Osteo-cementum Producing Odontogenic Myxomas. A Literature Review of a Distinctive Variant

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Received: 9 September 2013 / Accepted: 14 June 2014 / Published online: 10 July 2014
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Abstract

Introduction Odontogenic myxoma (OM) is a benign neoplasm of mesenchymal origin with growth characteristics, clinical behaviour and radiographic presentation similar to those of ameloblastoma. It is an intraosseous neoplasm characterized by stellate and spindle-shaped cells embedded in loose myxoid or mucoid extracellular matrix. Although sometimes bony islands that represent residual trabeculae are found throughout the lesion, the formation of osteocement-like calcified spherules within the tumour is an extremely rare phenomenon.

Review We report a very rare case of an OM of the left maxilla exhibiting osteo-cementous metaplasia within the substance of the tumour and beyond the facial skeleton, in the nasopharynx. A review of all four similar cases previously reported in the literature is also presented.

Conclusion Whether or not this property to produce significant amounts of bone can be associated with a

different biological behavior for this specific variant of OM remains to be proved with the study of more similar cases.

Keywords Odontogenic myxoma · Osteo-cementum metaplasia · Ossifying tumours

Introduction

Odontogenic myxoma (OM) is a benign neoplasm of mesenchymal origin with growth characteristics, clinical behaviour and radiographic presentation similar to those of ameloblastoma [1]. The incidence of OM ranges between 3 and 20 % of all odontogenic tumours and it appears to be the third most common one in most studies [2]. Although intraosseous myxoma has been reported in various anatomical sites [3, 4], the majority of these tumours occur in the mandible, followed by the maxilla [1, 2]. OM predominantly affects young adults with no obvious sex predilection [5, 6]. Its age distribution ranges between 5 and 65 years, showing a peak between 15 and 30 years [1]. Small lesions are usually asymptomatic [2], while large OMs most commonly manifest as painless, localized swellings which expand and may perforate the cortical plates. Sensory nerve symptoms are more infrequent [1, 2, 5–8]. OM is an intraosseous neoplasm characterized by stellate and spindle-shaped cells embedded in loose myxoid or mucoid extracellular matrix [6]. Inactive odontogenic epithelium may also be present but it is usually minimal and not required for the diagnosis [9].

Although sometimes bony islands that represent residual trabeculae are found throughout the lesion, the formation of osteocement-like calcified spherules within the tumour is an extremely rare phenomenon [10]. We present a very rare case of an OM of the left maxilla exhibiting osteo-

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Fig. 1 Preoperative photo showing left nasal obstruction

Fig. 2 Intraoral photo showing expansion of maxillary cortical wall

cementous metaplasia within the substance of the tumour and review all four similar cases previously reported in the literature.

Case Report

A 32-year-old male from Philippines was referred to the outpatient department, with main complaints of left nasal obstruction and epiphora, symptoms that had been present for a year (Fig. 1). The patient initially visited an Otolaryngologist, who performed a transnasal biopsy, which showed a stroma tumour and the differential diagnosis included fibroma, myxoma and choanal polyp. The ENT specialist subsequently referred the patient to our department for further investigation and treatment.

Fig. 3 Axial CT scan demonstrating the extent of the lesion (hard tissue window)

Fig. 4 Coronal CT scan showing a molar tooth repulsed to the left sphenoid sinus

Clinical examination revealed a polyp-like mass in the left middle nasal meatus, a firm, painless cortical bone expansion of the left maxilla in the premolar–molar region and a slightly distinct painless swelling of the left nasal and zygomaticomaxillary regions (Fig. 2). The overlying mucosa and the adjacent teeth were intact. No cervical lymphadenopathy was present. The patient's medical history was unremarkable.

Fig. 5 MRI revealed a possible hyperplastic reaction of the left frontal sinus mucosa

The CT-scan revealed a lesion of the left maxilla, which obliterated the left maxillary sinus permeating into the left orbital floor, expanded the left maxillary cortical bone and extended to the ipsilateral nostril and nasopharynx. Diffuse ossifications were present and an impacted molar was repulsed to the anterior wall of the left sphenoid sinus. The lesion showed no enhancement after intravenous contrast agent (Figs. 3, 4).

