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CASE REPORT

Odontogenic Fibromyxoma of Maxilla: A Rare Case Report

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ABSTRACT

Odontogenic fibromyxoma, a myxoma with abundant collagen fibers, is a rare benign tumor of jaw which mostly affects posterior part of mandible. It is ectomesenchymal in origin and probably arises from connective tissue of dental follicle or papilla. Commonly occurs in 2nd or 3rd decade with slight female predilection. Radiological investigations, such as conventional radiography, computed tomography (CT) scan or magnetic resonance imaging (MRI) can be used to differentiate it from other odontogenic tumors like ameloblastoma. Management is surgical by enucleation and curettage or by en bloc resection and segmental maxillectomy. Follow-up of patient is must for at least 2 years to diagnose any recurrence. In this article, we present fibromyxoma of maxilla which is a very rare site for occurrence of fibromyxoma and this relatively rare condition has been discussed in light of recent information from literature with a case report.

Keywords: Fibromyxoma, Mesenchymal maxillectomy, Myxoma, Odontogenic.

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INTRODUCTION

The odontogenic myxoma is a tumor of the jaws which apparently arises from the mesenchymal portion of the tooth germ, either the dental papilla, the follicle or the periodontal ligament. It is a rare, slow growing, noncapsulated benign but locally invasive tumor of jaw. It occurs commonly in 2nd or 3rd decade. Occurrence is rare in children below 10 of age and adults above 50 years of age.[1,2] A slight predilection for female with male to female ratio of 1:1.83 has been seen.[3] Incidence of new cases is approximately 0.07 cases per million population.[3] It is considered to be ectomesenchymal in origin. According to World Health Organization (WHO), odontogenic myxoma is defined as 'a locally invasive neoplasm consisting of rounded and angular cells lying in an abundant mucoid stroma'.[4] Odontogenic fibromyxoma is a myxoma with abundant collagen fibers. It mostly affects posterior part of mandible. Maxilla is a very rare site for fibromyxoma to occur. Only few cases of maxillary fibromyxoma have been reported so far in literatures till date.[5,6]

Clinically, it presents as a slow growing asymptomatic swelling. Pain and paresthesia can occur in advanced stages. Facial asymmetry and loss of nasolabial folds may occur due to large lesions. Displacement and mobility of teeth are relatively common. It may be associated with unerupted teeth. Cortical expansion can occur and large lesions can cause invasion of surrounding structures.[7,8] The exact pathogenesis of myxoma is still uncertain but origin from dental follicle seems to be most reasonable explanation.[9,10] Myxomas may present similar radiological features as that of ameloblastoma or central giant cell tumor which leads to difficulty in diagnosing a case of myxoma only on radiological basis. Biopsy is must to make an exact diagnosis.[11-13]

The aim of this article is to present the rarity of fibromyxoma of maxilla, the role of radiological investigation for differential diagnosis and the importance of meticulous treatment in order to prevent recurrence.

CASE REPORT

A 13-year-old girl presented with chief complaint of unilateral swelling over face on left side for 7 months. The swelling was initially small and it gradually increased in size over last 7 months to its present size. The swelling caused her no pain or numbness over face or mucosa (Fig. 1).

On extraoral examination, there was facial asymmetry as there was a diffuse nontender swelling of 3 × 4 cm over left anterior maxilla. No abnormality in skin over swelling seen (Fig. 2). On palpation, temperature over skin was normal. Swelling was nontender, firm in consistency, non-mobile and not adherent to skin. Nasolabial fold was obliterated on left side because of swelling. On intraoral examination, swelling was present in upper left buccal vestibule, extending from mesial part of right central incisor to mesial part of left first premolar. Left central incisor, lateral incisor and canine

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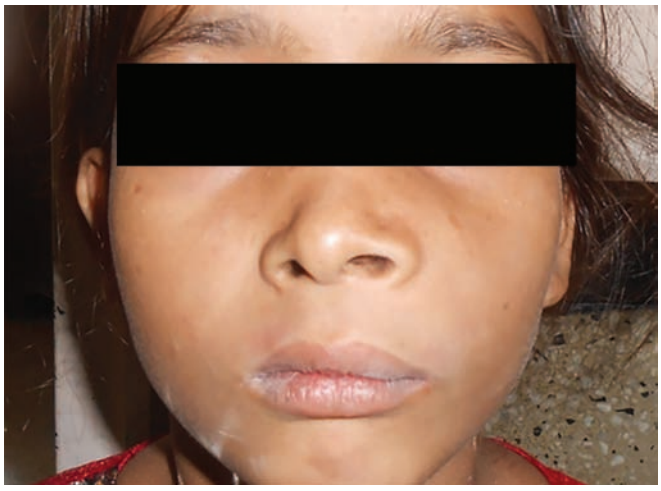


