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# Odontogenic Myxoma Of The Maxilla: A Clinical Case Report And Review Of Literature

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## Abstract

Odontogenic myxomas are rare benign mesenchymal tumours of head and neck with a potential for local infiltration and recurrence. They appear to originate from the dental papilla, follicle or periodontal ligament in mandible and less commonly the maxilla. These usually present in second or third decade of life as slowly progressive space occupying lesion in the jaw giving a mixed radiopaque-radiolucent appearance. The treatment is considered to be wide local excision in view of high recurrence with curettage alone. Here, we present a case of odontogenic myxoma of maxilla, in a 40 year old lady, with a brief review of literature, clinical, radiological, histopathological characteristics and therapeutic modality employed.

## INTRODUCTION

Myxomas are benign, slow-growing and locally aggressive mesenchymal neoplasms. Virchow coined the term in 1863 for a group of tumors that had histologic resemblance to the mucinous substance of the umbilical cord [1]. In 1948, Stout redefined the histologic criteria for myxomas as true neoplasms that do not metastasize and exclude the presence of recognizable cellular components of other mesenchymal tissues, especially chondroblasts, lipoblasts and rhabdomyoblasts.

Myxomas can be found in various body parts such as skin, subcutaneous tissue and the heart (left atrium). Myxomas of the head and neck region are rare. Two forms of myxoma have been identified in head and neck (1) those derived from facial bones which are further subdivided into true osteogenic myxoma and odontogenic myxoma and (2) those derived from facial soft tissue like perioral soft tissue, parotid gland, ear or larynx. [2]. Odontogenic myxoma represents an uncommon benign neoplasm comprising of 3-6% of all odontogenic tumors most commonly arising in the mandible (66.4%) followed by maxilla (33.6%) . They appear to originate from the dental papilla, follicle or periodontal ligament [3] .The evidence for its odontogenic origin lies in its almost exclusive location in the tooth-bearing areas of the jaws, its occasional association with missing or unerupted teeth and the presence of odontogenic epithelium. [4]

Histolopathologically, odontogenic myxoma is defined as non-encapsulated benign odontogenic tumor of mesenchymal origin that is locally invasive and consists of rounded and angular cells that lie in abundant mucoid stroma [5].

The purpose of the present article is to present such a case that was managed in the Department of Otolaryngology , Head and Neck Surgery at Dayanand Medical College and Hospital, Ludhiana, India emphasising the unique clinical presentations, differential diagnosis and management.

## CASE REPORT

A 40 year old lady was referred to our institute with a 2 month history of progressively worsening right sided nasal blockade, accompanying mucoid ,non foul smelling nasal discharge and history of heaviness in the right cheek region. There was no history of loosening of teeth or dental pain.

Right side anterior rhinoscopy revealed medialisation of turbinates with obliteration of nasal cavity by mucoid discharge. Facial examination was unremarkable with normal vision and extraocular movements. An intraoral exam revealed no palatal bulge nor dental malocclusion. Rest of the clinical head and neck examination was normal. There was no relevant past history.

Nasal endoscopic biopsy done elsewhere which was reported as spindle cell lesion (?neurogenic origin) with nuclear pleomorphism and hyperchromasia.

A computed tomography scan without contrast in axial and coronal sections of nose and sinuses was obtained .The axial section revealed a large expansile soft tissue mass occupying the right maxillary sinus and nasal cavity with erosion of anterior maxillary wall and extension into infratemporal fossa (Figure 1). Areas of bone remodelling were seen with dehiscence of the alveolar ridge.

