

Osteosarcoma of the Jaw: A Case Report

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ABSTRACT

The osteosarcoma is an uncommon malignant tumor characterized by the production of bone tissue. Maxillary localization is extremely rare and has a poor prognosis. The clinical and radiographic findings are similar to those seen in other malignant tumors of the jaw. The prognosis essentially depends on the forwardness of the diagnosis that must be the result of the vigilance of the practitioner and the dentist. Through this case report, our aim is to describe clinical, radiological and histological features, as well as the positive and differential diagnosis elements with the other maxillary pathologies.

KEYWORDS: Osteosarcoma, Sarcoma, Jaw

INTRODUCTION

Osteosarcoma is a malignant conjunctival tumor, characterized by the development of bone or osteoid substance of malignant tumor appearance. Mainly affecting the long bones, this tumor is rarely found in the jaws with an incidence of 4 to 7%.

Osteosarcoma of the jaws has a specific behavior. It presents an evolution characterized in most cases by severity, primarily in connection with its aggressive potential, its location in a complex anatomical region, its late diagnosis and difficulties for treatment.¹

The purpose of this work is to focus, more specifically, on the epidemiological, therapeutic and prognostic features of this tumor.

CASE REPORT

Mr.B.R 23 years old was followed and treated since November 2004 in the Oral Surgery and ENT departments, for a large left maxillary swelling, lasting for 6-8 months according to the patient.

The patient had no medical and surgical history, neither any alcohol or tobacco intoxication.

The Exo buccal examination (Fig. 1) showed a facial asymmetry due to the presence of a left cheek swelling. The recovery teguments are normal. The cervical lymph nodes are free, and skin sensitivity of the concerned area is retained. The intraoral examination revealed a budding swelling (Fig. 2) relevant to the buccal and palatal mucosa, extending from the 23 to the 28, firm in consistency, bleeding on contact. Related teeth are healthy and respond positively to vitality testing, but they are covered in part by dental calculus due to poor oral hygiene. The clinical table (budding aspect, contact bleeding, rapid growth) has suspected a malignancy.



Fig 1 : Left jugal Swelling with an important facial asymetry

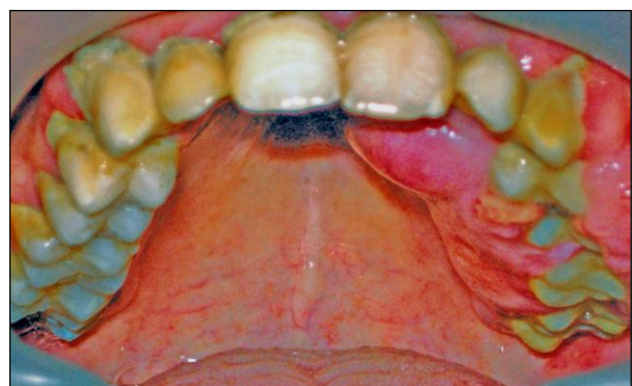


Fig 2 : Intraoral view: tumefaction to bud extending from the tooth 23 to the tooth 28

The radiographic assessment included:

- A panoramic radiograph that showed an osseocodensing image, extending from the incisor region to the molar region,
- A Blondeau incidence which revealed a filling of the

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left maxillary sinus (Fig. 3)

- A CT scan (CT) (Fig. 4), which confirmed the existence of a tumor process with heterodense aspect, containing a hyperdense component with an isodense component in the periphery, developed at the expense of the left maxillary bone. Tumor process reached the upper limit of the maxillary bone.

A biopsy was performed under local anesthesia, and histological examination showed that it was a sarcomatous process, of chondroid nature, suggesting the diagnosis of chondrosarcoma.

The staging (chest X-ray, bone scan, ultrasound abdominal) was negative, and laboratory tests were normal.