Fig. 1: Patient showing the presentation as any other maxillary sinus lesion

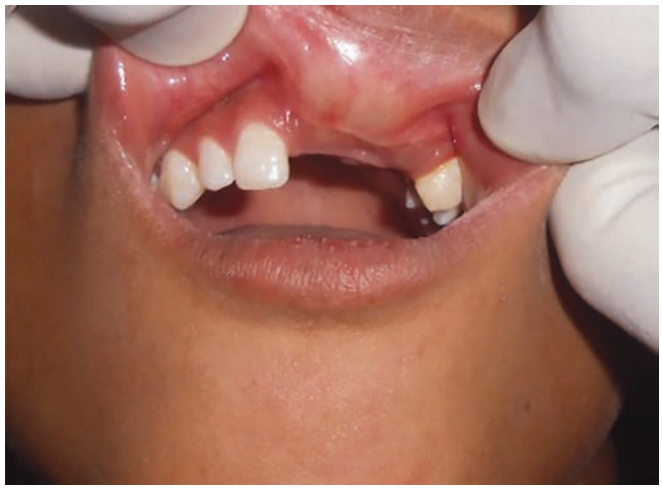


Fig. 2: On examination

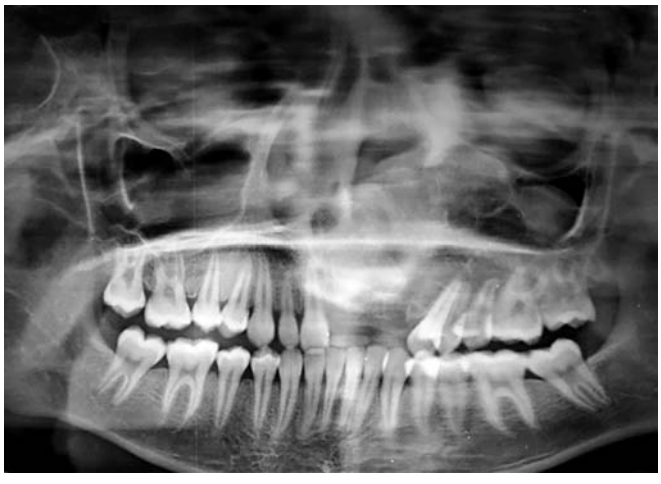


Fig. 3: Orthopantomograph

teeth were absent. Mucosa over swelling appeared normal. Effect of swelling on palate was less evident. Other general and systemic examinations were within normal limits.

Fine-needle aspiration cytology (FNAC) was done which was non-informative. Patient was subjected to radiological investigations by means of orthopantomograph (OPG) (Fig. 3), occipitomental (OM) view for paranasal sinuses and computed tomography (CT) scan (Fig. 4). Orthopantomograph revealed ill-defined radiolucent lesion extending from right central incisor to left first premolar and also involving left maxillary antrum. Occipitomental view showed haziness of left maxillary antrum. Computed tomography scan showed a hypodense, radiolucent lesion extending from area of central incisor to left first premolar. Lesion was obliterating the left maxillary antrum and expansile in nature causing erosion of anterior and medial wall of maxillary sinus. No erosion of palate or floor of orbit seen. Lesion did not enhance on intravenous contrast administration. Routine hematological and serological studies of calcium, phosphorus and alkaline phosphatase were normal.

The lesion was approached by sublabial incision and tumor was removed by enucleation and curettage (Fig. 5). It was white gelatinous lesion and had solid consistency. Excised tissue was sent for histopathological examination which revealed predominantly myxomatous ground substance with loosely arranged spindle shaped and stellate cells. Occasional vascular spaces and areas of calcification were also seen (Figs 6A and B). Histopathological examination results led to final diagnosis of fibromyxoma of maxilla. Postoperative period was uneventful. Patient was advised to visit regularly for follow-up. There was no sign of recurrence after 6 months of follow-up.

