Case Report

Odontogenic myxoma: A causality dilemma – Report of a nonpareil case and review of literature

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Abstract

Odontogenic myxoma is a benign locally aggressive neoplasm with a sparse prevalence and incertitude histogenesis. They constitute 3%–6% of odontogenic tumors in gnathic bones. It is ubiquitously seen between vicenarian to early quadragenarian group with female proclivity and fondness to the mandibular jaws. They are silent lesions clinically and show myxoid stroma amidst fibrous background. This report highlights central odontogenic myxoma in a 43-year-old male patient and focuses on concepts, differential diagnosis, molecular concepts and treatment aspect.

Keywords: Aggressive neoplasms, myxoma, odontogenic tumors

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INTRODUCTION

Odontogenic myxoma belongs to a rare group of odontogenic neoplasms affecting the jaw bones. They arise from the elements of embryonic dental anlage of mesenchymal tissue. The controversy on the pathogenesis of these tumors continues to entice the pathologists until date. The World Health Organization (WHO) grouped them as tumors of ectomesenchymal origin with or without odontogenic epithelium.[1] The term "myxoma" was originally coined by Virchow and was later defined by Stout.[2] WHO categorized myxomas as central and peripheral variants. The peripheral lesions are comparatively less aggressive and encapsulated. In contrary, central odontogenic myxomas are generally nonencapsulated tumors with infiltrative capacity into the adjacent medullary bone.[3] Histologically, myxomas show delicate fibrous to loose mucoid stroma, this is due to the presence of undifferentiated mesenchymal cells showing fibroblastic differentiation.[2-5] When dense collagenous stroma is evident the term-fibro myxoma/myxofibroma can be used interchangeably.[3] Due to this dual pattern, the odontogenic myxoma is believed to be a continuum of odontogenic fibroma.[3] The present report highlights a rare entity of odontogenic tumors, i.e., central odontogenic myxoma in a 43-year-old male patient who presented with painless swelling over left posterior cheek region. Based on clinical, radiological and histopathological findings, the diagnosis of odontogenic myxoma was made. A bird's eye view on concepts, differential diagnosis, molecular concepts, and treatment aspects were discussed.

CASE REPORT

A 43-year-old male patient reported with a chief complaint of painless swelling over left cheek region.

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Clinical examination revealed facial asymmetry; the swelling measured 6 cm × 3.5 cm, which was extending from the corner of mouth anteroposteriorly to middle portion of the body of the mandible. On intraoral examination, the lesion extended from the mesial end of 37–33 region, mobility was observed with 35 appears to be floating within the lesion [Figure 1]. On palpation, the lesion was soft to firm in consistency, nontender and with smooth margins. Correlating clinically a provisional diagnosis was made as "ameloblastoma." The further patient was advised for radiographic investigations and routine hematological examination before incisional biopsy. ortho pantamo graph (OPG), computed tomography (CT) scan was taken, OPG revealed multilocular radiolucency with soap bubble appearance extending from the distal root of 37–41 regions. The teeth involved in the lesional area showed displacement and mild root resorption, along with bony erosion in left mandible body region [Figure 2]. CT revealed radiolucency extending from the distal root of 37–41 regions with perforation of cortical plates [Figure 3]. Further to confirm diagnosis incisional biopsy was done under local anesthesia and sent for histopathological examination. On gross examination, the tissue bits were approximately measuring 2 cm × 2.5 cm, creamy white, round to oval in shape, soft gelatinous in consistency [Figure 4]. On histopathological examination, hematoxylin and eosin-stained sections showed spindle or stellate-shaped mesenchymal cells seen in loose myxoid stroma with few collagen fibrils, interspersed with odontogenic islands [Figure 5]. Histochemical staining of mucoid stroma with alcian blue stain at pH2.5 showed positivity of the stromal component indicating the presence of acidic glycosamino glycans within the mucoid stroma confirming the diagnosis of odontogenic myxoma [Figure 6]. Further immunohistochemical analysis using BCl2 marker showed negativity of tumor cells [Figure 7]. Poor Bcl-2 staining which is indicative of lack of tumor aggressiveness favored the complete resection of tumor in toto. *En bloc* resection of the tumor mass was done. Two-year follow-up showed no evidence of any recurrence.

