Odontogenic myxoma of maxilla: A review discussion with two case reports

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Abstract

Odontogenic myxoma (OM) is a rare entity of slowly growing benign neoplasm of ectomesenchymal origin, comprising of 3–6% of all odontogenic tumors that histologically presenting spindle-shaped, stellate and round cells within loosely arranged myxomatous tissue stroma. OM originates from the dental papilla, follicle or periodontal ligament with an exclusive location in the tooth-bearing areas of the jaws, association with missing or unerupted teeth. Clinically and radiographically the reported incidence and demographic information of this tumor has wide variability. Most common clinical variant is associated with the impacted tooth and shows local invasion with destruction of adjacent structures and displacement of teeth. Radiographically, common manifestations are multilocular radiolucent areas with well-defined borders and typical soap bubble or tennis racket appearances. This paper presents two rare case reports of OM of maxilla along with review discussion.

Keywords: Fibromyxoma, myxofibroma, myxoma, odontogenic myxoma, tennis racket appearance

Introduction

Myxomas are benign, slow-growing and locally aggressive mesenchymal origin neoplasm.[1] Myxomas can be found in various body parts such as skin, subcutaneous tissues, and heart. Myxomas of head and neck region are rare.[1,2] Two forms of myxomas have been identified those derived from "facial bones" and they are either osteogenic myxoma or odontogenic myxoma (OM) and another derived "soft tissue" like perioral soft tissue, parotid gland, ear or larynx.[2,3] OM represents an uncommon benign neoplasm comprising of 3–6% of all odontogenic tumors,[2-5] most commonly arising in mandible (66.4%) followed by maxilla (33.6%).[2,3] According to World Health Organization (WHO), OM is classified as a benign tumor of ectomesenchymal origin with or without odontogenic epithelium.[3,5,6]

Odontogenic myxomas are asymptomatic. Pain, paresthesia or asymmetries occur only when they take on larger sizes. Their growth is usually slow; however, they are locally aggressive. They may cause divergence of root, resorption, tooth shifting or movement. When involving maxilla, OM can expand inside the maxillary sinus and are then diagnosed later only after having grown to larger sizes.[5]

Radiographically, the tumor presents as a unilocular or multilocular radiolucent lesion with well-defined borders with fine bony trabeculae within its interior structure expressing a "honeycombed", "soap bubble" or "tennis racket" appearance. Unilocular appearance may be seen more commonly in children and in the anterior parts of jaws. Displacement of teeth is a relatively common finding, root resorption is rarely seen and the tumor is often scalloped between roots.[1,3,6]

A histological characteristic of this tumor resembles the mesenchymal portion of a tooth in development. OM consists of rounded, spindled, fusiforms and star cells arranged in a loose, abundant myxoid stroma with few collagen fibrils. Small islands of apparently inactive epithelial odontogenic rests may be scattered through the myxoid substance without any capsule, and they are important to establish the diagnosis.[7] Because of its high rate of recurrence, especially due to its gelatinous and mucous aspect and having no capsule, surgical treatment through bone resection is the most indicated treatment modality, followed by long-term follow up.[5]

This paper presents two cases of OM of maxilla with different clinical and radiographic pictures. The purpose of the present paper is to increase cognition of the radiographic appearances of OM and to provide diagnostic references for better diagnosis.

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Case Reports

Case I

A 36-year-old Hindu male patient presented with the chief complaint of painless swelling on 23 tooth region since 1-year.

Patient had undergone for excision for the same type of lesion at private clinic before 3 years. Since then, patient was normal, except peanut size swelling which remained present after surgery, but since 2–3 months, patient noticed increased in size of the swelling, and it reached up to present size.

Intraoraly, well-defined around 2 cm in size, oval shaped swelling was present at 23 tooth region. Swelling was soft, nonfluctuant, nontender and without any discharge. Distal displacement of 22 was present. No pain or discoloration was found in any tooth. Medical history was not significant. Vitality test was done and 22, 23 and 24 was found vital [Figure 1].

