Odontogenic Myxoma Involving the Maxilla: Case Report of a Rare Entity

Archana Shivamurthy, Geetha Vasudevan, Padmapriya Jaiprakash, Dipak Ranjan Nayak, Ajay M Bhandarkar

Assistant Professor, Department of Pathology, Melaka Manipal Medical College, Manipal University, Manipal, Karnataka, India, Additional Professor, Department of Pathology, Kasturba Medical College Manipal, Manipal University, Manipal, Karnataka, India, Associate Professor, Department of Pathology, Kasturba Medical College, Manipal University, Manipal, Karnataka, India, Professor and Head, Department of Otolaryngology-Head and Neck Surgery, Kasturba Medical College, Manipal University, Manipal, Karnataka, India, Assistant Professor, Department of Otolaryngology-Head and Neck Surgery, Kasturba Medical College, Manipal University, Manipal, Karnataka, India

Odontogenic myxomas (OMs) are rare, benign, slow-growing tumors that may arise in the maxilla. They are known to have a female predilection and present as a painless mass commonly in the second or third decade of life, comprising 3-6% of all odontogenic tumors. They show a locally aggressive behavior, are radioresistant tumor and hence the need for early recognition and surgical resection. A high rate of recurrence has also been noted owing to its infiltrative pattern of growth and lack of capsule. Clinical and radiological aspects of maxillary OMs are not conclusive hence it is necessary to have a histopathological exam for the final diagnosis. We present a case of OM involving the maxilla in a 51-year-old female patient. The various histopathological differentials are also discussed.

Keywords: Maxilla, Myxoma, Odontogenic, Painless

INTRODUCTION

Odontogenic myxomas (OMs) are relatively rare, slow-growing, locally invasive, benign mesenchymal neoplasms, seen exclusively in tooth-bearing areas that have a slight predilection for females. They comprise 3-6% of all odontogenic tumors, commonly occur in the second and third decade of life, presenting as a painless growth. The prevalence of myxomas is between 0.04% and 3.7% and the mandible is involved more commonly than the maxilla. It was Rudolf Virchow, who was the first to describe this lesion as myxofibroma in 1863. However, Thoma and Goldman in 1947 renamed it as OM. OMs are known to originate from the embryonic connective tissue associated with the tooth forming apparatus. Very few cases of maxillary gingival myxomas are reported in the literature till date. The surgical treatment for these myxomas vary from simple enucleation and curettage to segmental resection and hemimandibulectomy. However, the risk of recurrence appears to be higher in the former.

CASE REPORT

A 51-year-old female patient presented with complaints of swelling in the right cheek since 8 months. It was insidious in onset and progressive. There was no history of pain, fever or difficulty in swallowing. On examination, a proliferative mass was seen occupying the right buccogingival sulcus (Figure 1). On radiological examination, a well-defined minimally enhancing hypodense lesion measuring 5.2 cm × 2.4 cm was found with the destruction of right upper alveolar bone and extending superiorly into the floor of the right maxillary sinus.
sinus. The patient thus underwent right partial maxillectomy and the specimen was sent for histopathological examination.

Gross examination revealed a $6 \text{ cm} \times 4 \text{ cm} \times 2.5 \text{ cm}$ bosselated, polypoid growth involving the maxillary bone (Figure 2). On the cut section, the white lobulated myxoid appearance of the tumor was noted. On microscopic examination, the tumor was unencapsulated and composed of randomly dispersed stellate cells with eosinophilic cytoplasmic processes enmeshed in an abundant myxoid stroma (Figure 3a-c). The myxoid intercellular matrix stained positively with alcian blue (Figure 4), but periodic acid-Schiff (PAS) staining was negative. Ulceration of the overlying stratified squamous epithelium was noted along with granulation tissue. On immunohistochemistry, few stellate cells showed positivity for smooth muscle actin (Figure 5), but were negative for S-100. Based on the above observations, a diagnosis of OM was rendered.