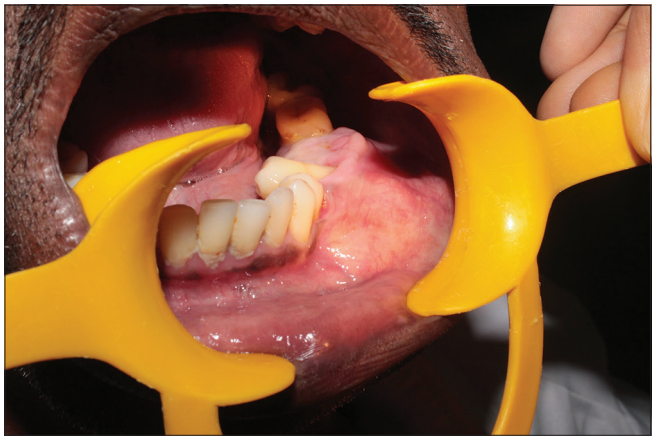

Figure 1: Intra oral extension of lesion with floating premolar within the lesion

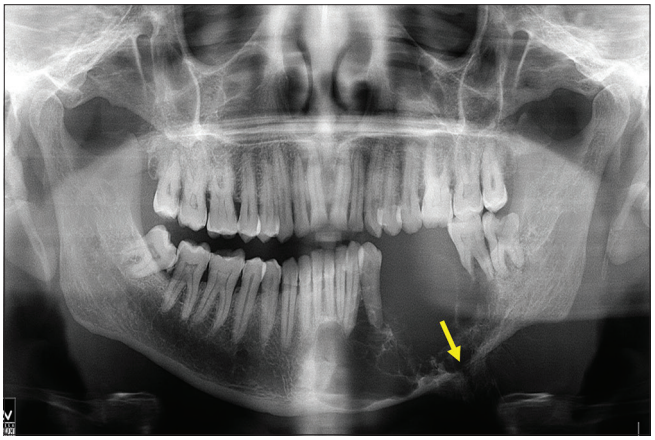

Figure 2: Ortho pantamo graph showing soap bubble appearance, and arrow mark showing eroded bone

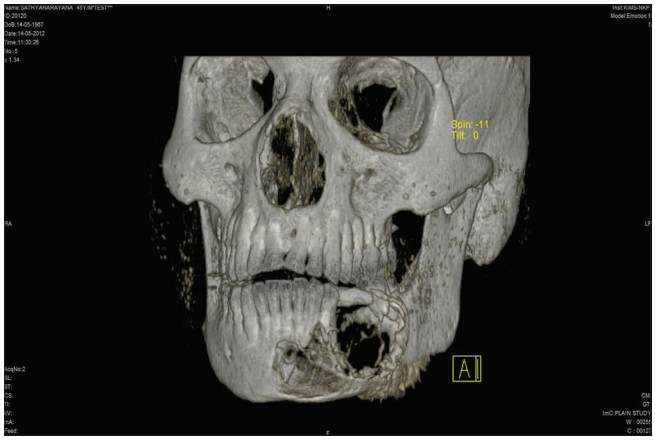

Figure 3: Computed tomography image showing perforation of cortical plates of mandible

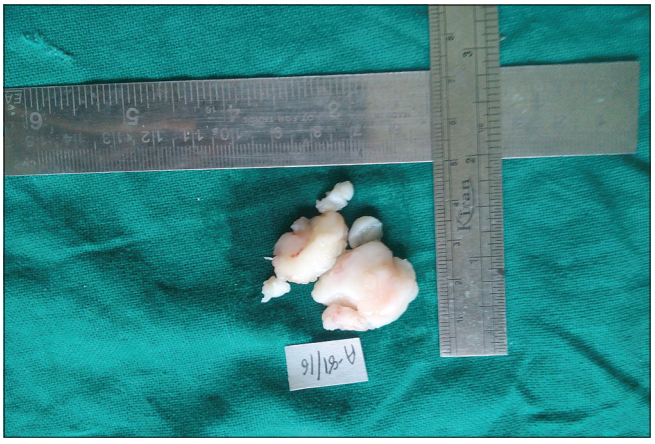

Figure 4: Gross image showing glistening gelatinous creamy white tissue bits

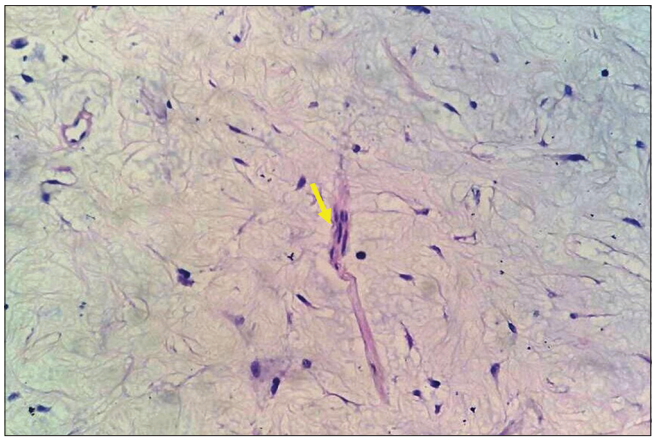

Figure 5: Spindle- or stellate-shaped mesenchymal cells within loose myxoid stroma. Arrow representing odontogenic epithelial islands (H&E, ×40)

