

Case report

Maxillary odontogenic myxoma involving the maxillary sinus: A Case report

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ABSTRACT

Odontogenic myxoma(OM) is a rare and locally invasive benign neoplasm found exclusively in the jaws. OM is predominantly found in young adults, and most of the time mandible is involved. OMs are lesions with extremely varied clinical and radiographic characteristic and the diagnosis is therefore not easy. We report an interesting case of OM with extensive involvement of maxillary sinus and upper jaw in a hemophilic patient. A brief review of relevant literature is included.No recurrence was found after one year follow up.

Key words: Odontogenic tumors, Odontogenic myxoma, maxillary Sinus, , case report

INTRODUCTION

Odontogenic myxomas (OMs) are uncommon, accounting for only 3% to 6% of odontogenic tumors [1]. OMs of the jaws are believed to arise from odontogenic ectomesenchyme [1-3]. It appears to originate

from dental pulp or periodontal ligament [3]. Presence of tiny epithelium remnants that resemble inactive odontogenic epithelium and absence of occurrence in other parts of skeleton suggests an odontogenic origin for this tumor [1,4].

The average age in patients with OM is 25 to 30 years [1-3] but recently infant patients with OM have been reported. [5,6] There is 1:1 male to female ratio [2,3]. The tumor may be found in almost any area of the jaws [2,3], and the mandible is involved more commonly than the maxilla [1,2]. This is a slow-growing and invasive benign tumor that can extend, so that a large area of bone may be occupied by this tumor [1,3 and 4]. Larger lesions are often associated with a painless expansion and

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perforation of the involved bone [1-3, 7]. Kaffe et al. found that 74% of OM's show expansion [8]. In maxillary cases, it may invade the maxillary sinus [1]. Kaffe et al. affirmed that entire sinus may be occupied. It leads to nasal obstruction and exophthalmus [8].

Radiographically, OM appears as a unilocular or multilocular radiolucency or mixed pattern [1-4]. The presence of septa gives the tumor a multilocular appearance. A characteristic septa identified with this tumor is a straight, thin-etched septa. These have been described as making a tennis-racket-like or stepladder-like pattern [1]. Zhang et al. described 6 radiographic patterns for OM:

- Unilocular
- Multilocular
- Involvement of local alveolar bone
- Involvement of maxillary sinus
- Osteolytic destruction and
- A mixed of osteolytic destruction and osteogenesis [9].

It may cause root displacement or resorption [1,3]. CT, and, in particular, MRI can help in establishing the intraosseous extent of the tumor [1]. In one research MRI could determine pathologic entity of OM [10] in that in contrast T1-weighted image, the peripheral portion of the lesions with a relatively large quantity of collagen bundles was enhanced, while the central portion with only mucoid component was not. The CT and MRI appearances correlated well with the histologic features and therefore were considered to be a useful tool for diagnosis of myxoma.

Microscopically, the tumor is composed of haphazardly arranged stellate, spindle-shaped, and round cells in an abundant, loose myxoid stroma that contains only a few collagen fibrils [1-4].

OMs are treated by extensive resection because these are not encapsulated and tend to infiltrate the surrounding bone [1-4]. Recurrence rates as high as 25% have been reported [1,2]. One case of malignant transformation of OM has been reported [11].

In this report, a case of maxillary odontogenic myxoma involving the maxillary sinus is presented.

Case report

A 26-year-old male was referred to Oral Medicine Department of Dental Faculty, Mashhad University of Medical Sciences, complaining of a painless swelling in the posterior right maxillary and buccal regions for a period of 2-3 months that has been caused asymmetry. Initially, the swelling was small in size so patient did not seek care until a sensible growth and enlargement of the lesion was observed. The patient was affected with severe hemophilia (type A) with a 4% level of factor VIII.

Intraoral examination revealed an expansion along the alveolar region. The swelling extended buccally from right first premolar region to the right first molar region. There was no palatal expansion (Figure 1). The lesion was bony-hard in consistency, painless in palpation and without mobility in teeth of that region. The teeth, right first premolar and right first molar, had old restorations and restoration of right second premolar had fracture. The patient didn't have clinical signs due to the involvement of sinus (e.g. nasal obstruction etc). No pain, paresthesia, or lymphadenopathy was noted. Extraoral examination showed diffuse swelling in the right infraorbital region. The skin over the swelling was normal.

