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CASE REPORT

Odontogenic Myxoma of the Maxilla

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ABSTRACT

Aim: To report a case report of odontogenic myxoma and to present a brief literature review to highlight the histogenesis of this pathology.

Background: Myxomas are benign tumors of mesenchymal origin that rarely appear in the skeleton. Odontogenic myxoma (OM) is an unusual (comprising of 3-6% of all odontogenic tumors) and locally invasive benign neoplasm of uncertain histogenesis, found exclusively in the jaw bones. Maxillary lesions are further uncommon and behave more aggressively, as it spreads through the maxillary sinus, reaching a considerable size before being detected.

Case report: We report a rare case of OM occurring in the maxilla of a 24 yr old male who presented with a relatively large swelling which had caused considerable destruction of the surrounding bone. The lesion was excised and the patient is on follow-up.

Conclusion: Co-relation of clinical, radiological and histopathological features is essential when trying to diagnose lesions with aggressive clinical behavior.

Clinical significance: Owing to its clinical behavior and high recurrence rate, early detection coupled with complete surgical removal and long term follow-up is highly essential.

Keywords: Aggressive, Maxilla, Odontogenic, Recurrence.

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BACKGROUND

Myxomas of the head and neck are rare tumors of uncertain histogenesis. They are benign, aggressive, locally invasive, and rarely appear in the skeleton. When they are found in osseous sites, they almost exclusively manifest in the jaws and were first described by Thoma and Goldman in 1947.[1] Odontogenic myxoma (OM) is a nonencapsulated benign tumor of the jaws with a very rare occurrence rate. The tumor grows gradually with accumulation of mucoid ground substance with minimal collagen. The origin of OM is believed to be the mesenchyme of a developing tooth or the periodontal ligament.[2] There are variable reports on the relative frequency of OM in the available literature. In Asia, Europe, and America, OM frequencies between 0.5 and 17.7% of all odontogenic tumors have been reported.[1,3] Others report the frequency to be around 3 to 6% of all odontogenic tumors. The tumor mostly affects young patients in their second and third decade of life.[4] It occurs across an age group that varies from 22.7 to 36.9 years. It is rarely seen in patients younger than 10 years or older than 50 years.[5] The OM is usually a slow and painless swelling occurring more often in the mandible than maxilla, especially in the molar region. Displacement of teeth and paresthesia are uncommon clinical features. When located in the maxilla, OM often involves the maxillary sinus.[6] Despite the benign nature of these lesions, there is a high rate of local recurrence after curettage alone and in certain cases it requires a resection of the surgical area.[7] It therefore reaches considerable size before being detected, and perforation of the cortices of the involved bone may be seen.[5,8] The radiographic features of OM are not pathognomonic, but the most common presentation is a multilocular radiolucency frequently described as honeycombed, soap bubble, or tennis racket appearance. Unilocular appearance may be seen more commonly in children and in the anterior part of the jaws.[9] Although OM presents as an asymptomatic expansion, due to the accumulation of mucoid or gelatinous gray-whitish tissue that replaces the cancellous bone, it sometimes results in perforation of the cortical borders of the affected bone. In view of its rarity, the present case of OM of the maxilla in a 24-year-old male infiltrating the maxillary antrum and adjacent tissues is herewith reported.

CASE REPORT

A 24-year-old male patient reported with a 2-month history of painless swelling in the maxillary right quadrant. On extraoral examination, swelling extended anteroposteriorly from the corner of the mouth to zygomatic buttress and superoinferiorly from the infraorbital margin