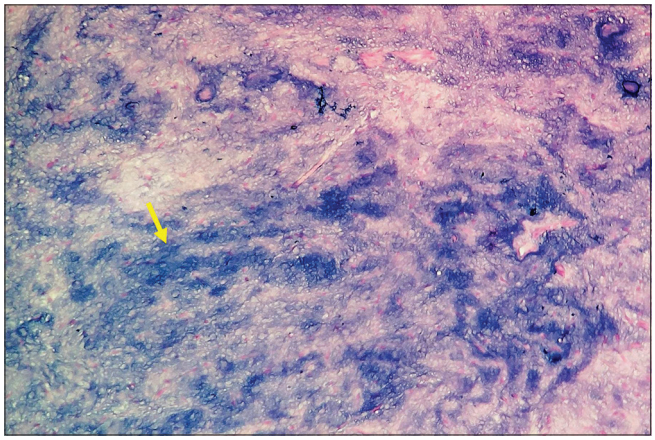

Figure 6: Arrow mark showing alcian blue positive mucoid material

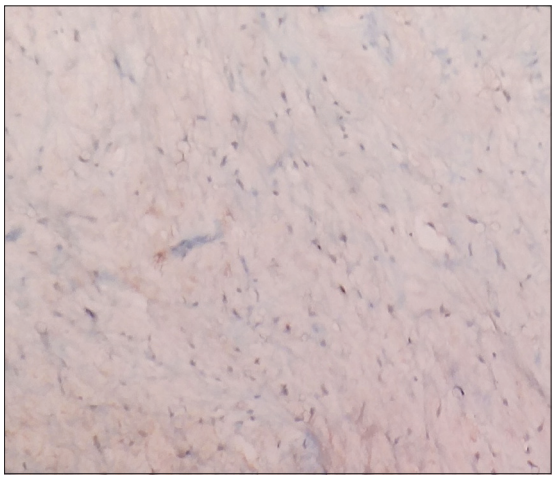

Figure 7: Photomicrograph showing negative reactivity for BCl2 marker

DISCUSSION

Odontogenic myxoma was first described in 1947 by Thoma and Goldman as a rare benign tumor of tooth-bearing areas.[6] Root resorption and displacement is uncommon unless the tumor grows to larger size. The present case showed mild root resorption and displacement of teeth involved in the lesion, these findings were similar to that of the case reported by Altug *et al*.[7] The overall rate of prevalence is about 0.04% to 3.7% in general race and ethnicity. Marked female predilection is the most common feature reported in several studies,[2,5,7,8] but the present case was reported in a 43-year-old male patient which contradicts the other reported cases. Radiographically, classic description of honeycomb/tennis racquet pattern is appreciable in many cases.[3-7] In the present case, multilocular radiolucency with soap bubble appearance was appreciated, and similar findings were reported by Manne *et al*.[5] The radiographic differentials include ameloblastic fibroma, ameloblastoma, odontogenic fibroma, central hemangioma and odontogenic keratocyst, glandular odontogenic cyst, cherubism, aneurismal bone cyst.[5,7-10]

The histogenesis of the myxoid tumors is debatable till date, i.e., whether they are of odontogenic or nonodontogenic origin. Sivakumar *et al*. have carried out immunohistology on the stromal components and stated that the duality in origin of myxomas is mainly attributed to its fibroblastic-histiocytic origin.[9] This causal duality arises either due to their fibroblastic origin or their ability to secrete excess mucopolysaccharides making them histogenetically related to myxoid tumors.[3] This put forward the very existence of the first event (the cause) and the second event (the effect), where the second event is a consequence of the first. This raises a familiar doubt of "Chicken and Egg" situation with histogenesis of these tumors. In 1948, Stout redefined the histopathology of myxomas as lesions that do not have cellular elements of skeletal muscle, adipose or cartilage.[2] The myxomas are classified into two types: (1) facial bone-derived which are subclassified into true osteogenic myxoma and odontogenic myxomas and (2) soft-tissue myxomas of larynx, parotid and the ear.[2] Histologically, the myxomas are bland appearing tumors with stellate-shaped cells with mucoid rich matrix and pale staining eosinophilic cytoplasm. Some areas may show mild pleomorphism which does not relate to the rate of recurrence of these tumors. In the present case to confirm mucoid reaction alcian blue staining was done which showed positive staining, this finding is in accordance with the results of Kiresur and Hemavathy.[11]

