J Bras Patol Med Lab. 2021; 57: 1-5.

10.5935/1676-2444.20210033

Multicentric osteoblastoma-like: report of a rare case in the craniofacial region

Osteoblastoma-like *multicêntrico: relato de um caso raro na região craniofacial*

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ABSTRACT

The osteoblastoma-like tumor is a rare condition with limited information about its treatment in the current medical literature. The tumor histologically resembles osteoblastoma, although the imaging features are similar to those seen in primary vascular lesions. Due to the uncertainty in the biological behavior of this tumor and because it is an unusual diagnosis, treatment can be aggressive, such as amputation, en bloc resection, and/or chemotherapy. This work reports a rare case of a patient with multicentric osteoblastoma-like in the craniofacial region, treated aggressively with total resection of the lesions.

Key words: osteoblastoma; hemangioendothelioma; angiomatosis; osteoma osteoid; pathology clinical.

RESUMO

O osteoblastoma-like *é uma condição rara, e a literatura médica atual tem informações limitadas sobre seu tratamento. Ele se assemelha histologicamente ao osteoblastoma, embora as características imaginológicas sejam semelhantes às observadas nas lesões vasculares primárias. Devido à incerteza do seu comportamento biológico e por se tratar de um diagnóstico incomum, o tratamento pode ser agressivo, com amputação, ressecção em bloco e/ou quimioterapia. Este trabalho relata um caso raro de osteoblastoma*-like *multicêntrico em região craniofacial, tratado de forma agressiva com ressecção total das lesões.*

Unitermos: *osteoblastoma; hemangioendotelioma; angiomatose; osteoma osteoide; patologia clínica.*

RESUMEN

El tipo osteoblastoma es una afección poco común y la literatura médica actual tiene información limitada sobre su tratamiento. Es histológicamente similar al osteoblastoma, aunque las características de las imágenes son similares a las que se observan en las lesiones vasculares primarias. Por la incertidumbre de su comportamiento biológico y por tratarse de un diagnóstico poco habitual, el tratamiento puede ser agresivo, con amputación, resección en bloque y/o quimioterapia. Este trabajo reporta un caso raro de osteoblastoma multicéntrico en la región craneofacial, tratado de manera agresiva con resección total de las lesiones.

Palabras clave: *osteoblastoma; hemangioendotelioma; angiomatosis; osteoma osteoide; patología clínica.*

First submission on 04/03/20; last submission on 04/05/20; accepted for publication on 04/05/20; published on 07/20/21

INTRODUCTION

Osteoblastoma is a rare benign neoplasm of bone formation. It was first described by Jaffe and Mayer in 1932 as an osteoblastic tumor that forms osteoid tissue. In 1956, Jaffe and Lichtenstein determined the final terminology: benign osteoblastoma[1]. This neoplasm comprises approximately 3.5% of all benign primary bone tumors, and only 1% of bone neoplasms are osteoblastomas[2].

The osteoblastoma-like tumor is an uncommon condition, with few reports in the current medical literature. The tumor is histologically similar to osteoblastoma, but due to the radiographic characteristics of a multifocal osteolytic lesion, predominantly intracortical, it can simulate a primary vascular lesion, such as hemangiomatosis or epithelioid hemangioendothelioma of bone[3]. Due to the uncertainty in the pathological diagnosis and the biological behavior of the lesion, the typical treatment consists of aggressive procedures[4].

The objective of this work is to report a rare case of osteoblastoma-like in the craniofacial region and to correlate it with the world literature. The diagnosis of this neoplasia is a challenge; therefore, it is essential to reconcile the clinical, imaging, and pathological evidence to determine it.

CASE REPORT

A female patient, 30 years old, leucoderma, sought the oncology surgery service to assess facial asymmetry due to an asymptomatic and firm in palpation volumetric increase in the right maxillary region, associated with partial obstruction of the ispilateral nasal fossa, with time of evolution of approximately one year and with overlying skin tissue with no changes and well-defined limits, visible on extraoral examination.

During the intraoral examination, we observed an exuberant mass in the upper right vestibule associated with teeth 13, 14, 15 and 17, with expansion of the cortical bone. Upon palpation, the lesion showed a hard consistency with intraosseous growth.

The computed tomography (CT) (**Figure 1**) revealed an expansive lesion with a heterogeneous attenuation coefficient, causing osteolysis in the anterior margin of the maxilla and in the right anthromaxillary. In addition, we noticed another tumefacient lesion with the same characteristics described in the left maxillary sinus. We also noticed an expansive lesion of mixed components (lytic and blast) in the left frontal sinus.

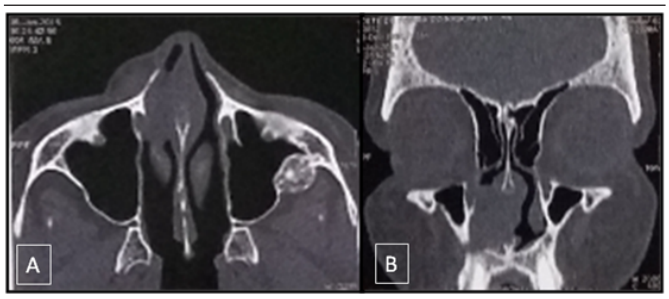

FIGURE 1 – *CT images*

Axial (A) and coronal (B) cross-sectional images showing the maxillary injury. CT: computed tomography.

