

Chondroblastic osteosarcoma of the mandible: case report

Osteossarcoma condroblástico em mandíbula: relato de caso

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ABSTRACT

We report the case of a 32-year-old male patient, who was subjected to cone-beam tomography for planning the removal of an unerupted tooth. Cone-beam CT scans revealed the presence of a mixed lesion with poorly defined limits in the left posterior mandibular region, with “sun-ray” periosteal reaction. An incisional biopsy was performed and the diagnosis was chondroblastic osteosarcoma. In this paper, a discussion of the differential diagnosis criteria and treatment of this unusual malignant neoplasm is performed. Thus, it is intended to provide information that will help in the study of management protocols for the increasingly effective treatment of gnathic osteosarcomas.

Key words: head and neck neoplasms; bone neoplasms; osteosarcoma.

INTRODUCTION

Osteosarcoma (OS) is the most common primary malignant bone tumor, occurring more frequently in long bones and rarely in the maxillofacial region⁽¹⁾. It represents less than 1% of all biopsies in the maxilla and mandible^(2, 3). The most common signs and symptoms of OS are pain or paresthesia, rapid growth, swelling and expansion of cortical bone, facial asymmetry, nasal obstruction, displacement and mobility of associated teeth⁽⁴⁾.

Radiographically, the tumors appear as a radiolucent, radiopaque or mixed image, with irregular and poorly defined margins and periosteal osteophytic reaction in an spiculated pattern perpendicular to the cortical bone (“sun ray” appearance)^(5, 6). From a histopathological perspective, the OS is characterized by the proliferation of ovoid, spindle-shaped cells or with stellar shaped irregularly morphologies, with varying degrees of atypia, associated with direct osteoid matrix deposition, by tumor cells⁽⁷⁻¹⁰⁾.

The treatment of choice for head and neck OS is aggressive surgical resection, with wide margin of safety, associated with adjuvant chemo and radiotherapy⁽¹¹⁾. Radiotherapy should be reserved for the treatment of OS of the jaws, with the possibility of positive margins for tumor cells after surgical resections or in cases

of unresectable local recurrences. The use of chemotherapy in OS of the jaws has also been reported, since it may be an important tool for the treatment of this neoplasm, but consensus is still to be reached concerning the timing of its application, whether before or after surgery^(2, 12, 13).

Based on the above, this article aims to report a case of chondroblastic osteosarcoma of the mandible, as well as to discuss the morphological criteria of differential diagnosis and factors involved in the prognosis of this lesion.

CASE REPORT

A 32-year-old mulatto male patient attended the private dental office for extraction of tooth 38 partially erupted. During the intraoral clinical examination, no noticeable changes were observed. Cone-beam computed tomography (CT) was ordered (**Figure 1**), and demonstrated, in a panoramic reconstruction, the presence of a mixed image with poorly defined limits, located in the posterior mandible area, on the left side. Furthermore, partial resorption of the distal root of tooth 37 was observed. In coronal sections, it was possible to observe perforation of buccal and lingual cortical plate and osteophytic reaction. In the axial sections, in

turn, change was observed in the density of the trabecular bone, which was hyperdense. In 3D reconstruction (**Figure 2**), the loss of lingual and vestibular cortical continuity was confirmed. Therefore, the diagnostic hypotheses established by the dentist at the clinical and imaging examination were osteosarcoma and osteomyelitis. The incisional biopsy was performed and followed by histopathological evaluation.

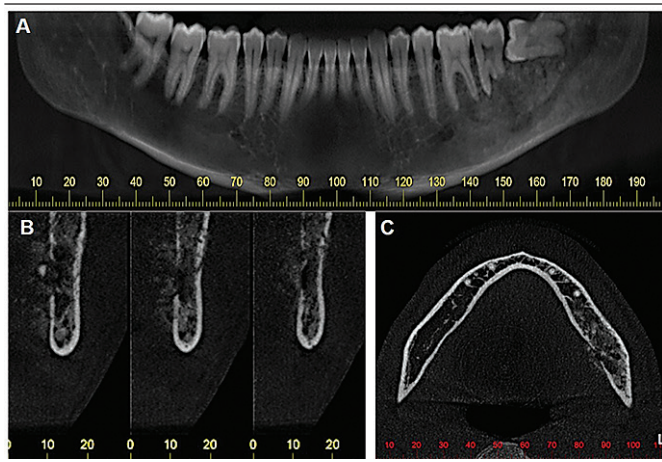


FIGURE 1 – Cone-beam CT scan of the patient

A) panoramic reconstruction, showing a mixed lesion in the mandible posterior region, with poorly defined limits, which caused partial reabsorption of the distal root of the tooth 37; B) coronal sections exhibiting periosteal osteophytic reaction with “sun ray” appearance in the mandibular lingual cortical; C) axial section showing hyperdense mandibular medullary bone, on the left side.

