

## Odontogenic myxoma of posterior maxilla – A rare case report

Soundarya Ramesh<sup>1</sup>, Poornima Govindraju<sup>1</sup>, Balaji Pachipalusu<sup>1</sup>

<sup>1</sup>Department of Oral Medicine and Radiology, RajaRajeswari Dental College and Hospital, Bengaluru, Karnataka, India

### ABSTRACT

Odontogenic myxoma is a benign, rare neoplasm of mesenchymal origin comprising of 3%–6% of all odontogenic tumors. Odontogenic myxoma occurs more commonly in the second and third decade and is more commonly seen in mandible compared with maxilla but behaves more aggressively in maxilla as it spreads through the maxillary antrum. It is usually associated with a painless swelling without any symptoms. The radiographic features are similar to other odontogenic tumors, and hence, it leads to diagnostic dilemma. Therefore, thorough knowledge regarding clinical, radiographical, and histopathological features are important to arrive at a proper treatment protocol as it shows a high recurrence rate. The aim of this paper was to present a rare case of 21-year-old male with a chief complaint of swelling in the left maxilla that infiltrated the maxillary sinus in a very short duration.

**Keywords:** Infiltration, maxillary antrum, mesenchymal tumor, myxoma, odontogenic

### Introduction

Myxoma is a rare benign mesenchymal tumor, which may involve hard and soft tissue (heart, subcutaneous, skin). When it involves the bony tissue, it affects the facial bones.<sup>[1-3]</sup> Myxoma of the jaws was first reported in the 1950.<sup>[4]</sup> Myxoma of the jaws can be classified as osteogenic or odontogenic.<sup>[4]</sup> Odontogenic myxoma (OM) is a rare intra-osseous neoplasm, which is benign but locally aggressive in nature<sup>[5]</sup> and is characterized by mucoid or gelatinous grayish-white tissue that replaces the cancellous bone and expands the cortex.<sup>[6]</sup> The tumor develops from the ectomesenchymal portion of the tooth germ and shows an inactive effect of nests of odontogenic epithelium on mesenchymal tissue or as a direct myxomatous change in fibrous tissue; hence, it is also called OM.<sup>[5]</sup> It commonly occurs in the second and third decades, and mandible is involved more commonly than the maxilla predominantly in the premolar and molar regions.<sup>[6]</sup>

Clinically, OM is a benign slow growing painless mass, which causes asymmetry of the face. Cortical expansion and perforation are common findings. Radiographically, OMs appear as unilocular or multilocular radiolucency, sometimes showing soap bubble, tennis racquet, or honeycomb appearance occasionally with fine trabeculations. However, other radiological appearances such as “Sun-ray” appearance have also been reported in the literature root resorption is rarely seen although displacement of teeth is relatively common. It frequently shows aggressive infiltration of the adjacent tissues as well as tendency to re-occur after excision. Various odontogenic tumors represent the similar radiological behavior such as OM and biopsy are, therefore, necessary to ascertain an accurate diagnosis.<sup>[7]</sup>

OM is very unspecific in nature and it should be diagnosed primarily after thorough history, proper clinical, radiographical evaluation, and histopathological examinations. It has high recurrence rate; hence, proper follow-up for few years is mandatory.<sup>[2,3]</sup>

In view of its rarity, the present case of OM of left maxilla in a 21-year-old male infiltrating the maxillary antrum and adjacent tissues is herewith reported.

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**Address for correspondence:** Dr. Soundarya Ramesh,  
#14, Ramohalli Cross, Kumbalgodu, Bengaluru - 560 074,  
Karnataka, India.

E-mail: soundaryagr@gmail.com

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## Case Report

A 21-year-old male patient visited the Department of Oral Medicine and Radiology, RajaRajeswari Dental College and Hospital, with a chief complaint of painless swelling in his left cheek region since 3 months.

Patient presents with the history of extraction in his upper front tooth region 2 months back, followed by swelling, which was initially small in size, which gradually increased to its present size. There was no history of pain, pus discharge, bleeding, and trauma. Medical history and family history were noncontributory. On general physical examination, patient was moderately built and nourished.

