

Eosinophilic granuloma: an unusual finding in adults

Granuloma eosinofílico: um achado incomum no adulto

Bruno F. B. Abreu¹; Marcio Luís Duarte²; Murilo G. Grande¹; Newton M. Lima Neto¹; Edimeire F. Ottoni¹

1. Hospital Militar de Área de Campo Grande, Campo Grande, Mato Grosso do Sul, Brazil. 2. Webimagem, São Paulo, São Paulo, Brazil.

ABSTRACT

A 45-year-old man presents with occipital headache for about one month, resistant to analgesic therapy, and palpable nodule at the pain site. At initial evaluation, the nodule was 1.5 cm in diameter without apparent skin alteration. Ultrasonography revealed a well-defined osteolytic lesion and doubtful communication with the dura mater. The study was complemented with computed tomography and contrast-enhanced magnetic resonance, both identifying lytic lesion with soft tissue component and contrast enhancement, without diffusion restriction, guiding the diagnostic hypothesis of eosinophilic granuloma. Surgical treatment with mass excision and anatomopathological evaluation was made, which confirmed the radiological diagnostic hypothesis. After three months of follow-up, the patient remained symptom-free and radiographic control showed no signs of relapse.

Key words: bone neoplasms; tomography X-ray computed; magnetic resonance imaging.

RESUMO

Paciente do sexo masculino, 45 anos de idade, apresenta cefaleia occipital há cerca de um mês, resistente ao tratamento com analgesia, e nódulo palpável no local da dor, medindo 1,5 cm de diâmetro, sem aparente alteração da pele. A ultrassonografia revelou uma lesão osteolítica com margens bem definidas e discutível comunicação com a dura-máter. O estudo foi complementado com tomografia computadorizada (TC) e ressonância magnética (RM) com contraste, ambos os exames identificando lesão lítica com componente de partes moles e realce pelo contraste, sem restrição à difusão, orientando a hipótese diagnóstica de granuloma eosinofílico (GE). Instituiu-se então o tratamento cirúrgico com excisão da massa e avaliação anatomopatológica, que confirmou a hipótese diagnóstica radiológica. Com três meses de seguimento, o paciente estava livre de sintomas, e o controle radiográfico não evidenciava sinais de recidiva.

Unitermos: neoplasias ósseas; tomografia computadorizada por raios X; imagem por ressonância magnética.

RESUMEN

Paciente varón de 45 años presenta cefalea occipital desde hace un mes, refractaria a tratamiento analgésico, y nódulo palpable en el lugar del dolor, que mide 1,5 cm de diámetro, sin alteración visible de la piel. La ecografía reveló una lesión osteolítica de bordes bien delimitados y comunicación dudosa con la duramadre. Se complementó el estudio con tomografía computarizada (TC) e imagen por resonancia magnética (IRM) con medio de contraste, ambos exámenes identificando lesión lítica con componente de partes blandas y realce del contraste, sin restricción a la difusión, planteando la hipótesis diagnóstica de granuloma eosinófilo (GE). Se instauró el tratamiento quirúrgico con escisión de la masa y examen anatomopatológico, que confirmó la hipótesis diagnóstica radiológica. A los tres meses de seguimiento, el paciente estaba libre de síntomas, y el control radiográfico no mostró señales de recidiva.

Palabras clave: neoplasias ósseas; tomografía computarizada por rayos X; imagen por resonancia magnética.

INTRODUCTION

The eosinophilic granuloma (EG) is characterized by abnormal proliferation of histiocytes, localized or multifocal; it mainly affects skull bones, ribs, pelvis, mandible, femur and spine. It is more frequent in children and teenagers⁽¹⁾.

Since 1940, this entity has been described in the literature by several authors. Lichtenstein and Jaffe⁽²⁾ suggested its inclusion in a spectrum that comprises two other conditions with systemic repercussion: Letterer-Siwe and Hand-Schüller-Christian⁽¹⁾ diseases. Those three entities were encompassed in the term histiocytosis X, as their etiology is unknown and in the histological analysis of their characteristic lesions the presence of histiocytes is constant⁽¹⁾.

Letterer-Siwe disease occurs in children aged below 2 years and has, in most cases, a rapidly fatal course⁽³⁾. It is characterized by hepatosplenomegaly, lymphadenopathy, skin rash, fever, anemia, thrombocytopenia and multiple bone lesions⁽³⁾.

Hand-Schüller-Christian disease is manifested later in childhood, or in adults, in some cases, and follows a more indolent and chronic course⁽³⁾. A classical triad of osteolytic skull lesions, exophthalmos, and diabetes insipidus (resulting from skull involvement) are some of its characteristics⁽³⁾.

