Odontogenic myxoma of the maxilla

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Abstract

Myxomas are mostly seen in the 2th and 3rd decades. They rarely occur in childhood and maxillofacial region is rarely involved. The recurrence incidence is high. We report this unusual case occurring in a 9-year-old girl in the maxillofacial region and recurrence four months after initial treatment.

Introduction

According to the World Health Organization (WHO) classification, odontogenic tumors are considered to be tumors of the odontogenic mesenchyme, with or without the presence of odontogenic epithelium.[1]

Myxomas of head and neck are divided into two forms:[2] i) deriving from facial bone which has also been subdivided into osteogenic myxoma and odontogenic myxoma; ii) deriving from soft tissue which originate from perioral soft tissues, parotid glands, ears and larynx.

Odontogenic myxomas are benign, slow-growing and locally invasive tumors. They have predilection for the molar and premolar region of the maxilla.[3] Generally, adolescents are affected. They are rare in children.[4] The most frequent symptom is slow facial swelling. Patients may also complain of malocclusion, loss of tooth and palatal swelling.[5]

From a histological aspect, odontogenic myxomas include spindle like cells, star shaped cells with elonged cytoplasms and, in some cases, small masses of inactive odontogenic epithelium.[6,7] Radiographically, the tumors present as unilocular or multilocular radiolucent lesions. The lesions usually have well-defined borders and are described as *honeycombed* or *soap bubble* shaped.[8,9] Differential diagnosis must be made with ameloblastoma dentiginous cysts, fibrous dysplasia, osteosarcoma, chondrosarcoma and odontogenic fibroma.[10] Although local trauma has been thought to be the cause, the etiology is unknown.[11] There are high recurrence rates after surgical excision. Therefore, wide local excision is mandatory for treatment.

In this case report we discuss these tumors with examples from literature because they are very unusual in childhood and have high recurrence rates.

Case Report

The patient was a 9-year old girl presenting with swelling on the right facial region. She had no other symptoms. The mass was painless and progressively enlarging. It had bulged out of the right nasolabial region in the last five months. Physical examination of the patient revealed that the mass had obliterated the right nasolabial groove and caused swelling of the right facial region. Medial wall of the right nasal cavity had displaced towards the median axis. Dentition was normal. The cranial nerves appeared to be intact. The computerized tomography (CT) examination revealed that the mass was originating from the right maxillar bone and the maxillary sinus antrum was filled by the tumor. Medial and anterior wall of the sinus was displaced because of the growing mass (Figures 1 and 2). The mass was hypodense and had microcalcification areas. These findings suggested fibrous dysplasia, ossifying fibroma or odontogenic cyst. An incisional biopsy was made through the nasolabial groove and was reported to be odontogenic myxoma. Surgical removal of the mass including wide periincisional tissue was performed via a right superior gingivo-buccal incision. During the operation it was seen that the mass was white in color, completely encapsulated and was completely filling the sinus antrum. The anterior wall of the maxillary sinus was thinned, partially destructed and pushed anteriorly.

Four months after the first operation, swelling of the right maxillar region recurred. The CT scan showed that the maxillary antrum was filled by a new mass. Partial maxillectomy was then performed. The patient was followed up for two years and there has been no evidence of recurrence.

Histological examination of both masses removed during the two surgical interventions revealed hypocellular tumoral tissues with myxoid stromas and scattered stellate fusiform cells with hyperchromatic nuclei (Figures 3 and 4). Therefore, the masses were assumed to be odontogenic myxomas.

Discussion

Virchow first described myxomas in 1863.[12] He described that these tumors resemble the mucinous substance of the umbilical cord. These tumors are most frequently seen in heart muscle. Myxomas of the mandible and maxilla are very rare. In 1947, Thoma and Goldman[13] separated myxomas of the mandible and maxilla from other myxomas. In 1948, Stout[8] redefined the histological criteria for myxomas as benign neoplasms of mesenchymal origin.

