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Giant Odontogenic Maxillary Myxoma: A Rare Case Report

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ABSTRACT
Odontogenic myxoma is an uncommon benign neoplasm comprising of 3-6% of all odontogenic tumours. Being a locally aggressive tumour, its diagnosis poses a challenge with overlap between benign and malignant neoplasms. Aggressive infiltration of the adjacent soft tissues with increased tendency of recurrence are its characteristics features. We present a rare case of Odontogenic Myxoma occurring in the maxilla of a 35 year old male patient. The clinical, radiological, histopathological and immunohistochemical characteristics of Odontogenic Myxoma are reviewed. Complete surgical excision is the treatment of choice but it can be challenging because of tumour's indistinct margins. Though there are still no clear guidelines for management of Odontogenic Myxoma in the head and neck region, the general consensus is that the surgical excision should be complete and patients treated in a conservative manner should benefit from regular follow-ups.

INTRODUCTION

Myxomas are a slow growing, benign, locally aggressive tumor of mesenchymal origin. It can arise from both the hard and soft tissues which includes skin, subcutaneous tissue and heart. Myxoma of the jaws can be classified as osteogenic or odontogenic. Odontogenic myxoma (OM) is a rare intra-osseous neoplasm that develops from the ectomesenchymal portion of the tooth germ and shows an inactive effect in the form of nests of odontogenic epithelium on mesenchymal tissue or as a direct myxomatous change in fibrous tissue. Myxoma of jaws is generally regarded as of tooth germ cell origin and specifically derived from dental papilla. Hence it is called as odontogenic myxoma and is associated with malformed or missing teeth[1]. Odontogenic myxoma frequently occurs during second or third decades of life. Myxoma can occur anywhere in the jaws but have a predilection for the molar and premolar regions of mandible and maxilla[2]. It is a benign, slow-growing and locally aggressive mesenchymal neoplasm[3]. Cortical expansion with perforation and replacement of cancellous bone are common findings in Odontogenic Myxoma[2].

Odontogenic myxoma is a rare benign neoplasm comprises of 3-6% of all odontogenic tumours. It most commonly arises in mandible (66.4%) followed by maxilla (33.6%). They may grow to a large size and lead to marked facial deformity. Odontogenic myxomas are locally invasive nonmetastasizing neoplasms of the jaws, almost exclusively seen in tooth bearing areas[2]. Characteristic gross features include mucoid or gelatinous grayish-white tissue which replaces cancellous bone and expands the cortex. Often, Maxillary Myxoma extend into the sinus. Radiographically, Odontogenic Myxomas appear as unilocular or multilocular radiolucent tumor. The various appearances on radiology include soap bubble, tennis racquet, honeycomb, sunray appearance occasionally with fine trabeculations. Microscopically, odontogenic myxoma contains loose stellate cells with long, branching, eosinophilic cytoplasmic processes that are set within a paucivascular myxoid stroma. Occasionally, strands of odontogenic epithelium which may represent rests of Malassez are found. This infiltrative tumour often entraps spicules of bone trabeculae[1].

A comprehensive diagnosis is attained by taking detailed history along with proper clinical and radiographical evaluation. However histopathological examination stays as the gold standard for definitive diagnosis. Its high recurrence rate mandates proper follow-up for few years in all cases.

In this article, we describe a rare case of odontogenic maxillary myxoma presented in a 35 years old male patient. To the best of our knowledge, this case is the largest one in terms of size measuring 17×8×6.3 cm reported in the literature.

CASE REPORT

A 35year old male came with complaints of painless swelling in the right cheek for 2 years causing facial deformity. The swelling was initially small in size and has slowly increased to reach the present size without any pain.

Intraorally, a well-defined swelling was present on right upper alveolus anteriorly from 1st premolar, posteriorly up to 3rd molar region medially extending over the hard palate for 2 cm and laterally involving the gingiva buccal sulcus. There is no ulceration. On palpation, swelling was firm to hard in consistency, nontender with smooth, well-defined margin.

On extraoral examination, diffuse swelling was present over the right cheek in maxillary region, extending superioinferiorly from infraorbital margin to upper lip and mediolaterally from ala of nose to outer canthus of the eye. Skin is pinchable over the swelling. Submandibular and submental lymph nodes are nonpalpable (Fig. 1).