Fig 3 : Buccal view of the Swelling extending from the 23 to the posterior part of the arch

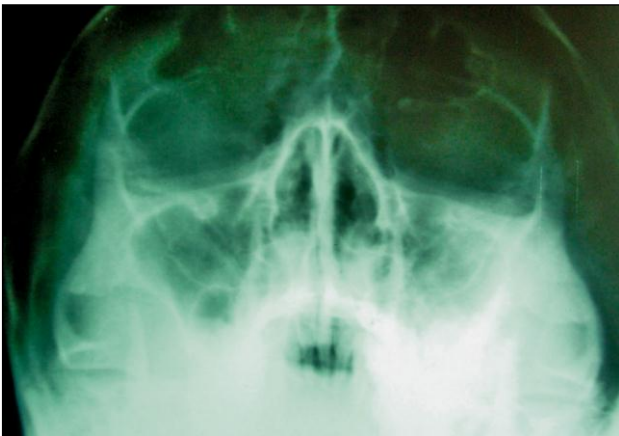


Fig 4 : Blondeau radiography: radiopacities of the maxillary sinus.

The excision of the tumor conducted with an external surgical approach (para-lateral-nasal surgical approach) (Fig. 5) consisted on a monobloc left maxillectomy, taking palatal dental tray but preserving the orbital floor.

Macroscopically the tumor resection margins were intact and past a minimum of 1 cm from the tumor (Fig. 6). Pathological examination of the surgical specimen (Fig. 7) showed a sarcomatoid stroma with skeleton bases arranged haphazardly and synthesized by atypical osteoblasts with malignancy characters.

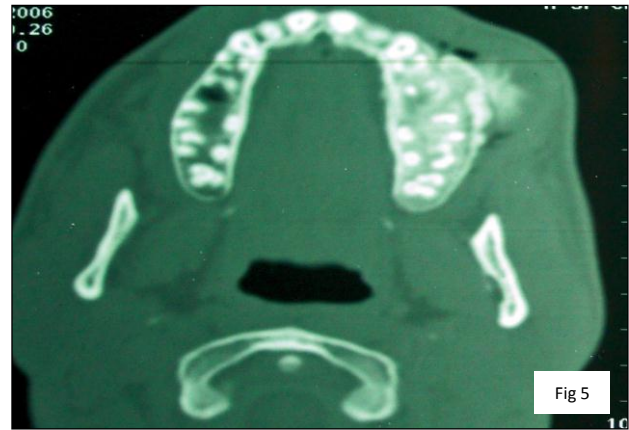


Fig 5



Fig 6

Fig 5 & 6 : Axial and coronal CT scan showing a tumoral process including a hyperdense component surrounded by a hypodense component



Fig 7 : Front view in per surgical : surgical resection tumoral per external way

This review concluded in osteoblastic steosarcoma (Fig. 8,9,10,11).

Surgical treatment was completed with chemotherapy (adriamycin, cisplatin, and methotrexate) in a period of 2 months and a half and adjuvant radiotherapy at a dose of 60 Gy. 18 months later, tumor recurrence was suspected and confirmed by CT scan: it showed a massive tumor recurrence, with extension to infratemporal fossa and the orbit. Palliative chemotherapy was initiated.



Fig 8 : Front view in per surgical: left maxillectomy in monobloc, the limits of resection were healthy and remote 1 cm of the tumour.