Fig. 1. Intraoral view: An expansive lesion with bony-hard consistency extending from right first premolar to the first molar region, no palatal expansion and no mobility of the teeth

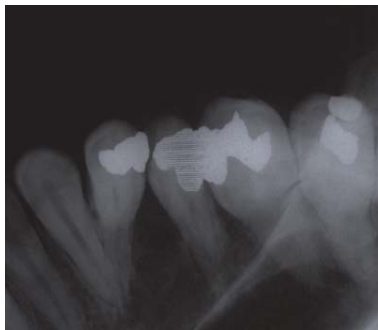


A panoramic radiograph revealed a poorly-defined radiolucency. Distal border of the lesion was obscured. The lesion extended from right first premolar region to posterior portion with involvement of maxillary antrum. Divergence of roots of first and second premolar was observed (Figure 2-A). In periapical radiograph root divergence of right maxillary premolars was found. The bone trabeculation was straight and coarse, making tennis-racket pattern. The lamina dura of second premolar was lost but no root resorption was evident (Figure 2-B).

Fig. (2 A). Orthopantomograph of the lesion: A poorly-defined radiolucency with obscured distal border extended from right first premolar to posterior portion with involvement of maxillary antrum.



Fig. (2 AB). Periapical radiograph : divergence of roots of first and second premolars and multilocular appearance around them. Look at straight coarse trabeculae making tennis-racket pattern.



Axial and coronal Computerised Tomography Scanning (CT-scan), with 5 millimeters slices revealed an expansive lesion involving most parts of right maxillary bone with extension to ipsilateral sinus (Figure 3). The lesion extended from right lateral incisor to second permanent molar causing expansion of buccal cortex. The internal pattern of the lesion consisted of trabeculae creating multilocular appearance. Maxillary sinus was

occupied by the lesion and medial wall of the sinus was eroded (Figure 3).

Fig. 3. CT-scan image, axial view: An expansive lesion involving right maxillary bone with extension to ipsilateral sinus, extended from right lateral incisor to second permanent molar. Medial wall of the sinus is eroded

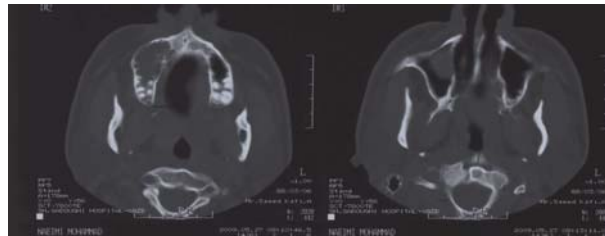


Fig. 4. Photomicrograph of Myxoma with a small strand of odontogenic epithelium (100X magnification, H&E staining).

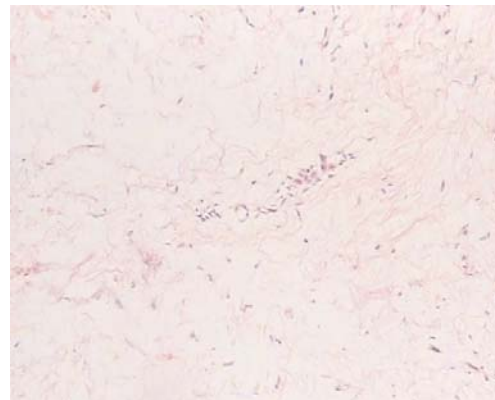
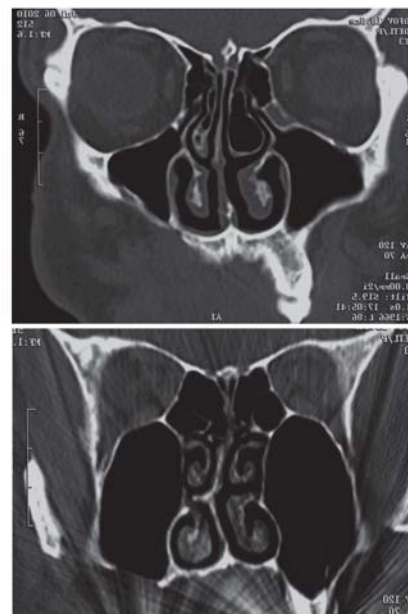


Fig. 5. 12 months follow up: A small surgical defect is observable in antero-inferior part of sinus



Clinical and radiographic presentation suggests the following differential diagnosis

- Odontogenic keratocyst
- Odontogenic myxoma
- Central giant cell granuloma
- Odontogenic carcinoma
- Ameloblastoma

An incisional biopsy was performed after IV transfusion of factor VIII. Histopathologic section composed of stellate, spindle shaped and round cells in an loosed myxoid stroma and a few collagen fibrils were observed. Small islands of odontogenic epithelial rests were found throughout the myxoid ground substance. Therefore, histopathological examination of incisional biopsy specimen led to diagnosis of OM (Figure 4).

