


Chondroblastic osteosarcoma of the maxilla: diagnostic challenges on incisional biopsy and report of a case

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Abstract:

Osteosarcoma is a rare mesenchymal neoplasm characterized by osteoid production. Its neoplastic cells can synthesize variable kinds of matrix which determines the sub types as the chondroblastic variant and its cartilaginous matrix. We report a case of a 30-year-old man who presented a painful swelling in the right posterior maxilla, associated to teeth mobility. Radiographic exam showed bone rarefaction with poorly defined margins and root resorption in the corresponding area. An incisional biopsy was performed and histopathologic examination revealed proliferation of mesenchymal spindle-shaped cells in a stroma with irregular, immature and basophilic cartilaginous tissue without osteoid tissue. The cell were diffusely positive for S100 protein. A provisional diagnosis of chondrosarcoma was rendered until the excisional biopsy revealed the presence of osteoid tissue possibiliting a final diagnosis of chondroblastic osteosarcoma. Differentiating between chondrosarcoma and chondroblastic osteosarcoma in incisional biopsies may be a hard task when material is scarce, often incurring in the need for further biopsy. Correct diagnosis is essential for the correct treatment.

Keywords: Osteosarcoma; Chondrosarcoma; Diagnosis, Differential.

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INTRODUCTION

Osteosarcoma (OSA) is defined as a primary malignant tumor with osteoid production, even in small amounts, which may or may not present mineralized tissue¹. This malignancy occurs more frequently in long bones. In the craniofacial skeleton, the jawbones are the most common affected sites, with approximately 6% of all cases arising in the mandible or maxilla².

Although OSA of the jaws can affect any age group, the majority of patients are older than 30 years^{2,3}, with a predominance of males^{3,4}. Some studies have shown a higher incidence of OSA in the mandible than in the maxilla^{2,3}, but the opposite is also found⁵. The body of the mandible and the alveolar ridge of the maxilla are the most commonly affected sites^{2,3}.

According to the type of matrix synthesized by neoplastic cells, OSAs can be subclassified as osteoblastic, chondroblastic and fibroblastic variants¹. Microscopically, approximately a half of OSAs of the jawbones are chondroblastic and are often mistaken for chondrosarcoma, especially in small incisional biopsies⁶. We report a case of a fatal chondroblastic OSA arising in the maxilla with emphasis on the challenge of histopathological diagnosis in an incisional biopsy.

CASE REPORT

A 30-year-old male patient, non-smoking and non-alcoholic, complained about a painful swelling in the right posterior maxilla that had been growing over a period of a month, associated with mobility of the teeth 1 and 2 (Fig. 1A and B). Intraoral examination showed a considerable erythematous mass lined by extensively ulcerated and necrotic tissue in the right posterior maxilla with involvement of the whole ipsilateral palate (Fig. 1C and D).

The radiographic exams showed a process with mixed aspect and ill-defined borders in the corresponding area (Fig. 1 E and F). There was also root resorption of involved teeth. The patient underwent a computed tomography (CT) that revealed right maxillary destruction by the lesion with invasion of the maxillary sinus and nasal cavity. An incisional biopsy was performed and the material was sent for pathological examination.

Histopathological examination of hematoxylin and eosin (H&E) stained sections revealed a neoplastic process characterized by immature tissue with chondroid differentiation presenting atypical chondrocytes

with variable morphology, many of which spindle or star-shaped, sometimes binucleated (Fig. 2A). Immunohistochemical reaction for S100 protein showed diffusely positive cells (Fig. 2B). Despite the meticulous microscopic inspection, no osteoid tissue or tumor ossification were found, which led to the diagnosis of chondrosarcoma, and a note to the clinician about the possibility of a chondroblastic osteosarcoma.

The patient underwent a complete tumor resection. Microscopic analysis of the surgical specimen revealed a high grade mesenchymal neoplasm characterized by sharply pleomorphic cells associated to partially mineralized eosinophilic material deposition interpreted as osteoid, as well as areas with differentiation in neoplastic cartilaginous tissue (Fig. 2C and D). The diagnosis of chondroblastic osteosarcoma was established. Even with radical surgery and chemotherapy sessions, the patient developed lung metastases and died eight months after diagnosis.

DISCUSSION

Osteosarcomas represent a heterogeneous group of mesenchymal malignancies sharing common characteristic as osteoid deposition and/or neoplastic ossification¹. About 5% to 10% of osteosarcomas arise preferentially in the mandible and maxilla. Usually the Head and Neck OSAs occurs in patients one to two decades older than OSA of long bones^{7,8}. The most frequently clinical appearance is a painful swelling along with others. Other common symptoms as: paresthesia, toothache, displacement or loosening of teeth, bleeding and nasal obstruction, depending on the location of the tumor^{2,4,9}.