The patient underwent radiographic investigations that suggested radiolucent area around 3 cm × 2 cm in size at interdental and periapical area of 22 and 23 region. A thin sclerotic bone reaction was seen at only superior border, rest of the area was blending to adjacent bone. Internal structure showed hazy radiolucency with one to two thin septa crossing lesion centrally. Mesial displacement of 22 was seen without any root resorption [Figure 2]. Clinico-radiographic features and history of recurrence was suggestive of either odontogenic keratocyst or lateral periodontal cyst. A routine hematological investigation was done before surgery. As negative aspiration was found, lesion was totally excised and specimen sent to the pathology department [Figure 3]. Histologically, section showed abundance of fusiform, stellate and round cells in myxoid tissue [Figure 4] and was diagnosed as myxoma.

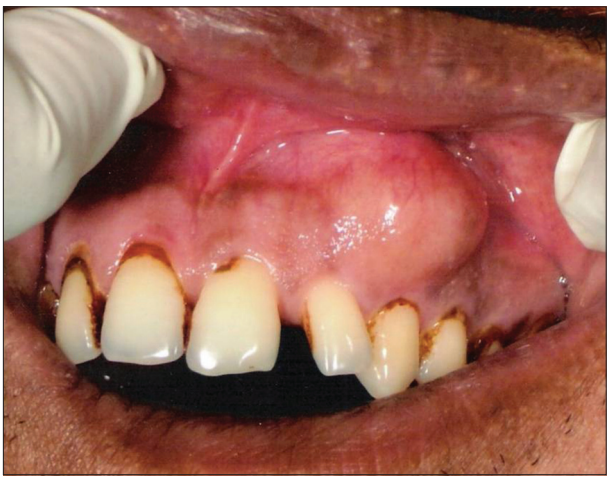

Figure 1: Intraoral photograph of case 1 showing dome shaped swelling present at 22.23 region with distal displacement of 22

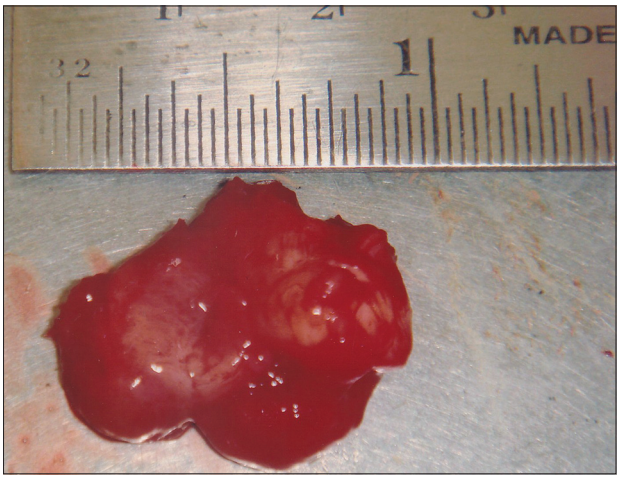

Figure 3: Gross specimen of excised soft tissue mass of case 1

Case II

A 28-year-old Hindu female patient came with painless swelling in right posterior maxilla since 1 year and 3 months. No history of pain, fever or trauma was found. Initially swelling was small in size and remained static for 1 year. Since last 3 months, swelling was rapidly increasing in size causing facial asymmetry without any pain or any other associated symptoms. Patient went to local private practitioner where she had been prescribed antibiotics, but the size of swelling

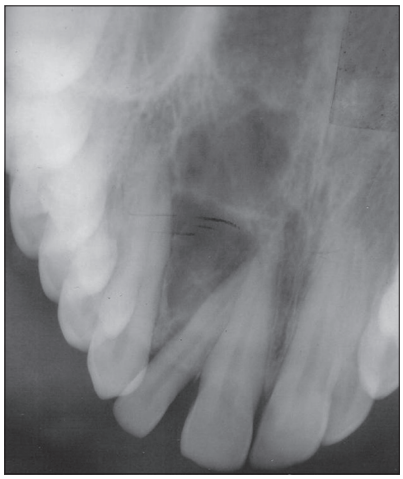

Figure 2: Maxillary occlusal radiograph of case 1 showing hazy radiolucency with one to two thin septa crossing lesion centrally at 22,23 region

