

## Maxillary Osteosarcoma: A Case Report

K. Mazouz\*, H. Eddaoualine, L. Benyounes, D. Nimbuna, A. Elmatlini, M. Saadoun, H. Abourazeq, S. Barkich, H. Sami, I. Lalya, A. Elomrani, M. Khouchani

Radiation Oncology Department, Mohammed VI University Hospital, CHU Mohammed VI BP2360 Principal Avenue Ibn Sina, Marrakesh, Morocco

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\*Corresponding author: Mazouz Khadija

### Abstract

### Case Report

Osteosarcoma (also called osteogenic sarcoma) is a primary osseous tumor developing from mesenchymal cells elaborating malignant osteoid tissue. Although this is the most common primary malignant tumor of the bone after myeloma, it rarely affects the face. Maxillary osteosarcoma is particular by its clinical and histological aspects. Evolution is often rapid because of the aggressive character and the late diagnosis. Clinical presentation is not specific; surgery is the mainstay of management. The effectiveness of chemotherapy and radiotherapy has not yet been shown because of the rarity of this entity. The prognosis of osteosarcoma remains poor and depends on several factors. Through our observation we discuss the clinical, radiological and histological aspects as well as therapeutic management of maxillary osteosarcoma.

**Keywords:** Osteosarcoma, Osteogenic sarcoma, maxillary, facial, radiation therapy.

### Résumé

L'Ostéosarcomes (Sarcomes ostéogéniques) est une tumeur primitive de l'os dont les cellules tumorales élaborent du tissu osseux ostéoïde. Bien que ce soit la plus fréquente des tumeurs malignes primitives de l'os après le myélome, il touche rarement le massif facial. L'ostéosarcome maxillo-mandibulaire est différent par ses aspects cliniques et histologiques. L'évolution est souvent grave en raison du potentiel agressif, du diagnostic souvent tardif et des difficultés rencontrées pour le traitement. La symptomatologie est peu spécifique. Son traitement est principalement chirurgical, l'efficacité de la chimiothérapie et de la radiothérapie n'a pas encore été montrée à cause de la rareté de cette entité. Le pronostic de l'ostéosarcome reste sombre et dépend de plusieurs facteurs. À travers notre observation d'un cas d'ostéosarcome maxillaire, nous discutons les aspects cliniques, radiologiques et histologiques ainsi que la prise en charge thérapeutique des OSM.

**Mots clés:** Ostéosarcome, maxillaire, Clinique, radiologie, traitement.

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## INTRODUCTION

Osteosarcoma (OS) is a malignant mesenchymal tumor that produces immature osteoid tissue [1]. It usually affects long bones, representing less than 1% of the tumors of the face [3-5]. The maxilla-mandibular (OS) differs from that of the long bones in its clinical and histological appearance [2-7]. The prognosis is often worst due to a more aggressive evolution, the complex anatomy, late diagnosis and the difficulties of local control. We report the case of a patient with a maxillary bone.

## OBSERVATION

Mr. El. O, 64 years old farmer, with long smoking history. The first symptoms date back to 6 months; a painless non-hemorrhagic tumefaction at the

level of the upper gum (upper maxillary) gradually increasing in volume. Clinical exam described indured left maxillary mass. The covering teguments are of normal appearance, without palpable cervical nodes or neurological deficiency. Endo-oral examination revealed a burgeoning mass of the left vestibular and palatal mucosa, of firm consistency, bleeding in contact. The related teeth are healthy and respond positively to the vitality test, but they are partly covered with tartar due to poor oral hygiene. The clinical signs: (budding appearance, bleeding on contact, rapid growth) suggests a malignant tumor.

The radiographic assessment included:

Panoramic x-Ray which showed an osteocondensing image extending from the incisor region to the molar region.

And a CT scan (Fig-1) which confirmed the existence of a heterogeneous tumoral process of the left maxillary (Fig 2 & 3) reaching the upper limit of the maxillary bone (Fig-4).

Without distant suspicious lesion at Abdomino thoracic imaging. A biopsy was performed under local anesthesia and the pathological examination revealing asarcomatous process.

Tumor excision by external approach (para-latero-nasal approach) consisted of a 5x4.5x4 cm monobloc left maxillectomy containing a 3x3x3 cm of vegetative tumor with clear margins.

The histological study revealed a high-grade malignant tumor proliferation of osseous origin (Fig 5 & 6). On a myxoid background, we note the presence of sarcomatous cell areas with some fusiform (Fig-7) and atypical round cells. The tumor proliferation includes an osteoid and chondroid matrix (Fig 8 & 9). The anatomopathologist concluded to a high grade maxillary osteosarcoma with a chondroitin component estimated at 20%.

