Juvenile Ossifying Fibroma Masquerading As Maxillary Sinus Malignancy: Case Report

Maksiller Sinüs Malignensisi Gibi Görünen Juvenil Ossifiye Fibrom

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Geliş Tarihi/*Received:* 23.10.2009
Kabul Tarihi/*Accepted:* 23.11.2009

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ABSTRACT Juvenile ossifying fibroma (JOF) is a rare locally aggressive fibro-osseous neoplasm that arises within the craniofacial bones, predominantly in younger individuals under 15 years of age. With the unique feature of local aggressiveness, JOF is considered to be a well-defined clinical and histological entity that has been separated from other fibro-osseous lesions including cemento-ossifying fibroma. As the aggressive clinical presentation mimicks malignancy, complete investigations play a vital role in accurate diagnosis. The term JOF is used in the literature to describe two distinct histopathological variants of ossifying fibroma and they are referred as psammomatoid and trabecular JOF (Ps JOF and Tr JOF). Reported here is a case of JOF of maxilla in a 36-year- old male patient which clinically masqueraded as maxillary sinus malignancy.

Key Words: Jaw; maxilla; fibroma, ossifying

ÖZET Juvenil ossifiye fibrom (JOF), çoğunlukla 15 yaşın altındaki gençlerde kraniyofasiyal kemiklerde ortaya çıkan, nadir lokal bir agresif fibro-osseöz tümördür. JOF, kendine özgü lokal agresif özelliği ile; iyi sınırlanmış klinik ve histolojik bir antite olup; semento-ossifiye fibrom dahil diğer fibro-osseöz lezyonlardan ayrılır. Agresif klinik prezentasyonu malignensiyi düşündürdüğünden, kesin tanı için tam bir inceleme yapmak gereklidir. Literatürde JOF terimi ossifiye fibromanın 2 farklı histopatolojik varyantını tanımlamak için kullanılır, ki bunlar psammomatois (Ps JOF) ve trabeküler (Tr JOF) olarak bilinir. Bu makalede, klinik olarak maksiller sinüs malignensisi düşünülen ancak maksillasında JOF tespit edilen, 36 yaşında erkek bir hasta sunulmuştur.

Anahtar Kelimeler: Çene, maksilla, fibrom, kemikleşme

Turkiye Klinikleri J Dental Sci 2010;16(3):305-10

Juvenile ossifying fibroma (JOF) is a rare fibro-osseous neoplasm of the craniofacial skeleton occurring predominantly-but not exclusively-in children.[1] The term JOF is used in the literature to describe two distinct histological subtypes, that is, psammomatoid juvenile ossifying fibroma (PsJOF) and trabecular juvenile ossifying fibroma (TrJOF). PsJOF is characterized by a fibroblastic stroma containing small ossicles resembling psammoma bodies. TrJOF consists of cell-rich fibrous tissue containing bands of cellular osteoid without osteoblastic rimming together with slender trabeculae of immature bone.[2]

The term psammomatoid ossifying Fibroma was originally used by Gögl in 1949.[3] Similar lesion was later termed as juvenile active ossifying fibro-

ma by Johnson et al[4], in 1952. In a monograph on fibro-osseous lesions of the craniofacial skeleton published in 1983, Makek[5] reviewed 69 cases of PsJOF from the world literature and added 17 new cases of his own from the files of Zurich University Hospital for a total of 86 cases. The author used the term psammous desmo-osteoblastoma for describing PsJOF cases. The latter was called trabecular desmo-osteoblastoma.

The term Juvenile Ossifying Fibroma was used in the second edition of WHO histological typing of odontogenic tumours as an actively growing lesion consisting of a cell-rich fibrous stroma, containing bands of cellular osteoid, trabeculae of more typical woven bone and aggregates of giant cells.[6] The third edition of WHO classification of odontogenic tumors has mentioned PsJOF and TrJOF as histologic variants of ossifying fibroma.[2]

JOF has been considered as a distinct disease entity from conventional ossifying fibroma (OF) and the other fibro-osseous lesions because of its tendency to occur at a young age and its locally aggressive behavior.[7] The lesion is non-encapsulated but well demarcated from surrounding bone.[6, 8] It is usually asymptomatic, achieving a larger size and exhibiting aggressive behaviour, and is often mentioned as "Aggressive ossifying fibroma" or "Active ossifying fibroma" in the literature.[9]

Owing to this lesion's aggressive nature and high recurrence rate, early detection and complete surgical excision is considered essential. Here is reported a case of JOF of maxilla in a 36-year- old male patient which clinically masqueraded as maxillary sinus malignancy.