Finally, the lesion was surgically excised (bone resection). After 6 and 12 months of treatment, we did not find any clinical or radiographic signs of recurrence at follow-up sessions. Only a small surgical defect was observed in antero-inferior part of sinus, in follow up CT-scan.(Fig 5).

DISCUSSION

We have reported a case of OM involving the maxilla and the maxillary sinus. OM is a rare aggressive odontogenic tumor derived from embryonic mesenchymal tissue associated with odontogenesis [12] which grows slowly but rapid growth rate leads to large size lesions and, when involving the maxilla, it can invade the entire maxillary sinus, as was the case here.

The radiographic features of this tumor are variable and prompt diagnosis may be challenging [13]. There are several radiographic patterns: unilocular, multilocular, and mixed pattern. Multilocular lesions can be seen in soap bubbles, tennis racket and honey combs patterns. There have been reports of a sun ray pattern [14]. In a retrospective radiographic analysis of 21 cases, unilocular forms were usually located in the anterior maxilla, [14] while the multilocular

forms involved the posterior region as was the case here.

In this patient, a poorly defined radiolucency was observed in orthopantomography. In C.T scan a mixed well defined lesion involving right maxillary sinus, was detected in axial sections. Macdonald jankowski et al. proposed that CT scan can display internal pattern and effects on adjacent structures more accurately than plain radiography [1,15].

Presence of ill-defined sinus occupying radiolucency in orthopantomography can be suggestive of malignant lymphoma, central or sinus originating SCC (Squamous cell carcinoma), bone malignancy and rarely desmoplastic ameloblastoma. Due to multiple superimpositions of anatomic structures in orthopantomography, spongy nature of Maxillary bone could be masked, and a false ill-defined border may be observed. Radiographic differential diagnosis was based on CT scan in our case. Several lesions such as OM, CGCG (Central giant cell granuloma) and desmoplastic ameloblastoma can be considered in well-defined mixed pattern of a maxillary lesion [1]. In one research on 25 cases of OM maxillary lesions showed ill-defined borders less frequently than mandibular lesions [16]. Malignant lymphoma was excluded due to mixed pattern. The straight and coarse trabeculae in periapical radiograph is characteristic of OM [1] and therefore desmoplastic ameloblastoma and CGCG were considered with less probability. In addition CGCG rarely cause tooth mobility and root divergence.

When extension is more than jaw expansion OKC and OM are most probable lesions but mixed pattern is not a feature of OKC.

Most commonly, the tumor occurs in the second and third decades [17]. Our case presented at the age of 26 years, which is in conformity with current data in literatures. The mandible is involved more frequently than the maxilla [1,17], although some authors report an equal jaw distribution [18]. OM of the maxilla behaves more aggressively than that of the mandible, as it spreads through the

maxillary sinus [19-21] as presented in our case.

OM is a benign neoplasm without encapsulation. CT can help in establishing the intraosseous extent of the tumor and thus guide the surgeon in planning the resection margins [1]. A spectrum of fibrous connective tissue stroma is present: from myxoid to densely hyalinized and from relatively acellular to cellular [20,22]. Biopsy of this case composed of spindle-shaped cells in an abundant, loose myxoid stroma. Small islands of odontogenic epithelial rest could be seen. A myxoma may be microscopically confused with other myxoid neoplasms, such as the rare chondromyxoid fibroma or the myxoid neurofibroma [2]. Myxoid change in an enlarged dental follicle or the dental papilla of a developing tooth may be microscopically similar to a myxoma [2,3].

Since it is an infiltrative aggressive disease with a high recurrence rate, *en bloc* resection was performed because besides allowing entire tumor enucleation, it removes bone tissue that could be microscopically involved and afterward may cause recurrence [14]. This method of treatment is particularly indicated in the maxilla due to proximity of vital structures [23].

CONCLUSION

Although OMs are rare in maxilla, an expert clinician must include OM in differential diagnosis of an extensive lesion in maxilla and maxillary sinus region; specially, if there is a tendency to grow within bone and the lesion have multilocular radiographic appearance (specially with coarse straight trabeculae) along with invasion to surrounding structures. Accurate surgical removal with wide margins is advised for treatment of OM.

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