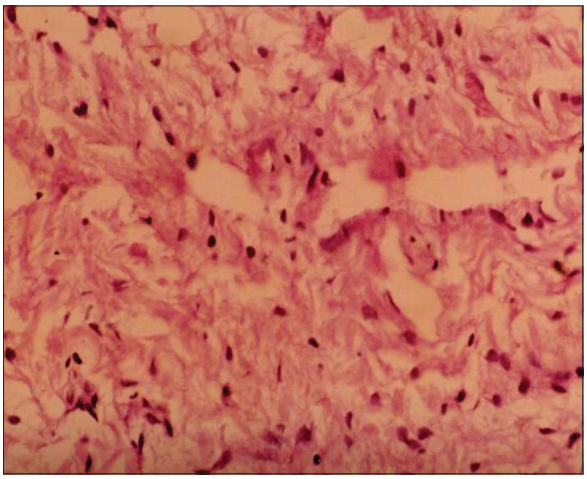

Figure 4: Histological picture of case 1 showing abundance of fusiform, stellate and round cells in myxoid tissue

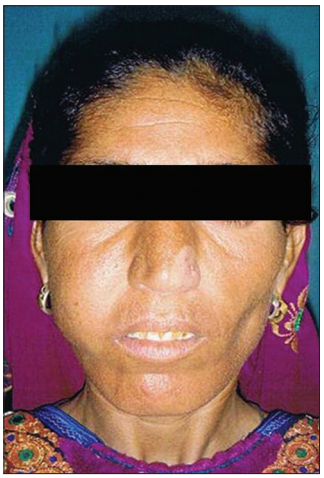

Figure 5: Extraoral photograph of case 2 showing swelling on right maxilla with obliteration of nasolabial fold

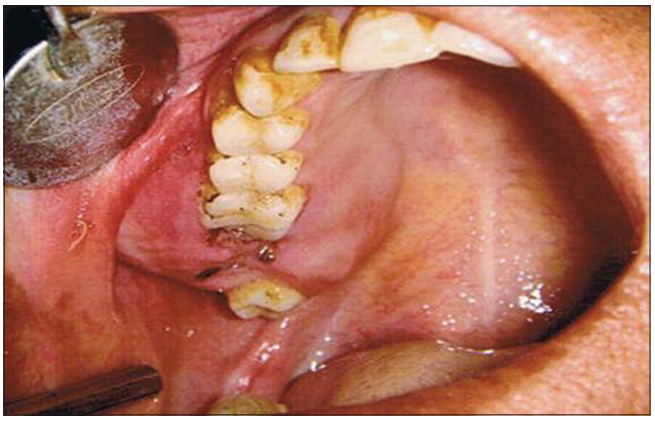

Figure 6: Intraoral photograph of case 2 showing swelling at posterior maxilla of 14–18 region with buccal cortical plate expansion

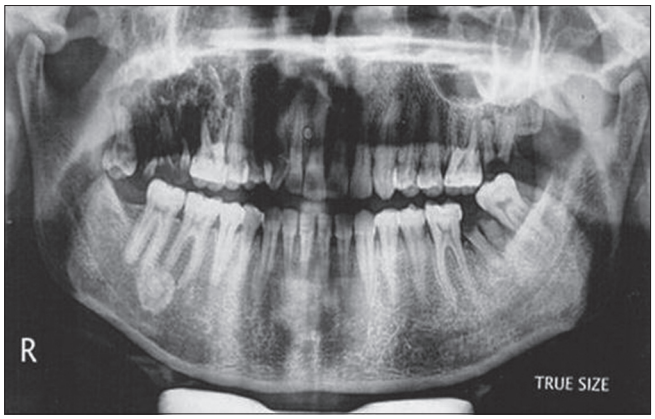

Figure 7: Orthopantomogram of case 2 showing poorly defined mixed radiolucent radiopaque area at right posterior maxilla with root piece of 17 and without root resorption of involved teeth