The patient received adjuvant chemotherapy: 6 cycles of Adriamycine-Cisplatine (Adriamycine 25mg / m<sup>2</sup>, cisplatin at a dose of 100mg / m<sup>2</sup> followed by external radiation therapy on the tumor bed according in 3D conformal technique for a dose of 60 Gy, with classical fractionation.

Currently the patient is in complete remission with a follow-up of 2 years. Last clinical examination does not find any signs of local recurrence or late radiation or chemotherapy related toxicity (Fig-10).

The last imaging describes a loss of substance in the anterior part of the bony palate with thickening of the soft parts nearby suggesting a post radiation origin (Fig-11).

The patient is always under regular surveillance.



**Fig-1: Maxillofacial CT Image: Osteocondensant process extending from the Incisive to the molar region**



**Fig-2: Axial CT scan, osseous process of the left maxillary**



**Fig-3: Frontal CT scan, osteolytic process of the left maxillary reaching the upper limit of the maxillary bone**



**Fig-4: Abdominal CT showing no suspicious lesion**



**Fig-5: Thoracic CT scan showing no suspicious lesion**

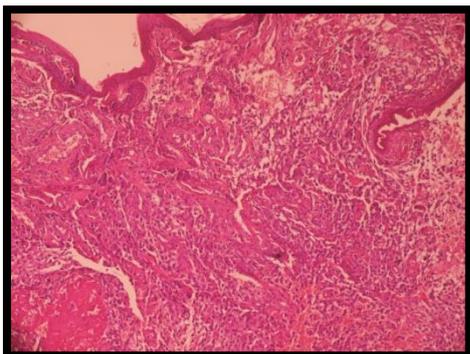


Fig-6: Poorly differentiated sarcomatous proliferation (HE coloration, x 4)

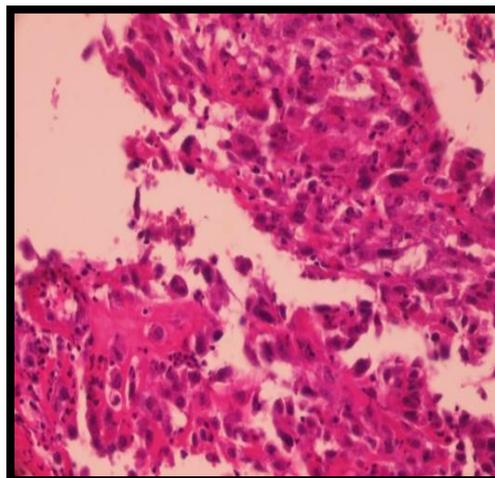


Fig-9: Neoplastic cells with malignant osteoid substance

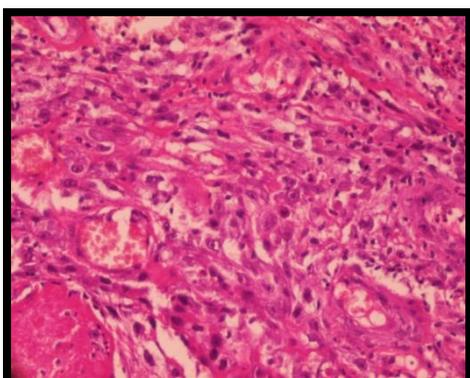


Fig-6: Atypical fusocellular tumor cells (HE coloration, x 40)



Fig-10: Loss of substance without signs of recurrence

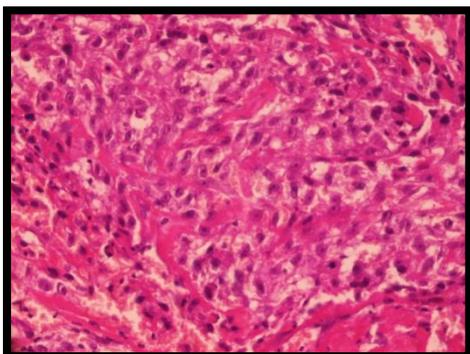


Fig-7: Production of an osteoid substance (\*) by tumor proliferation (HE Grs x 40)



Fig-11: Loss of substance in the anterior part of the bony palate with thickening of the soft parts next to the upper alveolar arcade

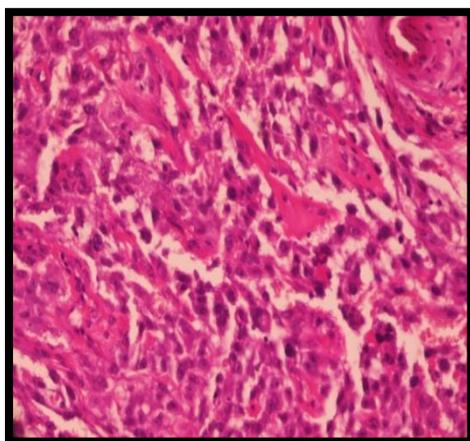


Fig-8: Atypical and giant cells

## DISCUSSION

The OS, formerly known as osteogenic sarcoma, is the most commonly encountered primary malignant bone tumor but remains like all sarcomas a rare disease [1], can affect all skeletal bones but most often develops on long bones sarcomas of the oral and maxillofacial region are even more rare: Gorsky and Epstein reviewed 11,250 head and neck tumors, of

which there were 139 cases of sarcomas (1,24%), and among those there were only 16 cases (0.14%) of intraoral soft tissue sarcoma [2]. This demonstrates the rarity of sarcomas in the maxillofacial system [3].

