

CASE REPORT

Odontogenic Myxoma: A Case Report and Retrospective Analysis of Five Cases

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ABSTRACT

Odontogenic myxoma is a rare, locally aggressive, benign tumor of the jaws representing less than 0.5% of all bone tumors and only 3 to 6% of odontogenic tumors. This article reviews five cases of the same treated in the Department of Oral and Maxillofacial Surgery, KLE's VK Institute of Dental Sciences between 1993 and 2010. One case of left maxillary sinus odontogenic myxoma in a 30-year-old female patient was of aggressive nature and has been described in detail in this report.

Keywords: Odontogenic myxoma, Weber-Ferguson, Maxillary sinus.

INTRODUCTION

Myxoma, also known as odontogenic myxoma and myxofibroma is described by WHO as a locally invasive neoplasm consisting of rounded and angular cells that lie in an abundant mucoid stroma.¹ It is locally aggressive and infiltrating in nature. Its histogenesis is uncertain but supposedly of odontogenic origin in view of its occurrence in the jaws and absence of any extragnathic central form of the tumor.² It is more common in second and third decades of life.¹ There is a slight female predilection with the female: male ratio stated as 1.5-1.83:1.¹⁻³ It has been reported to occur mainly in the jaws, mostly as central lesions, but peripheral variants have been reported.² The tumor may be first noticed as swelling or asymmetry of the affected jaw, rarely associated with pain or ulceration which may manifest when it interferes with occlusion.

CASE REPORT

A 30-year-old female patient reported to the Department of Oral and Maxillofacial Surgery, KLE's VK Institute of Dental Sciences with a chief complaint of swelling over left side of face since three months. She complained of inability to breathe from left nostril. She reported no toothache or any other complaints related to onset or progression of the swelling. Extraorally, diffuse swelling of 4 cm diameter was located over left malar region obliterating the nasolabial fold (Fig. 1). It was bony hard in consistency. Skin over swelling appeared normal and showed no signs of inflammation or adhesion. A bulge from the lateral wall was seen in the left nasal cavity. Intraorally, swelling obliterated the buccal vestibule in relation to upper teeth from the lateral incisor to the first molar. A palatal swelling was also present in relation to the same dental segment. The

molar teeth were grade I mobile. There was no evidence of dental infection or any other local causative factor. No lymph nodes were palpable, tender or fixed. Radiographs and CT scan revealed a hyperintense area in left maxillary sinus extending up to floor of the orbit and invading the left lateral nasal wall (Figs 2 and 3). An incisional biopsy of the lesion was reported as fibromyxoma. Patient was planned and taken up for surgical excision of the tumor. All investigations were within normal limits.

Surgery was performed under general anesthesia. Patient was painted and draped in usual manner. Marking was made for the Weber-Ferguson on incision for extraoral approach (Fig. 4). The incision was made and flap reflected to access the bony swelling. On deroofting the bone through a Caldwell-Luc approach, the underlying lesion was exposed. The tumor occupied the entire left maxillary sinus. It extended up to the floor of the orbit superiorly but did not invade the same. The inferior orbital rim was intact. Medially the lateral nasal wall had been invaded with extension of the tumor into the nasal cavity up to the nasal septum. Inferiorly, the alveolar processes were involved. It was soft to firm with glistening, stringy appearance and mucoid consistency (Fig. 5). Enucleation was carried out till the orbital floor above, the nasal septum to the right and the sinus was completely evacuated. Owing to the involvement of the palate, a near hemimaxillectomy was carried out from the left lateral incisor to second molar region sparing the third molar to support prosthesis for rehabilitation (Figs 6 and 7). An intranasal antrostomy was done and antibiotic pack was placed (Fig. 8). Pack was removed on third postoperative day. Postoperatively, the patient was kept nil by mouth and



