

Odontogenic myxoma with numerous keloidal-like collagen fibres and calcifications

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Abstract

Introduction

Odontogenic myxoma (OM) is a benign tumour which usually occurs in the jaw. Histologically, this tumour is composed of spindle - to stellate-shaped cells embedded in an abundant myucoid/myxoid stroma containing only a few collagen fibrils scattered throughout the tumour. We herein report a rare case of OM which showed, as unusual feature, the presence of numerous keloidal-like collagen fibres, some of which containing calcifications.

Case report

A 50-year-old male came to our institutions for the evaluation of posterior maxillary swelling. The lesion had been presented for six months and had gradually increased in size. After radiographic examination, the presumptive diagnosis of odontogenic cyst or non-odontogenic tumour was proposed.

Conclusion

Awareness of these features is crucial to avoid misdiagnosis with other fibro-myxoid tumours. Differential diagnosis is discussed. The authors suggest that cases of OM with a significant component of collagenous fibres should be better named "odontogenic myxofibromas".

Introduction

Odontogenic myxoma (OM) is a rare benign tumour, first described by Thoma and Goldman in 1947¹, which represents about 3% of all odontogenic tumours.

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It usually occurs in young patients in their second and third decade of life, with a slight female predilection,^{2,3,4,5} and involving the mandible more commonly than the maxilla.⁶ Clinically, it is a slowly growing, expansile, painless tumour, which may cause root resorption, tooth mobility, cortical expansion and perforation, but small lesions may be asymptomatic and diagnosed only during a radiographic examination. Tumour appears as unilocular or multilocular radiolucency with well-developed locules, consisting of fine trabeculae, arranged at right angles, known as the 'tennis-racquet' or 'step-ladder' pattern. A 'sun-ray' or 'sun-burst' appearance has also been reported in the literature.^{7,8} Tumour borders may be well or poorly defined. OM can be locally aggressive, although it is typically a slowly growing tumour with less than 1% of tumour cells positive for the proliferation marker Ki-67.⁹ Although small OMs are generally treated by curettage, larger lesions require extensive resection.¹⁰ As OMs tend to recur locally, it is mandatory that affected patients be carefully followed after surgical treatment. It is widely accepted that local infiltration reflects the aggressive nature of the tumour, suggesting a potential high recurrence usually associated with scattered deposits of residual bone and dystrophic mineralization in the tumour stroma.

Only rarely OM shows unusual morphological features which may represent potential diagnostic pitfalls for pathologists. In this regard, rare cases may contain diffusely dispersed globular or trabecular osteo-cementum-like calcified products which can mimic fibro-osseous lesions, as typically seen in the jaws.^{11,12} These lesions consist of small spherules and/or larger masses

of calcified products with peripheral cellular osteoid matrix or radiating collagen fibers.^{11,12} We herein report a rare case of OM exhibiting, as unusual morphological finding, the presence of numerous keloidal-like collagen fibres, some of which containing calcifications.

Case Report

A 50-year-old male came to our institutions for the evaluation of posterior maxillary swelling. The lesion had been presented for six months and had gradually increased in size. After radiographic examination, the presumptive diagnosis of odontogenic cyst or non-odontogenic tumour was proposed. At the first clinical visit, the patient did not refer systemic diseases, and extra-oral examination revealed no signs of swelling or inflammation. Regional lymph nodes were not palpable. Intra-oral examination highlighted a soft tissue mass extending from the distal first premolar to second molar zone. Panoramic radiography showed bone alteration and a large transparent area delimited just before the left maxillary sinus, associated with an unerupted tooth (Figure 1). Computed tomography (CT) confirmed the findings of the first radiographic examination (Figure 2). The modest extension of the lesion and the absence of other radiological and clinical findings suggested the complete removal of the lesion by an intra-oral approach. The anatomical limits of the tumour were marked with a dermic pen in order to be sure on extending the incision 3 mm over healthy tissue. A mucoperiosteal flap was elevated after local anaesthesia injection, and osteotomy of the thin buccal wall was performed (Figure 3). The lesion was totally enucleated and the empty area was cleaned by saline solution. Suture 3/0 no resorbable was used for closing the flap.