The difficulties with clinical, radiological and histopathological diagnosis of osteosarcomas of the jawbones are frequent, especially in small biopsy specimens¹⁰⁻¹². The material initially sent for histopathological exam was represented only by immature cartilaginous tissue with pleomorphic chondrocytes in a myxoid matrix. Due to the lack of microscopic features, the diagnosis of OSA could not be established immediately. However, the possibility of being a chondroblastic OSA was not ruled out then. The posterior excisional biopsy and revealed a chondroblastic OSA.

Since OSA of jawbones, including the chondroblastic variant, carries a worse prognosis than chondrosarcoma, the differentiation between these two entities is probably the most important issue to consider

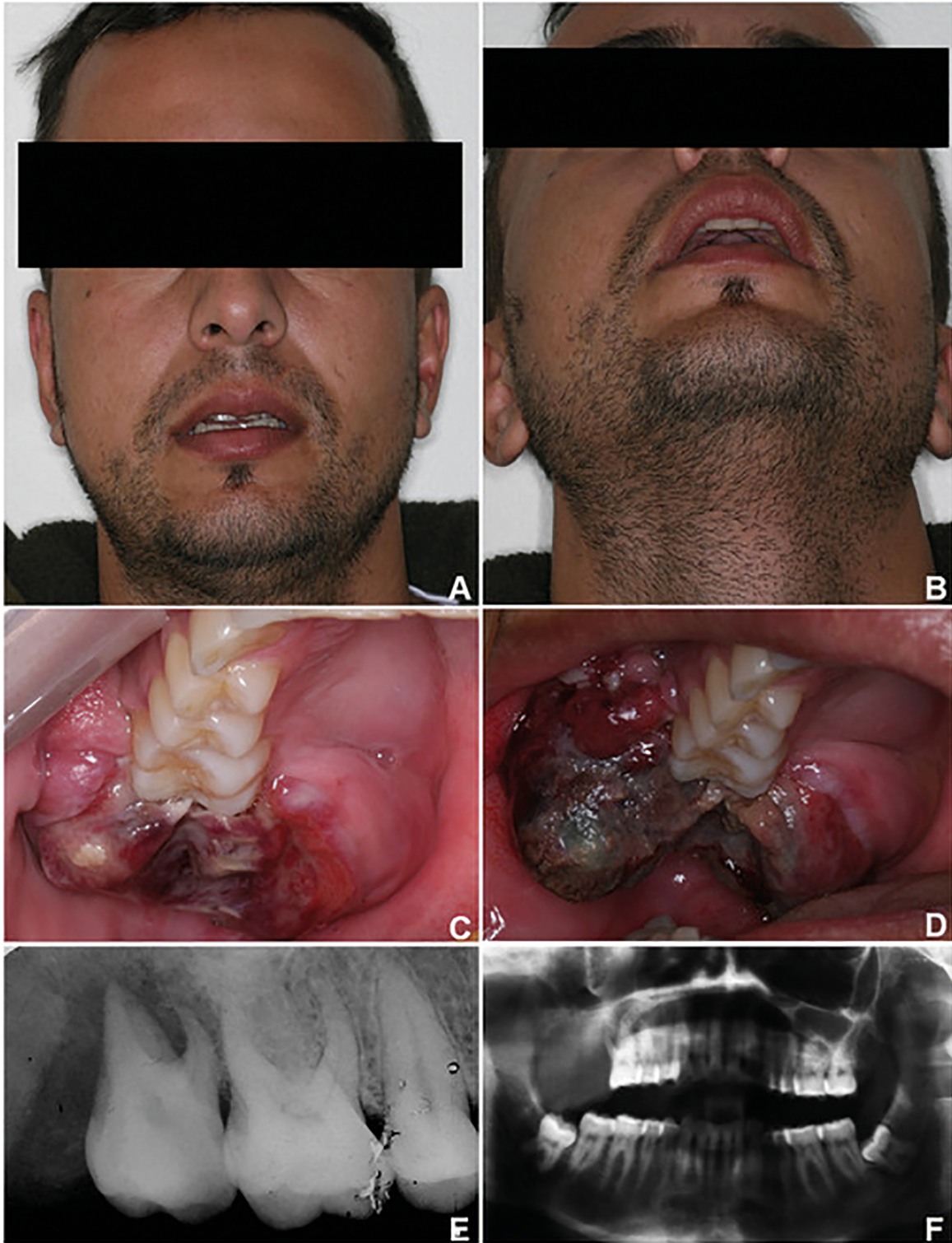


Figure 1. Clinical aspect of the patient showing difficulty in dental occlusion (A) and a swelling in the right maxilla (B); Intraoral view of neoplastic mass showing occlusion mark (C); Tumor appearance one week later (D); Radiography made after of tooth 1 excision and before loss of tooth 2, showing bone rarefaction and root resorption (E); Process in posterior maxilla with mixed aspect and ill-defined borders (F).