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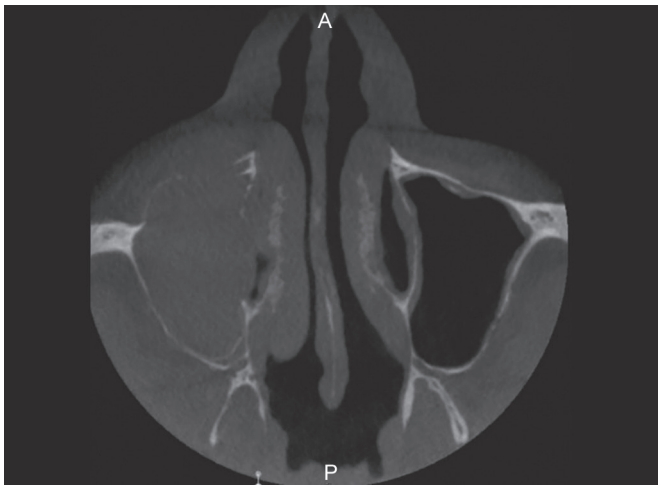

Fig. 1: The CBCT axial section showing soft tissue radiopacity involving the entire maxillary sinus with discontinuity in the mesial and anterior walls

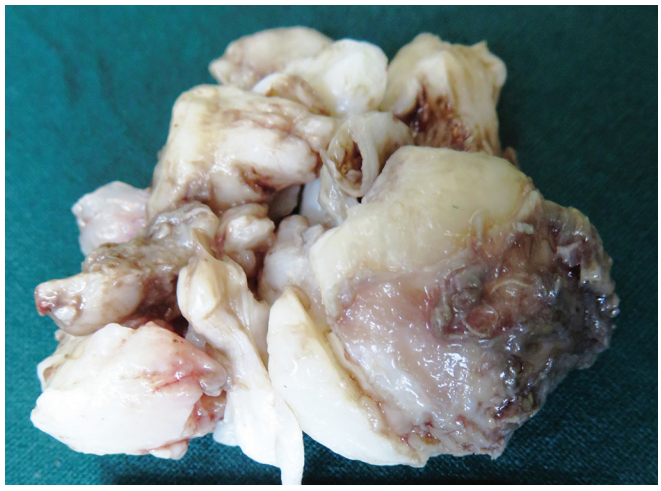

Fig. 2: Gross specimen showing a rubbery, gelatinous soft tissue mass

to upper lip with deviation of nasal ala. On inspection, a smooth-surfaced mucosal colored swelling of 4 cm × 3 cm × 2 cm was observed from mesial aspect of 13 extending up to 16, obliterating the buccal vestibule. On palpation, the swelling was nontender, nonfluctuant, firm in consistency, and fixed to underlying structures. Medical history, personal history, and family history were noncontributory. The hematological tests were within normal limits. The radiograph showed a poorly defined radiolucency with noncorticated margins extending between 13 and 16. The cone beam computed tomography (CBCT) image (axial and coronal view) showed an expansile soft tissue mass in the right maxilla. The maxillary sinus was completely obliterated with destruction of the anterior and medial borders (Fig. 1). The patient was treated surgically with total excision of the lesion. Gross pathologic examination revealed nonencapsulated soft tissue mass with ill-defined margins, gray white in color and rubbery gelatinous in texture (Fig. 2). The cut surface showed a typical glistening translucent appearance. On microscopy, hematoxylin and eosin (H&E)-stained sections exhibited haphazardly arranged stellate cells and spindle-shaped cells in an exuberant, loose myxoid stroma. Small islands of odontogenic epithelial rests were also noted (Fig. 3).

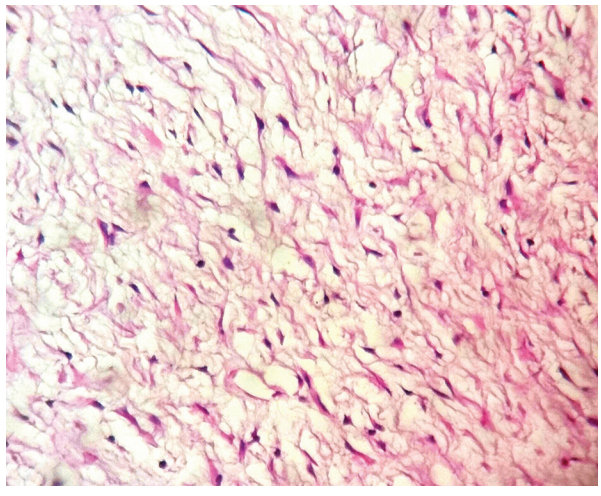

Fig. 3: Photomicrograph (40×, H&E) showing pale staining spindle cells against a myxoid background