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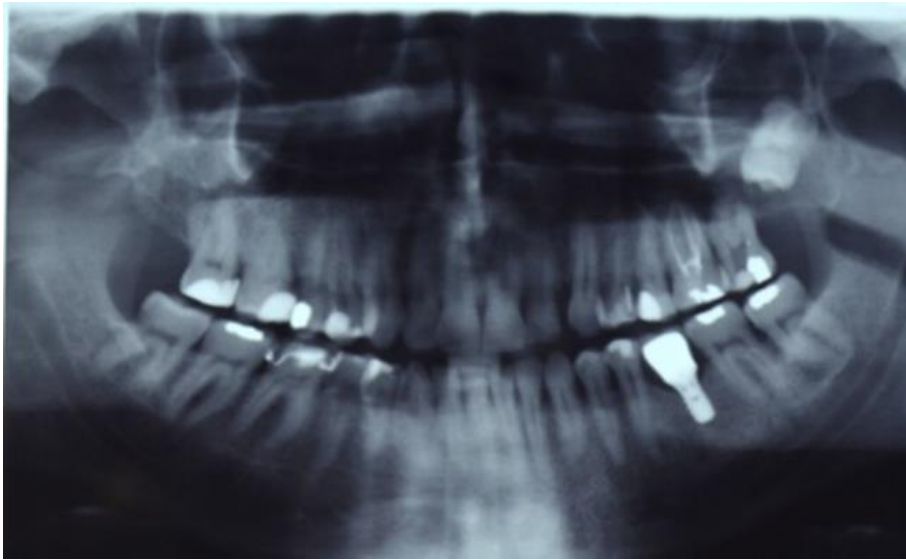


Figure 1: Panoramic radiography showed ill-defined radiolucent lesion associated with an unerupted tooth; bone alteration and a large transparent area delimited just before the left maxillary sinus.

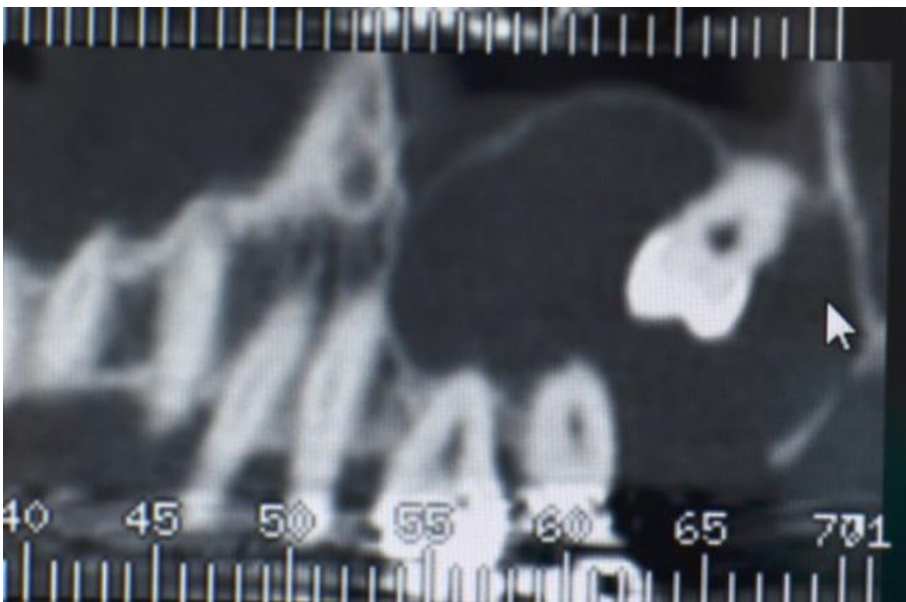


Figure 2: Computed tomography (CT) confirms the radiographic features.

