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CASE REPORT

A CASE OF RECURRENT ODONTOGENIC MYXOMA OF THE MANDIBLE: CLINICAL AND RADIOGRAPHIC DIAGNOSIS WITH REVIEW OF LITERATURE

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ABSTRACT

Odontogenic myxomas are benign tumors of ectomesenchymal origin and comprise of 3%-8% of all odontogenic tumors. They may be derived from cells of the dental follicle, dental papilla or periodontal ligament. Most of the tumors occur in the second to third decades of life and show a marked female predilection. They occur in the premolar-molar region of both jaws but show a preference to the mandible over the maxilla. Radiographically they may be identified due to their characteristic 'tennis racquet' appearance although other radiographic forms such as soap-bubble, honeycomb or wispy lace-like trabeculae also exist. Histological appearance of a myxoid stroma made up of 80% hyaluronic acid and 20% chondroitin sulphate with collagen fibres and spindle shaped fibroblasts is distinctive. Owing to the absence of a capsule and the gelatinous nature of the ground substance, a high degree of recurrence is seen especially in tumors that are conservatively managed. Therefore adequate surgical resection with other modes of tissue destruction such as cryotherapy should be followed. Long term follow-up with radiologic surveillance is advocated in order to recognize a possible recurrence and prevent massive tissue destruction and its associated morbidity.

Key words; Myxoma, odontogenic, tumor.

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Introduction

Myxomas are tumors that are universally found in the skin, subcutaneous tissue, heart and bones. Although myxomas of the head and neck are relatively rare, nearly all of the centrally originating myxomas occur in the jaws.[1] Leiser in 2009 described odontogenic myxoma as a benign, locally invasive and aggressive, non-metastasizing neoplasm of the jaw bones.[2] Later in 2013, WHO defined myxomas histologically as benign odontogenic neoplasms of ecto-mesenchymal origin consisting of rounded and angular cells embedded in an abundant myxoid stroma with few collagen fibrils probably originating from either the dental papilla, follicle or the periodontal ligament.[3]

The etiology of odontogenic myxoma (OM) is controversial. While Virchow considered a group of tumors that had histologic resemblance to the mucinous substance of the umbilical cord as myxomatous tumors, Stout redefined the histologic criteria for myxomas. He considered myxomas as true neoplasms which did not undergo metastasis and did not include any identifiable cellular component of other mesenchymal tissues, especially rhabdomyoblasts, lipoblasts or chondroblasts.[1,4]

The histochemical nature of the ground substance of OM has been described by Farman *et al* as about 80% hyaluronic acid and 20% chondroitin sulfate. The tumor cells possess low levels of oxidative enzymes and appear relatively inactive. However, they show slight activity of alkaline phosphatase. The intercellular matrix is characteristically myxoid, stains positive with alcian blue, but negative for Periodic acid-Schiff staining.[5]

While the prevalence of OM in Asia, Europe and America is 0.5%-17.7%, the global prevalence is slightly lower at about 0.04%-3.7%.[6,7] Most cases of OM occur in the second to third decades of life and show a marked female predilection. Mandibular myxomas are more commonly seen (66.4%) than maxillary myxomas (33.6%). In the mandible, 65.1% of the tumors occur in the premolar-molar region while 73.8% of the lesions occur in the same region of the maxilla.[1,8]

Case Report

A 19 years old female patient reported with a complaint of a slowly progressive, painless swelling on the left side of the face since 3 months. The patient claimed that she had been surgically treated for a similar swelling 3 years ago which had been confirmed histopathologically as myxoma.

Extra-orally the lesion did not cause any gross asymmetry of the face. On intra-oral examination a painless, bony hard, diffuse swelling was seen on the left mandibular alveolus measuring approximately 2 X 3 cm in size extending anterio-posteriorly from the distal aspect of the left mandibular first premolar up to the second molar. Superiorly it was seen to begin from the gingival margin of the existing teeth. Involvement of the buccal and lingual vestibules of the lower jaw was noticeable along with decrease in their depth. The

overlying mucosa appeared normal and there was no evidence of any discharge. Expansion of the buccal and lingual cortical plates was also seen (Figure 1). Electric pulp testing was performed on the teeth of the lower left quadrant and all teeth showed delay in their responses. Based on these features, a provisional diagnosis of benign odontogenic tumor; possibly a recurrence of the earlier odontogenic myxoma was made. Differential diagnoses such as ameloblastoma, keratocystic odontogenic tumor and central hemangioma were also considered.

