular lesions are seen in 10% of the cases.[2] It usually expands aggressively with cortical disruption and involvement of adjacent anatomical structures. Depending on the site of occurrence, symptoms such as pain, paresthesia, malocclusion, sinusitis, and proptosis can occur. Radiographically, lesion appears as radiopaque, radiolucent, or mixed radiopaque-radiolucent with a well-defined sclerotic border. Two forms of JOFs have now been distinguished: Psammomatoid JOF and trabecular JOF.

We report a case of aggressive ossifying fibroma because of its relative rarity, emphasizing its clinical, radiographic, histopathological features, and successful surgical management.

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CASE REPORT

A 45-year-old female patient reported to our outpatient department with the chief complaint of swelling on the right side of the face for 5 months. She stated that the swelling was neither preceded nor followed by trauma, toothache, pain, discomfort, or fever. The swelling was fresh and started suddenly and spontaneously adjacent to the right nostril and had gradually grown to its present size and was asymptomatic. There was no significant medical or family history was found.

On general physical examination, the patient was conscious, cooperative, healthy, moderately built, and well nourished. All vitals were within the normal range. On extraoral examination, facial asymmetry noted with a single diffuse swelling on the right middle face [Figure 1]. The swelling measured approximately 2.5 cm × 3.5 cm in size and extended mediolaterally from right ala of the nose obliterating the nasolabial fold to 4.5 cm in front of the lobule of the right ear and superioinferiorly, extended from the right infraorbital region to corner of the mouth. The skin over the swelling and the surrounding area appeared normal. On palpation, the swelling was well circumscribed, nontender, hard in consistency, and fixed to the underlying bone. There was no evidence of paresthesia.

Intraoral examination revealed a solitary, well-defined swelling obliterating the vestibule and extending from the right maxillary central incisor region (11) to the right maxillary second premolar region (15) posteriorly [Figure 2], associated with the swelling on palatal aspect. The surface was covered by normal mucosa. There was Grade III mobility and nontender on percussion with the associated teeth. Class III glass-ionomer cement restoration noted on 11 and 21. On palpation, the swelling was hard in consistency, nontender, causing the expansion of the buccal cortical plates.

Based on the history and findings of the clinical examination, a provisional diagnosis of a benign odontogenic neoplasm was made. The possibility of a radicular cyst, fibro-osseous lesion and a benign tumor of the bone was considered in the differential diagnosis.

Hematological findings were unremarkable, but there was elevation in serum alkaline phosphatase level.

Radiological investigations included intraoral periapical radiographs, orthopantomogram (OPG), and computed tomography (CT) scan (coronal and axial view). OPG revealed irregular, well-bordered mixed radiolucent-radiopaque lesion in the right maxilla extending from midline to the right molar region. Teeth displacement with root resorption noted (12,13,14). The CT scan showed well-defined soft-tissue solid hypodense lesions with osteolytic destruction involving the alveolar process of maxilla extending to the lower lateral aspect of the nasal cavity [Figures 3 and 4]. Intraorally, the lesion was extending into the buccal mucosa and measure 1.8 cm × 1.3 cm. Irregular erosion of the adjacent bone noted with intact dentin portion of the teeth. No extension to the adjacent right maxillary antrum. No calcification with in the lesion. Skin and subcutaneous tissue overlying the mass was intact [Figure 3].
Under local anesthesia, a cravicular approach was made and surgical excision of the tumour was performed [Figure 5]. The tumor was completely removed followed by extraction of involved teeth (11,12,13,14). Right orbital floor, nasal floor, and palatal mucoperiosteum were preserved. Closure was done with resorbable suture. Postoperative recovery was uneventful [Figure 6]. Regular follow-up was done with every 2 weeks’ interval.

**Microscopic appearance**

Hematoxylin- and eosin-stained section reveals epithelium and connective tissue stroma. Epithelium is stratified squamous parakeratinized in nature. Stroma is highly collagenous with dense collagen of bundles, plump proliferating fibroblasts and lots of areas of calcification resembling trabeculae of bone mostly and with few psammomatoid type of calcifications. Stroma also consists of numerous multinucleated giant cells, blood vessels, and inflammatory cells [Figures 7-9].

**Discussion**

Fibro-osseous lesions of jaws are usually benign, asymptomatic, and tend to grow slowly. However, if the lesion occurs in a younger patient, especially below the age of 15 years, an unusual clinical presentation with apparent aggressive and destructive growth may be expected.[3]

The JOF is a rare fibro-osseous lesion that occurs mainly in the facial bones.[4] It is also called aggressive ossifying fibroma due to its aggressiveness and the high tendency to recur, unlike other fibro-osseous lesions. According to the WHO classification 2017, JOF has been recognized as a separate histopathological entity among the fibro-osseous group of lesions due to its distinct histological features.

