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Odontogenic Myxoma Of Mandible:Report Of A Rare Case With Review Of Literature

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Abstract: *Odontogenic myxoma represents an uncommon benign neoplasm comprising of 3–6% of all odontogenic tumors.This articles presents a rare case of odonogenic myxoma occurring in the mandible with a brief overlight on its etiology,immunocytohistochemistry,radilogical and treatment fetaures of odontogenic myxoma.*

keywords: *odontogenic myxoma,fibro-myxoma,odontogenic tumours*

I. Introduction

In 2003, the World Health Organization (WHO) classified odontogenic myxoma (OM) as a benign neoplasm arising from odontogenic ectomesenchyme with or without odontogenic epithelium Goldman and Thoma were the first to describe OM of the jaws in 1947. [The alternative terms are myxofibroma or myxoma. [1] Odontogenic myxoma (OM) is a tumor thought to be derived from embryonic mesenchymal elements of dental anlage. It is a locally invasive benign neoplasm. The invasiveness is attributed to the biological nature of the tumor.[1]

II. Report

A 30-year-old female reported to the department of dentistry of Mandya institute of medical sciences .Mandya July 2014, complaining of a painless swelling in the right side of her face since six months. Initially, the swelling was small in size and showed a gradual increase to its present dimensions following which she consulted a local dentist for which extraction of lower right wisdom was suggested and was done in the month of Jan 2014(apparently orthopantamogram was not done instead IOPA was done).following which there was no reduction in the swelling on the contrary a gradual increase in the swelling was noticed. Extraoral examination showed a diffuse swelling involving the entire preauriclar region on the right side extending into submandibular region measuring roughly about 5cm x4cm. The skin over the swelling was normal, and there was no local rise of temperature. On palpation the swelling had variable consistency of firm to hard.

fig 1

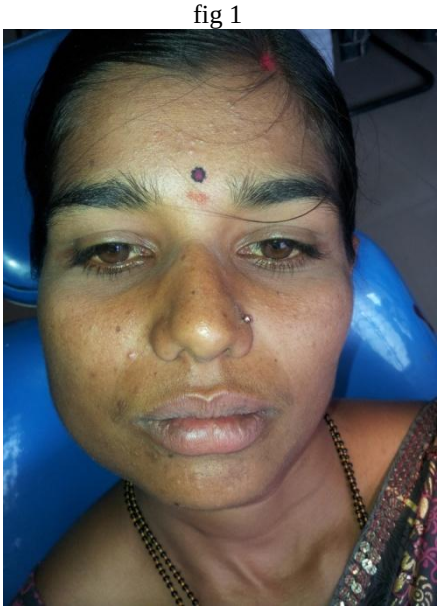
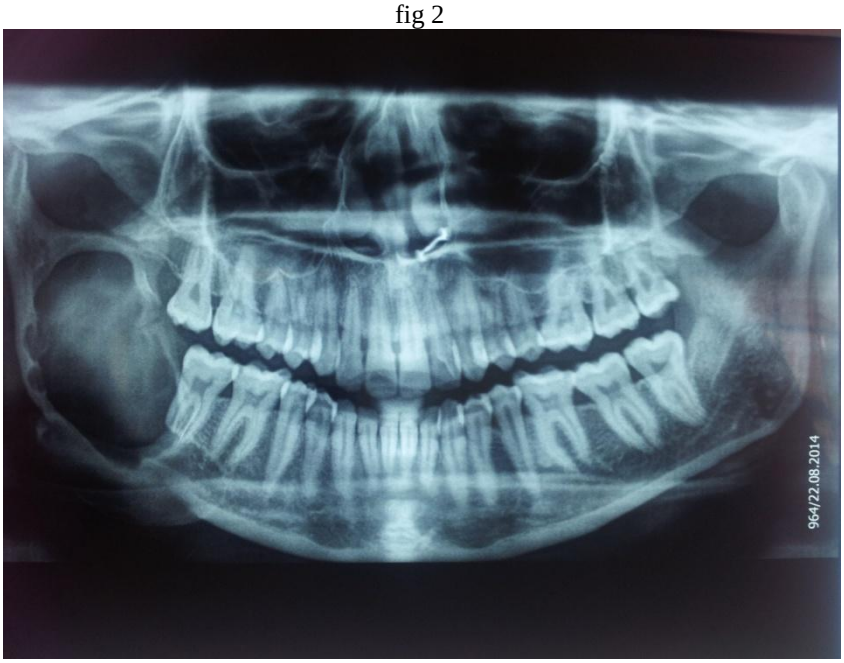


