

# ODONTOGENIC MYXOMA- A CASE REPORT WITH EMPHASIS ON DIFFERENTIAL DIAGNOSIS

## Dentistry

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### Corresponding Author

Dr. Suhani S. Khundia  
Dept. No. 133, Oral pathology  
and Microbiology,  
Govt. Dental College and  
Hospital, Nagsevan,  
Dhanvantri Nagar, Aurangabad  
(M.H) – 431001  
Mob. Number – 7568798854  
E-mail – suhani.mds@gmail.com

Suhani S. Khundia<sup>1</sup>, Jyoti D. Bhavthankar<sup>2</sup>, Mandakini S. Mandale<sup>3</sup>, Vaishali A. Nandkhedkar<sup>4</sup>

<sup>1</sup> - Post graduate student, Dept. of Oral pathology and Microbiology, Govt. Dental College and Hospital, Nagsevan, Dhanvantri Nagar, Aurangabad (M.H)

<sup>2</sup> - Professor and head of the department, Dept. of Oral pathology and Microbiology, Govt. Dental College and Hospital, Nagsevan, Dhanvantri Nagar, Aurangabad (M.H)

<sup>3</sup> - Associate Professor, Dept. of Oral pathology and Microbiology, Govt. Dental College and Hospital, Nagsevan, Dhanvantri Nagar, Aurangabad (M.H)

<sup>4</sup> – Assistant Professor, Dept. of Oral pathology and Microbiology, Govt. Dental College and Hospital, Nagsevan, Dhanvantri Nagar, Aurangabad (M.H)

### Abstract:

Odontogenic myxoma is a rare intraosseous neoplasm considered to be derived from the mesenchymal portion of the tooth germ. It is benign but locally aggressive. Clinically, it is a slow-growing, expansile, painless, non-metastasizing present chiefly in the mandible. Here we report the case of a odontogenic myxoma in a 14-year-old male patient, in which extraoral swelling present on left side of face intraoral swelling seen extending from 33 to 35 since 5 months.

**Keywords:** Odontogenic Myxoma, benign intraosseous tumor, Differential Diagnosis

### Introduction:

Odontogenic myxoma is rare benign intraosseous tumor.<sup>1</sup> It contributes only 3–11% of all odontogenic tumors.<sup>2,3</sup> Odontogenic myxoma most frequently occurs in second or third decades of life,<sup>4</sup> involves the mandible more commonly than the maxilla and has a slight female predilection.<sup>5</sup> Soft tissue myxomas are frequently found, but their intraosseous counterparts are rarely encountered, and those that are found are almost exclusively confined to the jaws. Myxomas have also been found in other body parts such as skin, subcutaneous tissue and the heart (left atrium) than the jaws.<sup>2</sup> This neoplasm is thought to be derived from mesenchyme of periodontal ligament.

Clinically, it is a slow-growing, expansile, painless tumor. Other clinical manifestations include, tooth mobility, bone expansion, cortical destruction and facial distortion.<sup>6,7</sup> Radiographically, the classic presentation is ‘Tennis-racquet’ or ‘Step-ladder’ pattern with well-developed locules, consisting of fine trabeculae, arranged at right angles and in some cases root resorption is also seen. A ‘sun-burst’ or ‘sun-ray’ appearance has also been reported in the literature.<sup>8,9</sup> Histopathologically, the lesion consists of loosely arranged spindle, stellate-shaped or round cells, in an abundant myxoid stroma.<sup>9</sup>

Odontogenic myxoma is locally aggressive and has potential for persistent local growth and bone destruction. This may result into recurrence if resection is inadequate. Herewe report this case in order to put forth

a case of a benign tumor, i.e., odontogenic myxoma in a 14 year-old male, involving mandible which grew, within a short span of five months.

### **Case Report:**

A 14-year-old male patient reported with the chief complaint of a painless gradually progressive swelling in the lower left side of face, which was initially noticed as a small swelling in the lower left vestibule and grew to the present size within a span of 5 months. The patient gave no history of trauma. Non-contributory past medical and dental history.