On contrast enhanced computed tomography a locally aggressive expansile hypodense lesion of size 6.6×5.3×5.9 cm was noted involving the alveolar and palatine processes of right maxilla with lysis of the surrounding bone. Right Subtotal maxillectomy with post segmental mandibulectomy was done. Gross examination of the resected specimenshowed a huge lobulated tumour measuring 17×8×6.3 cm involving right maxilla, upper alveolus, gingivae with adjacent soft tissue, crossing the midline of the hard palate. Cut surface of the tumour was myxoid and glistening. Histopathological examination showed spindle shaped cells with long branching cytoplasmic processes dispersed in an abundant, amorphous, faintly basophilic myxoid matrix (Fig. 2).

DISCUSSIONS

The term “Myxofibroma” was coined by Rudolf Virchow in 1863 for a group of tumours that had histologic resemblance to the mucinous substance of umbilical cord[2-6]. Odontogenic Myxoma was first mentioned in the literature by Thoma and Goldman in 1947[6]. 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[2-4]. Myxoma is a tumour that can be found in heart, skin and subcutaneous tissue and centrally in the bone[2].

A worldwide estimated incidence of Odontogenic Myxoma ranges from 0.5-17.7% for all odontogenic tumours of the jaw[7]. In majority of cases, almost 75%, by Singaraju *et al*.[2] and Kaffe *et al*.[8] these tumours occur in the second, third and fourth decade. The exact prevalence of Odontogenic Myxoma in children is deemed to be under 10% but is still considered higher

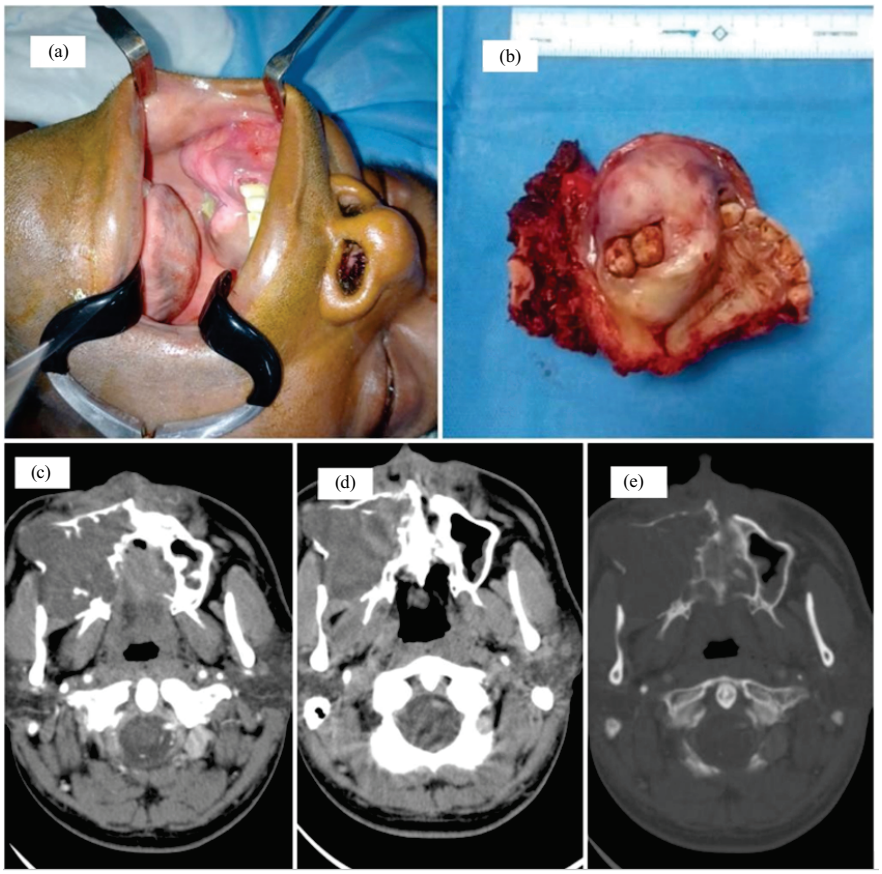

Fig. 1(a-e): (a, b) Infiltrative growth involving the right maxilla, upper alveolus, gingivae with adjacent soft tissue, maxillary sinus and crosses the midline of hard palate, (c-e) On radiology a locally aggressive expansile hypodense lesion of size 5.3×5.9×6.6 cm noted involving the alveolar and palatine processes of right maxilla

than that of other aggressive tumours[7]. Myxomas of the head and the neck are rare tumours. Two forms can be identified:

- Facial bone derived, which had been subdivided in the past into true osteogenic myxoma and odontogenic myxoma
- Soft tissue derived myxoma derived from the perioral soft tissue, parotid gland, ear and larynx[2]

Posterior mandible is the frequent location of Odontogenic Myxoma[7]. Odontogenic myxoma involving the maxilla, can expand to a significant size within the maxillary sinus and remain undetected until it invades the surrounding palate, orbit, or nasal cavity. Since Odontogenic myxoma is a painless slow growing tumour, it is often asymptomatic, discovered fortuitously on a radiograph, or sometimes results in a painless facial swelling or deformation, increasing regularly in volumesimilar to the presenting complaints of our patient[7]. The maxillary sinus and the hard palate can be infiltrated by the process[7]. Other symptoms include dental mobility, abnormalities in dental development, disturbance of mastication or speech, pain and parasthesia[7]. Singaraju *et al*.[2] and Liu *et al*.[9] supported the notion of odontogenic origin of myxomas by suggesting that fibroblasts that compose the tooth germ undergo modification to give rise to odontogenic myxoma.

The myxoma of the maxilla and mandible is considered as a neoplasm of odontogenic origin traditionally. The support of an odontogenic origin has been perpetuated by its almost exclusive occurrence in the tooth bearing areas of the jaw, its common association with an unerupted tooth or a developmentally absent tooth, its frequent occurrence in young individuals, its histologic resemblance to dental mesenchyme-especially the dental papilla and the occasional presence of sparse amounts of odontogenic epithelium[2].

The radiographic aspects of Odontogenic Myxoma vary from small unilocular radiotransparent lesions to large multilocular lesions, possibly displacing the corresponding teeth[7]. The specific diagnosis of Odontogenic Myxoma can be suspected in the

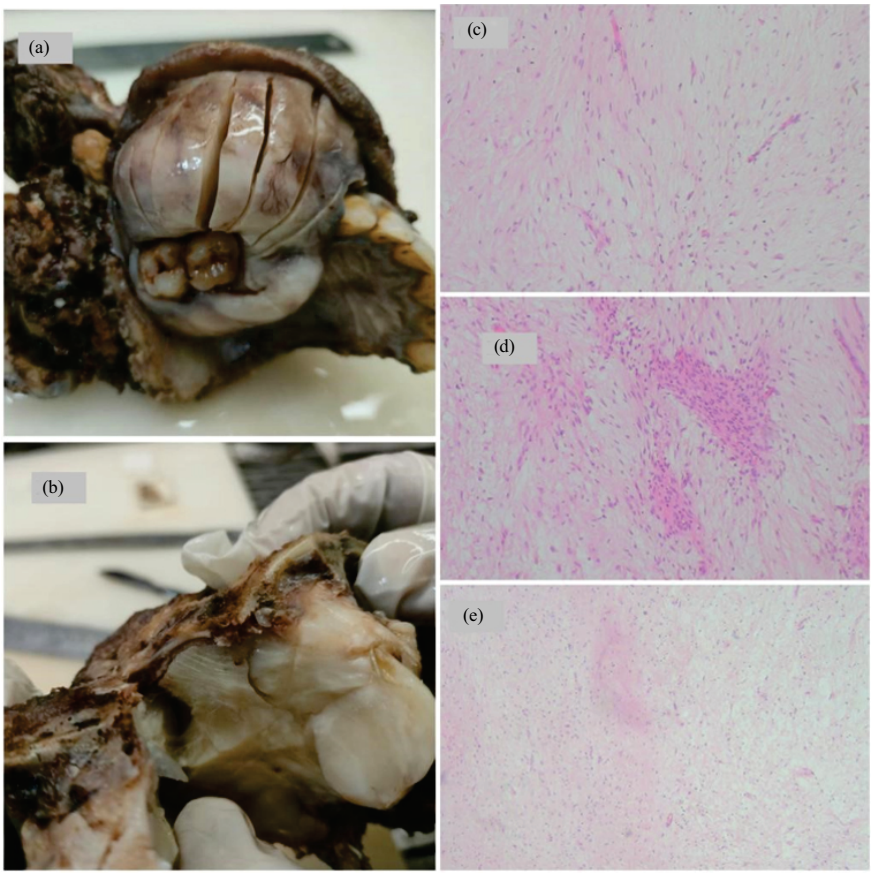

Fig. 2(a-e): (a,b) Gross appearance-The tumour is lobulated, whitish, myxoid and glistening in appearance, (c-e) Microscopic picture, H and E 4x, 10x and 40x showing stellate cells dispersed in an abundant faintly basophilic myxoid matrix