Surgical specimen was fixed in 10% buffered formalin, routinely processed, and embedded in paraffin. Sections (4-5 μ m in thickness) were stained with haematoxylin and eosin. Additional sections were cut for histochemical and immunohistochemical procedures. Immunohistochemical studies were performed with the labelled streptavidin-biotin peroxidase detection system using the Ventana automated immunostainer (Ventana Medical Sistem, Tucson, AZ). The

following antibodies were used: vimentin, CD34, -smooth muscle actin, desmin, pancytokeratins, EMA and S100 protein (all from DAKO, Glostrup, Denmark). Appropriate positive and negative controls were included. (data not shown). Gross examination revealed a well-circumscribed, unencapsulated, gelatinous tumour, measuring 23mm on its largest diameter. The tumour was soft in consistency and the cut surface revealed a gelatinous mass (Figure 4).

Histological examination revealed a tumour with pushing borders, predominantly composed of haphazardly arranged, bland-looking spindle- to stellate-shaped cells, embedded in an abundant myxoid stroma which was stained with Alcian blue at pH7. Only a few collagen fibrils were scattered throughout the myxoid stroma. Mitoses and necrosis were absent. Neither odontogenic epithelial rests, nor foci of residual bone trabeculae were seen. The most striking feature of the tumour was the presence of numerous keloidal-like collagen fibres scattered throughout tumour (Figure 5 and Figure 6), rarely containing calcifications (Figure 7). Immunohistochemical analyses, showing a diffuse and a focal staining, respectively for vimentin and α -smooth muscle actin, revealed the fibroblastic/myofibroblastic nature of the neoplastic cells (data not shown). No immunostaining was observed with any of the other antibodies tested (data not shown). The patient is under regular clinical and radiological follow-up and he is waiting for prosthetic rehabilitation.

Discussion

Although the present case shows the typical clinical and radiographic features of OM, morphology diverged from that reported in the literature. Unlike classic-type OM, the present case exhibited, as unusual and striking morphological feature, the presence of numerous thick eosinophilic collagen fibres scattered throughout the myxoid stroma. Notably, some of these fibres contained centrally-located calcifications, resulting in structures closely reminiscent of “osteo-cementum-like spheroid bodies”, as occasionally reported in OM.^{11,12} Although the myxoid stroma of OM may contain a noticeable fibrous component and/or a few thick collagenous bands,¹ to the best of our knowledge, the presence of numerous keloidal-like fibres, some of with calcifications, is a novel finding which has not been previously emphasized. Although the basic morphologic appearance of our case was typical of OM, it was the

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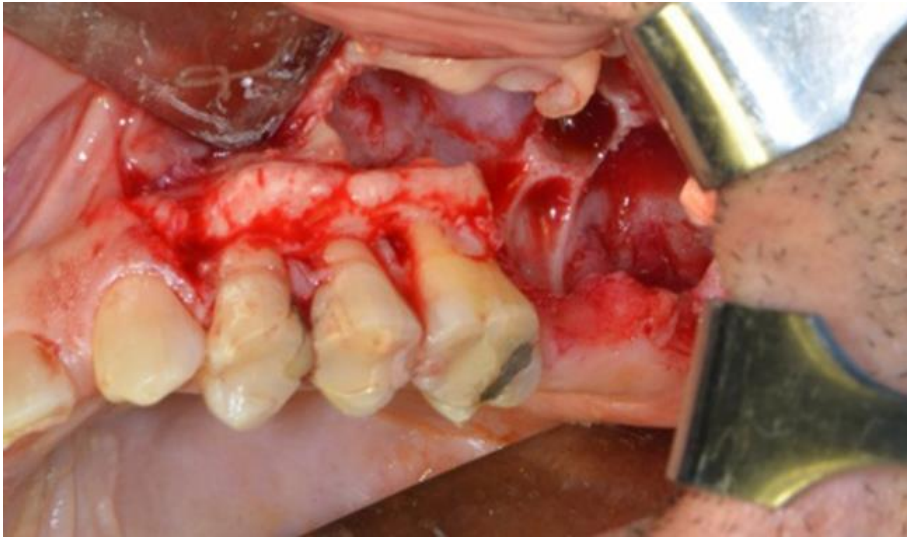


Figure 3: Osteotomy of the thin buccal wall performed.



