

Oral medicine case book 55: Odontogenic myxoma

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CASE REPORT

A 20-year old female was referred from her local community health clinic to the Oral Medicine Clinic at Groote Schuur Hospital for a swelling that started out as a small and painless lesion. She had attended the local clinic approximately two months earlier when the growth increased in size, her teeth loosened and the lesion became symptomatic. At that time, her upper left molar teeth were extracted but, despite this, the lesion continued to enlarge.

The patient reported no contributory medical history. Her oral hygiene was reasonable and the remaining dentition was in



Figure 1: This photograph of the patient shows an exophytic, dome-shaped soft tissue enlargement in the edentulous 25-28 region, extending into the buccal vestibule and palate, short of the midline.

a satisfactory condition. On intra-oral examination a 5x3 cm exophytic and dome-shaped soft tissue enlargement was seen in the edentulous 25-28 region. The lesion extended into the buccal vestibule and palatal area, short of the midline. It was in general the same colour as the surrounding mucosa, whilst a superficial pseudomembranous, erythematous and ulcerated surface was present in the area where the mandibular teeth impinged on the lesion, leaving an occlusal impression of the opposing teeth (Figure 1).

Radiographic evaluation revealed a large, destructive, but well demarcated lesion in the left posterior maxilla. The upper left third molar was markedly displaced into a postero-superior position (Figure 2). A computerised tomographic scan (CT) revealed a large, destructive and mixed radiopaque/radiolucent lesion involving the left maxilla, obliterating the entire ipsilateral



Figure 2: The pantomogram of the patient shows a large, poorly defined lesion in left posterior maxilla, displacing tooth 28 superiorly and posteriorly.



Figure 3: A computerised tomographic scan revealed a large, destructive and mixed radiopaque/radiolucent lesion involving the left maxilla.

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maxillary sinus. Multiple septa were present within the bulk of the lesion and destruction of the piriform rim medially was also evident. The lesion extended into the floor of the left orbit superiorly and into the left pterygoid plates posteriorly (Figure 3).

A punch biopsy was taken from the mid-palatal part of the lesion and the specimen sent for histopathological investigation. The patient was placed on anti-inflammatory medication and an antibacterial mouthwash, consisting of a 0.2% aqueous solution of chlorhexidine gluconate.

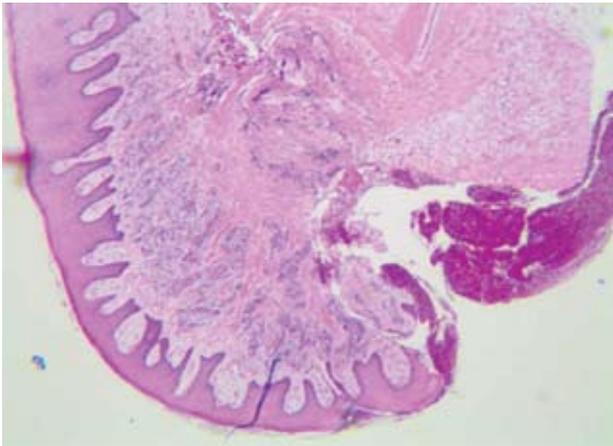


Figure 4: A low power view of the biopsy specimen shows lesional tissue in the top right part of image (X100, H & E).

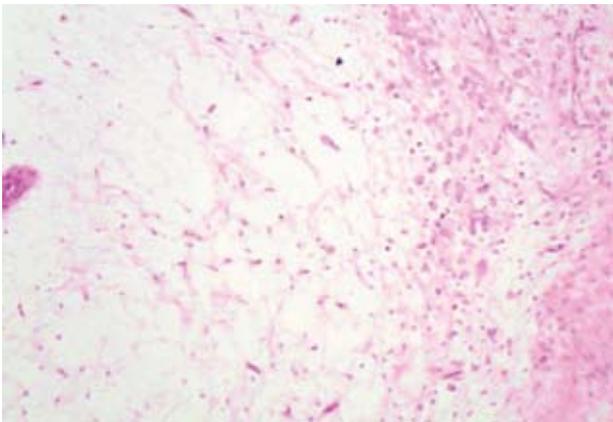


Figure 5: Higher magnification of the lesional tissue demonstrates an ill-defined lesion composed of loosely arranged spindle and stellate-shaped cells within a myxoid stroma. (x400, H&E).

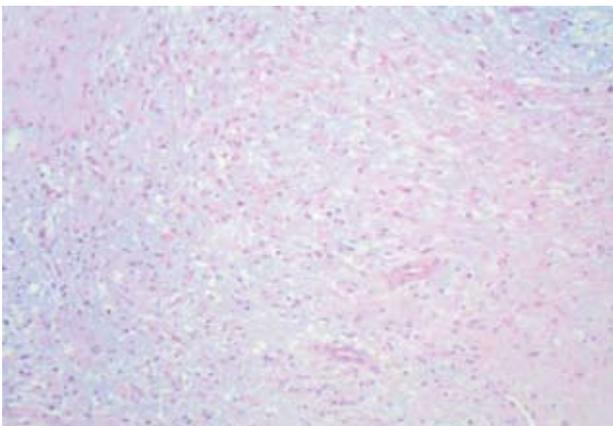


Figure 6: The mucoid-rich extracellular matrix of the lesion contains acid mucopolysaccharides, chiefly hyaluronic acid and chondroitin sulfate, as highlighted here by alcian blue staining. (x400).