The differential diagnosis included, fibrous dysplasia, ossified mycetoma, odontogenic tumours (OM, calcifying epithelial odontogenic tumour, ameloblastoma) and fibro-osseous tumours of the maxillary sinus. A T2-weighted magnetic resonance imaging (MRI) of the affected area was performed, in order to define the anatomic extent of the lesion and to plan the surgical intervention. The MRI confirmed the CT findings, and revealed a possible hyperplastic reaction of the left frontal sinus mucosa (Fig. 5). A second biopsy was obtained via a Caldwell-Luc approach and histopathologic examination showed spindle-shaped and round cells dispersed in mucoid or myxoid stroma admixed with collagen fibers. These findings were consistent with an OM.

The patient underwent surgery via a Weber–Fergusson approach with subciliary extension. The flap, at the area of the intraoral biopsy, was raised in a supraperiosteal plane, in order to avoid tumour spillage. The left nasal bone was osteotomised as part of the facial disassembly procedure. Left hemimaxillectomy was performed including part of the orbital floor and an en-block excision of the tumour was achieved (Fig. 6). The intraoral defect was reconstructed with a temporalis muscle flap and the floor of the orbit was

Fig. 6 Intraoperative photo showing the defect after the removal of the tumour

Fig. 7 Photo showing the specimen with greatest diameter reaching 12 cm

restored with a titanium mesh. The left nasal bone was repositioned in its original position. The specimen consisted of three parts connected through islets; the maxillary part of the tumour measured $6 \times 4.5 \times 0.8$ cm, the nasopharyngeal part measured $6.5 \times 4.2 \times 2$ cm and the nasal part measured $5.5 \times 2.4 \times 0.6$ cm. The greatest diameter of the tumour was almost 12.5 cm (Fig. 7). The patient's postoperative course was uneventful.

During cutting and preparation of the specimen in the pathology department a hard tissue element was retrieved from the nasopharyngeal portion of the tumour. The final histopathologic report confirmed the findings of the intra-oral biopsy i.e. OM. There was a proliferation of stellate and spindled cells in a myxoid stroma with collagenized areas (Fig. 8). In addition, the tumour cells showed pleomorphism and some binucleated cells were also noted.

Fig. 8 Proliferation of stellate and spindled cells in a myxoid stroma with collagenized areas, the tumour cells showed pleomorphism and some binucleated cells were also noted

Fig. 9 Calcium deposits leading to small foci of osteoblastic activity within the tumour

Remnants of odontogenic epithelium were found. Moreover, areas of calcified products were identified throughout the myxoid stroma. These calcifications were in the shape of spherules which resembled osteo-cementum formations (Fig. 9). The same histologic features were also prominent in the hard tissue element of the nasopharyngeal portion of the tumour.

There are no signs of recurrence after 2 years of follow up and the aesthetic outcome is excellent (Fig. 10).

Discussion

Myxomas are considered to be benign tumours that occur both in bone and soft tissues [11]. OM is a locally

Fig. 10 Postoperative photo of the patient 2 years after surgery

aggressive tumour of the jaws. The histogenesis of OM is thought to derive from the pluripotential mesenchymal stem cells of the odontogenic mesenchyme or the somatic mesenchyme of the jaws [1]. Islands of inactive odontogenic epithelium may be present throughout the tumour and some authors believe that OM is of odontogenic origin [12]. This is further supported by the almost exclusive occurrence of the tumour in the tooth-bearing regions of the jaws and its frequent association with an unerupted tooth. However, in other studies biochemical analysis of the tumour cells suggested the osteogenic origin of OM [13, 14]. At present, there is no universally accepted theory about the histogenesis of the tumour.

The most common radiographic presentation of the OM is a multilocular or a unilocular radiolucency [1]. In most cases the radiographic description of the tumour follows a “honeycomb” or a “soap bubble” pattern [15]. The radiolucent area frequently contains fine residual trabeculae, often arranged at acute angles to one another [7]. The radiographic borders of the tumour may be well or poorly defined and displacement of teeth is not uncommon [16]. Some authors have published cases of OMs with mixed radiopaque and radiolucent components while others reported myxomas with irregular radiolucencies traversed by fine crossing trabeculae [10, 17]. In our case a mixed radiopaque and radiolucent appearance was seen. This radiographic pattern was evident throughout the whole lesion, even in the area of the nasopharynx, where ossifications can be found only in fibro-osseous appearing lesions. Therefore, we suspected initially a bone forming lesion and not a myxoma. In addition, a molar-shaped tooth was seen in close proximity with the left sphenoid sinus, suggesting an odontogenic tumour. At that time, the differential diagnosis included calcifying epithelial odontogenic tumours and desmoplastic ameloblastoma. The data from the literature could not support the diagnosis of an