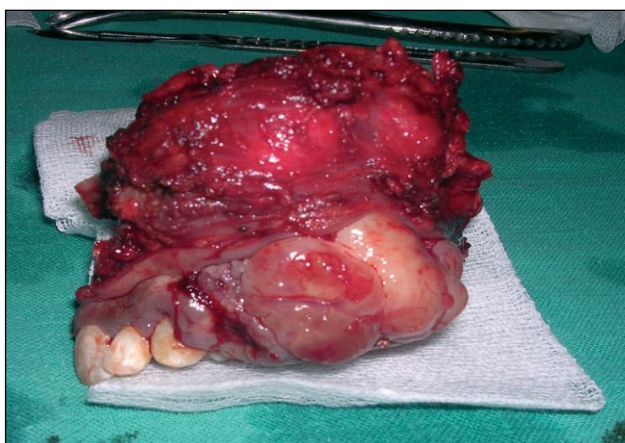


Fig 9 : Operative view: hemimaxillectomy in monobloc

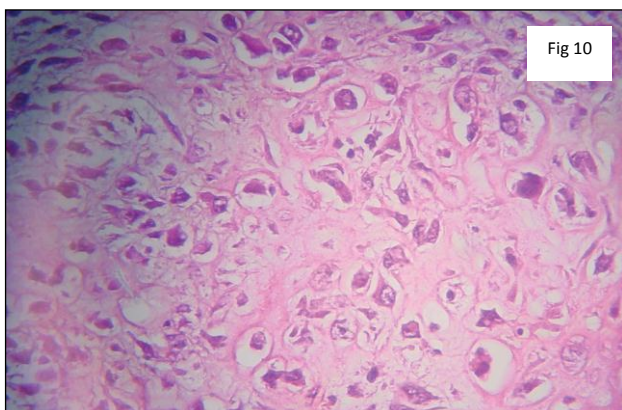


Fig 10

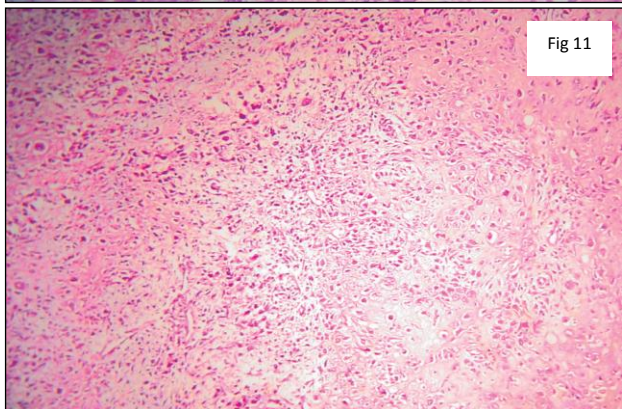


Fig 11

Fig 10 & 11: Histological aspect presence of a stroma sarcomatoide. This examination shows an osteoblastic osteosarcoma (hemateine - eosin, X 100 and 400).

DISCUSSION

Osteosarcoma was once called osteogenic sarcoma.^{1,2} These terms were gradually replaced in the International Nomenclature, the osteosarcoma.¹

For the first time in 1959, Hirota described the ultrastructural aspects of osteosarcoma. This tumor develops mainly in the long bones. In two-thirds of cases, it interests the knee and the shoulder blade; however, all skeletal bones may be affected.^{1,3,4}

Osteosarcoma is a frequent malignant primary bone tumor since it represents 47% of all malignant bone tumors.^{1,5} The craniofacial osteosarcoma is rare: about 1% of all tumors of the head and neck, and 10% of all osteosarcomas²; 4-7% of the osteosarcomas affect the maxillary bone^{4,5} and about 60% the mandible.² The elective site is the horizontal branch and symphysis region.^{1,2}

For the maxilla, all locations are found with the same frequency.^{1,3} The maxillary tumor, particularly common in the male subject, is observed at an older age than extra cephalic locations. The average age of patients between 30 and 40 years, ranging from ages 16 months to 74 years.^{2,6}

Most osteosarcomas appear primitive way.^{1,2,4} In some cases, a history of trauma was found in the anamnesis.