fig 2



Intra orally there was expansion of both the lingual and buccal cortical plate limited to only ramus of the mandible associated with a history of parasthesia from past 2 months. The radiograph showed a well-demarcated unilocular radiolucent lesion involving the entire ramus and extending into the body of the mandible upto 46 along with the presence of impacted lower wisdom teeth on the same side. The lower border of the lesion was expanding inferiorly in relation to 46 and 47. Perforations along the anterior border of the ramus and along the inferior cortical border of the border of body of mandible in relation to 47 was noticed

Aspiration biopsy lesion revealed air suggesting it to be a solid lesion. A provisional diagnosis of ameloblastoma was considered.

Complete excision of the lesion was done under local anesthesia following routine blood investigation which was found to be well within normal limits.

The excised lesion was found to be fleshy mass

The lesion was sent for histopathology.

Histopathological study revealed it to contain stratifies squamous epithelium with underlying connective tissue component. The coon tissue stroma is loose, fibrous and demonstrates numerous haphazardly arranged stellate spindle shaped cells scattered in myxoid stroma background.Mild inflammatory infiltrate predominantly composed of lymphocytes and few blood vessels lined by endothelial cells are also seen. Alcian blue stain was positive for intercellular areas.The clinical and histopathological features are suggestive of odontogenic myxoma.

III. Microscopy:

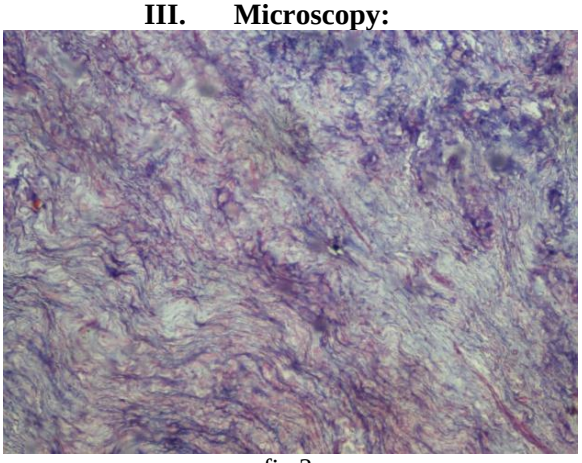


fig 3

Low power:

Microscopic examination of Hematoxylin and Eosin stained section shows a moderately fibrous connective tissue mass with a few areas showing myxoid change

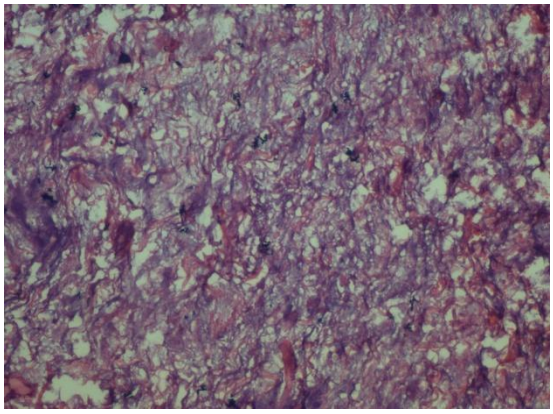


fig 4

High power:

The connective tissue stroma displays spindle or stellate cells in a myxoid background with focal areas of the connective tissue appearing basophilic. Moderately dense chronic inflammatory infiltrate composed predominantly of lymphocytes is seen. Blood vessels lined by endothelial cells and areas of hemorrhage are also seen

IV. Discussion

Odontogenic myxoma (fibromyxoma) is a benign neoplasm of uncertain histogenesis with a characteristic histologic appearance. It often shows infiltration and behaves in a locally aggressive fashion [3]

Odontogenic myxoma (OM) is thought to be derived from dental papilla and Hertwig's epithelial root sheath. Evidences supporting the odontogenic origin are (a) its occurrence is almost exclusive, only to the tooth-bearing areas in the jaws, (b) occasional association with an unerupted tooth or a missing tooth, (c) occurrence in younger individuals, (d) histologic resemblance to dental papilla, and (e) occasional presence of odontogenic epithelial island. The OM cells express extra cellular membrane molecules like fibronectin, type 1 collagen and tenascin resembling human immature dental papilla stem cells. [1]

The German pathologist Rudolph Virchow was probably the first to describe the histologic features of myxofibroma in 1863, although the lesions of jaws were not particularly mentioned. In 1947, Thoma and Goldman first described myxomas of the jaws. Since then odontogenic myxoma has been a subject of continuous scientific debate.[4]

There has been a great deal of controversy regarding the origin of myxomatous tumors. Virchow, in 1863, coined the term myxoma for a group of tumors that had histologic resemblance to the mucinous substance of the umbilical chord. 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. Myxoma is a tumor that can be found in heart, skin and subcutaneous tissue and, centrally, in the bone.[3]

Myxomas o f the head and neck are rare tumors. Two forms can be identified: (1) facial bone derived, which had been subdivided in the past into true osteogenic myxoma and odontogenic myxoma and "soft tissue"-derived myxoma, derived from the perioral soft tissue, parotid gland, ear and larynx.[3]

Traditionally, the myxoma of the maxilla and mandible has been considered to be a neoplasm of odontogenic origin. Although the evidence is mainly circumstantial, support of an odontogenic origin has been perpetuated by its almost exclusive occurrence in the tooth-bearing areas of the jaws, 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.[3] Slootweg and Wittkampf on the other hand showed that the matrix of myxomas of the jaw is entirely different from the matrix seen in the dental pulp and periodontal ligament. In addition, they also argued that myxomas may also develop in the sinonasal tract and other facial bones that originate from the nonodontogenic mesenchyme. According to them, even the presence of odontogenic epithelium is not necessary to make the diagnosis of myxoma of bone.[3]

Contrary to the findings of Slootweg and Wittkampf, McClure and Dahlin reviewed more than 600 bone tumors of patients at Mayo Clinic and concluded that there were no true myxomas of the bone except for those found in the mandible and maxilla.[3]

Most odontogenic myxomas are first noticed as a result of a slowly increasing swelling or asymmetry of the affected jaw. Lesions are generally painless and ulceration of the overlying oral mucosa only occurs when the tumor interferes with dental occlusion. Growth may be rapid and infiltration of neighboring soft tissue structure may occur. Both the buccal and the lingual cortical plates of the mandible may expand occasionally. Kaffe *et Al*. found expansion of the jaws in 74% of the cases. 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. Displacement of teeth has been registered in 9.5% of the cases. .[3]

Most commonly, it occurs in the second and third decades. The mandible is involved more frequently than the maxilla, and most reports show a slight predilection for females. OM are usually painless and displacement of teeth and paresthesia are uncommon clinical features. It therefore reaches considerable size before being detected, and perforation of the cortices of the involved bone may be seen. Kaffe *et al*. reviewed 164 odontogenic myxomas of the jaw and found that 75% occurred between the second and fourth decades (patient age range, 1–73 years; mean 30 years).[3]

Farman *et al*. differentiated between maxillary and mandibular odontogenic myxomas and suggested that the mean age at the time of diagnosis of maxillary odontogenic myxomas in men was 29.2 years and in women was 35.3 years, while the mandibular odontogenic myxomas in men occur at a mean age of 25.8 years and in women they occur at 29.3 years.Gunhan *et al*. and Regezi *et al*.. reported a higher incidence of these tumors in women (64–95%) than in men[3] which tallied with our patient.

