

ODONTOGENIC MYXOMA IN THE MAXILLA: A REPORT OF A RARE CASE

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SUMMARY

The aim of this paper is to determine the unique and very interesting case of odontogenic myxoma, which involved the right maxilla and its sinus. Odontogenic myxomas are considered to be a benign odontogenic tumor with locally aggressive behavior and non-metastasizing neoplasm of the jaw bones. It represents an uncommon benign neoplasm comprising 3–6% of all odontogenic tumors. It is usually derived from the dental mesenchyme or periodontal ligament. Despite the benign nature of myxoma, there is a high rate of local recurrence after curettage alone and thus in certain cases adequate resection is the only option.

KEYWORDS:

Odontogenic Myxoma(OM), Odontogenic Tumors, Maxilla, Sinus, Computed tomography.

INTRODUCTION

Odontogenic myxoma is a benign tumor with mesenchymal origin. A relatively rare but is almost exclusively seen in tooth-bearing areas. It is an asymptomatic lesion that shows an infiltrative growth pattern. Such lesion causes a destruction of the medullar bone and expansion of the cortical bone. Frequently, the lesion is able to invade and perforate the adjacent soft tissues and cortical bone. The lesion usually presents, approximately, 25% of recurrence, mainly when conservative treatment is focused. Truly speaking the origin of odontogenic myxoma is still obscure. Cells of the dental papilla, folliculous or periodontal ligament are blamed of being the origin of this lesion.

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Other investigators believe that the fibroblast is the responsible cell that allows the tumor to spread. Usually myxoma occurs in the second and third decades of life being rare in children and adults older than 50 years. The posterior region of the jaw is the most frequent site where it occurs, having an equal Distribution in males and females. A histological characteristic of this tumor resembles the mesenchymal portion of a tooth in development. The lesion is not encapsulated being characterized by a proliferation of a few rounded cells, fusiforms and star cells, being stranded in an abundant myxomatous estroma with a few collagen fibers. Small islands of odontogenic epithelial tissue can be found scattered across estroma and they are key cells to establish the diagnosis.

The treatment of choice is surgical, enucleation and curettage. Because of its infiltrative behavior, this lesion is difficult to be curretted, and this is rationale in wake of high recurrence rate. Cryotherapy as an adjunct modality of treatment to curettage can be used as such technique minimizes the risk of recurrence.

CASE REPORT

Case History and Clinical Findings:

A 16 years old female presented to the department of oral & maxillofacial surgery department with a chief complaint of swelling on the right side of the face for the last few months .The swelling was clinically limited to the right infraorbital region with obliteration of the nasolabialfold. There was no

fluid on aspiration. Incisional biopsy was undertaken, which showed rubber-like and jelly-like consistency. The computed tomography images revealed an expansive mass in the right maxilla.



Figure: Intraoral view, swelling extending up to lateral incisor anteriorly to the second molar posteriorly.

Imaging Evaluations:

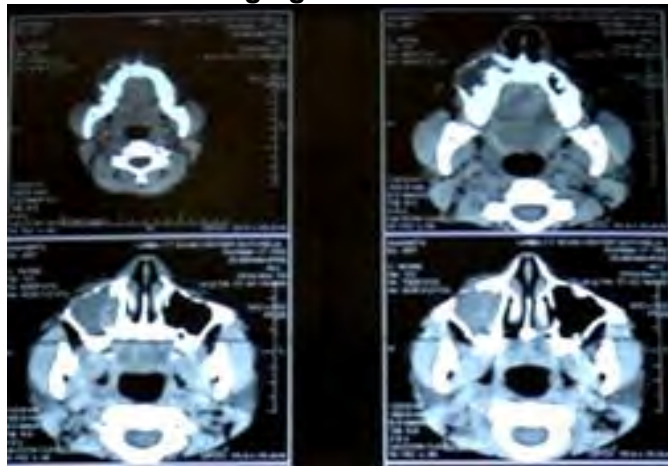


Figure: CT scan showing tumor expansion involving maxilla and maxillary sinus

Computed tomographic images showed a single large expansile radiolucent lesion with multiple radio-opaque foci seen on the right side of craniofacial region, involving the maxillary sinus with erosion of the alveolar bone and medio-latero-superior walls of the sinus.

MANAGEMENT

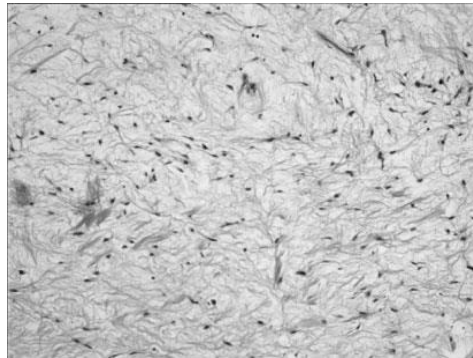
Under general anesthesia through an intraoral approach, the patient underwent total removal of the tumor with a partial en bloc resection of the maxilla. Bone skimming was done using a large round bur. Extraction of the lesion inflicted teeth # 12, 13, 14, 15, 16 was done to prevent the chances of recurrence. The tumor had a jelly like consistency which thus peeled easily from the bone. The patient was followed first after a week & then after one month duration. The healing was satisfactory with mild swelling and slight blood in the sputum in the first few days.