On extraoral examination, solitary diffuse swelling was present on left middle third of the face measuring approximately 4 × 3 cm in dimension, extending superoinferiorly from infraorbital margin to corner of the mouth and mediolaterally from ala of nose to line drawn from outer canthus of the eye. Surface over the swelling appeared smooth and slightly glossy, surrounding skin was not stretched and no visible pulsation seen [Figure 1]. On palpation, swelling was soft to firm in consistency and nontender. Submandibular and submental lymphnodes are nonpalpable.

Intraorally, a well-defined swelling was present on upper left back tooth region measuring about 2 × 3 cm, extending antero-posteriorly from gingival margin of 24, 25, 26 to buccal vestibule of 24, 25, 26, and mediolaterally from distal aspect of 23 to mesial aspect of 28. Mucosa overlying the swelling appeared normal [Figure 2]. On palpation, swelling was firm to hard in consistency, nontender with smooth, well-defined margin. Other findings include periodontal pocket present irt 24, 25, 26. Hard tissue examination revealed grade 1 mobility irt 24, 25 with tenderness on percussion, supragingival calculus, and generalized white intrinsic stains.

Based on history and clinical examination, a provisional diagnosis of Adenomatoid odontogenic tumor irt 24, 25, enamel

hypoplasia, and chronic generalized gingivitis was considered. Differential diagnosis of Pindborg's tumor and keratocystic odontogenic tumor was considered.

Radiographical investigations included orthopantomograph, intraoral periapical radiograph (IOPAR), occlusal radiograph, and CBCT of maxilla.

Orthopantomograph revealed ill-defined radiolucency involving left maxilla extending mediolaterally from distal aspect of 23 to mesial aspect of 28 and anteroposteriorly from alveolar crest into the maxillary sinus [Figure 3]. Intraoral periapical radiograph of 24, 25, 26 revealed mixed radiolucency and radio-opacity with displacement of roots of 24 mesially and 25 distally [Figure 4], while maxillary occlusal radiograph revealed buccal cortical plate expansion [Figure 5].

CBCT of maxilla revealed, a well-defined osteolytic lesion seen irt left maxilla, which was roughly circular in shape, extending from 23 to 28 region mesiodistally, from superior wall of left maxillary sinus to crestal bone of inter premolar region superoinferiorly and from medial wall of the maxillary sinus to buccal cortical plate mediolaterally. Borders are thin, corticated, scalloped, and internal structure is mixed radiolucent-radiopaque. Internal structure shows thin, wispy septae and incomplete loculations with multilocular appearance seen irt 23 and 24 region. Roots of 24, 25, and 26 show mild resorption. Two-third of the maxillary sinus was involved with antral cloudiness and thinning of the antral walls. Roots of 24 and 25 appeared diverged. Lateral wall of the left nasal fossa is thinned out in 24 region. Palatal cortical plate is thinned out irt 24 and 25 region yet is intact [Figures 6-8]. So, the radiographical differential diagnosis of ameloblastoma and OM was considered.

Based on the clinical diagnosis of AOT, biopsy was performed. The microscopic examination of hematoxylin and eosin-stained section showed lesional tissue fragments composed of fibrocollagenous stroma with hypocellular myxoid areas and scattered band appearing spindle shaped cells [Figure 9]. No frank



**Figure 1:** Diffuse swelling present on left middle third of the face



**Figure 2:** Well defined swelling was present on upper left back tooth region art 24 25 26

pleomorphism or mitosis or necrosis was noted and the section was negative for granulomas or malignancy. Subsequently, the lesion was diagnosed as OM and surgical resection followed by prosthetic reconstruction was proposed.

