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Case Report

Maxillary Odontogenic Myxoma: A Rarity

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Abstract

Odontogenic Myxoma is a relatively rare neoplasm which is almost exclusively seen in tooth bearing areas. They constitute around 3-6% of total odontogenic tumours. Odontogenic Myxoma occurs commonly in mandible and their presentation in maxilla is rare. Here we report one such rare case of odontogenic myxoma occurring in the maxilla of an 11-year-old male patient.

Keywords: Odontogenic Tumours; Myxoma; Myxofibroma; Fibromyxoma; Neoplasms; Connective Tissue; Maxilla.

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Introduction

Odontogenic Myxoma (OM) is a rare benign odontogenic mesenchymal tumour arising from embryonic connective tissue associated with tooth forming apparatus.[1,2] It was probably first described by Rudolf Virchow as myxofibroma in 1863.[2] In 1992, WHO defined OM as a locally invasive neoplasm consisting of rounded and angular cells lying in an abundant mucoid stroma.[3]

OM is slow growing but locally invasive neoplasm. It is a sporadic neoplasm with an annual incidence of 0.07 per million.[4] The occurrence of OM in the maxilla is rare when compared to the mandible (1:2) and slight predilection to females can be noted.[2,5] Commonly, OM occur in molar and premolar region in the mandible (65.1% of total mandibular cases), while in the maxilla the occurrence in these areas is around 73.8%.[2] OMs usually are unilateral lesions and they rarely cross the midline.[4] Farman et al, have put the mean age at the time of diagnosis of maxillary OMs in men at around 29.2 years.[6] An accurate histopathological interpretation and proper surgical management is the key to treat this entity. Here we report a rare case of OM occurring in the maxilla of a paediatric patient.

Case Report

An 11-year-old boy reported to a private clinic with a chief complaint of swelling on midface region since 4-5 months. On intraoral examination, painless swelling of the right maxilla extending from the maxillary lateral incisor to the molar area was noted with obliteration of both buccal and palatal cortical plates. The lesion was rubbery in consistency and the mucosa covering the lesion was normal is colour. All teeth from maxillary right lateral incisor to permanent first molar were mobile.

Serial axial sections of Computed tomography showed evident hypo-attenuated mass obliterating the maxillary sinus and also displacing the nasal septum (Fig 1 & 2). A 3-dimension computed tomography reconstructed image of the lesion clearly showed the massive involvement of the right maxilla and extension of the lesion up to the right infraorbital margin of the eye superiorly and lateral wall of the nose medially (Fig 3). However, the patient didn't realize any other sign or symptom even with the lesion involving this extensively.

An incisional biopsy was performed, intra orally, under local anaesthesia and the wound was sutured. Grossly, this lesion was gelatinous in nature, making curettage difficult. On histopathological examination of the lesion, loose and delicate fibrous connective tissue was noted (Fig 4). The myxoid tissue consisted of spindle and stellate shaped fibroblasts with small round nuclei suspended on a delicate network of collagen fibrils, and no signs of malignancy was detected (Fig 5). Small blood vessels were also present. No noticeable odontogenic islands were seen. A diagnosis of Odontogenic Myxoma was made.

Following the histopathological report the patient underwent partial enbloc resection of the maxilla and inferior border of the right eye. As all surgical margins were clear, no other chemotherapy was administered to the patient. Uneventful healing after the surgical removal of the tumour was recorded after 6 months. The excisional biopsy confirmed the

incisional biopsy report of Odontogenic Myxoma.

Discussion

OM generally presents as a painless, slowly enlarging expansion of the jaw with possible tooth loosening or displacement.[3,4,7] The destructive nature of the tumour can cause palatal enlargements, nasal blockage or even ocular changes.[4] Our case had a similar presentation, although our patient neither noticed his expansion of the jaw nor his loose teeth. OM is known to show infiltration and they behave in a locally aggressive fashion.[4,8]

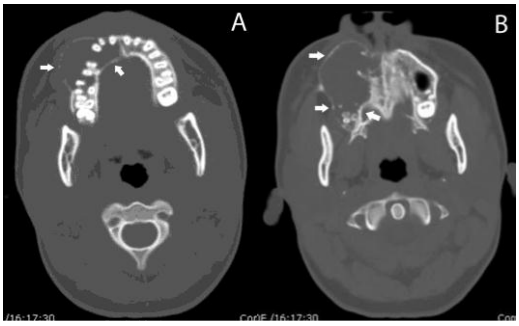

Figure 1: *A.* Axial CT Section showing hypo-attenuated image and, buccal and lingual distention of the cortical plates (arrows). *B.* Tumor mass causing the extra-oral swelling is evident.

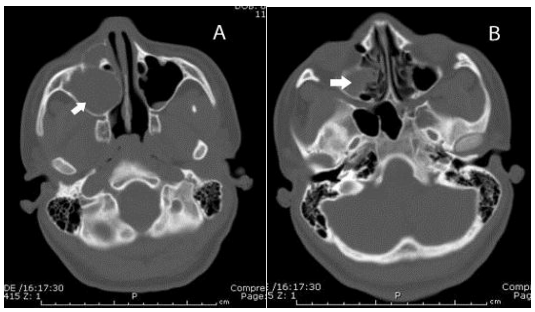

Figure 2: *A.* Complete obliteration of the right maxillary sinus by the tumor mass. *B.* Extensive involvement of the lateral wall of the nose.