FIG: Post-Op Picture & Maxillary Odontogenic Myxoma Dissected Mass.

HISTOLOGIC CHARACTERISTICS

The histologic specimen was stained using hematoxyline-eosine. Such staining showed a neoplasia of odontogenic origin formed by an intense myxomatous tissue proliferation which was well vascularized, contained spindle and small fusiform fibroblasts, with flabby deposits of winding and delicate collagen and some mononuclear inflammatory cells, features that clued the presence of an odontogenic myxoma.



DISCUSSION

Odontogenic myxoma is a rare aggressive intraosseous lesion derived from embryonic mesenchymal tissue associated with odontogenesis and primarily consisting of a myxomatous ground substance with widely scattered undifferentiated spindled mesenchymal cells. Though it is a benign neoplasm, it may be infiltrative and aggressive in nature with history of recurrence.

Odontogenic myxoma almost exclusively occurs in the jaw bones, comprising around 3-6% of all the hardly odontogenic tumors. The tumor affects the age group that spans from 22.7 to 36.9 years. It is hardly seen in patients younger than 10 years of age or older than 50. Our case presented at the age of 16 years, which is in conformity with that reported in literature. The mandible appears to be more frequently affected than the maxilla; especially the posterior region has more predilections. In our case posterior region of the maxilla is involved. Majority of myxomas are asymptomatic, although some patients present with progressive pain and with eventual neurological symptoms on the rise. Odontogenic myxoma of the jaw has a tendency for extensive devastation of and surrounding structures and a relatively high recurrence rate; however, metastasis is rare. OM of the maxilla is less frequent but behaves more aggressively than that of the mandible, as it spreads

easily through the maxillary spongy bone and its sinus as compared to tougher mandibular cortical bone as presented in our case.

Odontogenic Myxomas radiographically reflect multilocular or unilocular radiolucencies. unilocular are more frequently confined to the anterior region of the jaws, while multilocular occur mostly in the posterior region. Odontogenic myxoma should be included in the differential diagnosis of both radiolucent and mixed lesions, in both the jaws, for individuals of almost all age groups. When unilocular and without trabeculae, the tumor closely resembles periapical, lateral, periodontal and traumatic bone cysts. When multilocular, it must be differentiated from ameloblastoma, central hemangioma and odontogenic keratocyst. Odontogenic Myxomas is a neoplasm which lacks any distinct capsule. A spectrum of fibrous connective tissue stroma is existing from myxoid to densely hyalinized and from relatively acellular to cellular. Calcification may or may not be there. It is characterized by the presence of sparse cords and islands of inactive odontogenic epithelium. The variation in the histopathological diagnosis between the initial biopsy as odontogenic fibroma and the final histopathological diagnosis as odontogenic fibromyxoma in our case could be attributed either to the biological nature of this lesion or missing of myxomatous areas in the biopsy. The ultra-structural findings indicate that the odontogenic fibroma and the odontogenic myxoma share many common traits.

MORPHOLOGICAL FEATURES

An immunohistochemical panel of poly- and monoclonal antibodies was used to characterize and distinguish the nature of cells as of fibroblastic, histiocytic, myoblastic and neural origin. Three types of odontogenic myxoma cells were differentiated spindle cells, stellate cells and hyaline cells. Neoplastic cells of myxomas were positively stained for transferrin, ferritin, alpha-1-antichymotrypsin (alpha 1-ACT), alpha-1-antitrypsin (alpha 1-AT), S-100 protein, vimentin (pan-mesenchymal marker) and actin; however, neuron-specific enolase (NSE), S-100 alpha subunit, S-100 beta subunit, Factor VIII-related antigen (FVIII-AG) and cytokeratin (CK1) were negative. Antibodies directed against vimentin are used to identify mesenchymal cells, and keratin antibodies detect epithelial differentiation. Spindle cells were positive for transferrin, ferritin, alpha 1-ACT, alpha 1-AT, S-100 protein and vimentin. Stellate cells were strongly positive for transfer in, alpha 1-AT, S-100 protein and vimentin. Hyaline cells reacted with alpha 1-ACT and alpha 1-AT. Myxomatous matrix showed no reaction to all the antibodies used. These results confirm that odontogenic myxoma is a tumor of a dual fibroblastic-histiocytic origin and also suggest that the cells comprising odontogenic myxoma are of myofibroblastic origin.

The aggressive nature of OM is self-evident and well documented in the literature. The tumor is not radiosensitive, and surgery is the treatment of choice. The lack of a capsule and infiltrative growth pattern is responsible for high rate of recurrence when only conservative enucleation and curettage are chosen. Recurrence is controlled with extensive partial or total resection procedures, and this method of treatment is particularly indicated in the maxilla due to its proximity to the vital structures.

CONCLUSIONS

This case signifies the level of inclusion of myxoma in the differential diagnosis of radiolucent &/or radiopaque lesions of mandible or maxilla. Proper pre-operative radiographic examination is important in order to determine the bony involvement and the scale of the tumor. This should include plain radiographs obtained in different projections, and in challenging cases also three-dimensional techniques as CT-scans and MR-images. Input of an experienced pathologist is essential, since this diagnosis may be the turning point to either surgery or a more conservative treatment. Its follow-

up must be made mandatory, with imaging assessment to detect any early recurrence. In case of large tumor resections, the reconstruction procedures must be delayed until substantial disease free period has been observed.

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