CT: computed tomography.

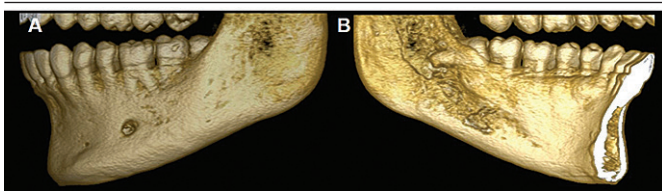


FIGURE 2 – Three-dimensional reconstruction of left posterior mandibular region, showing loss of continuity of buccal (A) and lingual (B) cortical plates

Histological sections stained with hematoxylin and eosin (HE) (**Figure 3**) revealed a proliferation of mesenchymal cells with ovoid to fusiform morphology, exhibiting moderate cytological atypia. The parenchymal component was associated with focal deposition of trabecular eosinophilic matrix, interpreted as osteoid, as well as abundant basophilic amorphous matrix, forming gaps that contain pleomorphic ovoid cells, consistent with neoplastic chondroid tissue. Both matrices suffered extensive areas of mineralization, forming irregular trabeculae of tumor bone tissue. The established diagnosis was chondroblastic osteosarcoma. The patient refuted the diagnosis and reported that he needed a

second evaluation with another professional. Four months after the incisional biopsy, the patient returned to the dental surgeon presenting, in the extraoral clinical examination, a noticeable increase in volume, affecting the middle and lower third of the left side of the face, with hard consistency on palpation (**Figure 4**). In the intraoral examination, the patient had an increase in volume with lining leukoerythroplastic mucosa, bleeding on touch, in the left posterior mandibular region (**Figure 5**). Faced with the new situation, the patient was immediately referred to the oncologist, who established pre-surgical chemotherapy as a treatment, in an attempt to reduce tumor size, and, later, mandibulectomy associated with adjuvant radiotherapy.

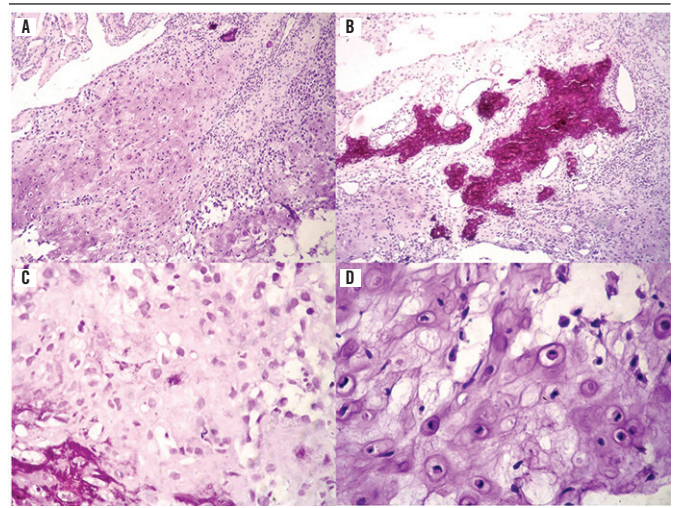


FIGURE 3 – Histological sections stained with HE

A) deposition of basophilic and eosinophilic material consistent with chondroid and osteoid matrix, respectively; B) intense deposition of interspersed mineralized trabecular tissue through a hypercellularized connective tissue; C) osteoid matrix and presence of atypical osteoblasts with ovoid to fusiform morphology; D) irregular gaps within the chondroid matrix (chondroplasts) containing cells predominantly compatible with atypical chondroblasts.

HE: hematoxylin and eosin.



FIGURE 4 – Frontal image of the patient, showing volume increase in the middle and lower third of the face, left side, with no cardinal signs



FIGURE 5 – Intraoral photography, showing volume increase with presence of leukoerythroplastic lining mucosa with ulcerated areas

About one year and a half after the initial diagnosis, the patient was diagnosed with brain metastasis and was referred to a new cycle of chemotherapy. However, he died two months later due to complications secondary to chemotherapy, culminating in multiple organ failure.

DISCUSSION

OS is a primary malignant bone tumor characterized by direct formation of immature bone and osteoid tissue by the tumor cells. Although it affects mainly long bones, about 6%-7% of these malignant tumors occur in the maxillofacial region⁽¹⁴⁾. The OS of the jaws most frequently affects males, with a mean age of 27.2 ± 13.6 years, with no significant difference between maxillary (30.7 ± 15.8) and mandibular (23.4 ± 10.1) lesions, affecting both gnathic bones equally⁽²⁾. Very similar data were also reported by Azizi *et al.* (2009)⁽¹⁵⁾. Since the reported case refers to a 32-year-old male patient, it is possible to state that he is in agreement with the demographic profile presented in the literature on this tumor.