OM cases at age of 3 months to 19 months have been reported in the literature.[4,9] A 75% of OM cases are detected between 2nd and 4th decade of life, with mean age of 30 years.[2,6] It is believed that only 7% of cases occur in patients aged less than 10 years.[10] Our case has been diagnosed at the age of 11, however the lesion must have started much early in the patient's life. On an average, the age of diagnosis of OM is around 1-5 years delayed from the onset of the lesion.[9] Exceptionally, OM can grow rapidly and that seem to occur in very young patients.[4] OM most commonly occurs in the mandible than in the maxilla. However, tumours of the maxilla tend to enlarge and often fill the maxillary sinus before presenting as a facial swelling.[9] Similarly, in our case the lesion spread in to the sinus before presenting as an intraoral swelling.

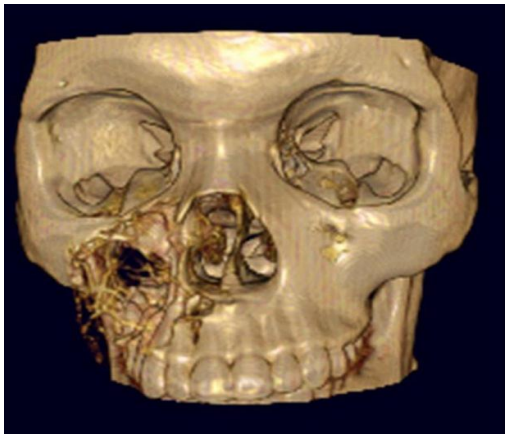

Figure 3: The 3-dimensional reconstruction of lesion showing massive destruction of the right maxilla and extensions to the lateral wall of nose and inferior border of right eye.

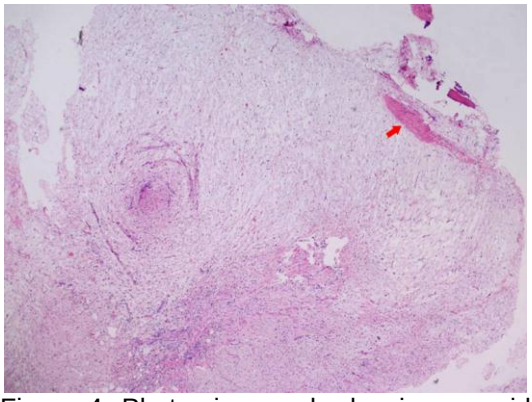

Figure 4: Photomicrograph showing myxoid stroma with bundles of collagen fibers (arrow). (Hematoxylin - Eosin, X 10)

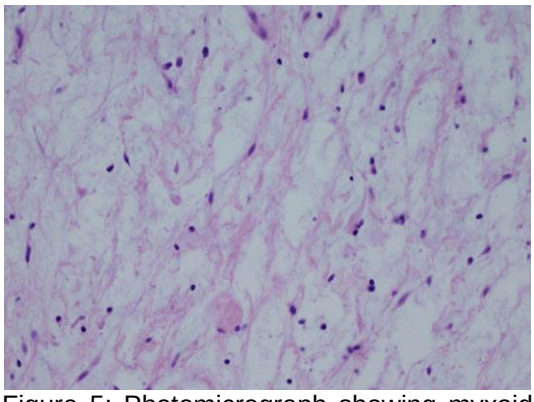

Figure 5: Photomicrograph showing myxoid tissue consisting of spindle and stellate cells with small rounded nuclei. (Hematoxylin - Eosin, X 40)

Radiographically, OM appears as unilocular or multilocular radiolucencies with an ill-defined border and the septa that cause the multilocular feature are usually thin and straight, producing a “tennis racket pattern” or they can be curved and coarse causing a “honeycomb appearance”.[8,11] When the lesion is multilocular, it must be distinguished from ameloblastoma, central giant cell granuloma, central haemangioma, and odontogenic keratocyst.[12] Advanced diagnostic imaging techniques like CT and MRI are being applied to this tumour in detecting the involvement of the adjacent bone and soft tissues.[13]

Farman et al, have reviewed the histological findings of OM.[6] Immunohistochemical studies show that the spindle-shaped cells present in this neoplasm have a combined fibroblastic and smooth muscle typing, suggesting that it is of myofibroblastic origin.[14] The tumour cells are mesenchymal in origin so they are positive for vimentin and muscle-specific actin. The ground substance of OM has been shown to consist mainly of hyaluronic acid (80%) and chondroitin sulphate (20%).[12] Other lesions that can be included in differential diagnosis of OM histopathologically are i) myxoid degenerated benign or malignant nerve sheath tumour; and ii) myxoid chondrosarcoma.[15]

The therapeutic management of OM is very controversial. OM is treated surgically as they are radio-resistant tumours.[1,5,6] Depending upon the size of the lesion a simple enucleation, wide resection or segmental bone resection is planned. Overall prognosis of OM is usually good; they commonly recur when the surgical margins are not clear. Recurrence can also be attributed to the lack of capsule and infiltrative growth in these neoplasms.[4,8,15] It is recommended that proper follow-up has to be done as few cases have been reported to turn in to malignancy.[15]

Conclusion

OM usually presents as an asymptomatic swelling of the jaw and their occurrence in the maxilla of a young patient as our patient, is rare. A correlation of clinical, radiological and histopathological findings is a prerequisite to treat the patient successfully. A proper surgery with clear margins is essential and long term follow-up of these patients is mandatory.

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All research work that involves the study of human subjects should state at the appropriate section, that the research was approved by the institutional review board or ethics committee in accordance with national and international guidelines and regulations, e.g., The Declaration of Helsinki.

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