Table 1 Cases of odontogenic myxomas with osteo-cementum calcifications reported in the English literature

Authors	Type of calcification	Year
Rennie et al. [17]	Cementum-like calcified material	1985
Oygur et al. [10]	Osteocement-like spherular calcified bodies	2001
Lin and Basile [9]	Osteo-cementum calcified products	2010
Lahey et al. [20]	Osteocement-like spherules	2013
Rallis et al. (current)	Osteo-cementum calcified spherules	2013

OM, since the radiopaque areas reported by other authors were seen within the limits of the bone affected by the tumour [10, 17]. To the best of our knowledge, this is the first case of OM with osteo-cementum products found outside the facial skeleton, reported in the literature.

Odontogenic myxomas are non-encapsulated, infiltrating, gelatinous tumours that are sparsely cellular [1]. These tumours consist of randomly oriented stellate, spindle-shaped and round cells with long, fine, slightly eosinophilic cytoplasmic processes extending from the centrally placed nucleus. The cells are evenly dispersed in an abundant mucoid or myxoid stroma that contains only a few fine collagen fibres [2]. Tumours that are more collagenous are called myxofibromas or fibromyxomas [1]. Remnants of odontogenic epithelium may be present, but are not required for establishing final diagnosis [5]. Bony islands representing residual trabeculae can be found scattered throughout the lesion [18].

In the present case, all the typical histopathologic characteristics of OM were described. However, apart from residual trabecular and lamellar bone located focally and at the periphery of the tumour, newly-formed calcified material was detected, not only throughout the bony part, but also in the soft tissue constituent of the tumour. Since no reliable histological criteria to distinguish bone from cementum exist [19], the nature of the calcified material that arose in our specimen requires further interpretation.

The calcified products that were identified in the bony and soft tissue component of the tumour did not exhibit properties of mature lamellar bone. Therefore, it is our impression that these calcifications are most likely of osteo-cementum nature, having arisen as an outcome of metaplasia. This hypothesis is further supported by the fact that no osteoblasts or cementoblasts were found in proximity with these formations.

The World Health Organization has classified OM as a mesenchymal odontogenic tumour that does not produce calcifications [2]. Obviously, cases of OM with the above mentioned histologic variation are extremely rare. In our case, calcified material was evident in the nasopharyngeal compartment of the tumour, beyond the facial skeleton.

Thus, there is strong evidence that this material should not be regarded as residual bone, but as newly-formed calcified tissue. One could suggest a new, bone forming, OM variant which includes the few cases with similar properties already reported [9, 10, 17, 20] (Table 1).

It is widely accepted that the local aggressiveness of OM, with infiltration of the adjacent tissues, is the most significant causative factor of its high recurrence rate [9]. Our patient presented with expansion of the maxillary bone, epiphora and extension into the left nostril. These signs left no doubt that the maxillary sinus and the nasolacrimal duct were involved and led to the decision of an aggressive surgical excision which could secure the patient from a recurrence. More conservative types of treatment have been described but are associated with higher incidence of recurrence [21] and should be preserved to treat less extensive tumours. The ipsilateral frontal sinus appeared with high signal intensity in the MRI. This could be the outcome of edematous thickness of the sinus wall, mucous retention or tumour extension. However, the radiologist advocated a hyperplastic reaction of the sinus mucosa therefore our excision was not extended to the frontal region.

There is a resemblance of the current case to fibro-osseous tumours of the jaws, which are characterized by newly formed calcified products. However, to support such a hypothesis we would have to accept that the initial fibrous stroma of the tumour underwent a myxomatous degeneration. We consider this is a rare possibility since it is very unusual that such tumours have the potential to infiltrate bone and transform all the initial fibrous stroma to a myxomatous one. Furthermore, the aggressive clinical behavior of the lesion can be justified by the presence of an infiltrative myxomatous stroma rather than a fibro-osseous growth.

Most of the reports of OM in the literature are consistent with tumours of uniform histology and the characteristic local infiltrative behavior. These lesions have demonstrated an obvious and well documented tendency to recur, therefore surgical treatment through bone resection should be considered the treatment of choice, especially for more extensive tumours. The formation of osteocement-like calcified spherules within the tumour is an extremely rare phenomenon. Whether or not this property to produce significant amounts of bone can be associated with a different biological behavior for this specific variant of OM remains to be proved with the study of more similar cases.

Conflict of interest None declared.

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