Mandibular myxomas accounted for 66.4%, with 33.6% in the maxilla. 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.[16] In the present case, the lesion was located in the premolar and molar area of the mandible.Goldblat in 1976 described two basic types of tumor cells i.e., secretory and nonsecretory. The secretory cell type was considered the principal tumor cell and resembled fibroblasts. Three types of odontogenic myxoma cells were discriminated: Spindle cells, stellate cells and hyaline cells. Spindle cells are positive for transferrin, ferritin, alpha 1-antichymotrypsin (ACT), alpha 1-antitrypsin (AT), S-100 protein and vimentin. Stellate cells were 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 is negative for all these antibodies. These results proved that odontogenic myxoma is a tumor of a dual fibroblastic-histiocytic origin and also suggest that the cells are of myofibroblastic origin.[1]

The immunohistochemical and ultrastructural studies done by Martinez-Mata G *et al* concluded that OM is a mesenchymal neoplasm. Similarity of protein profiles of extracellular matrix of dental follicle (DF) of a developing tooth and OM, supported the theory that OM could originate from DF. Miyagi SP *et al*., stated that the dental pulp stem cells, which are derived embryologically from the dental papilla cells may be the precursor of OM. The tumor is thought to originate from the mesenchyme of the developing tooth or the periodontal ligament because of the resemblance of these structures to the tumour tissue. Nestin, a marker for progenitor cells, was positive in the stromal neoplastic cells of the OM. So the authors suggested that the possible origin of tumor cells are from the dental papilla cells, fibroblasts or myofibroblasts. Using the notch signaling, Nakano *et al*., concluded that the differentiation level of the tumor tissue is similar to cap stage. The lower degree of differentiation is the reason for the degree of clinical behavior. [1]

Matrix metalloproteinases (MMPs) facilitate the invasion of the tumor cells through the normal tissues. Miyagi *et al*., analyzed the expression and activities of MMP 2 and 9 in the tumor. They concluded the invasiveness of OM is due to MMP 9. Also high degree of MMP 2 expression were found in cell lines derived from OM. Only 10% of the dental follicle cells and dental pulp cells expressed MMP-2, whereas 90% of the OM cells expressed this protein. [1]

Study done by Martinez-Mata G, showed expressions of Bcl-2 and Ki 67 were less than 1% in the tumor cells. However, Lezzi G *et al*., found that 4% of the stromal cells are positive for MIB-1 and only 1% in the epithelial cells. But Bcl-2 and p53 were negative in the stroma and were weak positive in the epithelial cells. Thus, the proliferation of both the epithelial and stromal components and lack of cell death protein expression could be the reason for growth of OM. Hypermethylation of pro-apoptotic genes p27, p53 and Rb1 downregulates protein expression and contributes to myxoma growth. [1]

On comparing the protein profiles of the normal DF and OM, the glycoprotein orosomucoid-1 (ORM1) was found to be overexpressed in OM. This protein was detected in the cytoplasm of the stellate, spindle-shaped cells and in the endothelial cells. The authors suggested the function of ORM1 may be angiogenic, immunomodulatory and play important role in the aggressiveness of the tumor. They also found the downregulation of carbonic anhydrase I (CA I) and glutathione S-transferase (GST). The downregulation of CA1, which may cause loss of balance in bone remodeling, resulting in bone resorption. The reduction in GST

could lead to genome damage as its function of protection from oxidative damage is lost. [1] Ultrastructural studies showed that the neoplastic spindle cells are fibroblast like cells called myxoblasts. They synthesize large quantities of mucopolysaccharides. The ground substance of OM has been shown to consist of about 80% hyaluronic acid and 20% chondroitin sulfate. Exuberant hyaluronic acid which is an extracellular membrane protein is responsible for the local invasiveness of this neoplasm. [1] Tumor cells are relatively inactive, showing low levels of oxidative enzymes and slight alkaline phosphatase activity.[1] On conventional radiographs, myxomas of the jaws often show multilocular radiolucencies representative of "honey comb," "soap bubble" or "tennis racquet" appearance, which helps 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. In the present case, the orthopantamograph revealed a single large expansile radiolucent lesion without any trabeculations in the area of bony destruction. However, few radioopacties were seen within the radiolucency.[1]