Fig. 1: Swelling in left malar region



Fig. 4: Marking for Weber-Ferguson incision

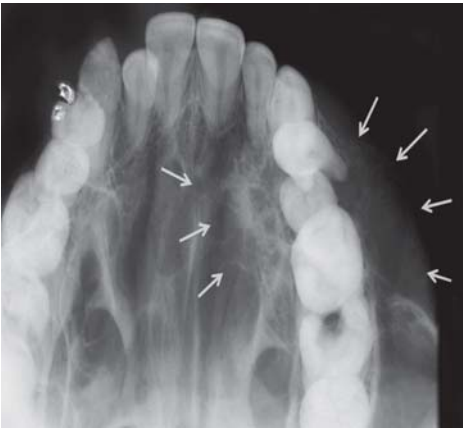


Fig. 2: Occlusal radiograph showing soap bubble appearance

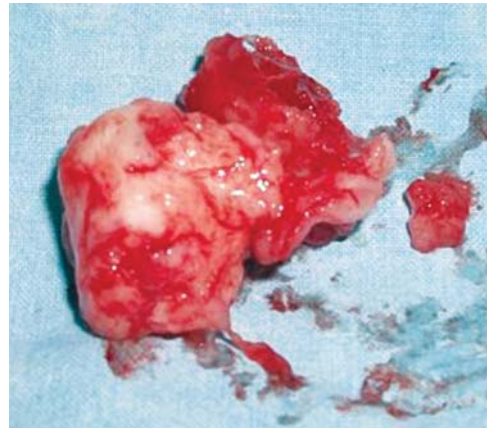


Fig. 5: Gross specimen, mucooid consistency

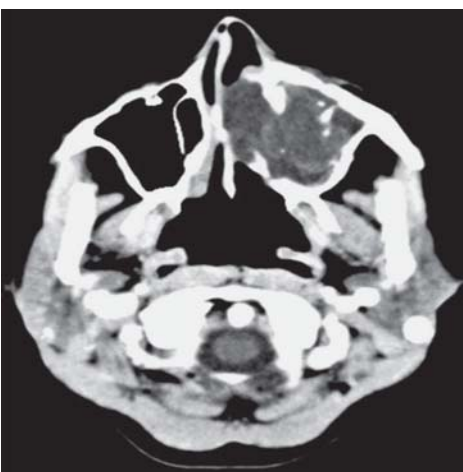


Fig. 3: CT scan showing tumor in left maxillary sinus



Fig. 6: Surgical defect

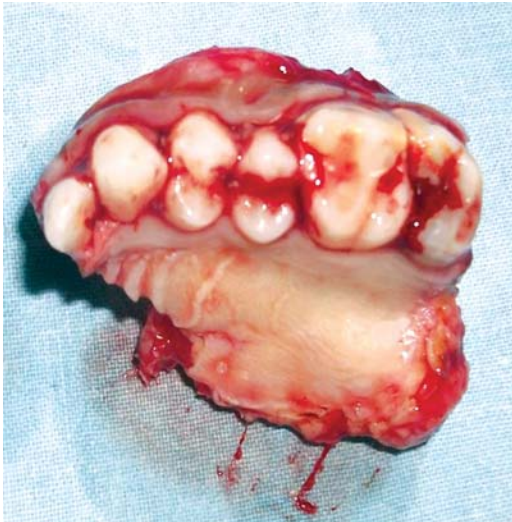


Fig. 7: Gross specimen, left maxilla



Fig. 10: Histological picture



Fig. 8: Antrostomy, suture line



Fig. 9: Obturator *in situ*

was on nasogastric tube feeding. On fifth postoperative day, an obturator was delivered with a hollow bulb to fit into the defect (Fig. 9). Patient recovered uneventfully.

The histopathological features were reported as stroma composed of myxoid tissue with spindle to stellate-shaped fibroblasts (Fig. 10). Many odontogenic cell rests were present all over the stroma with loose collagen fiber bundles. All these features are suggestive of 'odontogenic myxoma'.

REVIEW OF CASES

Five cases of odontogenic myxoma were operated in our institution between 1993 and 2010. There was a male predominance with male to female ratio of 3:2. In two cases, tumor was situated in the left maxillary sinus; in one case in region of left maxillary third molar and in two cases it was present in relation to left mandibular third molar. In three cases, the tumor appeared as a swelling of bony cortices in relation to the specified teeth with slight mobility of teeth. In two cases, the tumor was present occupying the maxillary sinus, the one described above being more aggressive and invading the lateral nasal wall and extending up to the orbital floor. All cases were treated with resection of the tumor. There has been no evidence of recurrence.

DISCUSSION

The myxoma is a rare bone tumor that represents less than 0.5% of all bone tumors and only 3 to 6% of odontogenic tumors.⁴ Histologic features were probably first described by the German pathologist Rudolph Virchow in 1863, though the mention was not specific to the jaws. Thoma and Goldman were the first to describe myxomas of the jaws in 1947.²

The origin of this tumor is debatable. It has been traditionally considered to be of odontogenic origin supported by the

following points: It almost exclusively occurs in the tooth bearing areas of the jaws, frequently seen in younger individuals, commonly associated with an unerupted tooth or developmentally absent tooth, histologically resembles dental mesenchyme specially of the dental papilla, and the presence of odontogenic epithelium occasionally.¹ This however is challenged by studies by Goldbat and Slootweg who proposed that the development of myxoma may be from nonodontogenic ectomesenchyme as it is observed to develop within sinonasal tract or nontooth bearing regions of jaws.⁵ They proposed that a diagnosis of myxoma of bone may be made in absence of odontogenic epithelium. Contradicting this, McClure and Dahlin concluded from their study of 600 bone tumors that true myxomas occur only in jaw bones.⁵

Kaffe and Farman et al separately reported that these tumors are generally more predominant in second to fourth decades of life. Kaffe et al state in their study that 75% occurred in second and third decades of life, 7% in the first decade, but can be seen in any age group.^{1,2} All cases treated in our institution were in the second and third decades except for one patient who was 45-year-old. Harder was of the view that the tumor rarely occurred before the age of 10.¹ This has been contradicted by Keszler,⁶ Brian W Rotenberg,⁷ Caleffi Edoardo,⁸ and Brewis⁹ in their individual case reports describing the tumor in pediatric population.