DISCUSSION

OMs are rare benign, invasive tumors that exclusively occur in the jaw bone and have a tendency for bone destruction. The mandible is more commonly affected than the maxilla. Although OMs of the maxilla are less frequent, they behave more aggressively than that of the mandible, tend to fill and cause enlargement of the maxillary antrum before clinically presenting as a facial swelling.\textsuperscript{1,3,7,8} In the present case, the patient presented with a facial swelling since 8 months. Several case reports have reported a higher frequency of occurrence in females than males. It is most frequently diagnosed between the second and the fourth decade.\textsuperscript{1,6,7} However, the present case is being reported in a 51-year-old woman, who had the destruction of right upper alveolar bone and the lesion was extending to the floor of the right maxillary sinus.

On radiography, OM may appear as radiolucent, unilocular or multilocular masses with either honeycomb or soap-
bubble pattern. Cortical expansion and tooth displacement can also be noted.1,2,5 In the present case, a well-defined minimally enhancing hypodense lesion measuring 5.2 cm × 2.4 cm was detected. The various clinical and radiographic differential diagnosis of OMs include: Odontogenic keratoctyts, simple cysts, traumatic bone cysts, intraosseous hemangioma, cherubism, aneurysmatic bony cyst, fibrous dysplasia, ameloblastoma, central giant cell granuloma, and metastatic tumor.1,9,10

OMs are aggressive unencapsulated lesions and said to have been derived from embryonic mesenchymal tissue associated with the development of the teeth/odontogenesis. They are primarily composed of widely scattered undifferentiated spindled to stellate mesenchymal cells in a myxomatous ground substance. The spindle cells have long eosinophilic fibrillar processes that intermesh with each other and do not show evidence of significant pleomorphism or mitotic figures. Interspersed capillaries and occasionally strands of collagen may be seen. Calcification may or may not be present.1,10,11 Odontogenic epithelium arranged in islands and cords may be present. However, it is not a histological criteria for the diagnosis of OMs. Tumor cells also may show slight alkaline phosphatase activity.1,11 Hyaluronic acid (80%) is the major component of the ground substance with a relatively small amount of chondroitin sulfate, fibronectin, and proteoglycans. The myxoid intercellular matrix stains positively with alcian blue, but PAS staining may be negative.2,5 In the present case, a bosselated, polypoid growth was seen, involving the maxillary bone with white lobulated myxoid appearing cut surface. On microscopy, the tumor was unencapsulated and composed of randomly dispersed stellate cells with eosinophilic cytoplasmic processes enmeshed in an abundant myxoid stroma, which was alcian blue positive.