The structure commonly mistaken histopathologically for odontogenic myxoma is the developing dental papilla, which has the immature mesenchymal tissue which develops into future pulp. Dental papilla is composed of plump, stellate, and fusiform fibroblastic cells set in a myxoid matrix with delicate collagen fibers. This tissue,

however, is lined focally, by a rim of odontoblasts at focal margins. This feature with radiographic appearance, distinguishes dental papilla from odontogenic myxoma. The dental follicle with a myxoid stroma can mimic the odontogenic myxoma. The presence of reduced enamel epithelium can distinguish the follicle from myxomas. Embryonal rhabdomyosarcoma can sometimes show myxoid areas, but the age of the patient, and lack of strap cells in the present case favored to give a diagnosis of odontogenic myxoma. The central myxoid neurofibroma shows prominent mast cell population, and zones showing parallel streams of collagen that organizes into fascicles. Other bony lesions that show myxoid component include chondromyxoid fibroma and myxoid chondrosarcoma. Both of these should show focal evidence of chondroid differentiation and pleomorphism in the sarcomatous counterparts. The myxoid variant of desmoid fibromatosis distinguishes from odontogenic myxoma by having dense collagenous bundles.

Many studies were conducted on the origin, presence and recurrence of myxomas using immune markers. Nonaka *et al*.[12] studied the role of matrix metalloproteinases-1 in the pathogenesis of myxomas but failed to get significant results. Martínez-Mata *et al*. studied extensively the role of Bcl-2 and Ki-67 in tumor growth and aggressiveness and concluded that the stromal reactivity is stronger in the myxoma for their growth potential due to high Ki-67 index. However, the proliferative activity was not strong enough to support the tumor growth; hence, the role of these markers was not significant.[13] Odontogenic myxomas showed three cell types as follows: spindle cells, stellate cells, and hyaline cells. Neoplastic cells and spindle cells of myxomas are positive for transferrin, ferritin, alpha-1-antichymotrypsin (alpha 1-ACT), alpha-1-antitrypsin (alpha 1-AT), S-100 protein, vimentin (pan-mesenchymal marker) and actin; however, neuron-specific enolase, S-100 alpha subunit, S-100 beta subunit, factor VIII-related antigen and cytokeratin 1 are negative. Stellate cells are 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 showed a negative reaction for all the antibodies used.[9-11] Bcl2 indicates the proliferative activity of tumor cells. Since myxomas are aggressive neoplasms, in the present case, this marker was used to assess the tumor activity. Poor Bcl-2 staining which is indicative of lack of tumor aggressiveness favored the complete resection of tumor in toto. Similar treatment protocol with Bcl2 marker synchronized with the studies by Farman *et al*.[14] and Martínez-Mata.[13] An extensive literature on the management of these tumors showed 45.5% of mandibular cases were treated surgically by enucleation, curettage or *en bloc* resection.[2,4,6] *En bloc* resection of the tumor mass was done. These tumors are radioresistant, and hence, radiotherapy is never a choice of treatment.[6] The rate of recurrences is attributed to the ability of the tumor to infiltrate into surrounding bone. Incomplete removal of tumor is responsible for recurrence more than the biological behavior. Boffano *et al*. suggested that lesions of size >3 cm are considered for radical resections and bloc resections and tumors of lesser diameter are better treated by enucleation or curettage. There should always be a follow-up period of 2 years postsurgically which represents the maximum activity of the tumor recurrence.[15] Two years follow-up showed no recurrence in the present case. The mere presence of myxomatous areas and mildness of these tumors should not hinder the clinician or the pathologist to move a step forward in carrying out ancillary methods of diagnosis that could have detrimental effect on the morbidity of the patient.

CONCLUSION

Odontogenic myxoma is a benign, painless, slow growing, and locally malignant tumor, with high recurrence rate. These are very aggressive tumors that are often mistaken as clement natured in the first outlook. Due to their permissive impressions, the surgeons avoid further investigation to plan the treatment. Unfortunately, due to their paucity of prevalence not many studies are done to prove their mighty destructive potentials. It is thus recommended to perform tests on its belligerence and take prudent surgical steps which can improve the outcome of patients. We take an opportunity to present one such rare myxoma in a male patient which was confirmed through the use of ancillary methods of detecting the stromal component and the tumor reactivity to aggressive nature. Although the present case showed negative aggressive nature, it seems imperative to carry out tests relevant to confirm the same. We performed aggressive treatment keeping in mind the tumor behavior. A thorough knowledge of other overlapping lesions is mandatory before diagnosing these unaccustomed myxomas. Due to its nonspecific nature and its diagnostic and operative dilemmas related to the myxomatous origin, proper knowledge over histopathology, behavior, and treatment of choice is recommended to avoid recurrence.

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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