At magnetic resonance imaging (MRI) (**Figure 2**), four expansive lesions were observed: two in the maxilla, one in the mandible, and one in the frontal sinus; the largest of them in the right maxillary bone, determining anteroinferior obstruction of the ipsilateral nasal cavity, measuring about 3 × 3 × 3.1 cm, with a solid-cystic aspect, with enhancement of the solid component, compromising the palatine and alveolar process of the maxilla. We observed another 1.7 cm solid expansive lesion, enhanced by gadolinium, medially affecting the left maxillary sinus, as well as the hard palate and the alveolar process of the left maxilla. In the head of the right mandible, we observed a small 1.3 cm solid-cystic lesion, with a slight enhancement by gadolinium; in the frontal sinus, an expansive lesion measuring 3.2 × 3 × 1.4 cm, with hypointense areas in T2, which may correspond to a sclerotic component. There was a slight accumulation of secretions in the left frontal sinus, adjacent to the lesion described above. We did not observe any change in the cerebral parenchyma.

In view of the clinical and imaging findings, the diagnostic hypothesis was metastatic implantation. The patient underwent an incisional biopsy of the lesion in the right maxillary sinus, and the surgical fragment was sent for histopathological examination. In addition, primary neoplasms were screened in other foci, which ruled out the hypothesis of metastatic implantation.

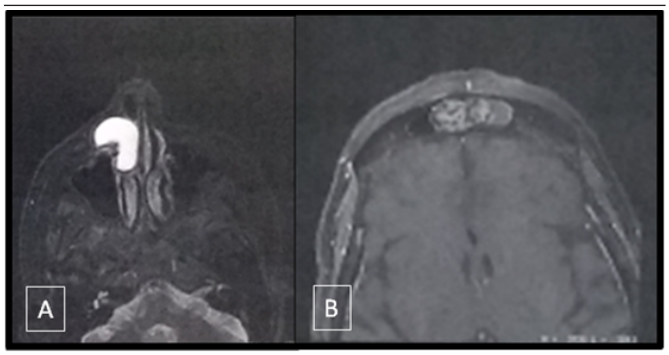

FIGURE 2 – *MRI images*

Axial section showing lesion of the maxillary (A) and frontal (B) sinus. MRI: magnetic resonance imaging.

Histological sections (**Figure 3**) revealed bone lamina covered by collagenized fibrous connective tissue with mild chronic inflammation and hemorrhage, without epithelial lining. We also observed giant cells with fibrous stroma, containing mononuclear cells, areas of hemorrhage, and reactive bone, with a diagnostic hypothesis of central giant-cell granuloma (CGCG).

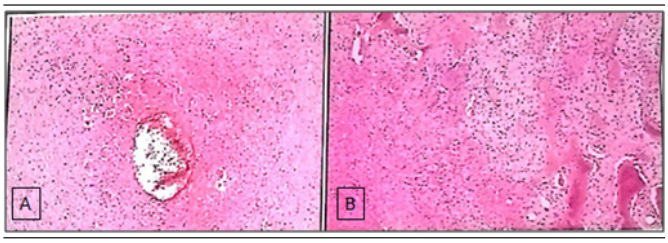

FIGURE 3 – *Microscopic examination of the anterior wall of the right maxillary sinus*
A) hemorrhage foci with giant cells; B) mononuclear cells with reactive bone.

Considering the histopathological aspect associated with the clinical and image findings of the aggressiveness of the lesion, we opted to perform total resection of the lesion in the right maxilla for further anatomopathological analysis. As the CGCG microscopically showed a similar aspect to a giant-cell tumor of hyperparathyroidism, serum levels of parathormone, calcium, and alkaline phosphatase were performed. All values were normal. Thus, we eliminate the hypothesis of GCCG.

We performed the resection of the lesion in the right maxilla using the Weber-Ferguson-Diefenbach approach, which enabled the excision of the right maxilla. This region involved almost the entire lesion, so we maintained the periosteum and a small amount of integument in the area to be resected. Osteotomies were performed using a drill, reciprocating saws, and chisels (**Figure 4**).

Then, the surgical fragment was sent to a pathology laboratory specialized in the musculoskeletal system. The histological sections (**Figure 5**) revealed a lesion consisting of mononucleated

FIGURE 4 – *Intraoperative*

Intraoperative incidence of the lesion and surgical dissection around it through the Weber-Ferguson Diefenbach approach (A) and intact lesion after surgical excision (B).

