CASE REPORT

Odontogenic myxoma of the maxilla – A rare clinical sighting

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Abstract

Odontogenic myxoma (OM) is a benign tumor originating from primordial mesenchymal tooth forming tissues. Myxoma is a locally invasive intraosseous neoplasm that usually occurs in tooth bearing areas of jaws. Clinically, it is a painless, slow growing, and non-metastasizing tumor of jaw. Here, we report the case of OM in a 21-year-old male patient which had a large, expansile lesion of the left jaw extending to zygomatic, and infraorbital rim. This caused grotesque facial deformity within a short span of 6 months. However, complexity of such tumors increases as several characteristics overlap with those of other benign and some malignant tumors. The treatment plan should consider the age and sex of the patient and the site and size of the lesion. Research shows that surgical treatment of OM varies from simple enucleation and curettage to hemimaxillectomy. Recurrence rates are also reportedly high at around 25% when conservative approaches are taken. Here, wide excision of the tumor with hemimaxillectomy was planned, but further reconstructive surgery was delayed to rule out recurrence.

Keywords: Invasive, mesenchymal, neoplasm, odontogenic

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Introduction

Myxomas are benign, locally aggressive, and slow-growing tumors originating from embryonic mesenchymal elements of dental anlage. According to the WHO, odontogenic myxoma (OM) is classified as a benign tumor of ectomesenchymal origin with or without odontogenic epithelium. Two types of myxomas have been identified – osteogenic or OM which are derived from the facial bones and the other – soft-tissue myxoma which derives from perioral soft tissues, parotid gland, or larynx. The undifferentiated mesenchymal cells are capable of fibroblastic differentiation. Depending on this pattern of differentiation, histologically, the tumors are complete myxomatous or mixture of fibrous and myxoid matter. Clinically, it usually presents as a painless and asymptomatic lesion, paraesthesia and displacement of teeth usually occurs in larger sized lesions. OM mostly occurs in the 2nd and 3rd decades of life and has slight female predilection. They comprise 3–6% of all odontogenic tumors, out of which mandibular tumors are most commonly seen. Updated analyses of only 1692 cases have been reported in the literature.[1]

Considering the rare incidence of myxomas in the maxilla, case studies such as these guide surgeons for better treatment planning. In this article, a rare case of OM of the maxilla in a Male patient is reported and the varied histopathological features are discussed.

Case Report

A 21-year-old male patient visited outpatient department with a chief complaint of swelling on the left side of his face. The patient noticed a swelling which was gradually increasing in size since 5 months and had mild intermittent pain associated with it. With the increase in size, the patient also developed epiphora subsequently. His past medical, dental, and family history showed no relevant significance.

Extraoral examination revealed 4 cm × 3 cm swelling in the left middle one third of the face which is extending from the left infraorbital region to the left commissure of the lip superoinferiorly, laterally extending to the zygomatic bone. Medially, it extends up to the left lateral wall of nose. Left nasobuccal fold was obliterated [Figure 1]. The skin over the swelling is normal and not attached to the underlying tissues. Mild tenderness on palpation observed and the swelling was firm on consistency.

Intraoral examination reveals diffuse swelling expanding labial and buccal cortical plate extending from left canine to left first molar. No mobility was noted in the overlying teeth but an occlusal cant was present, with a shift of 1-1.5 cm towards the left side of the patient’s occlusion.

Investigations

Orthophantamograph of the jaws revealed an ill-defined mixed with both radiolucent and radio-opaque lesion extending from
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distal side of 23 to the mesial root of 28. Superiorly, it extended to the floor of the orbit. Superior medially extended to the left lateral wall of nose and superior laterally to zygomatic bone. Computed tomography of the region showed irregular hypodense lesion with irregular thinned margins extending medially to lateral wall of nose, superiorly extended to inferomedial aspect of the orbit [Figure 2]. Posteriorly extended up to infra temporal fossa going across the posterior hard palate and soft palate. It also involved the posterior end of nasal cavity opening into nasopharynx and into nasolacrimal duct. It laterally extended to zygomatic arch involving left maxilla and posterior lateral wall of maxilla.

Routine blood investigations, along with thyroid profile, serum calcium, phosphorus, and alkaline phosphatize, were in normal limits. Incisional biopsy was performed under local anesthesia which showed as spindle shaped cells along with loose myxomatous tissue suggestive of OM.

The patient was admitted and pre-operative work-up was done for the left maxillectomy and tumor excision. Under general anesthesia tumor was exposed through intraoral vestibular incision from 11 to 27 regions. Osteotomy was performed in the 22 region and extended posteriorly with 1 cm clearance on all sides [Figure 3]. The left maxillectomy was done and tumor excised from the lateral wall of nose and posterior extensions also had been cleared [Figure 4]. The defect left after the maxillectomy was covered with pre-fabricated obturator and was secured in its position [Figure 5]. Post-operatively, the patient was shifted to intensive care unit uneventfully after extubation. The patient was inserted with Ryles tube for the feeding purpose post-operatively. The patient was discharged after 1 week and regular follow-ups done at the intervals of 1st month, 3 months, and 6 months. Healing was uneventful during the follow-up.