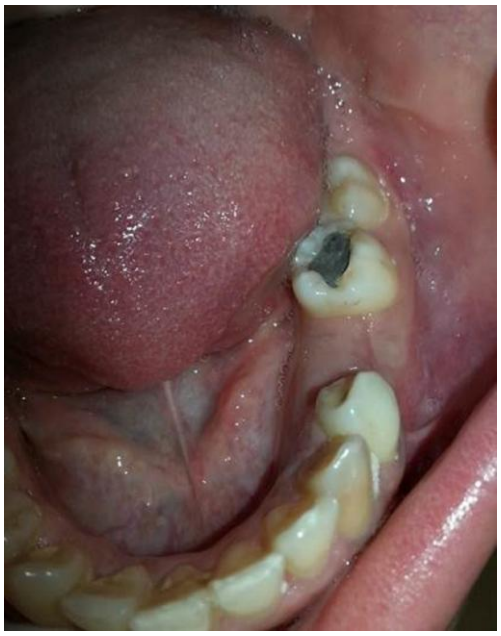

Figure 1. Intra-oral view showing the swelling with expansion of buccal and lingual cortical plates.

Radiographic investigations were carried out on the patient. Intra oral periapical radiograph of the posterior mandible revealed a well-defined multilocular radiolucency with a scalloped border in the region of the premolars and molars. Internal structure was made up of curved septae. Resorption of the mesial root of the mandibular first molar and displacement of the second premolar were seen (Figure 2).

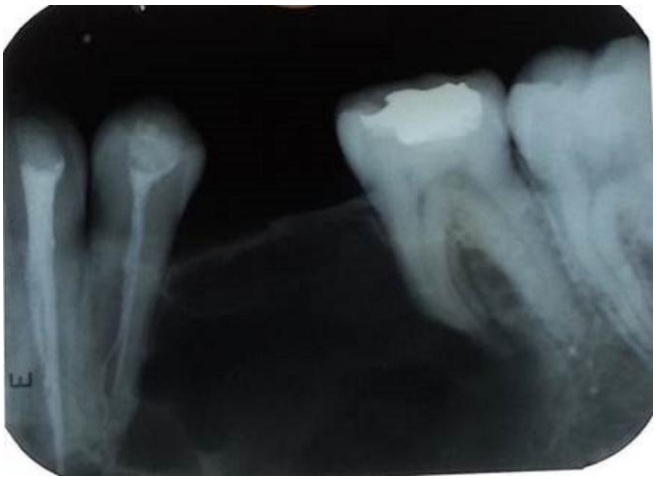

Figure 2. Intra oral periapical radiograph showing well defined multilocular radiolucency in the mandibular left premolar-molar region.

Mandibular occlusal radiograph showed a multilocular radiolucency in the left premolar-molar region and expansion of the buccal and lingual cortical plates (Figure 3). Orthopantamograph showed a well-defined multilocular radiolucency measuring approximately 5 X 6 cm in size, extending from the lower right lateral incisor, crossing the midline and extending up to the distal root of the left first molar. Superioinferiorly it extended from the crest of the alveolar ridge up to the inferior border of the mandible. Resorption of the mesial root of the first molar and displacement of the second premolar were also seen. Internal structure showed the presence of numerous trabeculae, some of which were arranged at right angles to the locules (Figure 4). Axial sections of cone beam computed tomography (CBCT) revealed a lytic lesion with expansion and thinning of the cortical plates and perforation of the buccal cortical plate adjacent to the second premolar (figure 5). Cross sections showed scalloping of the border of the lesion and fine wispy trabeculae in the internal structure (Figure 6 a,b). Three-dimensional reconstruction confirmed the extension of the lesion (figure 7). Following these investigations, radiographic differential diagnoses such as odontogenic myxoma, ameloblastoma and keratocystic odontogenic tumor were considered.