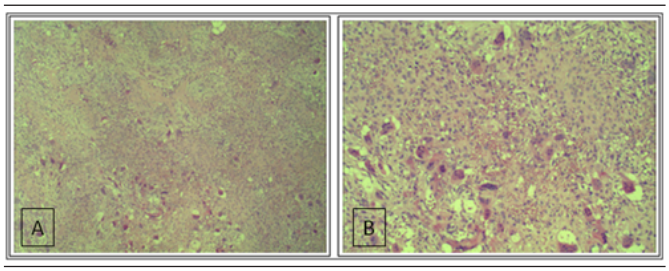

FIGURE 5 – *Microscopic examination of right anthro maxillary tumor*

A) panoramic view; B) higher magnification view.

spindle cells, with multinucleated giant cells, cavitated areas, and hemorrhagic areas infiltrating bone tissue, with free surgical limits in the examined sections. Therefore, these findings were compatible with osteoblastic neoplasia of low aggressiveness, of the osteoblastoma-like type, with secondary reactive and cystic changes.

Since it is an unusual event, whose radiological images also show lesions in other bones of the face, with signs of aggressiveness and multicentricity of the pathological process and histological findings of possible cell atypias, it is recommended to obtain samples of other craniofacial lesions for better clarification of the nature and the real aggressiveness of the disease.

Thus, an excisional biopsy of the lesions of the frontal sinus, left maxilla and right mandible was performed, whose anatomopathological analysis concluded that it was a lesion with an osteoblastic pattern, with occasional cystic cavities, confirming the osteoblastoma-like diagnosis previously proposed.

There were no complications in the surgeries; the patient had a good clinical evolution. Currently, she has been in regular follow-up for a year since the craniofacial surgical approach and shows no signs of recurrence in the operated area.

DISCUSSION

Osteoblastoma is an osteogenic tumor of intermediate grade and of local aggressiveness. It usually manifests as isolated lesions measuring more than 2 cm[5]. However, Fletcher *et al*. (2002)[3] reported multifocal tumors, which were referred to as osteoblastoma-like multifocal lesions. According to the authors, only five cases have been described in the medical literature.

The clinical presentation of an osteoblastoma varies according to the size and location of the lesion[2]. This tumor mainly affects the spine and long bones. The craniomaxillofacial region affects

only 11% of patients, with the mandible being the most common site; involvement of the paranasal sinuses and the maxilla is considered rare[6]. It has a predilection for male adolescents and young adults[7].

The multifocal osteoblastoma-like lesion is an uncommon diagnosis recently described, which should be considered when multiple well-defined lytic osteoblastic lesions are found in imaging studies, especially if the lesions exhibit an intracortical and multifocal pattern[5].

Among the differential diagnoses of multifocal lytic bone lesions, metastatic pathologies, multiple myeloma, and vascular neoplasms must be considered. Although the main differential diagnostic challenge is bone neoplasms, vascular neoplasms are common and can involve bones or soft tissues. Thus, vascular neoplasms such as epithelioid hemangioendothelioma (EHE) or epithelioid hemangioma (EH) are often mistaken for other neoplastic processes because they are radiologically indistinguishable from osteoblastoma-like[3]. For this reason, some authors have classified the lesion as osteoblastomatosis, which has been used in literature since 2007[5] although not yet included in the classification of the World Health Organization (WHO).

Histologically, although vascular endothelial cells in EHE and EH are similar in appearance, EHE differs from EH by the appearance of its vessels, since vascular differentiation in EHE is more primitive than in EH. Osteoblastoma-like, in turn, exhibits a pattern that consists of a stroma with spindle cells woven by bone trabeculae, lined by prominent osteoblasts and occasional osteoclasts. Besides, well-differentiated vascular structures can be seen in the stroma, but these are less visible than those seen in vascular bone tumors[5].

The other pathologies included in the differential diagnosis of osteoblastoma-like are similar to those of conventional osteoblastoma: central giant-cell lesion of hyperparathyroidism, Langerhans cell histiocytosis, multiple myeloma, fibrous dysplasia, lymphoma, and metastatic tumors, which can be easily excluded histologically[5].

Some authors recommend that, due to the uncertainty in the diagnosis of this type of injury, the treatment can be aggressive, such as amputation, en bloc resection and/or chemotherapy[3]. Still, other treatments are cited in the medical literature. Two cases reported by Kyriakos *et al*. (2007)[5] were successful with combined therapy with surgery and chemotherapy, while another case was treated with bisphosphonates. However, in the latter, there was no follow-up[5]. Mait *et al*. (2012)[8] reported successful multimodal treatment, which involved combined curettage and adjuvant cryotherapy for bone injuries. Additionally, they prescribed intravenous bisphosphonates for five years and radiofrequency ablation. After seven years of follow-up, there was no evidence of disease activity.

Recurrence after incomplete resection is not uncommon; presents a rate that varies between 14% and 23%, depending on the study, which reaffirms the importance of complete resection of the lesion[1, 9].

CONCLUSION

Osteoblastoma-like is a rare pathology with an uncommon diagnosis, which can have a silent evolution and variable presentation. Due to its rare presentation, with few cases described in the literature, it can be commonly confused with other neoplastic conditions. This way, the detailed study of the imaging and histopathological profile and available clinical data are essential elements for diagnostic confirmation.

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