# Odontogenic Myxoma: A Case Report

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**Abstract**

Odontogenic myxoma are rare, benign intraosseous lesions, arising from odontogenic ectomesenchyme. They are locally aggressive. It affects patients in their first to third decade of life. Two third of cases are in the mandible and the remaining third in the maxilla. We herein report a case of odontogenic myxoma in mandible in a 25 year old female.

**Key words:** Odontogenic tumor, odontogenic myxoma, benign, mandible,

Clinical relevance: Odontogenic tumors are rare and odontogenic myxomas are still rarer. This case enlists the importance of enlisting such lesions in differential diagnosis.

## Introduction

Odontogenic myxoma (OM) is a benign, intraosseous neoplasm that arises from odontogenic ectomesenchyme and resembles the mesenchymal portion of the dental papilla. These develop only in the facial bones. It is occasionally related to a tooth that failed to erupt or is missing, and in some cases odontogenic epithelium can be detected microscopically. It accounts for 3-6% of odontogenic tumors. It is more common in females, with an age range of 10 to 30 years. Mandible (premolar-molar area) is more commonly affected. It is a slow growing hard fusiform swelling which may cause facial asymmetry and pain. The involved teeth may be displaced and loosened but resorption is rare. OM rarely crosses the midline. According to Simon et al[1] the lesions occupy one side only (left or right), but Noffke et al[2] found six mandibular lesions that crossed the midline, being probably size related. Microscopically these lesions are characterized by stellate and spindle-shaped cells embedded in a richly myxoid extracellular matrix, with scarce collagen. On imaging, odontogenic myxoma may present variable features - mostly as radiolucent, multilocular or unilocular structures on plain radiographs. Diagnosis is most commonly established by biopsy. Differential diagnosis like ameloblastoma,

ameloblastic fibroma, odontogenic fibroma, central hemangioma, or odontogenic keratocyst along with OM could be listed as initial diagnostic hypothesis based on the clinical and radiological findings. [3,4]

Management varies depending on the location and size of the tumour, the age of the patient and individual experience. They range from minimally invasive excisional biopsies to an en bloc resection of multiple of relevant structures. Long-term follow-up is crucial as myxomas have a significant tendency of recurrence. Recurrence rates for myxomas in the literature vary from 5 to 10% but there is no reliable data in the existing literature.[5, 6]

**Case report:**

A 25 year old female reported to the department of oral medicine with the chief complaint of swelling with respect to lower left side of face since five months. There was history of occasional intermediate pain on mastication. Initially, the swelling was small in size and showed a gradual increase to its present dimensions. Clinical examination revealed a firm, non-tender swelling expanding the buccal and lingual cortices of the mandible, extending from left incisor region to mandibular ramus region, and it obliterated the buccal vestibule (figure 1). The skin over the swelling was normal, and there was no history of paresthesia.

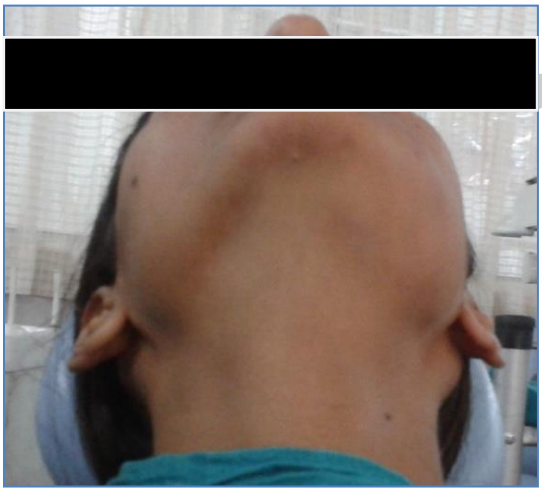

Figure 1: Clinical picture showing swelling involving left mandible.

The left mandibular lateral occlusal radiograph showed multilocular radiolucent lesion with expansions of buccal and lingual cortices (Figure 2).

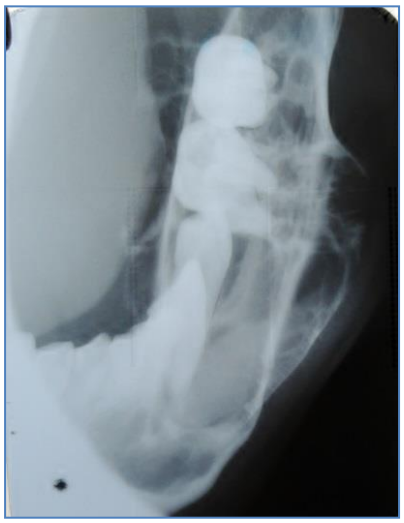

Figure 2: Cross sectional left mandibular occlusal radiograph showing buccal and lingual cortical plate expansion.