### Discussion

According to WHO's classification of odontogenic tumors (1992), the myxoma belongs to the tumor of the odontogenic mesenchyme with or without the presence of odontogenic epithelium.<sup>[1]</sup> Virchow coined the term "myxoma" in 1863 and defined it as group of tumors that had histological resemblance to the mucinous substance of the umbilical cord.<sup>[2,3]</sup> Thoma and Goldman mentioned it first in the literature in 1947. Stout, in 1948, redefined the histological criteria for myxomas as true neoplasms that do not metastasize and exclude the presence of recognizable cellular components of other mesenchymal tissue, especially chondroblasts, lipoblasts, and rhabdomyoblasts.<sup>[6]</sup>

OM is an uncommon, nonencapsulated,<sup>[1]</sup> locally invasive benign tumor, slow growing with well-defined pathological characteristics and generally associated with tooth germ.<sup>[8,9]</sup> It represents <10% of all odontogenic tumors.<sup>[9]</sup> Myxomas occur very rare in head and neck region and it can be grouped

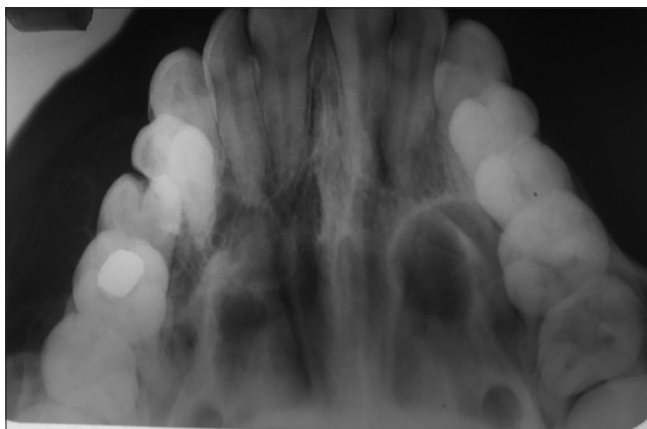
as (a) bone derived (osteogenic and OM) and (b) soft tissue derived (periolar soft tissue, parotid gland, ear and larynx).

The tumor most commonly occurs in the age group that varies from 22.7 to 36.9 years. It is rarely seen in younger and older individuals.<sup>[9]</sup> In the present case, the lesion occurred in a male patient of 21 year. Gunhan *et al.* and Regezi *et al.* reported a higher incidence of these tumors in women (64%–95%) than in men.<sup>[8]</sup> OM affects Mandible more than maxilla, especially the posterior region. According to Reichart and Philipsen, myxoma appeared more in mandible (66.4%) compared with maxilla (33.6%). Whereas 65.1% of the mandibular cases were located in the molar and premolar areas, 73.8% cases were seen in the same areas of the maxilla.<sup>[8]</sup> In this case, the lesion was located in the premolar and molar area of the maxilla.

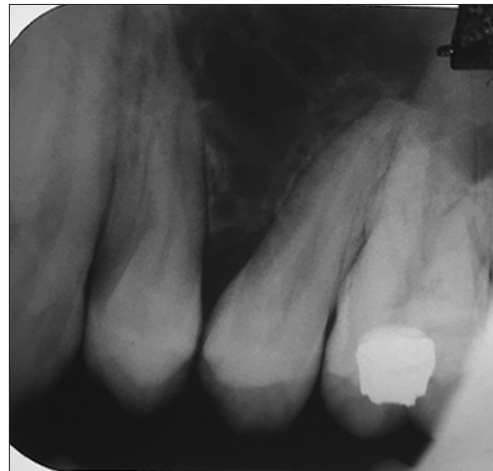
OM appears clinically as a slowly increasing swelling leading to asymmetry of the affected jaw. Lesions are generally symptomless



**Figure 3:** Orthopantomograph revealed ill-defined radiolucency involving left maxilla extending mediolaterally from distal aspect of 23 to mesial aspect of 28 and anteroposteriorly from alveolar crest into the maxillary sinus