Extra-oral examination revealed a diffuse, non-tender, bony hard swelling on the left side of face extending from lower lip to below the lower border of mandible, approx. 3-4 cm in size [Figure 1]. Color of the overlying skin was normal and no local rise in temperature and lymphadenopathy was observed. Slight facial asymmetry was seen. On intra-oral examination, a single diffuse localized swelling at left labial as well as lingual vestibular region, extending from 33,34 and 35 region, with displacement of 33,34 and 35 was observed. Obliteration of the buccal vestibule and floor of mouth. Limited movement of tongue, tongue-tie was present. There was no erythema, ulceration or pus discharge in the overlying mucosa [Figure 2].

Radiographically, the Orthopantomograph (OPG) showed a multilocular radiolucency, extending antero- posteriorly from the 33,34,35 and 36 and supero-inferiorly from the alveolar process to the lower border of the mandible. Poorly defined radiolucent lesion causing slight cortical expansion, with fine trabeculae, some lines intersecting at right angles, were also evident. Displacement of 34 and 35 was also apparent [Figure 3].

The lesion was biopsied and submitted to the Department of Oral Pathology & microbiology with the provisional diagnosis of central giant cell granuloma. On macroscopic examination, the biopsy specimen was in multiple pieces 1cmx0.5cm in size, whitish in color, transparent and slippery in nature and soft in consistency [Figures 4 and 5]. On microscopic examination, H and E stained section showed haphazardly arranged spindle and stellate shaped cells with long fibrillar processes in abundant loose myxoid stroma with fine collagen fibrils. Peripherally at focal places, fibrocellular connective tissue with bony trabeculae

and thickened and dilated blood vessels are evident [Figure 6 and 7]. Overall features suggestive of odontogenic myxoma to be the definitive diagnosis.

### **Discussion:**

Odontogenic myxoma is a rare benign intraosseous neoplasm, with exceptionally high local aggressiveness, high recurrence rate upto 25% is seen and non-metastasizing nature, on this account it is called 'locally malignant'. Majority of the investigators found that this lesion occurs in second or third decade of life. Zimmerman et al. reported that the average age for the odontogenic myxoma is 26.5 years.<sup>10</sup> There is a slight female preponderance and mandibular predilection suggested in many reports.<sup>11,12</sup> Aggressive lesion, involving most of the mandible within a short span of five months. Another interesting finding was that it did not cause much of a cortical expansion or gross facial deformity, displayed scalloping between the roots of the involved teeth and appeared to be invading the bone antero-posteriorly.

Radiographically, the appearance of an odontogenic myxoma may vary from a unilocular radiolucency to a multilocular lesion, with a well defined to diffuse margin. However, a unilocular appearance is more common among children and in anterior parts of the jaws. Sometimes giving a false appearance of an OKC as the tumor is often scalloped between the roots and root resorption may occur.<sup>13</sup> The bony trabeculae of a multilocular radiolucency intersect at right angles and the lesion causes root resorption, resulting in tooth mobility were the classical radiographic features of the odontogenic myxoma.<sup>14</sup> The typical soft, slippery and gelatinous nature of the specimen on macroscopic examination<sup>14</sup> and the histopathological findings were also in accordance with the literature. All these features were evident in our case except for male preponderance. Thus our case was of odontogenic myxoma.

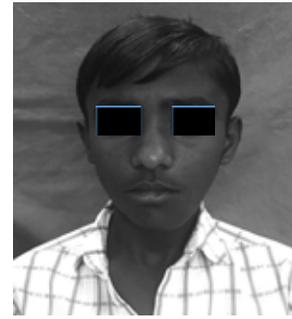
Clinical and radiographic differential diagnosis of odontogenic myxomas may include: intraosseous hemangioma, cherubism, aneurysmal bone cyst, fibrous dysplasia, ameloblastoma, central giant cell granuloma, traumatic bone cyst.<sup>15,16</sup> Based on the age of occurrence and the site a clinical diagnosis of odontogenic myxoma was made. Radiographically odontogenic myxoma appears as unilocular or multilocular radiolucencies with well-defined margins and fine bony septa. The current case had

multilocular appearance. Generally diagnosis is based on the histopathologic feature. Histologically, differential diagnosis must be made with rhabdomyosarcoma, myxoid liposarcoma, neurogenic sarcoma, neurofibroma, chondromyxoid fibroma and nodular fasciitis.<sup>17,18</sup> Failure to demonstrate nuclear contour of Schwann cells and neuritis, neurogenic sarcoma and neurofibroma was ruled out. As well as absence of elongated eosinophilic cells with cross striations, adipocytes, chondroblasts, chondrocytes and foci of calcification rhabdomyosarcoma, liposarcoma, chondromyxoma were also ruled out. The presence of loose spindle shaped and stellate fibroblasts with basophilic ground substance gives confirmatory diagnosis of odontogenic myxoma.