The panoramic radiograph showed a large well-defined, corticated margined,

multilocular radiolucent lesion with wispy indistinct trabeculae extending from the lower left incisor to ramus of mandible (figure 3).

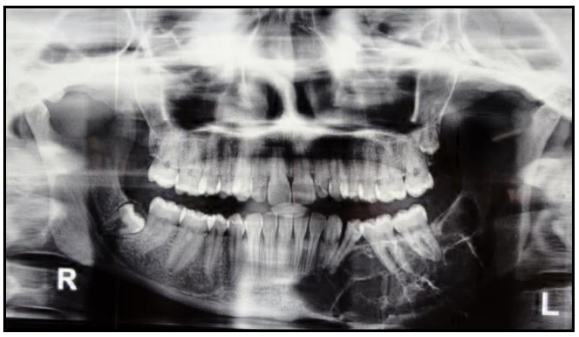


Figure 3: OPG showing multilocular lesion involving the mandible extending from apex of 41to left ramus

CT scan revealed a large osteolytic lesion irt left mandible which was suggestive of an expansile cyst (figure 4).

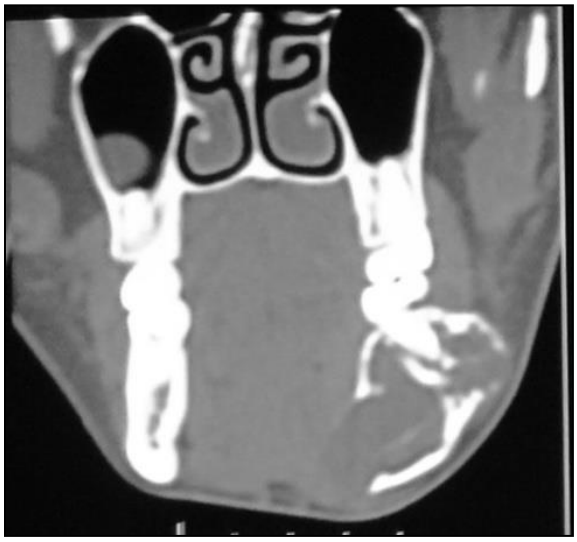

Figure 4: CT scan showing large osteolytic lesion involving left posterior mandiblular region

Fine needle aspiration was performed to rule out odontogenic cysts, and results were negative. Benign odontogenic tumors were considered, and incisional biopsy was made and a histopathological examination of the tissue sample exhibited rounded, stellate, and spindle shaped mesenchymal cells arranged in a loose, myxoid stroma with few collagen fibrils (Figure 5). These results were suggestive of OM.

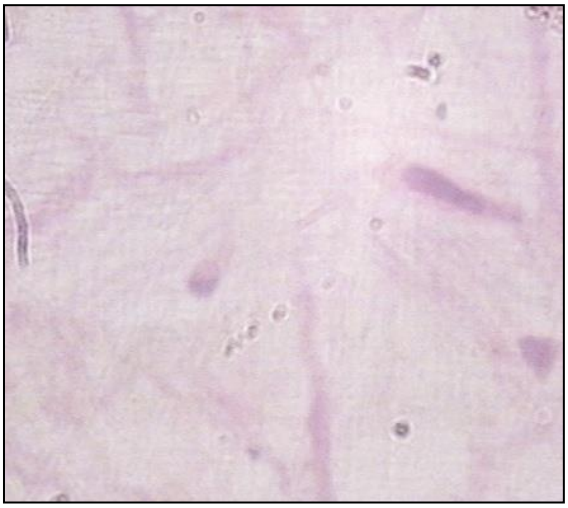

Figure 5: Photomicrograph showing spindle shaped cells arranged in a loose, myxoid stroma with few collagen fibrils

## Discussion

OM is a tumor thought to be derived from embryonic mesenchymal elements of dental tissues. It is a locally invasive benign neoplasm. The invasiveness is attributed to the biological nature of the tumor. OM is thought to be derived from dental papilla and Hertwig's epithelial root sheath.[7] Evidences supporting the odontogenic origin are (a) its occurrence is almost exclusive, only to the tooth-bearing areas in the jaws, (b) occasional association with an

unerupted tooth or a missing tooth, c) occurrence in younger individuals, (d) histologic resemblance to dental papilla, and (e) occasional presence of odontogenic epithelial island.[8] The OM cells express extra cellular membrane molecules like fibronectin, type 1 collagen and tenascin resembling human immature dental papilla stem cells.[9] The immunohistochemical and ultrastructural studies done by Martinez-Mata G et al., concluded that OM is a mesenchymal neoplasm.[10] Similarity of protein profiles of extracellular matrix of dental follicle (DF) of a developing tooth and OM, supported the theory that OM could originate from DF.[11] Miyagi SP et al., stated that the dental pulp stem cells, which are derived embryologically from the dental papilla cells may be the precursor of OM.[12]