Ghosh *et al.*[14] defined only 10 osseous myxomas in a review of 8723 primary bone tumors; 6 in the mandible and 4 in the maxilla. Although it is claimed that maxilla and mandible are equally involved, others noted a more frequent involvement of the mandible.[2,15,16]

The etiology of these tumors is not clear. However, there is a theory that they arise from odontogenic mesenchyme, especially from the molar and premolar region of the maxilla.[1,3,7,17]

Histological examination showed that there is no difference between osseous and soft tissue myxomas. Stromas of myxomas are hypocellular and they include mucopolysaccharides, hyaluronic acid, and chondroitin sulfate molecules. These substances are thought to be

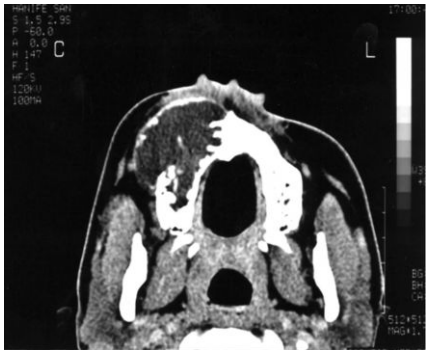

Figure 1. Axial tomographic image of the myxoma showing obliteration of the right maxillary sinus and bone destruction with finger like projections into maxilla.

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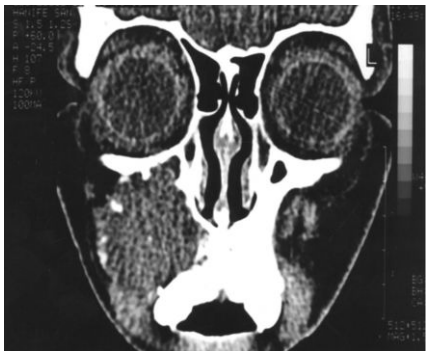
the cause of the locally aggressive behavior of the myxomas.[18] Histologically, these tumors may be confused with myxoid degeneration, malignant nerve sheath tumors and myxoid chondrosarcoma.[18]

Radiological examination may help to diagnose these tumors accurately. CT scan shows myxomas to be of two different types: unilocular or multilocular. They are described as being of a *honeycomb* or *soap bubble* shape.



Figures 2. Coronal tomographic images of the myxoma showing obliteration of the right maxillary sinus and bone destruction with finger like projections into maxilla.

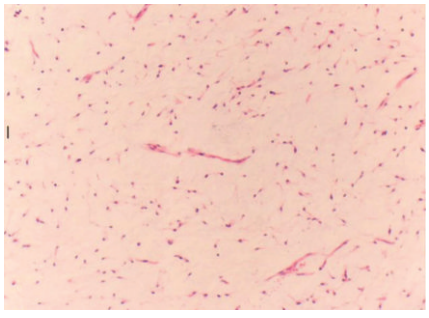


Figure 3. Hypocellular tumoral tissue with myxoid stroma and scattered stellate and fusiform cells with hyperchromatic nuclei (hematoxylin & eosin, ×200).

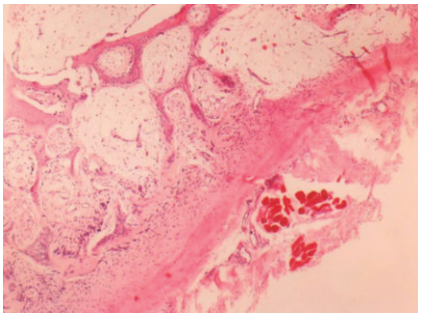


Figure 4. Tumor tissue showing the same morphology in every field including the bone trabeculae (hematoxylin & eosin, ×200).

Computerized tomography demonstrates the limits of the tumors and characteristics of the bony septa.

Radio- and chemotherapy are not effective. Surgery is the most effective therapy. Enucleation is performed for protecting the facial growth centers and this approach prevents facial asymmetry. It is suggested that enucleation may be performed in small tumors, but recurrence rates are higher than partial maxillectomy.[19-22] Therefore, wide surgical excision and close follow up is mandatory in osseous myxomas. In our patient, recurrence was confirmed four months after the operation and partial maxillectomy had to be performed.

Conclusions

There is no specific clinical and radiographic finding for odontogenic myxomas. Diagnosis can be made by biopsy. Differentional diagnosis has great importance for all tumors involving the maxillo-facial region. Recurrence rates are high and a long follow-up period over years is essential after treatment for patients with these tumors.

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