The aggressive nature of the lesion is well documented in literature. The absence of capsule and tendency of OM to permeate into marrow spaces makes effective enucleation and curettage difficult. Small lesions have been successfully treated in this way but the larger lesions require block resection with tumor-free margins. Recurrence rates from various studies average about 25%.[1]

Conservative treatment was defined as enucleation, curettage, and marginal resection; radical treatments was defined as segmental or block resection, and hemimandibulectomy requiring reconstruction.[2] Nonetheless, the risk of recurrence after more conservative surgery is greater as the myxoma is not encapsulated and its myxomatous tissue infiltrates the surrounding bony tissue without causing immediate destruction . Therefore, complete surgical removal can be challenging, which may explain the high recurrence rates (10–30%) after conservative surgical treatment for odontogenic myxoma [2]

Differences in recurrence rate appear to be entirely accounted for by treatment approach: the rate after simple enucleation and curettage has been reported to be as high as 25% .The main reason for recurrence is thought to be incomplete removal rather than the intrinsic biological behavior of the tumor .Several investigators have recommended that tumor size should determine whether a radical or more conservative surgical approach should be adopted .Boffano *et al*. suggested that conservative treatment by enucleation and curettage is recommended when the diameter of an odontogenic myxoma is less than 3 cm, whereas a segmental resection with immediate reconstruction is preferred in patients with larger tumors[5] .Our literature review found that recurrence was only reported after conservative treatment .Kancy *et al*. reported a polycystic lesion in the right retromolar mandible, for which they performed a resection with preservation of mandibular continuity, filling the defect with an autologous bone graft from the iliac crest a week later .Nevertheless, 5 years after surgery, a recurrence was detected. Lo Muzio *et al.* described two cases of recurrence after resection of large unilocular radiolucent lesions causing tooth displacement and nonhomogeneous bone reabsorption with extrusion of the third molar; both after enucleation and curettage . Recently, Zanetti *et al*. strongly suggested conservative treatment should involve enucleation of the lesion with a wide curettage of normal tissue or a generous amount of apparently uninvolved surrounding tissue, or even peripheral osteotomy, as this has the advantage of preserving vital structures and maintaining oral function .They also reported that this technique could be used to treat odontogenic myxoma should it recur after more conservative surgery[6]. In our case, the tumor was relatively large (approximately 35× 15 × 35 mm), but even though its diameter was greater than 3 cm, we chose a more conservative approach after obtaining informed consent from the patient. As odontogenic myxoma is so rare it is not possible for a single center to accumulate sufficient expertise to examine whether a more conservative approach is also suitable for larger tumors. Indeed, the tumor in our patient was >3 cm diameter and there has been no recurrence after enucleation and wide curettage of normal surrounding tissue. If a patient should ultimately develop a recurrence having being treated according to a more conservative strategy, careful consideration should be given to the subsequent treatment. Although there is little evidence upon which to base management decisions, we recommend that further, radical surgery is warranted after recurrence of conservatively-treated odontogenic myxoma, a view that concurs with reports of three cases in the literature [2]

A follow-up period is clearly also necessary. It has been recommended that patients should be followed closely for at least the first 2 years after surgery, which represents the period during which the neoplasm is most likely to recur [20]. Rocha *et al.* suggested that 5 years of surveillance is needed to confirm successful excision, but that ideally follow-up should be maintained indefinitely [2]

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