Figure 4: Gross features of the tumour: a well-circumscribed, nonencapsulated, gelatinous tumour.

presence of abundant keloidal-like collagen fibres, some of which containing calcifications, that caused some diagnostic problems. Differential diagnosis of OM mainly included benign tumours which can exhibit abundant myxoid stromal changes, such as odontogenic fibroma, chondromyxoid fibroma, myxoid neurofibroma, myxoid solitary fibrous tumour and myxoid mammary-type myofibroblastoma. Similarly to OM, odontogenic fibroma is a benign odontogenic tumour composed of stellate or spindled fibroblasts with an

associated stroma ranging from collagenous to fibromyxoid in nature. However, the present case, lacking odontogenic epithelium, does not fit the current WHO criteria for the diagnosis of odontogenic fibroma.¹³ Chondromyxoid fibroma is usually composed of lobules of stellate- to spindle-shaped cells embedded in a variable myxoid to chondroid matrix, rimmed by hypercellular areas containing spindle-shaped cells set in fibrous stroma. Unlike chondromyxoid fibroma, our case lacked both lobular architecture and

chondroid matrix. Neurofibroma can be composed of a proliferation of spindle cells embedded in an abundant myxoid stroma. The spindle cells are slender and often exhibit wavy nuclei, in contrast to the plump spindle cells of OM. Unlike OM, neoplastic cells of myxoid neurofibroma are typically positive for S-100 protein. Other soft tissue tumours which may concurrently contain myxoid matrix with scattered keloidal-like collagen fibres are solitary fibrous tumour and mammary-type myofibroblastoma. The former is a tumour which can rarely occur in oral cavity,¹⁴ but histological features are different from those of OM. Infact solitary fibrous tumour shows alternating hypercellular and hypocellular areas and variably fibromyxoid stroma in which numerous blood vessels with a hemangiopericytoma-like pattern are seen.¹⁴ Unlike our case, neoplastic cells of solitary fibrous tumour are diffusely CD34-positive. Mammary-type myofibroblastoma is a spindle cell tumour with a variable fibro-myxoid stroma in which numerous keloidal-like collagenous fibres are embedded.^{15,16,17} This tumour, unlike OM, exhibits significant expression of desmin, α -smooth muscle actin and CD34^{15,16}.

Conclusion

The present paper contributes to widen the morphological spectrum of OM. Awareness of the possibility that this tumour may contain numerous keloidal-like collagen fibres with associated microcalcifications is crucial to avoid misdiagnosis. Although tumour stroma of OM is usually myxoid in nature, the rare cases which exhibit a fibro-myxoid matrix could be better labelled as "odontogenic myxofibroma".

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the editor-in-chief of this journal.

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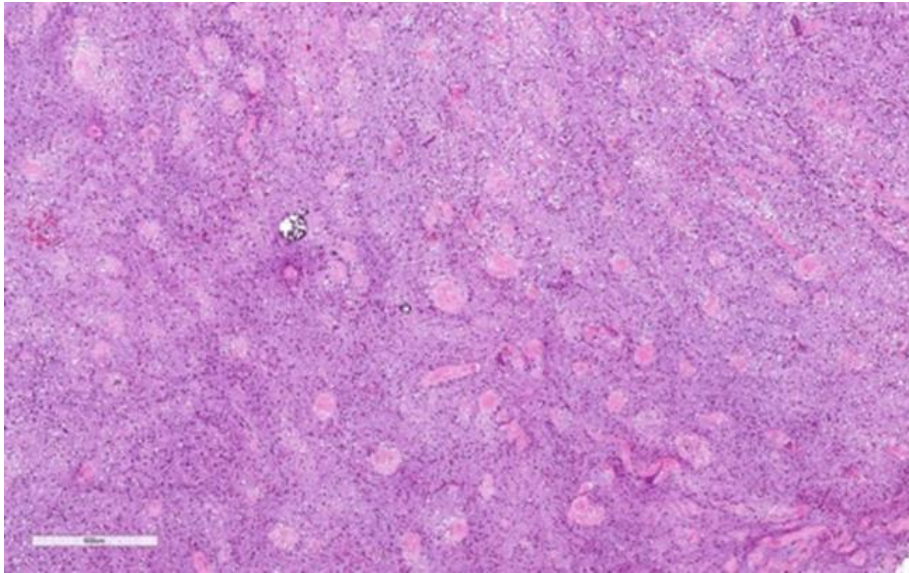


Figure 5: Low power magnification showing a myxoid tumour containing numerous keloidal-like collagen fibres (haematoxylin and eosin; x60).