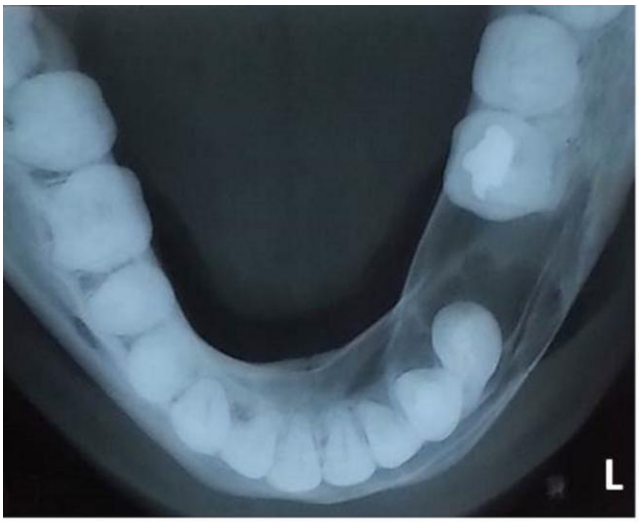

Figure 3. Mandibular occlusal radiograph showing a multilocular lesion along with expansion of the buccal and lingual cortical plates..

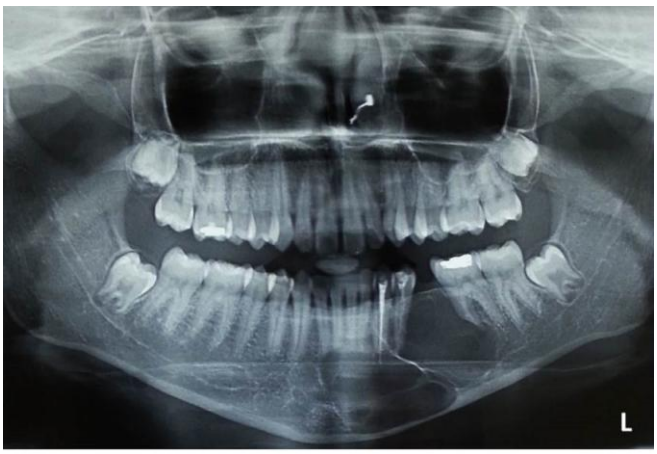

Figure 4. Orthopantamograph showing a well-defined multilocular radiolucency with displacement of the left mandibular second premolar and resorption of the mesial root of the first mandibular molar.

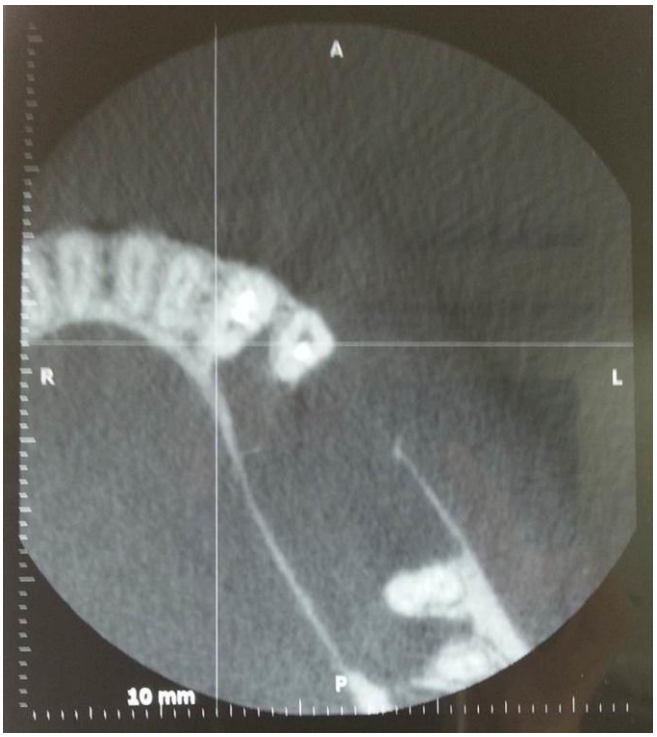

Figure 5. Axial section of CBCT showing thinning of the lingual cortical plate with perforation of the buccal cortical plate.