**Figure 5:** While maxillary occlusal radiograph revealed buccal cortical plate expansion

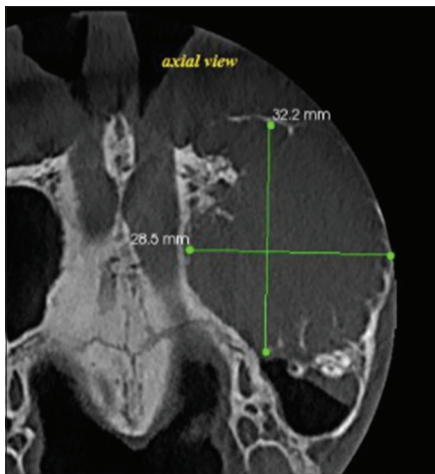


**Figure 4:** Intraoral periapical radiograph of 24, 25, 26 revealed mixed radiolucency and radio-opacity with displacement of roots of 24 mesially and 25 distally

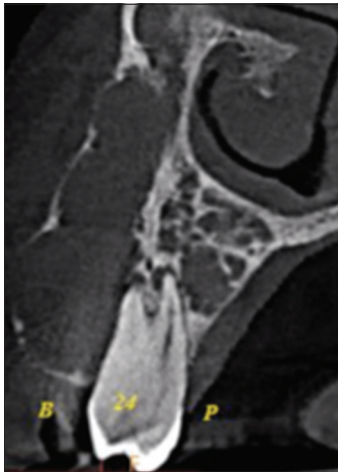


**Figure 6:** Lesion is roughly circular in shape, extends from 23 to 28 regions mesiodistally (4.5 cm), from superior wall of left maxillary sinus to crestal bone of inter premolar region superoinferiorly (4 cm), from medial wall of the maxillary sinus to buccal cortical plate mediolaterally (2.8 cm)

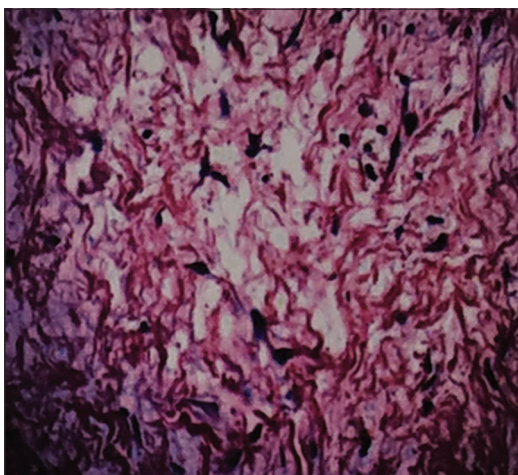




**Figure 7:** Lesion is roughly circular in shape, extends from 23 to 28 regions mesiodistally (4.5 cm), from superior wall of left maxillary sinus to crestal bone of interpremolar region superioroinferiorly (4 cm), from medial wall of the maxillary sinus to buccal cortical plate mediolaterally (2.8 cm)



**Figure 8:** Internal structure shows thin, wispy septae and incomplete loculations; multilocular appearance seen in 23 and 24 region



**Figure 9:** The microscopic examination of hematoxylin and eosin-stained section showed lesional tissue fragments composed of fibrocollagenous stroma with hypocellular myxoid areas and scattered band appearing spindle-shaped cells

and ulceration of the overlying oral mucosa only occurs when the tumor interferes with dental occlusion. OMs can grow rapidly but are rare and have been reported in the literature. Some patients present with progressive pain in lesions involving maxilla and maxillary sinus, with eventual neurological disturbance. Growth may be rapid and may infiltrate into the neighboring soft tissue. Both the buccal and the lingual cortical plates of the mandible may expand occasionally. OM of the maxilla behaves more aggressively than that of the mandible, as it spreads through the maxillary sinus as presented in our case. When there is involvement of maxillary sinus, the OM often fills up the entire antrum, which leads to nasal obstruction or exophthalmus as a leading symptom. Displacement of teeth has been registered in 9.5% of the cases. Similar features were appreciated in the present case.<sup>[8,10]</sup>