The tumor is thought to originate from the mesenchyme of the developing tooth or the periodontal ligament because of the resemblance of these structures to the tumour tissue.[10] Nestin, marker for progenitor cells, was positive in the stromal neoplastic cells of the OM. So the authors suggested that the possible origin of tumor cells are from the dental papilla cells, fibroblasts or myofibroblasts.[11] Using the notch signaling, Nakano et al., concluded that the differentiation level of the tumor tissue is similar to cap stage. The lower degree of differentiation is the reason for the degree of clinical behavior.[12]

Clinically, odontogenic myxoma is a benign, painless, invasive, slowly enlarging mass. Large lesions can cause marked asymmetry of the face. It causes expansion of bony cortices, displacement and loosening of teeth.[8] Zhang et al., classified radiographic appearances of OM into six types- Type I: Unilocular well-defined radiolucency, Type II (multilocular): Two or more compartments with multiple interlaced osseous trabeculae described as honey comb, soap bubble or tennis racquet radiolucency, Type III: Lesion located in alveolar bone, Type IV: Lesion involving the maxillary sinus, Type V (moth eaten appearance): Larger radiolucent area with irregular borders, Type VI: Combination of bone destruction and bone formation giving sun ray appearance.[13] The tumor often shows scalloping between the roots; root resorption can occur but is rare.[14]

Histologically the WHO (1992) defined OM as “A locally invasive neoplasm consisting of rounded and angular cells lying in an abundant mucoid stroma”.[15] They show loose myxoid stroma containing spindle-shaped, angular or round cells. Polymorphic cells or nuclei are rarely encountered. Stroma shows very few fine fibrillar material and minimum vascularity. Inflammatory infiltration and remnants of odontogenic epithelium are occasionally

seen. Rarely narrow zone of hyalinization can be seen around odontogenic epithelium. Hyperchromatic nuclei and mitotic figures are rare. The myxoid intercellular matrix stains positively with Alcian blue, but periodic-acid Schiff staining may be negative. The malignant counter part of OM is odontogenic myxosarcoma is very rare.[15,7]

The spindle-shaped cells are undifferentiated mesenchymal cells, which have the capacity to undergo fibroblastic differentiation. Depending on the degree of differentiation it is named as myxoma, odontogenic fibromyxoma or odontogenic myxofibroma.[8] Goldblat in 1976 described two basic types of tumor cells i.e., secretory and nonsecretory. The secretory cell type was considered the principal tumor cell and resembled fibroblasts.[9] Three types of odontogenic myxoma cells were discriminated: Spindle cells, stellate cells and hyaline cells. Spindle cells are positive for transferrin, ferritin, alpha 1-antichymotrypsin (ACT), alpha 1-antitrypsin (AT), S-100 protein and vimentin. Stellate cells were strongly positive for transferrin, alpha 1-AT, S-100 protein and vimentin. Hyaline cells reacted with alpha 1-ACT and alpha 1-AT. Myxomatous matrix is negative for all these antibodies. These results proved that odontogenic myxoma is a tumor of a dual fibroblastic-histiocytic origin and also suggest that the cells are of myofibroblastic origin.[16-17]

Matrix metalloproteinases (MMPs) facilitate the invasion of the tumor cells through the normal tissues. Miyagi et al., analyzed the expression and activities of MMP 2 and 9 in the tumor. They concluded the invasiveness of OM is due to MMP 9.[18] Also high degree of MMP 2 expression were found in cell lines derived from OM.[12] Ultrastructural studies showed that the neoplastic spindle cells are fibroblast like cells called myxoblasts. They synthesize large quantities of mucopolysaccharides. The ground substance of OM has been shown to consist of about 80% hyaluronic acid and 20% chondroitin sulfate. Exuberant hyaluronic acid which is an extracellular membrane protein is responsible for the local invasiveness of this neoplasm.[19] Tumor cells are relatively inactive, showing low levels of oxidative enzymes and slight alkaline phosphatase activity.[20] The aggressive nature of the lesion is well documented in literature.

The absence of capsule and tendency of OM to permeate into marrow spaces makes effective enucleation and curettage difficult. Small lesions have been successfully treated in this way but the larger lesions require block resection with tumor-free margins. Recurrence rates from various studies average about 25%.[8] A minimum of five years of surveillance is

required to confirm that the lesion has healed, and periodical follow-up of clinical and radiographic studies should be maintained indefinitely and irrespective of the treatment modality.[21,22]

## Conclusion

Odontogenic myxoma is a rare, benign, locally invasive tumor of jaws. It may have an insidious onset, usually in the second and third decade of life. The patient may report to the dentist due to swelling and associated facial asymmetry. A biopsy of the involved bone can aid in its diagnosis. Complete resection of the tumor as well as long term follow up is recommended.

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## Conflicts of Interest

There are no conflicts of interest.

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