CASE REPORT

A 36-year-old male patient admitted to the department of Oral Medicine and Radiology, Rajah Muthiah Dental College and Hospital, with a complaint of swelling on the left side of the face for 3 months. Patient related history of gradual increase in size of the swelling, unilateral nasal stuffiness in the left side and there was no paresthesia associated with the swelling. Family and medical history was otherwise normal and he was a smoker since 15 years.

Extraoral examination revealed facial asymmetry related to diffuse swelling on the left side of face localized to the maxilla. (Figure 1).On inspection, the swelling is single, oval in shape, measuring about 4x5cm in diameter extending superiorly to the level of infraorbital rim, inferiorly to the level of left commissure, medially involving the left ala of nose and laterally to malar prominence. On palpation, the swelling was non-tender, firm in consistency, immobile with normal temperature and exhibited no pulsation. Bilateral submandibular lymph nodes were palpable, single, mobile and tender.

Intra-oral examination revealed diffuse swelling with gross expansion of buccal cortical plate with obliteration of vestibule extending anteroposteriorly from left upper canine to second permanent molar (Figure 2). There was minimal expansion of the palatal cortex, as well. On palpation, the swelling was non-tender, firm in consistency and premolars and molars exhibited grade 2 mobility. There was no pus discharge, pulsation or any surface colour change.

Considering the duration, aggressive nature, gross expansion with destruction of bony cortex associated with tooth mobility and nasal-obstruction, the case was provisionally diagnosed as malignancy associated with the maxillary sinus. The differential diagnosis considered were adenoid cystic carcinoma, maxillary sinus carcinoma, os-

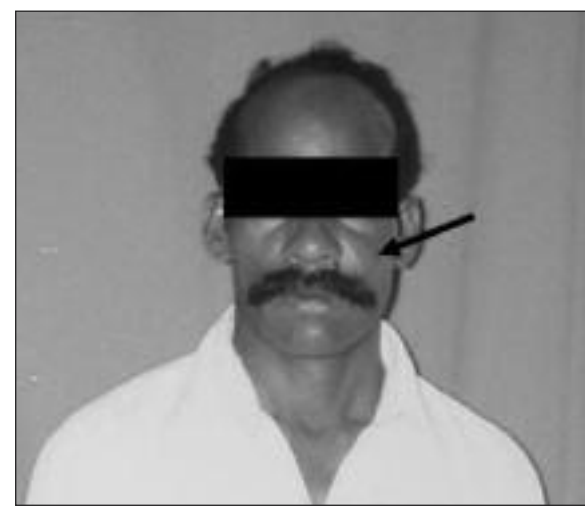


FIGURE 1: Extraoral swelling on left maxilla (arrow).

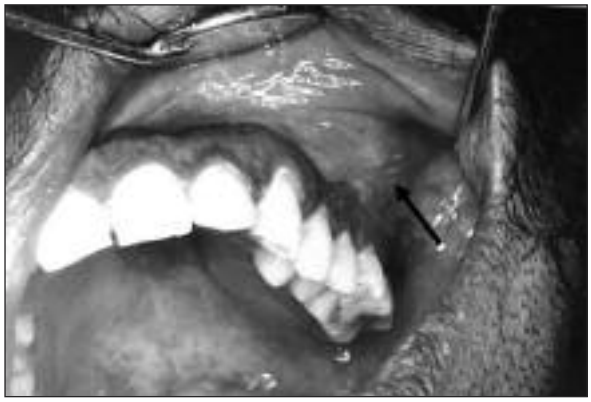


FIGURE 2: Gross expansion of the left maxilla and alveolar process (arrows).