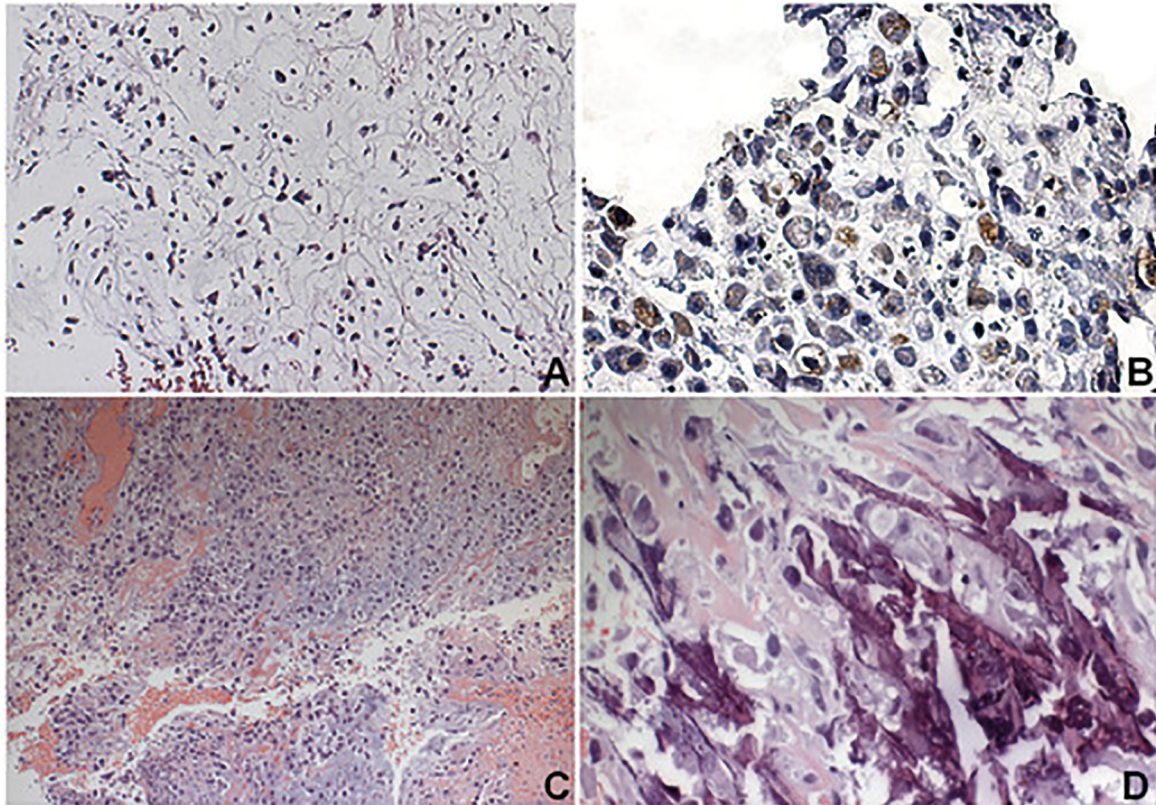


Figure 2. Histopathological aspect of the incisional biopsy (A and B). Immature tissue with chondroid differentiation and atypical chondrocytes and no osteoid tissue (H&E x 10 objective)(A); Neoplastic cells positive for S100 protein (x 40 objective). Histopathological aspect of the excisional surgery (C and D); Extensive areas of chondroblastic differentiation (H&E x 10 objective); Basophilic areas of tumor ossification and osteoid (H&E x 40 objective).

when dealing with a sarcomatous cartilage-forming tumor¹³. In the absence of a specific marker, differential diagnosis is sometimes impossible. Gomez-Brouchet et al.¹⁴ related this difficulty and proposed Galectin-1 as a useful marker in distinguishing chondroblastic OSA and conventional chondrosarcoma. Analyzing 165 bone sarcomas by immunohistochemistry and western blot, Galectin-1 was abundant in normal human osteoblasts from benign proliferations and in OSA, including chondroblastic OSA, but not in chondrosarcomas, which could help to differentiate these two neoplasms.

The prognosis of the jawbones OSA varies among the series, and it seems to have a better outcome than those arising in long bones, especially the chondroblastic variant⁴. Other studies have shown no prognostic differences between these topographies³. The prognosis seems to be more favorable for osteosarcomas in the mandible than in the maxilla, as in the presented case².

This may be related to the time course and resectability of the tumor. Therefore, the authors describe a case of chondroblastic OSA affecting the right

maxillary bone with fatal outcome. When faced with an equivocal diagnosis, a pluridisciplinary discussion among pathologists, surgeons, radiologists, and stomatologists should be performed to better conduct the diagnostic and optimize treatment and survival of the patient.

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