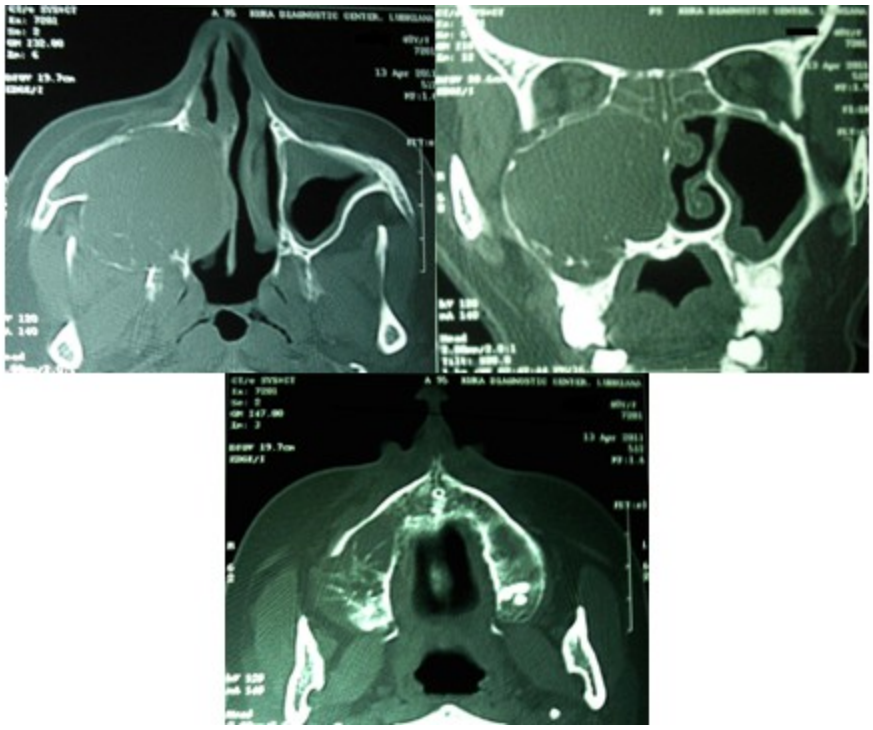

Figure 1: Computerized tomography images of the mass with dehiscence of alveolar ridge.

Measurements for insertion of a prosthetic dental obturator at the site of resection were carried out preoperatively by the dentistry services.

It was planned to undertake a  Caldwell Luc approach to the maxillary mass and convert into Weber-Fergusson, if required.

Under general anesthesia, a sublabial incision was made on the right side extending from lateral incisor to the maxillary tuberosity.  Periosteum was elevated from the anterior wall of maxilla. The canine fossa was found to be eroded .The defect was widened and a smooth soft well defined mass was visualized occupying the entire maxillary sinus. It was found to be attached inferiorly  to alveolar arch as the elevator could be passed superiorly and medially but not inferiorly.

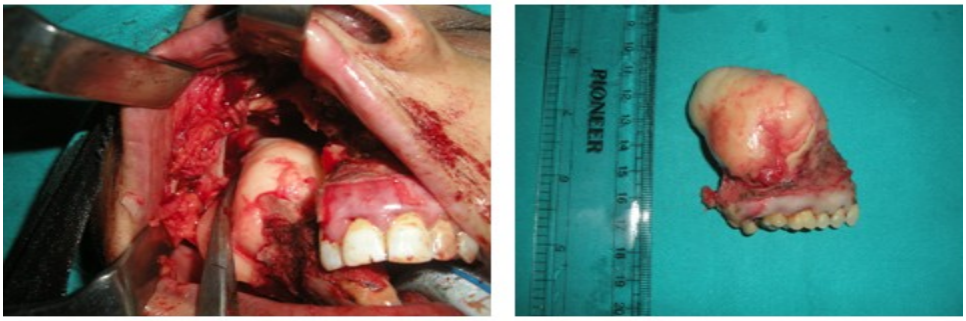


Figure 2 : In toto excision of maxillary sinus mass by Caldwell luc approach

The initial incision was extended across the maxillary tuberosity towards midline at the junction of hard and soft palate .Another incision was made just lateral to midline on the hard palate till the incisors. Wide middle meatus antrostomy was created using 0° endoscope. The mass was dissected off from the orbital floor and the lateral wall of maxillary sinus.

The entire mass was removed in toto along with the superior alveolar arch, hard palate and part of medial wall of maxilla. After complete removal, the margins of the cavity were carefully osteotomized with a large spherical drill, to minimize the chances of recurrence(Figure 2).

There was no need for a Weber Fergusson facial incision. Spit thickness skin graft from thigh was used to line the inner aspect of the cheek and a bolster dressing was done. Nasogastric feeding tube was inserted from left nasal cavity. The post operative course was uneventful .Dental obturator was fitted and the patient was discharged on fifth day after surgery. 16 months after surgery the patient is doing well without any radiological recurrence.

Histopathological examination revealed an odontogenic myxoma with clear margins. The tumour was composed of spindle-like cells, with egg-shaped or elongated nuclei, without cellular atypia. The cells

were found immersed in loose and myxomatous matrix, including scarce blood vessels and irregular calcification(Figure 3).