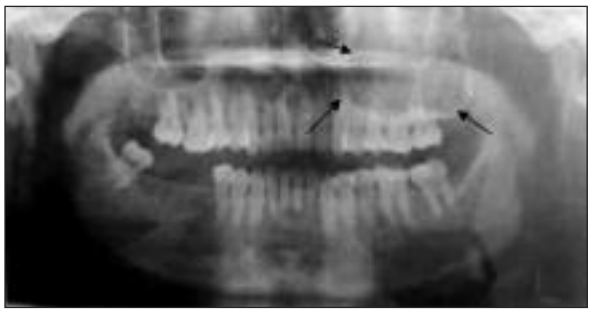


FIGURE 3: OPG reveals well defined radiolucent lesion involving the left maxilla with associated tooth root resorption (arrows).

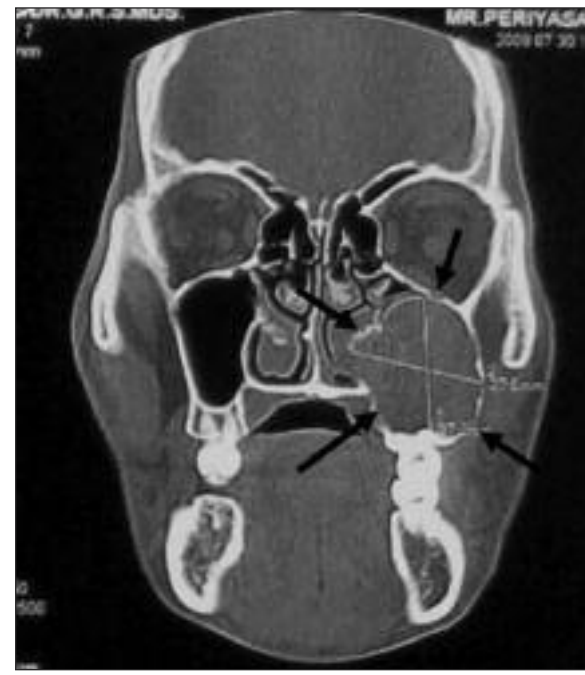
teosarcoma of maxilla, aggressive variety of fibro-osseous lesions and central giant cell granuloma.

Plain film radiographs (intraoral periapical, occlusal, panoramic radiograph) revealed, well defined radiolucent lesion involving the left maxilla extending to the maxillary sinus with associated tooth root resorption (Figure 3).

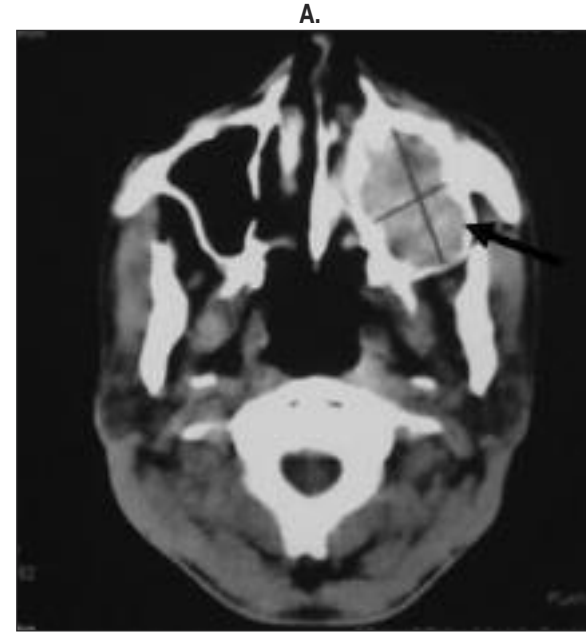
CT scan demonstrated a well-defined, mixed-density mass filling the left maxillary sinus extending up to the orbital floor with perforations. The eyeballs & cranial nerves were not involved. Medially there was destruction of lateral wall of nose with infiltration of mass into the nasal cavity. No evidence of nasal septum deviation was seen. There was destruction of buccal and palatal cortex of the maxilla. (Figure 4 A and B).

Considering a well defined, mixed-density mass associated with cortical perforations and root resorption, the radiographic differential diagnosis included are cemento ossifying fibroma, adenomatoid odontogenic tumour, calcifying epithelial odontogenic tumor and central giant cell granuloma.

