CASE REPORT

An unusual odontogenic myxoma in mandible and submandibular region: a rare case report

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An otherwise healthy 14-year-old male was referred to our hospital for the evaluation of a mass that was noticed 2 months previously. The mass was located in the left submandibular area. Comprehensive imaging examinations including panoramic radiography, CT and positron emission tomography-CT were performed. Appropriate surgical management and histopathological examination were taken for the patient. Histopathological examination demonstrated an odontogenic myxoma.

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Introduction

Odontogenic myxoma (OM) is a benign but locally aggressive tumour. It manifests with slow and painless swelling and tends to recur. OM is considered as the second most common odontogenic tumour in some countries,¹ while in China it is relatively uncommon.^{2,3} OM is rare in children below the age of 10 years and in adults above the age of 50 years. It shows a little predilection for the mandible. Duration of the lesion always shows a relatively long period.^{1,4,5}

Image examinations of OM are of great value to establish the differential diagnoses and accurately map the lesion.^{4,6} Generally speaking, CT and MRI are superior to plain radiographs when establishing the confines of the tumour, cortical perforation and involved adjacent structures.⁷ Tooth displacement and root resorption, however, can be observed more reliably on conventional radiographs, often on panoramic radiographs from a whole aspect.^{5,6} Ultrasonography is a useful tool to assess salivary glands, but is not always used in OM.⁸ More recently, a whole-body positron emission tomography-CT (PET-CT) scanner was developed to provide both functional and anatomical information in a single diagnostic

test.⁹ Diagnosis of OM is most commonly established by biopsy.^{10,11}

In this article, an unusual OM is presented. The myxoma located in the mandible and submandibular region was characterized by large size, short duration and bone and soft-tissue involvement. To the best of our knowledge, this is the first case of OM with a large lesion invading soft tissues that is exclusively being reported in the literature.

Case report

An otherwise healthy 14-year-old male was referred to the Department of Oral and Maxillofacial Surgery at the Peking University School and Hospital of Stomatology, Beijing, China, in June 2011 for the evaluation of a mass in the left submandibular area. With a history of progressive swelling for 2 months, the lump caused little discomfort and was painless.

Physical examination revealed a hard, oval-shaped entity situated in the left submandibular area. The mass was about 3 × 3 × 3 cm in size, well demarcated, sessile and not tender to palpation. No enlarged lymph node was palpated. Intraoral examination showed that

Figure 1 Panoramic radiograph confirms a giant, poor-defined, irregular radiolucent lesion in the left mandible from the first molar to the angle where the damaged inferior cortex is expanding outwardly. Bone septa are noticed. The shape of the lesion is analogous to a flame appearance, suggesting an aggressive neoplasm. Although the second mandibular molar is quite close to the lesion, no precise displacement or root resorption of a tooth was captured.

a swelling extended from the retromolar pad to parapharyngeal space and was also situated in the buccal region of the mandibular ramus. The overlying mucosa was normal. No symptom of inferior alveolar nerve involvement or tooth mobility or sensitivity was found.

Ultrasonography, in this case, was adopted to justify that the mass was rooted in the mandible but not in the submandibular gland. Panoramic radiography confirmed a giant, poor-defined, irregular radiolucent lesion in the left mandible from the first molar to the mandibular angle, where the damaged inferior cortex was expanding outwardly. The shape of the lesion was analogous to a flame appearance, suggesting an aggressive neoplasm (Figure 1). CT sections displayed an irregular giant lump involving the left submandibular, pterygomandibular and parapharyngeal spaces. The lump was mainly sited in soft tissues, and a clear soft-tissue border was found in the anteromedial direction. In some image sections, the lesion showed an oval appearance with a size of about $6.3 \times 5.5 \times 4.4$ cm, and in other sections, the destruction of the left mandible was

obvious in the medullary and cortical bone from the mandible ramus to the first inferior molar. Integral ectosteal lesion was connected with the minor intrabony part via discontinuous lingual cortical bone. Plain CT revealed that the average CT value was 34 HU. Slight enhancement, with a CT value of 50 HU, at the periphery of the lesion was noticed after an iodinated contrast agent was given (Figures 2 and 3). The fluorine-18 fludeoxyglucose-PET-CT examination was taken for the exclusion of distant metastasis to the jaws. Images were inconsistent with what was shown in the CT scan (Figure 4), and no foci were found in the other parts of the body. Incisional biopsy was taken before surgical treatment. The diagnosis was OM. Removal of the tumour and adjacent submandibular gland with a partial *en bloc* excision of the mandible was performed. The gross specimen of the mandible is shown in Figure 5. Immediate reconstruction of the mandible contour was carried out with fibular osteo-cutaneous flap, and one immediate post-operative CT scan was performed for later comparative analysis (Figure 6).