DISCUSSION

Odontogenic fibromyxoma is slow growing, non-encapsulated, benign lesion of jaw with rare occurrence. Virchow used term myxoma first of all in 1863. In 1964, Marcove et al described the term fibromyxoma.[14,15] Earlier in 1947 Thoma and Goldman first described myxomas of the jaws.[16] The tumor is consisting of accumulation of mucoid ground substance with collagen. It is amount of collagen which determines if it can be called as fibromyxoma.[3] Odontogenic myxoma is believed to originate from mesenchyme of developing tooth or periodontal ligament.[3,17] It is interesting to note that odontogenic myxomas only occasionally shows the presence of odontogenic epithelium.[3,18] However, its histological similarity to the stellate reticulum of a developing tooth, its exclusive occurrence in close proximity to the tooth bearing parts of the jaws, the occasional association with a missing or unerupted tooth, the presence of odontogenic epithelium in a minority of cases and the fact that it rarely appears in other parts of the skeleton, offer support to an odontogenic origin.[3,19]

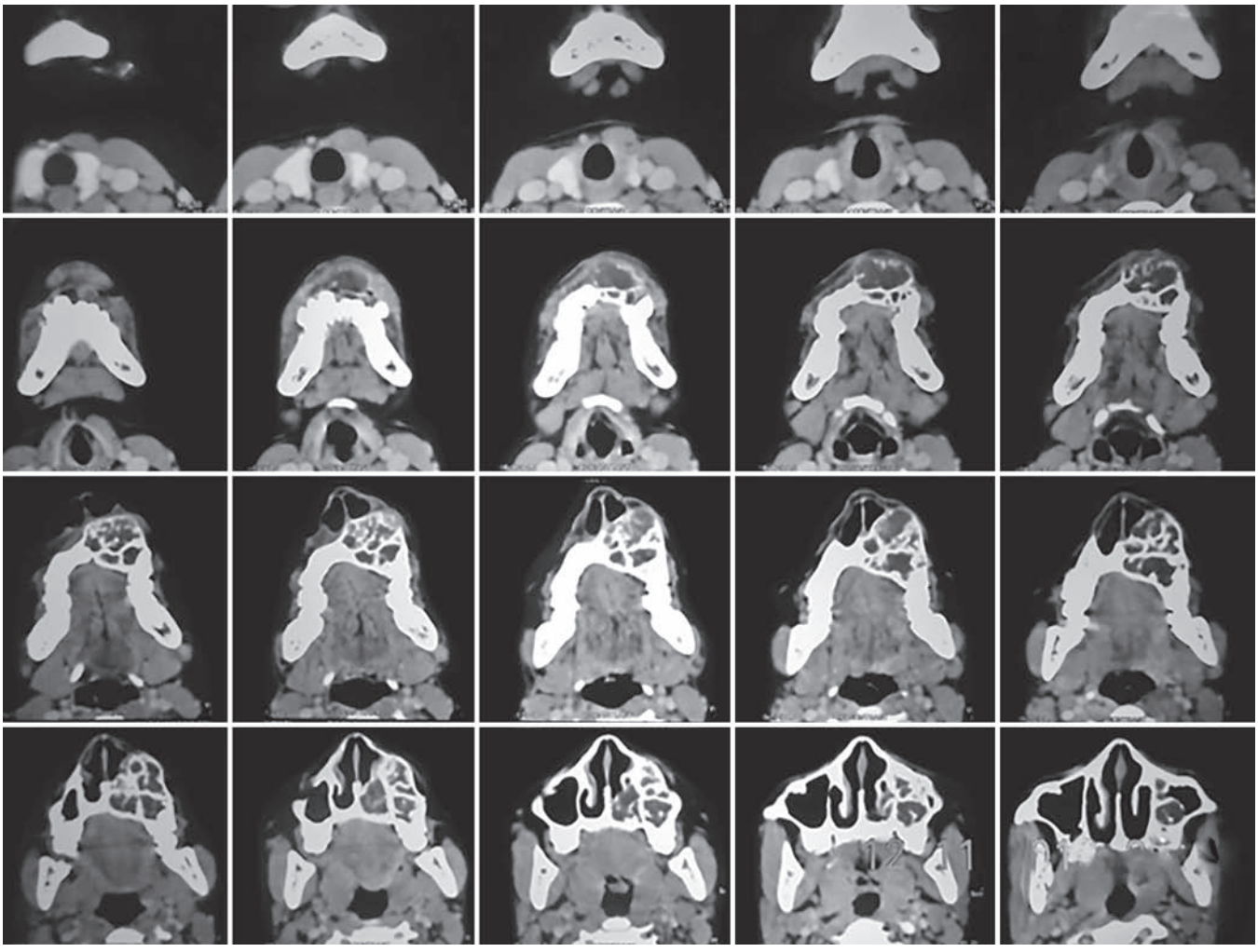


Fig. 4: Computed tomography scan showing the extent

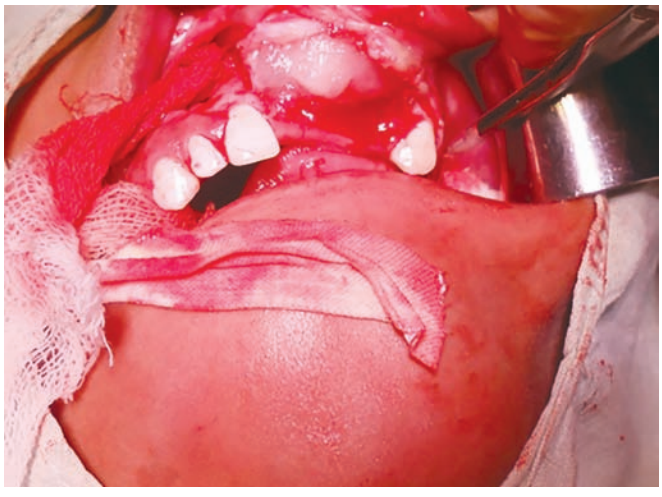