presence of fine, angular septa, visible on the panoramic radiograph and realising an aspect of a soap bubble, a honeycomb or tennis racket strings[7]. Though CT (Computed Tomography) imaging is non-specific in the diagnosis of Odontogenic Myxoma, it is of great topographic interest, as it allows the assessment of the extent of the lesion as well as the presence of cortical perforation which is essential in the planning of surgery[7]. Magnetic Resonance Imaging is more efficient and superior to CT imaging in the positive diagnosis of Odontogenic Myxoma, especially in the evaluation of soft tissue involvement and in the differential diagnoses between Odontogenic Myxoma and Ameloblastoma[7].

Gross examination of the specimen shows gelatinous loose structure of myxoma[2]. Liu *et al*.[10] described the size of the surgically resected mass macroscopically as ~15×16×16 cm. Tumour size in our patient was 17×8×6.3 cm. Microscopically, the Odontogenic Myxoma is bland in appearance and composed of loosely arranged evenly dispersed spindle shaped, rounded and stellate cells many of which have long fibrillar processes that tend to intermesh with a lightly eosinophilic cytoplasm in a rich mucoid intercellular matrix[7]. Although, some degree of mild nuclear pleomorphism or hyperchromatism may exist, including an occasional mitosis or binucleate cell, there is no proven correlation between the presence of these particularities and the recurrence of Osteogenic Myxoma[7]. The tumour is interspersed with a variable number of tiny capillaries and occasionally strands of collagen[2]. In case of fibromyxoma, the amount of collagen in the mucoid stroma is more prominent. The fibrils are shown by silver impregnation to be reticulin. The composition of the ground substance is found to be 80% hyaluronic acid and 20% chondroitin sulphate according to Singaraju *et al*.[2]. Tumour cells appear to be relatively inactive, with low levels of oxidative enzymes. Tumour cells also show slight alkaline phosphatase activity. The myxoid intercellular matrix stains positively with Alcian blue but PAS staining may be negative. Odontogenic myxoma tumour cells are mesenchymal in origin and express vimentin and muscle-specific actin. Conflicting description of S-100 and GFAP positivity is reported[2]. The matrix exhibits different proteins mostly type I and type IV collagen, fibronectin and proteoglycans[2].

The differential diagnosis includes: Odontogenic tumours, such as ameloblastoma, odontogenic fibroma, dentigerous cyst, odontogenic keratocyst and Non-Odontogenic tumours, such as central giant cell granuloma, ossifying fibroma, haemangioma, fibrosarcoma, chondrosarcoma and osteosarcoma[7].

Till date, there is no consensus for the management of Odontogenic Myxoma. Being resistant to radiation, the recommended treatment is surgery, either radical resection or a conservative approach depending on the size of the tumour[7]. If diameter is less than 3 cm, conservative treatment by enucleation and curettage is done but since myxomas are not encapsulated and tend to infiltrate the surrounding bone, a more extensive resection than curettage and peripheral osteotomy is often needed for larger lesions[7]. Other therapeutic options include radiotherapy and cryotherapy but these are markedly less effective than surgery[7].

The recurrence rate of Odontogenic Myxoma seems to be largely related to the treatment method, rather than an inherent behaviour of the tumour[7]. Although conservative treatments are less invasive and better tolerated by the patient, they present a high risk of recurrence of 10-30%, because complete resection of the myxomatous tissue can be difficult to achieve.[4] Maxillary Odontogenic Myxoma is also more likely to recur compared to mandibular Odontogenic Myxoma[7]. A clinical and radiographic surveillance is recommended for atleast 5 years after treatment, with a close follow up during the first 2 years after surgery, when Odontogenic Myxoma is most likely to reappear[7].

CONCLUSION

Maxillary Myxoma is a benign but locally aggressive tumour, positive diagnosis is received on histologic specimen analysis. Since it is rare, there is still no clear guidelines as to the management of Odontogenic Myxoma. Early detection and intervention with careful periodic evaluation may help avoid aggressive treatment methods and their morbidity[7]. In respect of biological behaviour and extensiveness of the lesion, correlation of clinico-radiographic appearance with histologic counterpart are mandatory for such lesions to avoid controversies and to reach the final diagnosis and to prevent further recurrences[3].

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