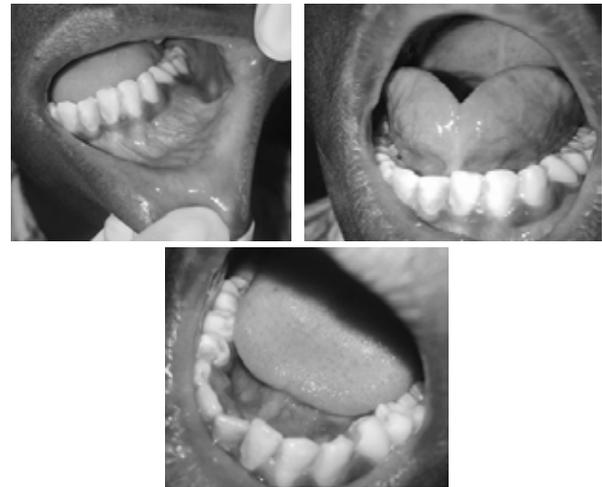
To establish a precise diagnosis immunohistochemical is most reliable for differentiating odontogenic myxoma from other lesions. The tumoral cells usually express SMA, vimentin, desmin and Ki-67. Vimentin as a fibroblastic marker, desmin stained for the confirmation of differentiation of spindle-shaped cells and Ki-67 utilized as an indicator of biological behavior of disease and an important biomarker related to prognosis. A possible recurrence will be seen later on and treatment will be impaired. For all these reasons, a resection with broad margins is the most indicated treatment.<sup>19</sup> Post-operative preservation of patients with odontogenic myxomas is indefinite, especially in the first two years as it is considered to be period of greatest recurrence rate.

### Conclusion:

The WHO and many authorities consider this neoplasm to be of odontogenic mesenchymal origin. On the basis of its common site of occurrence (tooth-bearing areas of jaws), the sporadic presence of odontogenic epithelial islands and the striking histological resemblance to dental mesenchyme usual age of occurrence second or third decade of life and female predilection is seen. Our case showed male predilection. Recurrence rate of 25% is seen so it is considered to be aggressive and hence the prognosis is compromised.



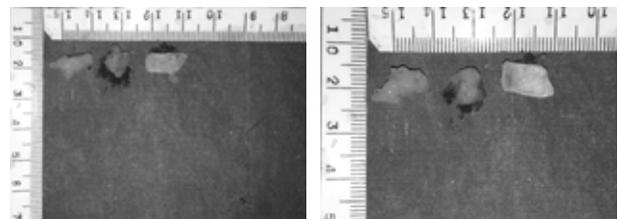
**Figure 1: Extraoral view showing a diffuse swelling on the left anterior region of mandible resulting in a slight facial asymmetry**



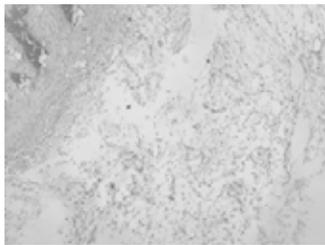
**Figure 2: Intraoral view showing swelling at left buccal as well as lingual vestibular region, extending from 33 to 35 region, with displacement of 34 and 35**



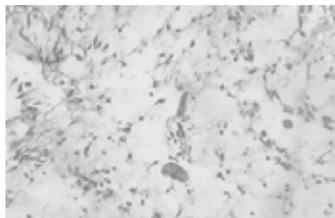
**Figure 3: OPG showing a poorly defined multilocular radiolucency, extending from the midline to left ramus region causing cortical expansion and displacement of 33,34 and 35 as well as root resorption of 34 and 35**



**Figure 4 and 5: On macroscopic examination, the biopsy specimen in multiple pieces 1cmx0.5cm in size, whitish in color, transparent and slippery in nature and soft in consistency**



**Figure 6:Low-power photomicrograph illustrating loosely arranged stellate- shaped cells with few collagen fibrils and capillaries (H and E ×10)**



**Figure 7:High-power photomicrograph showing loosely arranged stellate- shaped cells with intermingled fibrillar processes in a homogenous mucoïd ground substance (H and E ×40)**

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