Giant Myxofibrosarcoma of the Mandible

SUMMARY

Myxofibrosarcoma is one of the most common sarcomas in elderly patients, but in the head and neck region it is a very rare entity. They are only few reports in the literature of myxofibrosarcomas of the head and neck. We found only 2 cases in the literature with mandible involvement and we present a case of a huge low-grade myxofibrosarcoma of the mandible, in a 23-year-old man. These few cases provide unreliable dissimilar clinico-pathological parameters for the prognostic information of local recurrence, metastasis and patients survival. Even in large series and reviews for myxofibrosarcomas of the extremities and the trunk in the literature, there is no consensus about concrete clinico-pathological prognostic parameters. Although there is no consensus about the pathogenesis and the clinical behaviour of myxofibrosarcomas, there exists clearly an agreement about their treatment.

Close collaboration between surgeon, radiologist, histopathologist and clinical oncologist in making accurate diagnosis and appropriate management of this rare tumour are mandatory. The patient is under narrow follow-up and without any signs of recurrence 39 months postoperatively.

Keywords: Myxofibrosarcoma; Mandible; Head and Neck

Introduction

Myxofibrosarcoma (MFS) is one of the most common sarcomas in the elderly patients, but in the head and neck region it is a very rare entity. There are only few reports in the English literature of MFS of the head and neck1-7, but only 2 cases present MFS of the mandible1,6.

Significant risk factors of survival and metastasis are size larger than 5 cm, tumour necrosis, and < 75% of myxoid areas within the tumour8.

We present a case of a huge low-grade MFS of the mandible, in a 23-year-old man and the result after 2 years seems to be satisfying, without recurrence, but still without reconstruction by bone graft.

Case Report

A 23-year-old man presented with a 2-months-history of a swelling on the left side of the lower face, enlarging slowly (Fig. 1). The physical examination revealed a palpable mass, painless, with hard consistency. Intraoral examination showed elimination of the left lower vestibular sulcus. No nerve palsy or nerve deficit was present. His medical history was uneventful. Imaging scan with computer tomography showed an ossifying mass of the left mandible, 85 x 60 mm in size (Fig. 2).

The malignant nature of the lesion was confirmed through a fine needle aspiration biopsy and an incision biopsy. Clinical signs of inflammation arose immediately after these procedures and the mass started growing. White spectrum antibiotics were given to treat the infection.

To facilitate the removal of the tumour, during the surgical intervention, through a curving transverse left cervical incision and a lip splitting approach, a double osteotomy of the mandible, along with coronoidectomy, was performed (Figs. 3 and 4). The resulting defect was bridged by a reconstruction plate. The patient recovered uneventfully (Fig. 5).
Figure 1. Clinical view of the patient in the operation room

Figure 2. An axial computer tomography scan, showing the extension of the lesion

Figure 3. The intraoperative picture of the tumour

Figure 4. The specimen were the extension of the tumour can be seen

Figure 5. A panoramic x-ray one month postoperatively

Figure 6. The spindle cells are minimally pleomorphic and separated by copious amounts of myxoid stroma
Microscopically, the tumour was characterized by a biphasic pattern: a benign epithelial component and a malignant mesenchymal component. The epithelial part was composed of well circumscribed nests of odontogenic tissue, surrounded by a hypocellular tumour within abundant myxoid stroma. The tumour cells had generally spindle or stellate shape, with eosinophilic cytoplasm and indistinct cell borders. The nuclei were hyper-hromatic, mildly pleomorphic, with mitotic figures (Figs. 6 and 7). On the basis of the histological findings, the diagnosis of the MFS of the mandible was made.

The patient is under narrow follow-up and without any signs of recurrence 39 months postoperatively. We are planning to reconstruct the mandible with a fibular free flap after a legitimate time interval of follow-up.

Discussion

MFS was originally described as the myxoid variant of malignant fibrous histiocytoma, which is uncommon in the head and neck region. But the term histiocytoma seems unjustified in light of modern histopathologic studies, and MFS has been accordingly defined as a distinct fibroblastic sarcoma in the latest WHO classification.

There exist limited reported cases for MFSs of the head and neck region, which represent only 3% of all MFSs. They provide unreliable dissimilar clinico-pathological parameters for the prognostic information of local recurrence, metastasis and patients survival. Even for the MFSs of the limbs and the trunk, prior series could not provide concrete clinico-pathological prognostic parameters. After local recurrence, however, MFS tends to progress in grade.

Clinically, the primary lesion may present as a predominantly deep or subcutaneous growth, or may involve the dermal layer and present as a cutaneous lesion. A completely infiltrative growth pattern along fascial planes, without the formation of a discrete nodular lesion, has been described in some cases.

Cytomegalovirus infection has been implicated in the oncogenesis of an infantile form of the tumour and has been proposed as probability in transplant recipient. Further evaluation is necessary to estimate the influence of viral infection in the oncogenesis of MFS.

We were able to find only 2 cases in the literature with involvement of the mandible, and the presented case seems to be the third one.

Although there is no consensus about the pathogenesis and the clinical behaviour of MFS, there exists clearly an agreement about the treatment. Close collaboration between surgeon, radiologist, histopathologist and clinical oncologist in making accurate diagnosis and appropriate management of this rare tumour are mandatory.

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References


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