However, it is very difficult to establish an etiopathogenic link.⁴ These tumors may be secondary to malignant transformation of a benign bone lesion as fibrous dysplasia, Paget's disease or ossifying fibroma.^{2,6} The Radiation therapy may increase the risk of malignant transformation, thus radiation-induced osteosarcoma; time to onset is variable, ranging from 1 year to 40 years; they can grow from a dose of 30 Gy.^{1,7} The radiotherapy and chemotherapy significantly increase the relative risk of developing osteosarcoma compared with radiotherapy or chemotherapy alone.^{8,9} This relative risk is multiplied by 10 after radiotherapy alone and 27.5 after radiotherapy and chemotherapy.^{7,8}

Clinically, the symptoms are nonspecific and include the common signs of all maxillary tumors.^{1,5} This partially explains that the average time between the appearance of the first manifestations and diagnosis is relatively long from 6 to 12 months⁴, as in our case. The two most frequently clinical signs found are pain and swelling.

The swelling is elastic in consistency, firm or hard, painless, covered in the outbreak of the evolution by a normal or slightly congestive large sized mucosa, which can deform the face or limited, characterized by filling the vestibular fornix.^{1,4,10} In fact, the initial symptoms are frustrated and, at this stage, no clinical sign allows the practitioner to suspect the sarcomatous origin.^{1,10} In advanced cases, other signs occur and are also considered: dental manifestations (mobility, pain, spontaneous loss), sensory disturbances (hypoesthesia or anesthesia) in the territory of the infraorbital nerve. There

may also be a nasal obstruction, epistaxis, sometimes exophthalmos.^{1,3,4}

Conventional radiography (panoramic radiography) evokes the diagnosis, but it still retains a limited value in

the balance sheet expansion due to the superposition of bony structures. It can show osteolytic images in the mandible or osteosclerosis images, especially in maxillary lesions. Both aspects can also coexist.^{1,11} Typically, there is a characteristic appearance, although not pathognomonic in "grass fire" or "sunshine", due to periosteal reaction and production of osteoid tissue in the periphery of the tumor.^{1,11,12}

Other radiological signs may be associated with osteosarcoma as:

- A periodontal ligament expansion secondary of the tumor invasion,
- Root resorption of the teeth in relation to the tumor.^{1,4}

CT is used to evaluate the size of the tumor, its limitations, the presence or absence of intratumoral calcifications, tumor density before and after intravenous injection of contrast medium. It also clarifies invasion of the cortical bone and in most cases, the surrounding of soft tissues.^{12,13}

The magnetic resonance imaging (MRI) is preferable to CT for evaluation of tumor spread to soft tissues, the intracranial structures, and the orbital cavity.¹³ Bone Technetium-99 scintigraphy and chest radiography are part of the balance sheet for metastases.^{1,14}

In any case, even before a very suggestive radiological appearance, the certain diagnosis of the osteosarcomatous nature of the lesions will only be made by histological examination of the tumor biopsy.^{2,14} Histologically, the tumor presents a heterogeneous appearance with osteogenic areas and other non-osteogenic (parenchymal appearance or encephaloid). Osteogenic areas are harder and less vascularized. There may be association with the appearance of cartilaginous areas, areas of necrosis or hemorrhage.^{1,14,15} The surface portion of the tumor is usually softer and more suitable for biopsy.¹⁶ The essential histologic character of osteosarcoma is the existence within a skeleton sarcomatoid tissue stroma arranged haphazardly and synthesized by osteoblasts atypical, with malignancy characters.^{17,18} Depending on cell differentiation, three histologic types of osteosarcoma are considered: osteoblastic, fibroblastic and chondroblastic. Most often, these different histological types are found within the same tumor.^{19,20} In the maxillofacial region, the osteoblastic and chondroblastic osteosarcomas seem equally divided between the mandible and maxilla.^{1,9,14} For some authors, the chondroblastic type is the most common (50% of osteosarcoma) while for others, the osteoblastic type is instead.^{9,16} Histologically, the diagnosis of osteosarcoma is sometimes difficult to confirm. It can be confused with other tumors including chondrosarcoma when biopsy

interests the chondroid component of chondroid osteosarcoma, with a chondroma when it is a chondroid osteosarcoma, or osteoma, when it concerns a bone juxta well-differentiated osteosarcoma.