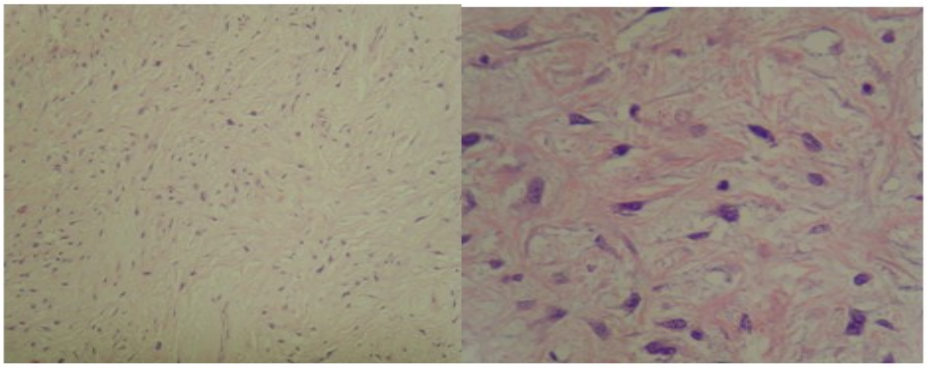

Figure 3 (a) 100 X low power view showing spindle shaped tumour cells separated by myxoid stroma (b) High power view 400 X

## DISCUSSION

According to the recent WHO classification of odontogenic tumors, the myxoma is considered a tumor of the odontogenic mesenchyme, with or without the presence of odontogenic epithelium. Odontogenic maxillary myxomas were first mentioned in the literature by Thomas and Goldman in 1947.

They most commonly present between age of 10 to 40 years although they have been reported in children as young as 3 months [6].The incidence is equal in both sexes with some studies suggesting a slight female preponderance[7,8].

The premolar–first molar region is the site of predilection in the maxilla [9].OM of the maxilla may be asymptomatic and detected as an incidental finding on routine dental or sinus examination or it may present as heaviness, swelling of cheek or palate, malocclusion or loosening of teeth. Displacement of teeth has been registered in 9.5% of the cases. None of these features were appreciated in the present case. Lesions are generally painless and ulceration of the overlying oral mucosa occurs only when the

tumor interferes with dental occlusion. Growth may be rapid and infiltration of neighboring soft tissue structures may occur. When the maxillary sinus is involved, the odontogenic myxomas often fill the entire antrum.

In severe cases, nasal obstruction or exopthalmus may be the leading symptoms [10].

On conventional radiographs, myxomas of the jaw often show multilocular radiolucencies giving a honey comb, soap bubble or tennis racquet appearance, which helps in distinguishing this entity from malignant tumors which produce massive bone destruction [11]. Lesions greater than 4 cm tend to be multiloculated, and smaller lesions tend to be uniloculated [ 10].

CT findings in odontogenic myxoma are varied and may reveal osteolytic expansile lesions with mild enhancement of the solid portion of the mass. Some cases may show bony expansion and thinning of cortical plates with strong enhancement of the mass lesion. A soft tissue mass with bone destruction and fine lace-like density representing ossifications in the maxillary sinus may be seen in some cases. [12]

Radiological diagnosis of odontogenic myxoma is difficult because of overlapping features with other benign and malignant bone lesions. Some odontogenic myxomas may show a mixed radiopaque-radiolucent appearance which is ascribed to the presence of foci of calcification .Thus diagnosis should be considered in mixed radiolucent-radiopaque lesions [ 6,10] . Differential diagnoses including ameloblastoma, central giant cell granuloma, intraosseous haemangioma, aneurysmal bone cyst, glandular odontogenic cyst, cherubism, metastatic tumor and, in cases of unilocular lesions, simple cysts and odontogenic keratocyst. In older patients, the possibility of a malignancy should not be ruled out.

On gross examination of the specimen, the gelatinous, loose structure of the myxoma becomes obvious [13]. Microscopically, the myxoma is made up of loosely arranged spindle-shaped and stellate cells. The intercellular substance is mucoid. The tumor is usually interspersed with a variable number of tiny capillaries and occasionally strands of collagen [14]. In case of fibromyxoma, the amount of collagen in the mucoid stroma is more prominent. Remnants of odontogenic epithelium have occasionally been noted, sometimes being surrounded by a narrow zone of hyalinization. The microscopic features in this case were compatible with that of odontogenic myxoma, which is comprised of spindle or stellate cells in a mu-

coid material. Since it is an infiltrative, aggressive disease, with a high recurrence rate, treatment should aim at complete surgical excision of the mass along with excision of bony margins .In present case the entire tumour mass was excised in toto without resorting to a Weber Fergusson facial incision in the female patient.

Surgical excision is the recommended treatment ranging from conservative curettage [7,15] to radical excision [16]. However, owing to its potential for local infiltration, simple enucleation and curettage alone have been associated with a high recurrence rate of 10 to 33% [8,17].Thus radical excision with burring of the cavity borders with a drill should be performed with maximum preservation of surrounding structures.

## CONCLUSION

Clinical and radiographic characteristics of the odontogenic myxoma are variable, thus it should be always considered in the differential diagnosis of mixed and radiolucent lesions in maxilla of all age groups.

The treatment should consist of a wide local excision with burring of the cavity borders with a drill to minimize the chance of local recurrence along with regular follow up.

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