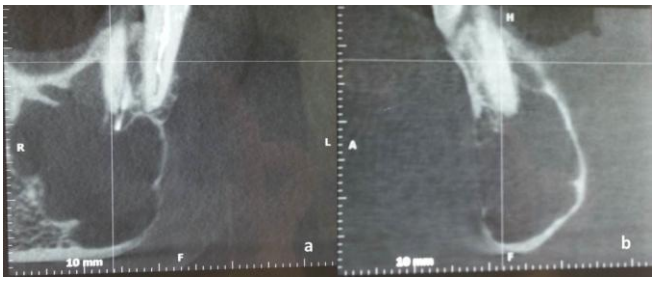

Figure 6. (a) Cross sections of CBCT showing scalloped borders and (b) wispy internal structure.

Incisional biopsy was made comprising of both bone and the mucosa. Histologically the sections showed spindle shaped fibroblasts in myxoid background with delicate, haphazardly arranged collagen fibres giving an impression consistent with features of myxoma (Figure 8). The lesion was surgically treated with a solo intra oral approach. Full thickness mucoperiosteal flap was raised with vertical releasing and gingival crevicular incisions. Tumor was excised as a single mass along with the first molar. The surgical site was irrigated with saline and betadine solution and cryotherapy was performed using liquid nitrogen. Antibiotic coverage was given to protect the patient against possible infections and analgesics were prescribed for pain control. The immediate post-operative period was uneventful. The patient complied with the advice for long term follow-up. The patient has been monitored for two years with recall visits of every month for the first six months and thereafter every four months. She appears to be healthy and there appears to be no evidence of recurrence either clinically or radiologically (Figure 9).

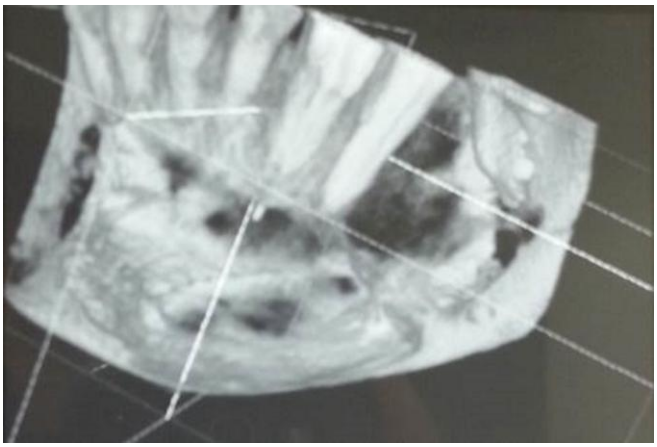

Figure 7. Three-dimensional reconstruction showing extent of the lesion.

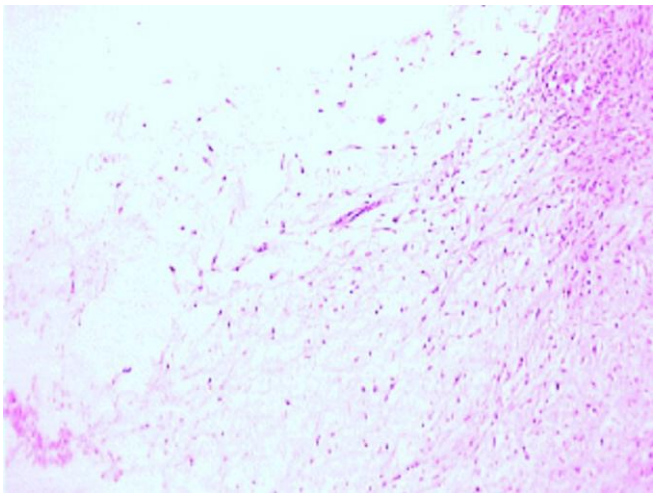

Figure 8. Photomicrograph showing myxoid background with spindle shaped fibroblasts and collagen fibers.