A biopsy was performed under local anesthesia and the specimen was sent for histopathological examination. Microscopically, the lesion was non-encapsulated showing infiltration of surro-



A.



B.

FIGURE 4: (A and B)- Computed tomography (CT) showing mixed density mass involving the left maxilla, maxillary sinus extending up to the floor of the orbit and nasal cavity (arrows).

unding bone structures and reactive new bone formation at the periphery. The lesion exhibited highly cellular stromal tissue with numerous spindle fibroblasts arranged in a whorling pattern and small spherical masses of osteoid imparting a psammoma body like appearance (Fig.5 A and B).Most of the cells showed oval to spindle shaped large nuclei. Numerous hematoxyphilic calcified mass of variable sizes are seen throughout the specimen. Many blood vessels and areas of haemorrhage were evident.

On the basis of above microscopic findings, the final diagnosis was Psammomatoid Juvenile ossifying fibroma of the maxilla. Complete surgical excision was performed through a transoral approach and the surgical wound healed satisfactorily.

DISCUSSION

Even though, JOF is seen predominantly in children, it can occur in adults as well.[1, 9-11] But in adults also the adjective 'juvenile' is indicated as age distribution and mean age in 60-80% of cases are between 5 and15 years and only 21% of the patients are aged over 15 years.[6, 9, 12] More over, the diagnosis of juvenile forms of ossifying fibroma is made based on the distinctive histology.[2, 11]

It is believed that JOF arise from cell of odontogenic origin, probably from periodontal ligament or alternatively from a primitive mesenchymal cell nest or from cells remaining after incomplete migration of the medial part of the nasal anlage.[13] Genetic alterations like nonrandom chromosome break points at Xq26 and 2q33, resulting in (X;2) translocation, were identified in some cases of JPOF.[14]

Among the two histologic variants of JOF, Ps JOF is reported more commonly than trabecular JOF.[1] It affects patients with a wide age range, from 3 months to 72 years (Mean age range of 16 y-33 y) compared with Tr JOF which has a age range between 2 to 33 years (Mean age range of 8.5 y-12 y).[1] There is slight male preponderance in both forms. Another significant difference between them is their site of occurrence. PsJOF occurs predominantly in the sinonasal and orbital bones, but TrJOF predominantly affects the jaws with a predilection for the maxilla.[1]

Clinically, JOF shows far more aggressive growth rate than COF.[11, 15] The other differentiating features from conventional COF are occurrence in relatively younger age group, the location of the tumour and tendency to recur after excision.[8] Most cases of maxillary JOF are asymptomatic. The first clinical manifestation is rapid painless expansion of the maxilla. When the orbital bone, nasal cavity and paranasal sinuses are involved, the patients may develop exophthalmos, bulbar displacement, nasal obstruction and epistaxis.[1,8] Due to presence of these signs the condition can clinically mimick malignancy, as with our case.

But JOF is often difficult to diagnose based on the clinical findings alone, so radiographic and his-