Myxomas of the jaws appear as multilocular or unilocular radiolucency on conventional radiographs. Unilocular radiolucencies are mostly found in the anterior region of the jaws, while multilocular lesions occur mainly in the posterior region.<sup>[6]</sup>

On computed tomographic images, OM can either appear as osteolytic expansile lesions with mild enhancement of the solid portion of the mass in the myxoma of the mandible, bony expansion and thinning of cortical plates with strong enhancement of the mass lesion in the anterior maxilla or a soft tissue mass with bone destruction and thinning and strands of fine lace-like density representing ossifications in the maxillary sinus.<sup>[6]</sup>

Magnetic resonance imaging (MRI) revealed a well-defined, well-enhanced lesion with homogeneous signal intensity on every pulse sequence. The lesion showed intermediate signal intensity on the T1-T2-weighted images. Unfortunately, MRI was not performed in this case.<sup>[6]</sup>

OM should be included in the differential diagnosis of both radiolucent and mixed lesions, in both the jaws, for individuals of all age groups. If it appears unilocular and without trabeculae, the tumor closely resembles periapical, lateral periodontal, and traumatic bone cysts and if it appears multilocular, it must be distinguished from ameloblastoma, central hemangioma, and odontogenic keratocyst. OM with multilocular radiolucencies represent “honey comb,” “soap bubble,” or “tennis racket” appearance, which help in distinguishing this entity from malignant tumors arising centrally within the jaw bones, because the latter usually cause massive bone destruction without compartments formed by bony trabeculations or bony septa.<sup>[8]</sup>

On gross examination of the specimen, the gelatinous, loose structure of the myxoma was obvious.<sup>[5,6]</sup> Microscopically, the myxoma is made up of loosely arranged spindle-shaped and stellate cells; many of which have long fibrillar processes that tend to intermesh. The intercellular substance is mucoïd. The tumor is usually interspersed with a variable number of tiny capillaries and occasionally strands of collagen.<sup>[7]</sup>

Farman *et al.* reviewed the histochemical findings in OMs. The ground substance of OM s has been shown to consist of about 80% hyaluronic acid and 20% chondroitin sulfate. Tumor cells appear to be relatively inactive, with low levels of oxidative enzymes. Tumor cells also show slight alkaline phosphatase activity. The myxoid intercellular matrix stains positively with alcian blue, but PAS staining may be negative.<sup>[7]</sup>

The myxomatous component of OM has been compared with the primitive mesenchyme that is found throughout the body. It has also been compared with the dental papilla and the dental follicle. OM tumor cells are mesenchymal in origin and express vimentin and muscle-specific actin. Conflicting description of S-100 and GFAP positivity has been reported. The matrix exhibits different proteins, mostly type-I and type-IV collagen, fibronectin, and proteoglycans.<sup>[6]</sup>

An extensive study on the ultrastructure of OM was published by Goldblat in 1976. Two basic types of tumor cells were described, secretory and nonsecretory. The secretory cell type was considered the principal tumor cell and resembled fibroblasts.<sup>[6]</sup>

The tumor is not radiosensitive, and surgery is the treatment of choice. Treatment of OM vary from local excision, curettage, or enucleation to radical resection.<sup>[10-12]</sup> The aggressive nature of OM is well documented in the literature. While generally considered a slow-growing neoplasm, OM may be infiltrative and aggressive, with high recurrence rates. Because of poor follow-up and lack of reports, a precise analysis of recurrence rates is still missing. Recurrence is considered to be directly related to the type of therapy, with conservative surgery resulting in a higher number of recurrences.<sup>[5,6]</sup>

## Summary and Conclusion

OM is an uncommon tumor of uncertain behavior. OM and other odontogenic tumors share common features on conventional radiographs that lead to a diagnostic dilemma. In order to establish a treatment protocol, various radiographical modalities can be used to determine the extent of the lesion. Histopathological examination is essential to provide conclusive diagnosis and treatment planning.

## Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other

clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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## Conflicts of interest

There are no conflicts of interest.

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