In the present case, the patient was asymptomatic at the time of diagnosis. This is in agreement with the study developed by Ajura & Lau (2010)⁽⁸⁾, in which 59 cases of gnathic OS were analyzed and observed that most cases (77%) did not report any type of symptomatology, while only 15% of tumors cases presented pain and 8% paresthesia. Moreover, these authors still mentioned that in 26% of the cases the initial suspicion of the lesion was, as in the present case, based on eminently imaging findings. These data suggest that routine imaging examinations may play a key role in the early diagnosis of central lesions of the jaws.

From the imaging perspective, the symmetrical thickening of the periodontal ligament space and the loss of continuity of the lamina dura of involved teeth have been pointed out as one of the earliest signs of development of this malignant neoplasm^(16,17). Furthermore, Arora *et al.* (2013)⁽¹⁸⁾ argue that, as observed in the present case, the formation of periosteal reaction as bone spicules radiating from the cortical bone, commonly referred to as “sun-ray”, is a frequent feature in Gnathic OS, especially in mandibular tumors. This latter characteristic can be better visualized in occlusal radiographs or axial and coronal CT scans⁽⁹⁾, which again corroborates the findings described in this case report.

The histopathological aspects of this study led to the classification of the tumor as a chondroblastic variant of OS. In fact, a number of studies have pointed to this histological subtype as the most common form of OS in maxillary bones^(2, 8, 15, 19). However, large-scale production of the chondroid matrix can make stressful the distinction between chondroblastic OS and chondrosarcoma⁽⁷⁾. In the morphological perspective, the identification of foci of osteoid matrix being deposited by tumor cells, as occurred in the present case, independently of forming or not the bone trabeculae tumor by endochondral ossification of the cartilage matrix, it is the main criterion to establish the diagnosis of chondroblastic OS to the detriment of chondrosarcoma⁽²⁰⁾. The identification of malignant osteoid matrix associated with the integration of clinical and imaging data continues to be the cornerstone for the diagnosis of osteosarcoma and, to date, no immunohistochemical marker has equaled the diagnostic value of this important morphological marker⁽²¹⁾. Furthermore, it is important to emphasize that the differential diagnosis between these two entities does not constitute a purely academic exercise. In fact, chondrosarcomas are tumors that are resistant to radiotherapy, whereas osteosarcomas are radiosensitive, that makes the precise distinction between these two entities a particularly important issue capable of greatly influencing the treatment and the prognosis of the lesion⁽²²⁾.

The option for the radical surgical approach, represented by mandibulectomy, adopted in this case, is in line with that suggested by the specialized literature for cases of mandibular OS⁽²³⁻²⁵⁾. Chemotherapy and, in particular, postoperative radiotherapy are also complementary therapeutic procedures commonly used in the treatment of head and neck OS⁽²⁶⁾. The present case is also in agreement with the literature, in view of the option for radiotherapy after surgical removal of the tumor.

Pre-surgical chemotherapy has been a procedure used for large tumors in an attempt to minimize tumor volume and provide a less aggressive surgical approach⁽²⁷⁾. In view of this possibility, this strategy was adopted in the present case. However, the tumor

was resistant to chemotherapy. Although it represents an unusual outcome, resistance to pre-surgical chemotherapy is a phenomenon well-documented in the literature, probably due to the specific selection of chemoresistant aggressive tumor clones⁽²⁸⁾. Accordingly, the possibility that such a biological phenomenon contributed to the fatal outcome, in the present case, it can not be ruled out^(2,29).

In the light of the above, case reports bringing information about diagnostic criteria, therapeutic response and clinical course of OS are extremely relevant tools for the better knowledge of these tumors. Thus, it is possible to bring in subsidies that will contribute to the establishment of increasingly effective protocols for OS treatment, which will reflect a better prognosis for patients.

RESUMO

Relatamos o caso de um paciente do sexo masculino, 32 anos de idade, que realizou tomografia cone-beam para planejamento de remoção de dente incluso. O exame revelou presença de lesão mista de limites mal definidos na região posterior mandibular esquerda, com reação periosteal em “raios de sol”. Foi realizada biópsia incisional, e o diagnóstico foi de osteossarcoma condroblástico. Neste trabalho, será realizada uma discussão dos critérios de diagnóstico diferencial e do tratamento dessa neoplasia maligna. Pretende-se, dessa forma, trazer informações que auxiliem no estudo de protocolos de conduta para o tratamento cada vez mais efetivo dos osteossarcomas gnáticos.

Unitermos: neoplasias de cabeça e pescoço; neoplasias ósseas; osteossarcoma.

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