Histopathology

Multiple greyish bits of 65 g weight ranging from 4.5 cm × 4 cm to 1 cm were excised. Segmental maxillectomy measuring

![Figure 1: Pre-operative profile picture demonstrating the shape and size of the lesion](image1)

![Figure 2: Computed tomography imaging showing the extent of the lesion](image2)

![Figure 3: Hemi-maxillectomy with tumor excision](image3)

![Figure 4: Excised tumor specimen collected](image4)
4.5 cm in length with attached 5 teeth was sent separately. The excised specimen revealed large areas of necrosed tissue including geographic type of necrosis. Collagenous or myxoid matrix was seen. In the areas with collagenous matrix, the cells are spindle shaped with fusiform nuclei arranged as interlacing fasciculi. At places, the nuclei are vestibular, pleomorphic, and show prominent nucleoli. Many inflammatory cells predominantly lymphocytes were seen closely to the surface. Slit such as and staghorn type of blood vessels was seen. Myxoid areas were sparsely cellular, vaguely lobulated, and polypoid and have many thin walled blood vessels. Thick walled fibrous capsule was noted.

**Discussion**

Myxomas are locally destructive benign odontogenic neoplasm. In 1863, Virchow coined the term which he later in 1871 defined as “Schleim geschwulste” meaning “Myxomata” as he considered only about soft-tissue myxomas. Soft-tissue myxomas are frequently found, but their intraosseous counterparts are rarely encountered, and those that are found are almost exclusively confined to the jaws. Very few intraosseous myxomas have been found in locations other than the jaws, as reported in the clavicle by Gupta et al. or in ribs, or femur, but most of the investigators have found that the intraosseous myxomas are most commonly found in jaws and are odontogenic in origin. Zimmerman et al. reported that the average age for the OM is 26.5 years, although majority of the investigators found that this lesion occurs in second or third decade of life. Most of the reports suggest that there is a slight female preponderance, mandibular predilection, and the lesion has a silent locally destructive nature.

On gross examination, OM appears as a grayish white, nodular heterogeneous mass of variable consistency with a glistening gelatinous cut surface (Landa et al., 2002). The tumor may have a minimal true capsule or may be unencapsulated and poorly demarcated from surrounding tissues (Rius et al., 2013). In the present case, there was capsule surrounding the tumor.

Radiographically, large multilocular radiolucency is more common in the posterior areas of the jaws and unicocular lesions are mostly located in the anterior. The lesion acquires its classic radiographic appearance, consisting of multilocular radiolucency with well-developed locules, composed of trabeculae tending to intersect at right angles, forming locules straight, thin, elongated and lacy, Eversole called this as “Lichen planus of jaw bone.” The radiographic tumor margins may be either well-defined or poorly defined. Many reviews 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. The above lesion which we had described was ill defined margins.

Sometimes the peripheral margin of the septa may be arranged at right angles to the margin, giving a ‘hair brush’ or “sunburst” appearance. It has varied radiographic presentations: Soap-bubble or honeycomb, spider web, and tennis racket appearances. Other shapes include small or large triangles, diamonds, squares, rectangles, and X, Y, and V figures.

The cause of histogenesis of the tumor remains controversial, but the tissue of origin is considered mesenchymal. Two theories of region have been proposed. First, the tumor results from myxomatous degeneration of fibrous stroma. Second, it is derived from the mesenchymal portion of the tooth germ, that is, dental papilla, follicle, or periodontal ligament. Even enlarged dental follicle or a dental papilla of a developing tooth with myxoid change may be microscopically similar to a myxoma.

An exact diagnosis is necessary to plan appropriate surgical and adjuvant therapy. Various modalities have been suggested for treatment of myxoma. Small myxomas are generally treated by curettage, but careful follow-up is necessary for at least 5 years. Larger lesions need extensive resection as myxomas are non-encapsulated and tend to infiltrate the surrounding bone. Complete removal of a large tumor by curettage is often difficult to accomplish, and lesions of posterior maxilla, in particular, should be treated more aggressively. The current treatment of OM includes resection with bony margins of at least 1.0 cm–1.5 cm and leaving behind one uninvolved anatomic boundary. Maxillectomy and sometimes resection of orbital floor are required from OM in the upper jaw. In the current scenario, maxillectomy was done and excised the tumor from the infraorbital margins and lateral walls of the nose. Defects present after maxillectomy was covered using the pre-fabricated obturator or reconstruction procedure. The above treated patient was given a prefabricated obturator, and after 2 months, it was replaced with interim obturator [Figure 6]. The patient had come for 6 months follow-up and removable partial denture with clasps was given. Period of greatest recurrence is seen in first 2 years. The studies have shown no cure with radiotherapy alone, and hence, OM is considered to be radioresistant. As OM is not encapsulated and its infiltration does not lead to any destruction of the immediate hard tissues, the risk of recurrence increases when conservative approach is applied during its treatment. These factors help to explain high recurrence rate of OM that ranges between 10% and 33% with 25% considered to be the average recurrence rate. Hence, our patient was managed with total excision of the lesion and is under follow-up at present.
Conclusion

OM is highly uncommon benign tumor that occurs in the jaws. Clinical correlation, radiographic evaluation, and histopathological reports will help in proper diagnosis and the extent of the lesion to decide the appropriate treatment plan. With the proper treatment, the prognosis will be good and long-term follow-up is necessary. Chance of recurrence will be more in the first 2 years, so the patient should be recalled for regular follow-up and both clinical and radiographic evaluation has to be done. This case was evaluated at intervals of 1st, 3rd, and 6 months and 1 year with no signs of recurrence.

References