Table 1: Differential diagnosis of OMs

<table>
<thead>
<tr>
<th>Entity</th>
<th>Clinicoradiographic features</th>
<th>Histopathological features</th>
</tr>
</thead>
<tbody>
<tr>
<td>Myxoid nerve sheath tumors</td>
<td>Localized facial or intraoral swelling, rare involvement maxilla, well defined and radiolucent.</td>
<td>Circumscribed but unencapsulated lesion, interfacing bundles of elongated cells with tapering, wavy nuclei.</td>
</tr>
<tr>
<td>Central neurofibroma</td>
<td>Wide age range (2-65 years), rarely involve mandible and maxilla.</td>
<td>Wire-like strands of collagen, IHC: S-100 positive.</td>
</tr>
<tr>
<td>Chondromyxoid fibroma</td>
<td>Very rarely involves maxilla/mandible.</td>
<td>Usually encapsulated masses composed of nodules cartilage found in between fibromyxoid areas.</td>
</tr>
<tr>
<td></td>
<td>Males between the age of 10 and 35 years, Lytic ovoid lesion with sharp sclerotic margins and pseudo loculated on radiography.</td>
<td>The chondrocytes are disposed in sparsely cellular lobules of myxoid or chondroid matrix.</td>
</tr>
<tr>
<td>Rhabdomyosarcoma</td>
<td>Most common malignant tumor of soft tissue in children under 15 years of age.</td>
<td>Embryonal form is the most frequent, Sheets of small, spindled or moderate to poorly differentiated round cells with scant or deeply eosinophilic cytoplasm and eccentric, small oval nuclei.</td>
</tr>
<tr>
<td></td>
<td>Are known to occur in the maxillary sinus, Pain of the cheek is the major facial symptom.</td>
<td>Myogenin/desmin positive, Lobules of bland, relatively uniform, spindled to ovoid to round cells in an abundant light pink to blue-gray myxoid/mucoid matrix.</td>
</tr>
<tr>
<td>Myxoid liposarcoma</td>
<td>Extremely rare in the head and neck, Can very rarely present as a periodontal mass.</td>
<td>Necrotic nuclei have a uniform chromatin pattern and generally inconspicuous nuclei. Prominent delicate plexiform capillary network.</td>
</tr>
<tr>
<td>Myxoid fibrosarcoma</td>
<td>Major symptoms are pain, swelling, and sometimes loosening of the teeth, Usually affects subcutis of distal extremities in elderly patients, rarely found around the maxillary region.</td>
<td>Spindle-shaped cells, deposited in a variably fibrous and myxoid stroma with significant mitotic activity and nuclear pleomorphism. Positive for vimentin and MIB-1, and focally positive for CD68. Negative for: 1-SMA, desmin, S 100, EMA.</td>
</tr>
<tr>
<td>Myxoid chondrosarcoma</td>
<td>Usually occurs in the soft tissue of extremities of adults, Only rarely originate in the head and neck in children, Show a permissive pattern of osseous destruction on radiography associated soft tissue mass.</td>
<td>Abundant chondroid matrix and malignant chondroblastic cells arranged in cords resembling chordoma.</td>
</tr>
<tr>
<td>Nodular fascitis</td>
<td>Up to 20% of cases may occur in the head and neck region, Most common in the third decade, Males and females are equally affected.</td>
<td>May be circumscribed but not encapsulated, Spindle-cell lesion composed of loose, plump, bipolar fibroblastic cells arranged in a storiform pattern with loose myxoid background. Mitotic activity may be present, Extravasated red blood cells and a mixed inflammatory infiltrate is present. IHC: Vimentin-positive, Actin, keratin, SMA, S-100, and desmin are negative.</td>
</tr>
</tbody>
</table>

SMA: Smooth muscle actin, OMs: Odontogenic myxoma, IHC: Immunohistochemistry, EMA: Epithelial membrane antigen.
Some of the microscopic differential diagnoses include myxoid nerve sheath tumors, chondromyxoid fibroma, rhabdomyosarcoma, and nodular fasciitis. Rapidly growing neoplasms showing myxoid degeneration such as fibrosarcomas, chondrosarcomas, and liposarcomas also fall into the differential diagnosis (Table 1).\textsuperscript{3,10,11}

OM tumor cells are mesenchymal in origin and express vimentin and muscle-specific actin. They are negative for, neuron-specific enolase, S-100 alpha and beta subunits, factor VIII-related antigen, and cytokeratin.\textsuperscript{1,2,11}

Effective nucleation and curettage difficult as OMs tend to permeate and destroy marrow spaces. Larger tumors require complete excision with free margins. Recurrence rates from various studies average about 25%. Despite the fact that OM shows aggressive local growth, it is believed that it never undergoes malignant transformation or gives rise to metastasis.\textsuperscript{4,6,11}

**CONCLUSION**

Clinical and radiological aspects of maxillary OMs are not conclusive, it is necessary to have a histopathological exam for the final diagnosis. Because of its high rate of recurrence, especially due to its gelatinous and mucous aspect, surgical treatment through bone resection is the most indicated treatment modality, and the patient must be followed-up closely for years.

**REFERENCES**