4 to 7% of osteosarcomas affect the maxillary and about 60% of the mandible [1, 4] Maxillary involvement is more readily on the alveolar ridge; the maxillary sinus is more rarely reached. The involvement of other bone structures (ethmoid, frontal, sphenoid) is exceptional [5, 10]. The maxillary localization is particularly frequent in the male subject [6] The average age of the patients varies between 30 and 40 years later than in the bones of the long bones [6, 7].

The exact etiology of OS is still unknown; however, some predisposing factors are involved: history of trauma, existence of a benign bone lesion that can be transformed [8, 9]. Radiation therapy: a dose of 30 Gy can promote the occurrence of a malignant transformation (radiation-induced OS) [8, 9]. The relative risk of developing osteosarcoma significantly increases with the combination of radiotherapy and chemotherapy [9]

Clinical presentation is not specific. Endobuccal swelling is the usual sign of discovery, some authors report a prevalence of 100% [10, 11]. It is usually painless, non-bleeding, sometimes associated with other symptoms including dental manifestations, sensory disturbances, nasal obstruction, epistaxis or exophthalmia. The duration between the first symptom and the diagnosis is about 3.4 months as our observation [10].

The radiological images of osteosarcomas are very diverse, often in relation to the different histological types [13]. The suspicion of the diagnosis is based on the highlighting of the appearance of "grass fire" or "sun rays": characteristic but non-pathognomonic. OS [12] panoramic radiography provides less information than CT and MRI, indeed CT scan and MRI allow to refine the assessment of local extension at the level of the soft parts, but especially to appreciate the intramedullary extension (MRI) and of thus guide the management decision [12, 13].

The OS is characterized by an anarchic osteogenic sarcomatous proliferation produced by malignant osteoblasts [10, 17]. It's an heterogeneous group with 3 histological types according to cell differentiation: predominant chondroblastic (the most common, such as in our case), osteoblastic, and fibroblastic [4]. Depending on the importance of cellular atypia, low grade (grade I), intermediate (grade II) and high grade (grade III and IV) lesions can be distinguished. Doval et al. also reported that patients with chondroblastic bone had a better prognosis than those with other histological components [5].

Maxillary OS are rare entity, so it is difficult to establish standard treatment. Complete surgical excision with clear supracentrimetric margins is the only effective treatment; This excision, often mutilating and difficult as the oral and maxillofacial regions include complex and vital structures [9, 14, 15]. Explaining the postoperative morbidity. In the presence of cervical lymphadenopathy, ipsilateral lymphadenectomy should be combined [9].

Pre- and postoperative chemotherapy is more controversial in maxillomandibular locations than in bone long [9, 10, 16]. Recent studies suggest that adjuvant chemotherapy improves overall survival and disease-free survival in patients with localized maxillary tumors in whom positive resection margins or high-grade tumor are present. Patients with local recurrence also benefited from the addition of chemotherapy [16].

The review of the literature confirms that postoperative radiotherapy of aggressive bone: tumor residue, positive margins, large tumor or high histological tumor at the dose of 60 Gy, significantly reduces the local recurrence rate [9, 15, 17].

Most series report an overall survival rate of 80%, and a 72% recurrence free survival [18], the metastatic potential of OSJ is low, but its growth is rapid explaining the high risk of local recurrence [9, 18].

The prognosis of the OS depends on: the delay between the first symptoms and the therapeutic management [9], the size of the tumor [20], the mandibular locations have a better prognosis than the maxillary localizations [19, 20], the quality of tumor resection that determines the risk of local recurrence [9, 19, 20], the response or to neo-adjuvant chemotherapy [9-21], the presence of osseous metastasis and the degree of differentiation; the histological type has no prognostic value [9, 20, 21].

## CONCLUSION

Maxillary osteosarcomas is a rare sarcomatous entity with a high degree of local aggressiveness. The diagnosis is not always easy and requires a confrontation of clinical, radiological, histological, and possibly biological data. Perioperative chemotherapy with clear margins is the mainstay of management; radiotherapy is most often indicated in palliative setting [9]. The evolution of the OS is characterized by rapid growth, a tendency to local recurrence and a low distant metastatic potential.

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