

Odontogenic myxoma of the maxilla – A case report

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Abstract:

Myxomas are considered to be of mesenchymal origin and occur both in bone and soft tissue. The sites most commonly involving the head and neck region are the jaws with a higher incidence in the mandible which have traditionally been considered to have an odontogenic origin and are slow growing, locally destructive neoplasms. We present here a case of odontogenic myxoma involving the maxilla with emphasis on clinical, radiographic, surgical and histopathological features.

Key words: Odontogenic tumour, Odontogenic myxoma, Maxilla.

Introduction

Myxoma is an uncommon mesenchymal tumor seen in superficial soft tissues, skin and bones. In the head and neck these tumors are rare, usually affecting young to middle aged adults but they most frequently involve the posterior mandible and less commonly the maxilla. These myxomas are slow growing, locally invasive neoplasms derived from mesenchymal elements. Since some myxoid lesions can be malignant, a precise surgical management and accurate pathological interpretation is crucial to direct appropriate therapy. We present here a case of a myxoma of the maxilla and review the clinical presentation, histopathology, and treatment of this unusual lesion.

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Case report

A 27 year old female presented with a slow, progressive mid facial mass on the right side of 2 years duration. Clinical examination revealed an extraoral hard, non-tender fixed mass measuring 5x5 cm located over the maxilla that approached the lateral aspect of the nose and inferiorly one cm below the ala tragus line on the right side (fig.1). Eye position, extra ocular movements and intranasal examination were normal. The texture and colour of the overlying skin was normal. Intraoral examination revealed a swelling obliterating the right posterior vestibule of 13 to 17 of size 5x5cm and also involving a part of the buccal mucosa (fig.2). Borders were well defined with no surface changes; swelling was nontender and hard in consistency. Mildly tender bilateral submandibular lymph nodes of size 1x1cm were palpable. Radiographs (PNS & OPG) revealed haziness involving the entire right maxillary sinus and posterior maxilla with no extension to the orbital floor or nasal cavity and displacement of roots of 15, 16 (fig.3). Occlusal radiograph showed multilocular radiolucency with fine septae and expansion of buccal cortex on the right side (fig.4).

Based on the clinical and radiographic findings a clinical diagnosis of ossifying fibroma was made with a differential diagnosis of odontogenic myxoma and osteoma involving the right maxilla. An incisional biopsy of the mass was performed. The hematoxylin and eosin-stained sections showed abundant pale stained myxomatous areas with more of intercellular substance and predominantly spindle and stellate shaped cells with no evidence of ossification. Also islands of odontogenic epithelium were noticed and a histopathologic diagnosis of odontogenic myxoma was made (fig .6).

Surgical intervention was recommended and the patient underwent surgical excision and curettage (fig.5). Macroscopically, the mass was gelatinous in consistency and covered by eggshell patches of bone and a final histopathologic diagnosis of the excised mass was odontogenic myxoma consistent with the earlier incisional

biopsy report. Post operatively the patient recovered uneventfully with no recurrence over a period of one year follow-up.

Discussion

Myxomas are neoplasms often presenting in bones, soft tissues and more frequently in the myocardium. Despite a wide range of possible tumour locations^{1,2}, studies reviewing the locations of osseous myxomas have shown that the facial bones remain the most common site in the head and neck region^{3,4}. Some authors report a higher incidence of these odontogenic myxomas in the mandible⁵ particularly in the premolar-molar region, while most authors have described an equal distribution of the tumour site in both maxilla as well as mandible¹. Equal distribution in males and females has been reported¹. The typical age of presentation is around third and fourth decades of life with less prevalence in children.

Midfacial myxomas most commonly presents as a slow growing painless mass of the maxilla or mandible. A delay in onset of diagnosis often occurs between onset and clinical presentation because tumours of the maxilla often grows undetected for a considerable duration of time as they expand innocuously into the maxillary sinus. While most myxomas are asymptomatic, despite its slow growth (which is reflected by studies^{6,7} showing less than 1% of the cells positive for the proliferation tumour marker Ki- 67) it is often locally aggressive invading the adjacent structures and cause symptoms referred to these structures. Odontogenic myxomas of the maxilla may invade the adjacent structures such as palate, nasal cavity and orbit and cause symptoms like malocclusion, tooth mobility, pain, nasal obstruction and diplopia. Our case had presented with a slowly enlarging mass of almost two years duration as a midfacial mass on the right side causing mobility of 15, 16 & 17 with no other symptoms.