JOF affects both males and females equally without any significant gender predilection. However, some researches showed that it is more common among men. In contrast, Johnson et al. stated that mandibular tumors are more frequently common in girls between the age of 5–11 or during the second to fourth decades of life.[5] In the present case report, a 45-year-old female was presented.

JOF commonly seen in the maxilla than in mandible and usually expand aggressively with cortical disruption and involvement of adjacent anatomical structures. Depending on the site of occurrence, symptoms such as pain, paresthesia, malocclusion, sinusitis, and proptosis can occur. In the present case, the patient presented with a solitary, well-defined swelling obliterating the vestibule and extending from the right maxillary central incisor region (11) to the right maxillary second premolar region (15) posteriorly [Figure 2], associated with the swelling on palatal aspect. There was Grade III mobility and nontender on percussion in associated teeth. On palpation, the swelling was hard in consistency and nontender.

The term Juvenile Aggressive Ossifying Fibroma (JAOF) describes two distinct histopathological entities of ossifying fibromas known as JPOF and JTOF as identified by El-Mofty in his study.[6]

JPOF has been reported under many names, including osteoid fibroma with atypical ossification” of the frontal sinus by Benjamins in 1938, psammomatoid fibroma of the nose and paranasal sinuses by Gogl in 1949, juvenile active ossifying fibroma by Johnson et al. in 1952, psammomatous desmo-osteoblastoma by Makek and juvenile psammomatoid cement-ossifying fibroma which was used in the 2005 WHO classification of odontogenic tumors and reclassified as a separate entity as JPOF under fibro-osseous lesion in the 2017 WHO classification of odontogenic tumors.[1] JPOF is commonly seen in children and young adults with a mean age 16–33 years and shows male predilection. JPOF commonly involves paranasal sinuses, orbit, and fronito-ethmoidal complex.

In 1965, Reed and Hagy suggested JTOF under the designation of JAOF,[4] Makek, in 1983,[7] published the largest series and he named these tumors as trabecular desmo-osteoblastoma.

![Computed tomography scan (coronal cut)](image-url)
Subsequently, the tumor was named as JTOF in the 2017 WHO classification of odontogenic tumours. JTOF commonly affects maxilla than mandible. It is commonly seen in young individuals with a mean age 8–12 years and shows slight male predilection. JTOF is usually confused with osteosarcoma due to rapid growth and with fibrous dysplasia due to similar demographical and histopathological features.

According to the literature plane radiography, CT, magnetic resonant imaging can be used, but they are neither specific to JAOF nor diagnostic. The radiological features are variable depending on the site and the extent of the ossification. It can be radiolucent, mixed, or radiopaque, depending on the degree of calcification and extent of the cystic changes. Root displacement is common and resorption, though rare, can occur. In the present case, OPG revealed irregular, well-bordered mixed radiolucent-radiopaque lesion in the right maxilla extending from midline to the right molar region. Teeth displacement with root resorption noted (12,13,14). On CT JAOF appears as an expansive but circumscribed lesion surrounded by a thick shell of bone and internal content of varying density, while in our case, the CT scan showed a well-defined soft-tissue solid hypodense...
lesions with osteolytic destruction involving alveolar process
of the maxilla extending to lower lateral aspect of nasal cavity
[Figures 3 and 4].

Histopathologically, JOF appears in two distinct forms,
psammomatoid and trabecular types. In our case, connective
tissue stroma was highly collagenous with dense collagen
bundles, plump proliferating fibroblasts, and areas of
calcification resembling trabeculae of bone with few
psammomatoid types of calcifications. Correlating with
clinical findings with histological features was suggestive of
trabeculated type, aggressive ossifying fibroma.

There is no definitive treatment of JOF. Radical resection,
local excision conservatively or enucleation with curettage
are among the treatment alternatives. Slootweg and Müller
suggested that there were no differences between the
cases that have limited surgical treatment and those with
major surgery in terms of results, and they recommended
conservative surgery.[7] On the other hand, Waldron et al.
suggested that local excision and curettage should be a more
preferable method and added that local surgical excision
can be applied for recurrent tumor treatment. However,
rate of recurrence after conservative treatment was
reported in 30%–58% of cases. Incomplete resection causes
recurrence in aggressive tumors. Therefore, some authors
were recommended en block resection as an adequate
treatment. Curettage together with peripheral osteotomy
is suggested in prevalent or recurrent cases. Sarcomatous
degeneration is reported to develop in lesions that have
recurrence in long-term. However, in the present case,
conservative treatment done includes complete surgical
excision with curettage. No paresthesia and esthetically
significant changes noted postoperatively. There was no
recurrence or complication during 6 months of follow-up
period [Figure 6].

**Conclusion**

JOF is a rare fibroosseous entity characterized by its aggressive
nature and high recurrence rate. Hence, it is essential to make
an early diagnosis, provide the appropriate treatment and
especially the long-term follow-up to prevent recurrence.

**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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