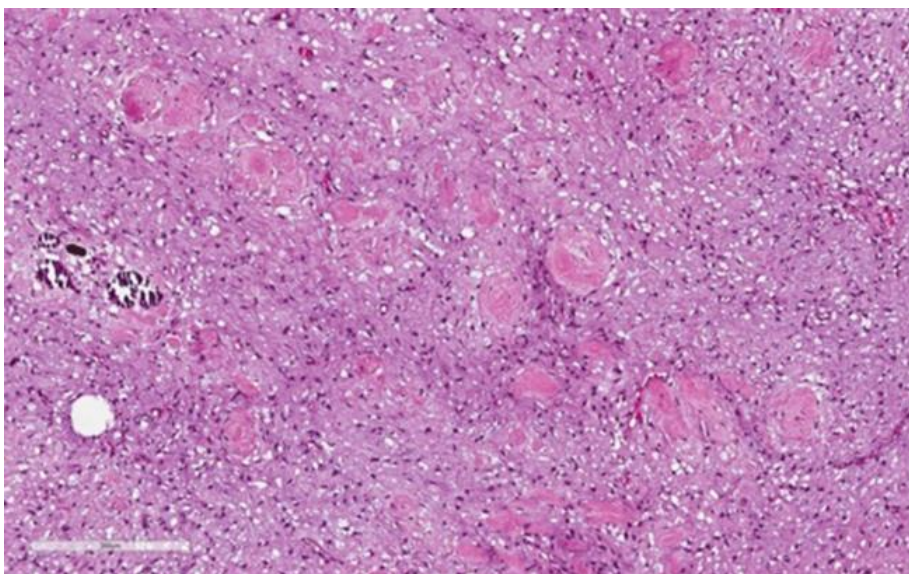


Figure 6: Higher magnification showing a myxoid tumour composed of stellate and spindled bland-looking cells. Thick collagen fibres are seen (haematoxylin and eosin; x100).

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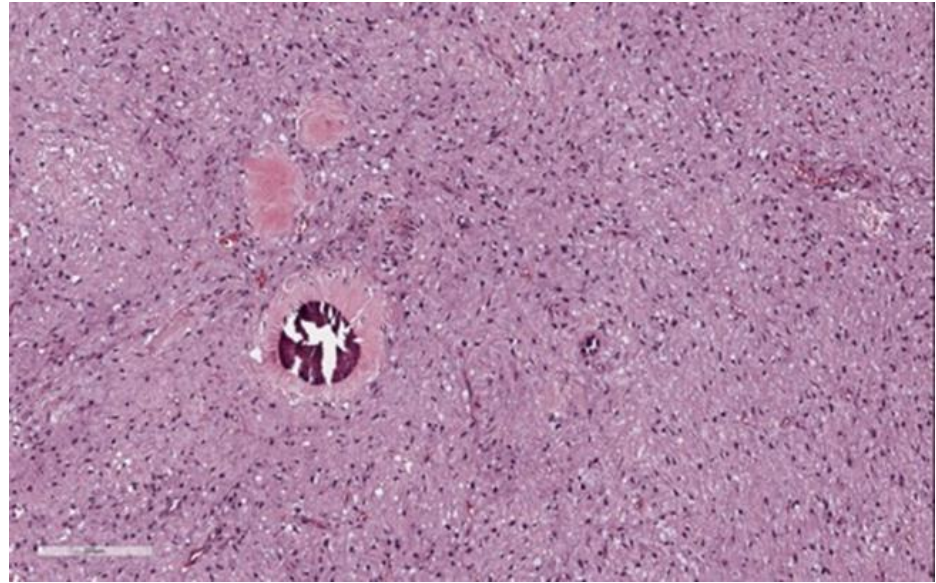


Figure 7: Centrally-located micro calcifications are seen within some collagen fibres (haematoxylin and eosin; x100).

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