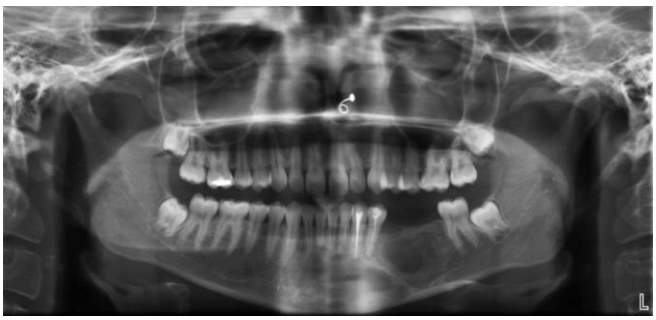

Figure 9. Orthopantamograph showing healing of the lesion 18 months after surgery.

Discussion

Odontogenic myxomas have created considerable controversy regarding their origin. Some researchers regard OM as a modified form of fibroma in which myxoid intracellular substance separates the connective tissue while others suggest that they are benign tumors derived from the embryonic mesenchymal elements of the dental anlage,

usually the periodontal ligament, dental follicle or the dental papilla. This origination led to its classification as 'benign tumor of ectomesenchymal origin with or without odontogenic epithelium'.[6] OM can occur both in the bone and in the soft tissue. Two distinct types can be identified; first as a facial bone derived form which has been further divided into true osteogenic myxoma and odontogenic myxoma. The second form may be derived from the perioral soft tissue, larynx, ear or the parotid gland and is classified as soft tissue derived myxoma.[9]

There exist several theories which attempt to explain the histogenesis of OM. Most of them are explicit about its origin from the odontogenic ectomesenchyme of a developing tooth or from the undifferentiated mesenchymal tissue of periodontal origin. These theories are supported by factors such as the uncommon occurrence of OM in other parts of the skeleton, contiguity of the tumor with the tooth-bearing regions of the jaws, periodic association of OM with unerupted or missing teeth, histological resemblance of OM to the ectomesenchyme of the pulp and sporadic presence of inactive odontogenic epithelium.[10] Some authors suggest that the tumor results from the myxomatous degeneration of fibrous stroma while others propose that there is no secondary change in the tissue but the tumor appears from the primitive mesenchyme. Another theory propositions that odontogenic epithelium induces the secretion of myomatous ground substance due to the aberrant development of mesodermal cells into myxoblasts.[11]

Shivakumar *et al* suggested that OM is a tumor of dual histocytic-fibroblastic origin.[12] While the cells that make up odontogenic myxoma are of myofbroblasic origin, the characteristic myxoid extracellular matrix (ECM) is composed of proteoglycans, fibronectin, glycosaminoglycans, tenascin and collagens.[10,12] The edematous appearance of the ECM is dependent on the amount of each of its components. These components help in the diffusion of metabolites and permit cell growth and migration through the creation of a network.[12] OM is not an encapsulated tumor and shows a high amount of infiltration into the surrounding structures. This plays a major role in the recurrence of the tumor.[6] The gelatinous nature of OM enhances its ability to infiltrate through thin layers of tissue planes.[13] Several theories have been put forward to explain the invasive nature of the tumor. Literature consigns this to genetic alterations, expression of antiapoptotic proteins and matrix metalloproteinases (MMP), alterations in receptor activator of nuclear factor kappa β ligand (RANKL) and its receptor (RANK) and alteration of the osteoprotegerin (OPG) system.[12] RANK-RANKL-OPG system plays a key role in the regulation of osteoclast formation, activity of osteoclasts, and osteoclast differentiation. The activation, fusion and differentiation of osteoclasts depend on the ligation of RANKL to RANK while OPG impedes this interaction. The relative content of RANKL in the mesenchyme of OM exceeds the content of OPG thereby facilitating bone destruction.[12,14]