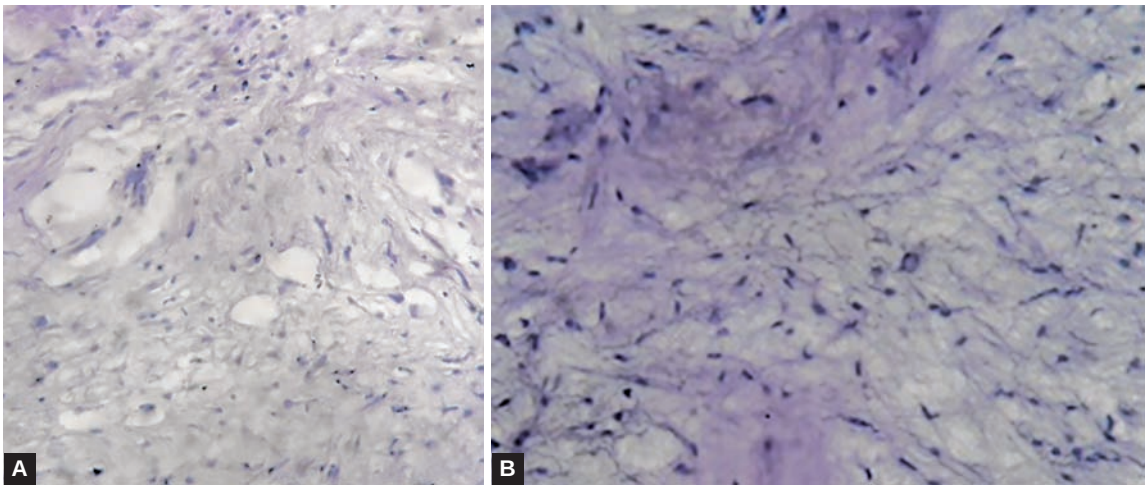


Fig. 5: Intraoperative photograph

In some parts of world like Africa, odontogenic myxoma is supposed to be the second most common odontogenic tumor after ameloblastoma with relative frequency of occurrence between 1 and 19%[3,20] while in other parts of world like Asia, Europe, America relative frequency of occurrence between 0.5 and 17%,[3,21] has been reported. Though no studies were done to calculate the incidence rate of this entity before Ellison NM Simon and Matthias AW et al conducted their study in Tanzania over a period of 21 years to calculate the population based incidence rate in Tanzania.[3] The incidence rate in their study was calculated to be 0.07 cases per million populations. It occurs commonly in 2nd or 3rd decade. Occurrence is rare in children below 10 of age and adults above 50 years of age.[1,2] But in a retrospective as well as prospective study of 33 cases done by Ellison NM Simon and Matthias AW et al in Tanzania over a period of 21 years, the age range was from 3 months to 64 years with a mean of 26.1 years. In their study, majority of cases were noted in patients between 10 years and 39 years.[3] A slight predilection for female with male to female ratio of 1:1.83 has been seen.[3]