The histopathological evaluation revealed a tissue fragment lined by stratified squamous epithelium with deposits of a myxoid material seen in the deeper aspects of the stroma (Figure 4 and 5), highlighted by the use of alcian blue special stain (Figure 6). The surrounding stroma demonstrated fibro-muscular tissue and a scattered lymphoblastic infiltrate. Special stains for AE1/AE3 cytokeratins were negative, thus excluding an epithelial origin of the tumour.

The clinico-histopathological correlation led to a diagnosis of odontogenic myxoma and the patient was consequently referred to the Maxillofacial Unit for treatment that would include total removal of the tumour and subsequent tissue reconstruction.

DISCUSSION

The odontogenic myxoma (OM) is a benign and non-encapsulated tumour, found almost exclusively in the jaws, especially the mandible and is most commonly found in females in the third and fourth decades of life.¹ Although it is relatively rare, in the African setting it is the second most common odontogenic jaw tumour, after the ameloblastoma.^{1,2,3} This neoplasm can resemble an ameloblastoma, peripheral giant cell granuloma or even a metastatic tumour. Radiographically, the OM may assume a unilocular or multilocular presentation with cortical expansion and tooth displacement and the image is classically described as having a soap bubble appearance.⁴ It occurs most often in the posterior part of the mandible, but may present in the posterior maxilla where it shows more aggressive behaviour. In rare cases it has been reported to appear on the gingiva as a peripheral OM, in which case it can present a diagnostic challenge as, in its early phase, it can be confused with a reactive epulis.^{5,6}

Histologically, the OM is characterised by the presence of sparse collagen found amidst an extensive accumulation of a mucoid ground substance, in which odontogenic epithelium may be an incidental rather than a usual finding.³ The tumour typically begins as a painless, slow growing swelling, but later can exhibit destructive local growth, resulting in bony perforation and soft tissue invasion. Despite this, it does not undergo malignant transformation nor does it metastasise. The destructive growth may be attributed to the fact that the tumour is not encapsulated and in addition, to the fairly rapid increase in mucoid material in the tumour. Mitoses are, however, infrequently seen.^{1,3}

Our case is consistent with other published reports which show a female gender predilection for OM. However, the location of the lesion was not in keeping with the predominantly mandibular presentation reported in the literature.¹ It does, however, concur with current evidence that maxillary lesions are unilateral, rarely cross the midline and are particularly destructive, even though remaining benign.² The tumour is locally invasive and may spread rapidly through the maxillary antrum, eventually leading to involvement of the orbit. It can thus reach extensive proportions by the time the patient seeks medical intervention.⁴ In fact, late treatment of the lesion is a common finding in cases of OM, with a mean period of 3.4 years between the tumour being noted and the patient seeking treatment.³ It is thought that the reasons for this delay is due to limited access to healthcare and the supposition that the lesion poses no immediate health threat.^{2,3,4} In the present case, the localised nature of the soft tissue swelling, in a patient with such

a good dentition, presented a clear signal for early referral and intervention.

The exact origin of the odontogenic myxoma has fueled much debate as to its cellular lineage and classification as a true odontogenic neoplasm. The evidence supporting its odontogenic lineage includes its histological semblance to the stellate reticulum of the developing tooth; its selective presentation in the tooth-bearing areas of the jaws; infrequent appearance in other parts of the skeleton and the occasional finding of odontogenic epithelium.³

Odontogenic myxomas necessitate surgically management and the size of the lesion at the time of presentation dictates whether a radical or conservative surgical approach is indicated. Therefore clinicians must practice caution when presented with exophytic lesions presenting in the tooth bearing tissues, particularly when concomitant alveolar bone expansion is present. In addition, mobile teeth in the absence of infection should always raise suspicion of an underlying bony pathology. Clearly a comprehensive radiological examination prior to tooth removal is mandatory in such cases and a final diagnosis can only be reached after biopsy and histopathological evaluation. This should help to effect early referral and ensure prompt intervention in order to optimise patient management and reduce morbidity.

CONCLUSION

Attending clinicians in private practice or primary healthcare dental clinics need to be aware of the clinical manifestations of OM and other lesions presenting in the tooth bearing tissues. They should also carefully note the progression after any treatment, i.e. if a lesion fails to heal or shows no signs of regression, it is imperative that the patient be referred for specialist attention in order to prevent and decrease patient morbidity.

Declaration: No conflict of interest declared

References and *recommended reading

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