OM represent 3%-8% of all odontogenic tumors and occurs as a slow growing lesion. It has a tendency to reach dramatic proportions before the patient seeks medical attention due to its almost universal asymptomatic nature.[7] The present case also did not show any symptoms associated with the swelling of 3 months duration. Radiographically the tumor presents a varied appearance ranging from unilocularity to multilocularity. The borders may be well-defined or diffuse and the internal structure is characteristically made up of fine bony trabeculae arranged in a soap-bubble, honeycomb or tennis racquet pattern.[5,7] Asaumi *et al* described three different types of radiographic appearance based on computed tomographic (CT) features.[15] First was the mandibular myxoma that is seen as an osteolytic expansile lesion with mild enhancement of the solid portion, second was the lesion in the anterior maxilla that occurs as a bony expansion and thinning of cortical plates with strong enhancement of the mass and third, the lesion in the maxillary sinus which is seen as a soft tissue mass with bone destruction and thinning along with ossifications that create fine strands of lace-like density.[15] Magnetic resonance imaging (MRI) reveals a well-defined, well-enhanced lesion with homogenous signal intensity on every pulse sequence.[1] The case reported here showed a well-defined multilocular radiolucency with internal septae and fine wispy trabeculae, few of which were arranged at right angles. Owing to the clinical and radiographic features, similar lesions such as like dentigerous cyst, ameloblastoma, keratocystic odontogenic tumor, ameloblastic fibroma, odontogenic fibroma, central hemangioma, unicystic calcifying epithelial odontogenic tumor, central giant cell granuloma and osteosarcoma may be considered in the differential diagnosis.[1,6,7]

Odontogenic myxomas are aggressive in nature and the tumor is not radiosensitive.[6,7,13] Therefore surgery is the mainstay of treatment. Conservative management techniques such as enucleation, curettage and cryotherapy offer several advantages such as decrease in morbidity, shorter duration of hospitalization, intra-oral access, absence of a donor site, low procedural cost and less interference with facial growth in case of a child patient.[6] However, a recurrence rate of 25% has been reported which has been attributed to its infiltrative nature, gelatinous components and lack of capsule.[1,6,7] Therefore, radical surgery such as partial or complete resection with liquid nitrogen cryotherapy followed by reconstruction using vascularized free tissue transfer, iliac crest, radial forearm and fibula free tissue transfers has been advocated by a majority of researchers.[6,13] Ayranci *et al* have proposed a protocol of conservative approach of enucleation and curettage when the lesions were less than 3 cm in size and segmental resection with immediate reconstruction when the lesions were larger.[7] Jewer *et al* opined that since most of the OM patients were of the younger age group, reconstruction and full esthetic as well as functional rehabilitation were of foremost importance.[16] They suggested that deep circumflex iliac artery-based free osseous flap was the ideal reconstruction method since it provided favourable shape and quantity of bone along with a reliable blood supply thereby making graft survival excellent. It also ensures that the quality of bone is adequate for the subsequent placement of endosseous implants.[13,16] An intra-oral approach with liquid nitrogen cryotherapy was used in the present case. Liquid nitrogen offers the advantage of bone devitalization thereby eliminating all neoplastic cells but does not affect the inorganic structure thereby allowing subsequent new bone formation.[6]

Due to the propensity for recurrence of the tumor in the first two years after treatment, close monitoring is mandated. Thereafter, the patient can be recalled less often but should be maintained on an indefinite follow-up since there have been reports of late recurrence in literature.[6,13] The present patient is currently on her third year of follow-up and appears healthy both on clinical and radiographic review.

Conclusion

Despite efforts to delineate the molecular genesis of odontogenic myxomas, their invasive behaviour is yet to be explained unambiguously. Their propensity for the posterior jaw, cortical expansion with perforation and radiographic appearance of uni- or multilocularity places them in the differential diagnosis of a variety of odontogenic cysts and tumors. However, the presence of the characteristic myxoid stroma on histopathological examination confirms the diagnosis of OM. The inclination towards occurring in young patients, an infiltrative nature and high degree of recurrence makes adequate treatment and long term surveillance a prime requisite.

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