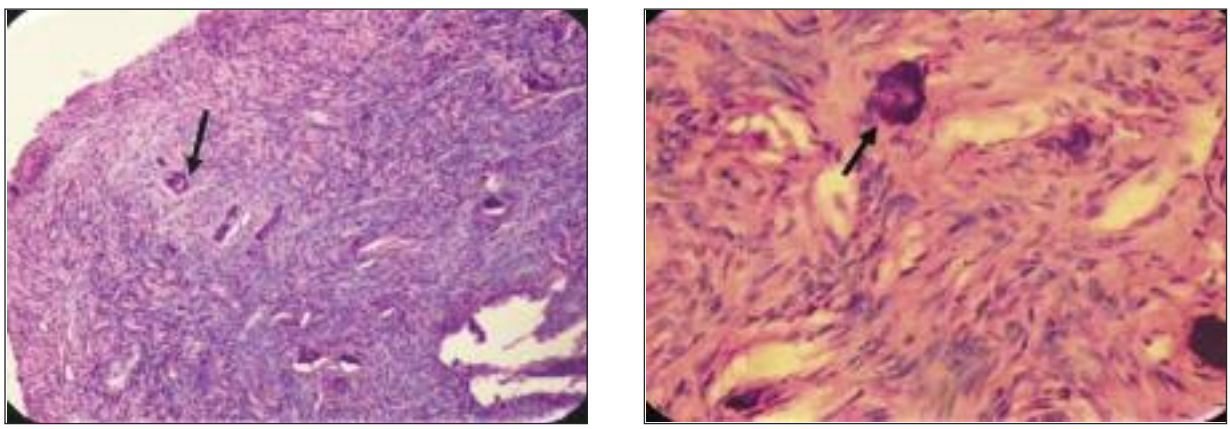


FIGURE 5: (A and B) - Microscopically, the lesion was non-encapsulated showing small spherical masses of osteoid imparting a PSAMMOMA body like appearance (arrows).

tologic investigations plays a vital role in the accurate diagnosis.

The radiologic features are variable and depend on the tumour location, extension and the amount of calcification produced. CT scan is helpful for the further evaluation of the lesion and for detecting subtle calcifications which is not evident in plain film radiography.[16] Radiographically the internal structure can be radiolucent, mixed or radiopaque depending on the degree of calcification. Root resorption, though rare, can occur. Cortical plate expansion is common and can cause perforations as well.[1,15]

Radiographically JOF can resemble OF and Fibrous dysplasia (FD). JOF and OF both displays a well defined, round and oval lesions. But, JOF often shows aggressive signs like cortical perforation and larger size, which can be used to discriminate it from OF. FD usually manifests with ill-defined borders which gradually blends with adjacent normal bone, and exhibit a slower growth pattern.[11, 15]

Other lesions considered in the differential diagnosis are benign lesions like Adenomatoid odontogenic tumor and Calcifying epithelial odontogenic tumor which can present as well defined mixed density radiographic appearance.[17] In case of JOF with ill-defined borders, Osteosarcoma also has to be considered in the differential diagnosis.[18]

Microscopically both the patterns are typically non-encapsulated but well demarcated from the surrounding bone. Ps JOF exhibits spherical or ovoid ossicles resembling psammoma bodies with or without osteoblastic rimming and some ossicles may show calcification with a basophilic center and an eosinophilic fringe with lamellation. The ossicles are surrounded by a thick irregular collagenous rim and occasionally fused together.[1,2,7,19] Haemorrhage is usually not prominent but when present it will be scattered rather than seen as foci.[19] TrJOF shows a mixture of cellular osteoids without osteoblastic rimming, and there are trabeculae of immature bone with osteoblastic rimming. The trabeculae are long and slender, with an anastomosing pattern resembling paintbrush strokes.[1,2,7,19]

Histologically it is difficult to differentiate JOF from other fibroosseous lesions like OF and FD. But the cementum-like deposits seen in OF show a smooth contour with a radiating fringe of collagen fibers, but the ossicles in JPOF has a thick irregular collagenous rim.[1,2,19] Moreover, JOF may show a zonal pattern with central cementum-like ossicles and peripheral mature bony trabeculae but OF seemed to show a mixed, random pattern. In FD osteoblastic rimming in osseous components will be usually absent or scanty.[19] The fibrous components of JOF, OF and FD are relatively similar but in JOF fibrous stroma tends to be extremely cellular, which may be attributed to the aggressiveness of JOF. [7]

The clinical management and prognosis of JOF is somewhat uncertain. The recurrence rate, ranges from 30% observed by Johnson et al[20] to 58% reported by Makek[21]. Although many authors favour conservative surgery rather than radical-en-bloc resection, immediate recurrence characterized by a high aggressive growth rate and the absence of a distinct separation between tumour and the adjacent bone are reported, and requires extensive surgery with wide demolition of the involved bone.[22] This makes enbloc resection with free surgical margins more preferred line of treatment over conservative curettage.[23]

JOF is an uncommon aggressive fibro-osseous neoplasm with non-specific clinical presentations. Proper clinical examination supplemented by appropriate investigations is essential for accurate diagnosis and proposal of the treatment plan. Careful follow-up is required because this condition can be recurrent.

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