Odontogenic myxoma comprises of 3 to 6% of all odontogenic tumors. Posterior mandible is most commonly affected with. It rarely affects maxilla with a ratio of mandible to maxilla involvement being 3:1. Anterior maxilla in region of incisor and canine is even rarer site of involvement.[3] Ours is a case of fibromyxoma involving anterior maxilla in region of incisor and canine. Fibro-



Figs 6A and B: Histopathological micropictograph of the tissue

myxoma of maxilla behaves more aggressively than that of mandible as it spreads through maxillary sinus. It is generally asymptomatic but some patients may present with varying degree of progressive pain over affected region. Displacement and mobility of teeth are relatively common. It may be associated with unerupted teeth. Cortical expansion can occur and large lesions can cause perforation.[3] Patients with posteriorly located tumors presented late with bigger lesions than those with anteriorly located tumors. This is probably due to more visible disfigurement and disturbance of bite function when the lesions are located in the anterior area.[3]

Fibromyxoma is diagnosed by radiological, histological and histochemical investigations. Radiologically, it shows homogeneous unilocular or multilocular radiolucencies or sclerotic trabeculations with 'honeycomb' or 'soapbubble' appearances.[3] In our case lesion appeared as large radiolucent area with trabeculations. The radiolucency may have clearly defined borders or poorly defined diffuse borders. Radiological examination plays a crucial role for the differential diagnosis of myxoma/fibromyxoma and also between benign myxomas and malignant neoplasms with myxomatous tissue. Immunohistochemical examination uses antibodies against specific biological substances of neuronal, muscular, epithelial, and mesenchymal tissues. The evaluation of the presence of vimentin, an intermediate filament of the cytoskeleton characterizes mesenchymal tissues, thus also myxomas. The differential diagnoses include: ameloblastoma, fibrous dysplasia, intraosseous hemangioma, aneurysmal bone cyst, central giant cell granuloma, metastatic tumor and, in cases of unilocular lesions, simple cysts.[3] The main pathological condition that may lead to difficulties in diagnosis is the ameloblastoma, especially when the bony septa are curviform. An important characteristic for differential diagnosis is the fact that when a contrast agent (Gd-DTPA) is being administered, in case of the ameloblastoma the magnetic resonance imaging (MRI) shows strong enhancement of the solid portion of the tumor, in contrast to the myxoma that shows homogenous high signal intensity. It is also important to mention that root displacement and resorption is not unique in ameloblastoma. Biopsy is, therefore, compulsory for an accurate diagnosis. In our case the lesion occupied one side only, left side. This is in accordance with current knowledge that odontogenic myxoma rarely crosses the midline.[3] Histopathologically, fibromyxoma is characterized by presence of stellate and spindle-shaped cells in loose myxoid extracellular matrix with cells presenting with thin, long cytoplasmic prolongations that give to the tissue characteristics of immature mesenchyme. The fibromyxoid lesion may present loci of calcification or ossification and a higher amount of collagen fibers and vessels than a typical myxoma.

Despite the fact that odontogenic fibromyxoma shows aggressive local growth, it is believed that it never undergoes malignant transformation or gives rise to metastasis.[3]

The recommended treatment of choice is resection and peripheral osteotomy. For smaller tumors conservative excision and curettage can be done. Ellison NM Simon and Matthias AW et al suggest that radical resection with a margin of 1.5 to 2 cm of healthy bone is the treatment of choice.[3] Small bony defects of the maxilla, under 5 cm, can be reconstructed by means of a pedicled buccal fat pad (BFP) flap. Larger bony defects require the positioning of an obturator prior to the reconstruction with a graft, most commonly iliac crest bone graft.

Fibromyxoma has a very high recurrence rate up to 25%. The absence of capsule and infiltrative growth of the myxomatous tissue to the adjacent bone determine the high rate of recurrence. Recurrence is the result of local

invasion into cancellous bone beyond radiographically visible margins in the absence of encapsulation. Recurrence is minimized with a more aggressive treatment by performing a partial or complete segmental bone resection, which is particularly indicated in the maxilla. Patient should be advised for follow-up at least for 2 years to find out any recurrence.

CONCLUSION

Odontogenic fibromyxoma is a rare benign tumor of jaw. The radiological examination by means of CT plays an important role in the diagnosis of a fibromyxoma and in the differential diagnosis from other pathological entities, such as the ameloblastoma. Its management is surgical and ranges from enucleation and curettage to complete resection and peripheral osteotomy according to its size. Patients must be monitored for at least 2 years postoperatively in order to diagnose possible recurrence.

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