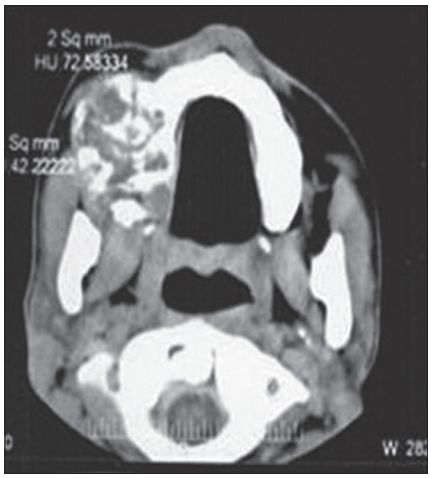

Figure 8: Computed tomography image of case 2 showed a well-defined expansile mass with calcified matrix arising from the alveolar process of maxilla, extending into the maxillary sinus with destruction of the posterolateral wall of the sinus

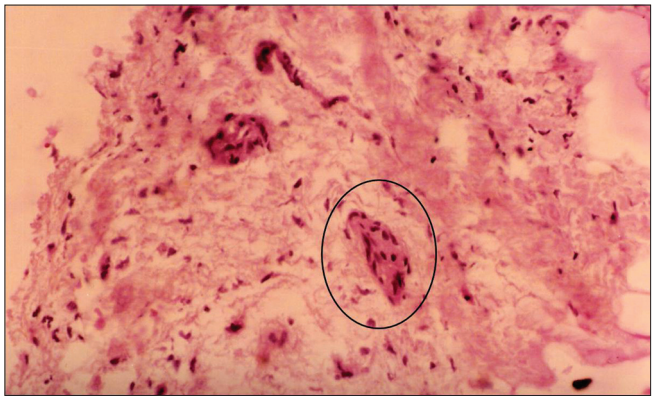

Figure 9: Histological picture of case 2 showing typical spindle-shaped cells in a myxomatous stroma

was not reduced. Finally, patient came to our hospital with painless asymptomatic swelling in the right maxilla.

Extraorally, swelling was around 4 cm × 3 cm in size at right maxilla with obliteration of nasolabial fold. Swelling was diffuse, nontender, hard in consistency, without any discharge. No history of nasal discharge, difficulty in breathing, paresthesia or anesthesia on the affected site. Overlying skin was normal in color with regional lymphadenopathy. No deviation of the lateral wall of the nose or no difficulty in mastication or trismus was found [Figure 5]. Intraoraly, swelling was well defined around 4 cm × 3 cm in size, extending from 13 to 18 teeth region with expansion of both cortical plates around 3 cm buccally and 2 cm palatally up to midline of palate. On palpation, swelling was hard, nontender, nonfluctuant without any discharge. Overlying mucosa was slightly red and inflamed. Tooth 17 was grossly carious with displacement of 16 palatally and 18 distally [Figure 6].

Maxillary occlusal radiograph showed a mixed radiopaque-radiolucent area with a granular appearance and ill-defined boundaries. No root resorption was seen of adjacent teeth. Orthopantomogram showed around 4 cm × 3 cm mixed radiopaque-radiolucent lesion at

periapical area of right maxillary molars approaching maxillary sinus. Lesion was having poorly differentiated boundaries. Internally, irregularly arranged thin trabeculae with bone resorption were present. No resorption of teeth found [Figure 7]. Computed tomography (CT) images showed a well-defined expansile mass with calcified matrix around 4.4 cm × 2.9 cm in size, arising from the alveolar process of maxilla, extending into maxillary sinus with destruction of posterolateral wall of sinus [Figure 8]. Benign odontogenic tumor, that is, central giant cell granuloma, fibro-osseous lesion, and OM were considered in the differential diagnosis.

Routine hematological investigations showed only slight eosinophilia. Incisional biopsy was performed, and it was diagnosed as myxofibroma [Figure 9]. Tumor was totally excised with surgical curettage and specimen was sent to the pathology department.