Radiographically these odontogenic myxomas appear radiolucent and may have a unilocular or multilocular presentation with well or ill defined borders⁸. Also arrangement of fine bony trabeculae within its interior structure giving a



Fig.1&Fig.2:Extra oral photograph showing a mid facial mass on the right side and an intraoral swelling obliterating the buccal vestibule of 13 to 17 region.



Fig.3: OPG revealing haziness on the right premolar-molar region with displacement of roots 15



Fig 4: Occlusal radiograph (right) showing fine wispy septae within the radiolucent lesion along the buccal and palatal aspect of 13 to 18.

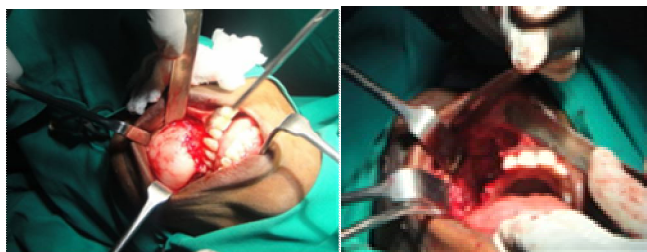


Fig 5: Surgically exposed lesion appearing as a lobulated mass arising centrally from the maxilla

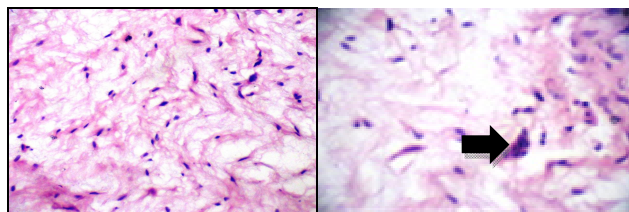


Fig 6: Histopathological picture showing the characteristic stellate shaped cells and thin interconnected cellular projections in a myxomatous background (H&E 20X) and (arrow) an inactive appearing odontogenic epithelial island (H&E 40X)

‘honey-comb’ or ‘soap-bubble’ or ‘tennis racket’ appearance characteristic of ameloblastoma may be seen probing a differential diagnosis based on clinical and radiographic findings as ameloblastoma, odontogenic myxoma, central giant cell granuloma, odontogenic fibroma, fibrous dysplasia, osteosarcoma and chondrosarcoma.

Gross pathologic examination often reveals an unencapsulated mass with the surrounding condensed tissue often mistaken for a capsule. Resected tumour mass feels as a mucoid or gelatinous mass, white to grey coloured.

Microscopically⁹, odontogenic myxoma is a benign neoplasm without encapsulation and exhibits a spectrum of fibrous connective tissue stroma from myxoid to densely hyalinized and from relatively acellular to cellular nature. Majority of the tumour consists of stellate or spindle cells similar to the stellate reticulum of a developing tooth bud in an amorphous, avascular and hypo cellular ground substance composed primarily of glycosaminoglycans, hyaluronic acid and chondroitin sulfate. Calcification may or may not be seen. Cords and islands of sparse and inactive looking odontogenic epithelial islands may be seen.

Controversies exist regarding the odontogenic origin of these jaw myxomas, as some studies^{10,11} have found that the cells in odontogenic myxomas are different from the ectomesenchymal cells of a developing tooth as well the occurrence of myxomas in extragnathic bones are also seen to occur which has prompted some pathologists to consider an osteogenic origin. An odontogenic

origin of these jaw myxomas was suggested because of their location in tooth-bearing areas of the jaws, their close resemblance to dental mesenchyme microscopically, and the more common association of these myxomas with unerupted teeth¹. While myxomas occurring in extragnathic sites are reportedly derived from modified fibroblasts that secrete excess glycosaminoglycans, which in turn inhibit polymerization of collagen¹².

The standard treatment for odontogenic myxomas is wide surgical excision¹³. But some surgeons recommend simple enucleation and curettage for fibrotic lesions¹⁴, and in pediatric cases, because of the concern for facial disfigurement and potential interference with growth centers. However a reported recurrence rate of 25% after enucleation and curettage has led surgeons to employ a wider margin of resection¹⁵. Regardless of the surgical method, it is mandatory that patients with odontogenic myxomas should be followed for extended post-operative duration of time.

Conclusion

Odontogenic myxoma is a rare benign odontogenic tumour that represents 3% of odontogenic tumours. The relative rarity of the occurrence of these tumours in addition to the lack of ultrasructural studies regarding the nature of the tumour, controversies still exist among pathologists regarding the histogenesis of these odontogenic myxomas as odontogenic or osteogenic thereby emphasizing the need for extensive studies on this slow growing but destructive lesion.

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