Odontogenic myxoma of the maxilla

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Abstract

Myxomas are mostly seen in the 2nd 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 forms2 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 elongated cytoplasm and, in some cases, small masses of inactive odontogenic epithelium.5,7 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 dentigerous 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 maxillary 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 periacinal 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 maxillary 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 mucous 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 Goldman13 separated myxomas of the mandible and maxilla from other myxomas. In 1948, Stout14 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.13,17

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.](image-url)
the cause of the locally aggressive behavior of the myxomas. Histologically, these tumors may be confused with myxoid degeneration, malignant nerve sheath tumors and myxoid chondrosarcoma. Radiological examination may help to diagnose these tumors accurately. CT scan shows the myxomas. Histologically, these tumors are diagnosed 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.

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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