The treatment of osteosarcoma most often associated three therapeutic approaches^{17,18,19}: Surgery remains the treatment of choice, chemotherapy has significantly improved treatment outcomes in carcinology, especially for osteosarcoma, and radiation which remains to be discussed because these tumors are known by their radiation resistance.^{20,21}

For maxillary tumor, tumor resection requires partial or total maxillectomy sometimes extended to neighboring anatomical structures. The surgical approach is usually external incision with para-lateral nasal type. This resection should be performed if possible, in one piece with a margin of safety. However, in most cases, the removal is difficult due to the anatomical complexity of the jaw bone and tumor spread to adjacent structures often found in these tumors.^{1,3,14,21} Postoperative sequelae can be very important both functionally (slurred speech, the flow of fluids and food in the nasal cavity) and esthetically (cheek depression, skin scar).

These effects can be corrected with prosthetic rehabilitation and / or reconstructive surgery.^{22,23} Surgical treatment of lymph nodes areas should consider the presence or absence of cervical lymph nodes in the clinical and radiological examination. In the presence of lymphadenopathy, cervical lymph node dissection should be performed and include different cervical lymph nodes: submental, submandibular, jugular-carotid, spinal and supraclavicular.^{1,14,21}

Chemotherapy has spectacularly improved the prognosis of osteosarcoma of long bones, but its role in maxillary osteosarcoma remains controversial. No study has clearly demonstrated that it has efficacy in the cephalic locations. Several therapeutic protocols are proposed and it is now accepted that the protocols combining preoperative chemotherapy (neoadjuvant) to a postoperative (adjuvant) chemotherapy have more benefits than conventionally carried out only adjuvant chemotherapy.^{14,21,22} Postoperative radiotherapy at a dose of 60 Gy, retains his place for the prevention of local recurrence of osteosarcoma. Recent studies suggest that postoperative radiation therapy can significantly reduce the frequency of relapses. It is especially indicated when the resection margins pass in tumor tissue, for large tumors and to all high histologic grade tumors.^{1,14,21}

The osteosarcoma prognosis depends on several parameters:

- The time interval between first symptoms and therapeutic care.^{1,3}
- The tumor volume: the prognosis is even worse when the tumor volume is important.^{1,16}
- Seat: prognosis of osteosarcoma of the maxilla is more favorable than osteosarcoma of other locations since the overall survival rate varies from 35% to

53% for the earliest while it is only 5 to 30% for the latter.^{2,17}

- Mandibular locations have a better prognosis than the maxillary locations since the overall survival rates were 41% and 25%, respectively.^{10, 14, 17}
- The quality of excision determines the risk of local recurrence.^{1, 14}
- The answer to whether neoadjuvant chemotherapy can be evaluated on the piece of tumor resection.^{4, 8}
- The presence or absence of metastases: although they occur more frequently, the maxillary osteosarcoma metastasizes later than those of different locations, and usually through blood. These metastases are an order of decreasing frequency, lung, spine, rib and pelvic. There is rarely a spin through the lymph.^{4, 14}
- The degree of differentiation: poorly differentiated osteosarcoma has a poor prognosis.²⁰

The histological type of osteosarcoma has no prognostic value.^{2, 20}

Evolutionarily, osteosarcoma is characterized by its rapid growth, its strong tendency to local tumor recurrence and distant metastatic low power.^{1, 14, 17}

CONCLUSION

Osteosarcoma is a sarcomatous tumor high degree of malignancy. This partly explains the poor results still reported in many series.

The diagnosis is not always easy and requires a comparison of clinical, radiological, histological, and possibly biologic data.

The treatment is mainly based on the surgical excision of the tumor associated with pre- and postoperative chemotherapy; Radiotherapy is usually given in palliation.

The dentist is the first practitioner viewed in most cases. The discovery of a tumor lesion in the systematic realization of thorough clinical examination of the oral cavity should never be trivialized: it must lead to investigations in order to clarify its nature.

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