Discussion

Rudolf Virchow coined the term myxofibroma in 1863,[2-5] for a group of tumors that had histologic resemblance to the mucinous substance of the umbilical cord.[2,3,5] OM was first mentioned in the literature by Thoma and Goldman in 1947.[5,8,9] 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.[2,3]

In 1992, WHO classified OM for histological typing of odontogenic tumors: "A benign tumor, which is of ectomesenchymal origin with or without the presence of odontogenic epithelium.[3,5] Traditionally, OM is mainly circumstantial, involving teeth bearing areas associated with unerupted tooth with histologically resemblance to dental mesenchyme (dental papilla, follicle or periodontal ligament) and the sporadic presence of islands of odontogenic epithelium.[3] The Mucoid tumors of soft tissue represent significant differences in biologic behavior, ranging from harmless to malignant neoplasms. As an osseous entity, however, OMs of the jaws are considered slow-growing tumors with the potential for extensive bone destruction, cortical expansion, and a relatively high recurrence rate.[10] In presented paper, both cases had different clinical picture and biological behavior. First case had slow growing lesion with asymptomatic condition but with a recurrent history and another case had faster growth, facial asymmetry, the destructive and expansile nature, affecting surrounding vital structures resembling malignant tendency.

The tumor occurs across an age group that varies from 22.7 to 36.9 years.[6,9] It is rarely seen in patients younger than 10 years and older than 50 years of age.[2,5,6,9] According to Farman *et al*., suggested mean age of maxillary OM for men was 29.2 years and 35.3 years for women while in mandibular OM in men occur at mean age of 25.8 years and 29.3 years for women.[11] Gunahan *et al.* and Regazi *et al*. reported a distinct predominance in females (64–95%) and a predilection for the mandible.[12,13] Keszler *et al*. noted that 8% of OMs occurred in children of <16 years.[14,15] Posterior Mandible is more frequently affected than the maxilla.[3-6] According to Reichart and Philipsen, mandibular myxomas accounted for 66.4%, with 33.6% in the maxilla. Whereas 65.1% of mandibular cases located in molar-premolar region, 73.8% were located in same areas of the maxilla.[2,3,16] In presented, both cases, one patient was male with 36 years of age with lesion found on anterior maxilla and another was female with 28 years of age and lesion was found in the posterior maxilla.

Most often OMs are first noticed as a result a slowly growing increasing swelling or asymmetry of the affected jaw.[3] Growth may be rapid with infiltration of neighboring soft tissue structures. OM of the maxilla may be asymptomatic or on examination, it may present as heaviness, swelling of cheek or palate, malocclusion or loosening of teeth.[16] Displacement of teeth has been registered in 9.5% of the cases. Lesions are generally painless, and ulceration of the overlying oral mucosa occurs only when the tumor interferes with dental occlusion. When the maxillary sinus is involved, OM often fills the entire antrum.[2,3,16] They may still involve the palate, orbit, and nasal cavity, causing symptoms associated with this structures.[4,5] In severe cases, nasal obstruction or exophthalmus may be the leading symptoms.[2,3,16] In the mandible, OM may involve the neurovascular bundle in the mandibular canal.[17] Our both cases were of maxilla in which first case was totally asymptomatic with slow growth, and another had all classical features accordance to review.

Radiographically, larger multilocular OM is more common in the posterior areas of the jaws and unilocular lesions are mostly located in the anterior.[3,6] The radiographic tumor margins may be either well-defined or poorly defined.[17] On conventional radiographs, OM presented varying radiographic appearances, ranging from unilocular to multilocular (including honeycomb, soap bubble and tennis racquet patterns); with involvement of local alveolar bone or maxillary sinus and sometimes osteolytic destruction with or without osteogenesis.[15] The tennis racket appearance where the bony septae appear as triangular, square or rectangular compartments with very fine trabeculation within them is the most common.[17] Many review suggest that there may be no correlation between the borders of the lesion and the amount of bony trabeculae within the lesion, but maxillary tumors were more likely to be ill defined in nature compared with mandibular lesions.[10]