DISCUSSION

The OM 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.[5] According to the World Health Organization, OM is classified as a benign tumor of ectomesenchymal origin with or without odontogenic epithelium.[10] Histogenesis of OM has been a subject of debate since long. They are postulated to originate from the dental papilla, follicle, or periodontal ligament by various authors.[2,5,11] Some regard OM as a modified form of fibroma in which the myxoid intercellular substance separates the connective tissue.[5] According to Lucas, the classification of the OM as an odontogenic tumor has been justified by its frequent occurrence in adolescence; its association with missing or unerupted teeth; and the sporadic presence of odontogenic epithelium within the neoplastic, myxomatous tissue.[12] The OM is usually described as a slow-growing tumor and is generally asymptomatic. The OM of the jaw has a tendency for extensive bone destruction, invasion into surrounding structures, and a relatively high recurrence rate.[5] Due to poor follow-up and lack of reports, a precise analysis of recurrence rates is still missing. However, an average recurrence rate of 28% has been reported in the literature.[12] The OM is a rare finding in maxilla but behaves in a more aggressive fashion than that in mandibular lesions, since it is known

to spread through the maxillary sinus, as observed in our case. Radiologically, two main presentations are reported in the literature. Commonly, a multilocular radiolucency is found, with cystic-like areas of variable radiolucency, separated by many bony trabeculae, usually of a well-defined nature. This appearance is frequently referred to numerously as "soap bubble", "tennis racket", and "honey combed". A less characteristic or rare presentation is the unilocular appearance. It is suggested that the unilocular lesions tend to behave less destructively than the multilocular variety.[8] A number of lesions should be included in the differential diagnosis including ameloblastoma, intraosseous hemangioma, aneurysmal bone cyst, central giant cell granuloma, metastatic tumor, and, in cases of unilocular lesions, simple cysts.[6] Dental follicle or papilla may be misinterpreted as an OM[8] due to variable degree of myxoid changes and presence of odontogenic epithelial rests; but unlike myxomas, follicles are partially or totally lined by epithelium. Based on this type of cellular differentiation, the histological pattern of OM also varies. It may exhibit complete myxomatous tissue or mixed myxomatous and fibrous tissue, according to which terms, such as "myxofibroma" or "fibromyxoma" appear in the literature. Histologically, myxomas are composed of spindled and stellate cells in a mucoid-rich (myxoid) intercellular matrix.[8] Odontogenic epithelial nests are present in a few cases, but they do not necessarily dictate diagnosis. Since OM tumor cells are mesenchymal in origin, they readily express muscle-specific actin and vimentin. Conflicting description of S-100 and glial fibrillary acidic protein positivity has been reported.[4] The matrix exhibits different proteins, mostly type I and type IV collagen, fibronectin, and proteoglycans.[4] The treatment of OMs depends on a variety of factors, such as the tumor size, nature of the lesion, and behavioral pattern. And it varies from local excision, curettage, enucleation, to radical resection. Lack of encapsulation and infiltration into the adjacent tissues and marrow spaces makes complete surgical curettage of these lesions a highly difficult task.[4] Recurrence is minimized with extensive partial or total resection procedures, and this method of treatment is particularly indicated in the maxilla due to the proximity of vital structures.[5]

CONCLUSION

The OMs are slow growing in nature but tend to be aggressive and locally invasive. The surgeon must correlate the clinical, radiological, and histopathological findings, since this tumor presents itself with a wide variety in clinical and radiological appearances. Resection with wide margins is the preferred treatment of choice and long-term follow-up is highly recommended due to the high recurrence rate.

CLINICAL SIGNIFICANCE

Owing to its clinical behavior and high recurrence rate, early detection coupled with complete surgical removal and long-term follow-up is highly essential.

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