The central odontogenic myxoma has been described to occur in the tooth bearing and nontooth bearing areas of the jaws, more common in the mandible than the maxilla in premolar and molar regions.¹ In our cases, three of five were found in the maxilla and two in the mandible, all involving the premolar and molar regions. It has been observed that these tumors rarely cross the midline.¹⁰ Peripheral variants on gingiva have also been reported.¹¹ Reports describe the occurrence of myxoma in association with other conditions like tuberous sclerosis and condyloma acuminatum.^{12,13}

Radiographically, myxomas may be unilocular or multilocular mostly radiolucent lesions. Kaffe et al reported mixed appearance in 12.5% and radiopaque appearance in 7.5%.¹ In the cases reviewed by us, four cases were radiolucent and one case was mixed radiolucent and radiopaque. This tumor has been described as honeycomb, soap bubble, wispy and spider web in appearance. J Zhang and H Wang suggested a classification for radiologic appearance of myxomas as follows: Type I—unilocular; type II—multilocular (including honeycomb, soap bubble and tennis racquet patterns); type III—involvement of local alveolar bone; type IV—involvement of maxillary sinus; type V—osteolytic destruction and type VI—a mix of osteolytic destruction and osteogenesis.¹⁴ In our cases, three involved the alveolar bone and can be classified as type III, one involved the maxillary sinus and can be called type IV, whereas the one described showed osteolytic destruction and can be placed under type V. Koseki and Asaumi et al have studied the CT and CT/MRI findings in myxoma respectively and observed that the CT and MRI findings correlate well with

the histopathology of the tumor.¹ In the described case, CT proved to be a valuable aid in defining the extent of the tumor before surgery (Fig. 3).

Surgical management of the tumor involves complete excision of the tumor. A wide excision and even resection is suggested in view of high recurrence rates of up to 25%.² Curettage may be insufficient but conservative resection in the pediatric age group has been suggested.⁷ All our cases were treated with complete resection of the tumor and none of them reported back with recurrence.

Odontogenic myxoma is a nonencapsulated benign tumor. Histologically, it shows a bland appearance of a mucoid matrix with loosely arranged spindle shaped and rounded cells, which are evenly dispersed. 'Cavities' lined by poorly defined fibers as well as epithelial islands have been described by JD Harrison.¹⁵ In the case report by Sivakumar G, the cells were differentiated into hyaline, stellate and spindle shaped for immunohistochemical analysis.¹⁵ Spindle cells were positive to ferritin, transferrin, alpha-1-AT, alpha-1-ACT, S-100 protein and vimetin. Stellate cells showed strong positive response to transferrin, alpha-1-AT, S-100 protein and vimetin. Hyaline cells reacted with alpha-1-AT and alpha-1-ACT. Myxomatous matrix did not react with any antibodies. These findings were suggestive of myofibroblastic origin of the tumor.¹⁵ G Iezzi from the research conducted suggests that the odontogenic epithelium may represent either an inactive form or have an inductive effect leading to formation of the myxoma.¹⁶ This study presents the epithelium to be reactive to bcl-2, p 53 and MIB-1 contradicting previous views it may be inert. The stromal cells also showed higher positivity to MIB-1¹⁶ suggesting contribution by both epithelium and stroma towards increase in tumor mass. They also suggest a dysregulated apoptotic mechanism involved in neoplastic growth.

Reconstruction of the defect can be done with prosthetic means or with an autogenous graft. Literature reveals reconstruction of defects postexcision of myxoma done using vascular free fibula graft for mandibular defects¹⁷ and maxillary defects restored using endoscopically harvested temporalis flap,¹⁸ reverse facial artery – submental artery mandibular osteomuscular flap.¹⁹ Nardy Kasap has described the use of a surgical navigation system for implant surgery after excision of an odontogenic myxoma.²⁰

CONCLUSION

Odontogenic myxoma is a rare, benign, locally invasive tumor of jaws. It may have an insidious onset, usually in the second and third decades of life. The patient may first report to a dentist due to association of bony swelling with the alveolar segment of the jaw or being accompanied by mobility of teeth. If ignored, it might assume a larger size and might invade surrounding bone. A biopsy of involved bone can aid in its diagnosis. Complete resection of the tumor as well as long-term follow-up is recommended.

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