According to some studies, root displacement rather than resorption is the rule of jaw myxomas.[3,6,9,10,17] Radiographically, OM may present similar features of

an ameloblastoma or a central giant cell granuloma or sometimes may mimic an osteosarcoma. These facts do pose potential difficulties in reaching a proper diagnosis merely on radiographic studies. A biopsy is, therefore, necessary to ascertain an accurate diagnosis.[9,10]

Recently, CT and magnetic resonance imaging examinations were applied in many case reports and it may present as osteolytic expansile lesions with mild enhancement of the solid portion of the mass in the myxoma of the mandible or bony expansion and thinning of cortices with strong enhancement of the mass lesion in the anterior maxilla. In the case of maxillary sinus, bone destruction and thinning and strands of fine lace like density representing ossifications may be seen.[2,3] Our second case showed internal calcifications with destruction of the posterolateral wall of maxillary sinus whereas first case had only soft tissue mass with mild osteolytic areas in the anterior maxilla. Furthermore, lesion was small, unilocular with fine one to two straight septa internally, involvement of local alveolar bone, poorly defined boundaries and only displacement of teeth without resorption, while second case was of posterior maxilla with larger in size, multilocular variety and involvement of maxillary sinus. Owing to the existence of the maxillary sinus, part of the lesion image would overlap it. This influenced the image definition. Therefore, the lesion image may be obscure and may often be easily mistaken for other tumors or sometimes malignancy.

Differential diagnoses include ameloblastoma, odontogenic keratocyst, central giant cell granuloma, intraosseous hemangioma, aneurysmal bone cyst, glandular odontogenic cyst, cherubism, fibrous dysplasia, metastatic tumor in multilocular cases and in cases of unilocular lesions, periapical, lateral, periodontal and simple bone cysts.[4,5,9,17] In older patients, the possibility of a malignancy should not be ruled out.[16] The radiographic appearance of unilocular type of OM was similar to that of the odontogenic cyst and unilocular ameloblastoma, but the fine bone septa can sometimes be seen in the unilocular type of OM and the latter two lack this feature.[15] In our first case, lesion was small with fine one or two septa internally.

Microscopically, it resembles the mesenchymal portion of a tooth in development.[7] The lesion is not encapsulated and exhibits the abundant extracellular myxomatous stroma of ground substance and thin fibrils characterized by a proliferation of a few rounded cells, fusiforms or star cells and spindle-shaped cells. It may have a complete myxomatous tissue or varying proportions of myxomatous and fibrous tissue. In the latter case it can be designated either as odontogenic fibromyxoma, in which the myxomatous element predominates; or odontogenic myxofibroma, with a predominance of fibrous tissue.[3,6] Small islands of odontogenic epithelial tissue can be found scattered in stroma, sometimes being surrounded by a narrow zone of hyalinization.[2,6] Immunocytochemicallly, all tumor cells were found to be positive for vimentin and muscle specific actin and negative for desmin, neuron-specific enolase, glial fibrillary acid protein, and S100.[2,3,10] Histologic features of our both cases were accordance to review.

Recommended therapy varies from curettage to radical excision. Complete surgical removal can be difficult as the lesion is not encapsulated especially in the maxilla because the myxomatous tissue infiltrates adjacent bone tissue as well as close proximity of vital structures and more complex anatomy.[1,10] The prime reason for recurrence is thought to be related to incomplete removal rather than the intrinsic biologic behavior of the tumor.[10] These characteristics may explain the high rate of recurrence of myxomas, which ranges from 10% to 33% with an average of 25%.[1,9,10] Although there are few studies regarding this, radiotherapy and chemotherapy appear to be ineffective in controlling the recurrent lesions. It is suggested that patients be followed closely for at least 2 years because this is the most likely time for recurrence.[10] Our first case had a recurrence history. Previous incomplete surgical removal may be probable cause.

Summary and Conclusion

Two rare cases of OM of maxilla were presented. One was with unilocular variety in the anterior region and another large lesion with multilocularity and aggressiveness. In respect of biological behavior and extensiveness of such lesion, better knowledge, 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.

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