Histopathological findings of the excised mass supported the diagnosis of OM (Figure 7). The course has been uneventful for 12 months after surgery (Figure 8). By phone call follow-up, an uneventful course of 30 months was confirmed.

Discussion

The giant expanded OM occurring in this 14-year-old male possessed a short clinical history of about 2 months. The short clinical history was rare and quite different from the consensus. According to many cases reported in the literature, the duration of the lesion always lasts a relatively long period owing to its slow growth and pain-free nature.^{1,4,5} The short clinical history of the presented case may be owing to the fact that the lesion broke the cortices and expanded in to the adjacent soft tissue at an early stage.

Figure 2 Plain CT sections. This image displays an irregular giant lump involving left submandibular, pterygomandibular and parapharyngeal spaces. The lump is mainly sited in the soft tissues and a clear soft tissue border is found in the anteromedial direction. In this section, the lesion shows an oval appearance with a size of about $6.3 \times 5.5 \times 4.4$ cm. 3D, three dimensional; DFOV, display field of view; STND, standard.

Figure 3 Enhanced CT sections. From this section, the left mandible is destructed in the medullary and cortical bone from the mandible ramus and body to the first inferior molar. Integral ectosteal lesion is connected with the minor intra-bony part via the discontinuous lingual cortical bone, and they share the same density. Plain CT revealed the average CT value was 34 HU. Slight enhancement, with a CT value of 50 HU, at the periphery of the lesion was noticed after iodinated intravenous contrast was given. DFOV, display field of view; STND, standard.

According to the literature, myxomas in the maxillofacial area may be divided into three types: central, peripheral and soft-tissue myxomas.^{4,5,12–14} Different types have different characteristics (Table 1). Intraoral periphery myxomas tend to be quite a benign tumour with a long history, more limited extent, rare bone involvement and no recurrence when compared with central myxomas of the jaws.^{12–15} Recently, one report demonstrated four cases of OM occurring in infants

less than 2 years old.¹⁶ Since these cases shared similar clinical manifestations and imaging characteristics, the authors proposed a name of infant OM for these cases.

It is generally accepted that conventional radiography and CT should be recommended to estimate OM.^{5,6,17} However, in a recent study, MRI was also suggested to be a routine examination method for OM because of its good ability to display patterns of growth of the tumour and to differentiate between various soft tissues.⁷ A

Figure 4 Positron emission tomography-CT images (axial and coronal sections) are inconsistent with what was shown in the CT scanning. The soft-tissue lesion was highlighted and gives a relatively smooth border to the adjacent tissues. DFOV, display field of view; STND, standard.

Figure 5 The gross specimen of the excised part of the mandible and the lesion. The lingual side of the bone was destroyed by the lesion.

scholar suggested that the varying radiographic appearances on conventional radiographs of OM could be divided into six types, that is, unilocular, multilocular, involvement of local alveolar bone, involvement of the maxillary sinus, osteolytic destruction and a mix of osteolytic destruction and osteogenesis.¹⁶ However, a number of cases reported in the literature evaluate radiographic images from the following three aspects: borders, internal characteristics and the effect on adjacent structures. Borders can be classified into three subtypes, namely, well defined, poorly defined and diffused; sometimes, the condition of the mandible cortex is recorded to explain the borders. Internal characteristics contain unilocular or multilocular, radiolucent or radiopaque and septa features. The adjacent structures usually refer to teeth displacement and other important anatomical structures.^{5,6,18,19} The above

Figure 7 Histopathological examinations show randomly oriented stellate, spindle-shaped and round cells with long, fine, anastomosing pale or slightly eosinophilic cytoplasmic processes extending from the centrally placed nucleus. Cells are evenly dispersed in an abundant mucoid or myxoid stroma that contains some fine collagen fibres. It supports the diagnosis of odontogenic myxoma. The bar in the image is 100 μ m.

classification is based on studies of intraosseous tumour of the jaws.

Problems emerged when we critically analysed the reported case. As the main part of this OM was in soft tissue and a minor part in the bone, what confused us was that which site, soft tissue or bone, was the primary site. Or was it a distant metastasis to the mandible? Thus, for this case, salivary gland tumour, malignant tumours from soft tissue, osteosarcoma or a distant metastasis need to be ruled out.

As ultrasonography is sensitive to differentiate a tumour within the salivary gland from a lesion outside the gland, it was adopted in this case.^{7,20} It turned out that the tumour was not from the submandibular gland. CT

Figure 6 CT image taken just after the operation together with a three-dimensional reconstructed image. DFOV, display field of view; STND, standard.

Figure 8 The image shows that there was no recurrence for 12 months after surgery.

images displayed that the mass perforated the cortical bone and invaded soft tissue. PET-CT revealed no other focuses or metastasis in the body. Differential diagnosis was thus limited to those lesions that originate from the mandible but may invade soft tissues. Osteosarcoma is the first possibility. However, the radiographic pattern of bone destruction in the mandible is not a typical sunray or hair-on-end appearance. Although haemangioma can occasionally present a sunray or sunburst image, the lesion should be radiopaque when an enhanced CT image is performed.

It is believed that OM is well delineated from the surrounding tissues and rarely invades soft tissues because the periosteum acts as a barrier and prevents extension of the tumour into soft tissues.^{7,21} However, this is not true for the present case, the lesions heavily invaded the adjacent soft tissues. This phenomenon will help a clinician put OM in the differential list when a similar case is presented.

Although the main purpose of a PET-CT examination was used for ruling out a distant metastasis to the mandible, the image of OM from a PET-CT scan was also obtained. From the PET-CT images (Figure 4), we can see that the soft-tissue lesion was highlighted and gives a relatively smooth border to the adjacent tissues.

This hints of a benign nature of the lesion. We also notice that the uptake of fluorine-18 fludeoxyglucose within the mass is not even. This may be a suggestion of a different histological composition within the tumours. To the best of our knowledge, this is the first PET-CT image of OM in the literature.

Usually, the management of OMs is surgical removal of lesions. According to the size of a lesion, contiguous structure infiltration, biopsy display and the general state of a patient, enucleation with curettage and peripheral ostectomy or segmental up to radical resections are the surgical options.^{10,12,15,17,22–24} In this case, removal of the tumour and the adjacent submandibular gland with a partial *en bloc* excision of the mandible was performed. By phone call follow-up, an uneventful course of 30 months was confirmed.

In the search of literature, we found that the recurrence rates of OM were about 25–43%, and closely relate to the location, extension and invasion character of the lesions and surgery strategy.^{15,23} Radical resections can guarantee the positive border of pathological tissues, while conservative enucleation may not remove lesions thoroughly.^{1,5,15,16,18,23} Most recurrences were identified in the close follow-up of the first 2 years.^{15,23}

Table 1 Three types of myxomas in the maxillofacial area

Item	Central myxomas of the jaws	Peripheral myxomas of the jaws	Soft-tissue myxomas
Prevalence rate	0.5–19.0% of oral tumours	Rare	Rare
Gender	Male > female	Male = female	Male > female
Age of patients	Often between 10 and 50 years	NA	Old people
Location	Jaw, with mandible predilection	Soft tissue overlying dentulous bone area	Skin, fascia, parotid gland, tonsil etc.
Growth rate	Quite slow (bone expansion as the chief complaint)	Slow (soft tissue mass)	NA
Growth pattern	Expansive growth	Expansive growth	Expansive growth
Invasiveness	Locally invasive	Infrequent bone invasion	Without bone involvement
Recurrence	Often within 2 years after operation	Rare	Rare
Histological origin	Odontogenic epithelia	Fibroblast or myofibroblast	Corresponding soft-tissue cells

NA, not available.

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