The possible factors involved in EG are: inflammatory response to an unknown etiological agent, immune system dysfunction and metabolic changes; however, so far, there is no definitive proof^(3,4). The disease is just known not to be inherited or familial, it is not contagious and does not present racial predominance⁽⁵⁾.

We report a case of EG with photographic recording of the diagnostic methods and the surgery, besides the anatomical pathological analysis and literature review.

CASE REPORT

Male 45-year-old patient presented with a 1-month history of occipital headache, resistant to analgesic therapy, and a palpable nodule at the pain site. He reported past history of trauma in the region, with no other complaints. He denied previous diseases and traumas. At the initial evaluation, the patient presented a palpable nodule with 1.5 cm of diameter, without apparent skin alteration. No other relevant findings in the physical examination.

The ultrasonography study revealed an osteolytic lesion with well-defined margins and doubtful communication with dura

mater (**Figure 1**). The study was complemented with a non-contrast computed tomography (CT) and a contrast-enhanced magnetic resonance (MR) imaging (**Figures 2 and 3**), both exams identifying a lytic lesion with soft-tissue components and contrast enhancement, without diffusion restriction, guiding the diagnostic hypothesis of EG.

The patient was then submitted to a bone scintigraphy (**Figure 4**), which verified a calvarial lesion suspicious of being a neoplastic process.

Surgical treatment was provided with excision of the mass (**Figure 5**) and anatomical pathological evaluation (**Figure 6**), which confirmed the radiological diagnostic hypothesis.

After three months of follow-up, the patient was symptom-free, and radiographic control showed no signs of relapse.

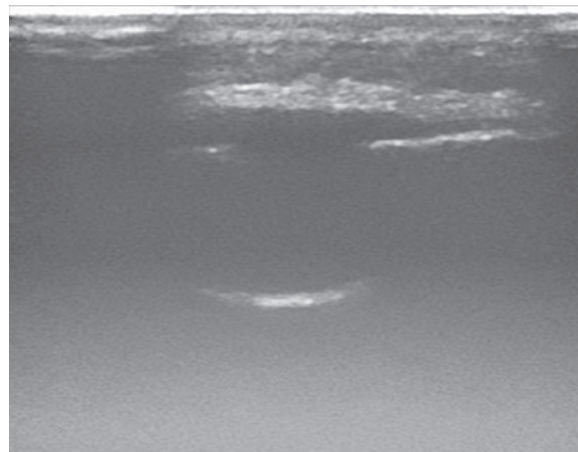


FIGURE 1 – Ultrasonography with high-frequency linear transducer, identifying cortical bone discontinuity and soft-tissue component, with doubtful contact with the diploe

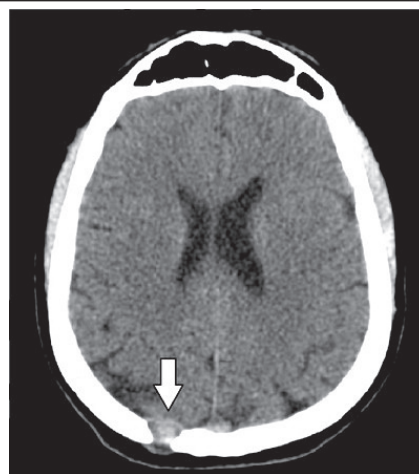


FIGURE 2 – Axial section of non-contrast CT scan showing a lesion with soft-tissue attenuation touching the dura mater (white arrow)

CT: computed tomography.

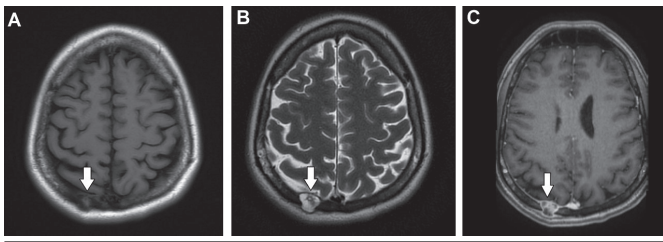


FIGURE 3 – A) axial T1-weighted MR sequence showing a lesion with predominantly low heterogeneous signal, and regular well-defined contours (white arrow); B) axial T2-weighted MR sequence showing a lesion with predominantly high heterogeneous signal, and regular well-defined contours (white arrow); C) axial T1-weighted MR sequence, after administration of contrast, showing a lesion with predominantly low heterogeneous signal, and strong contrast enhancement (white arrow)

MR: magnetic resonance.

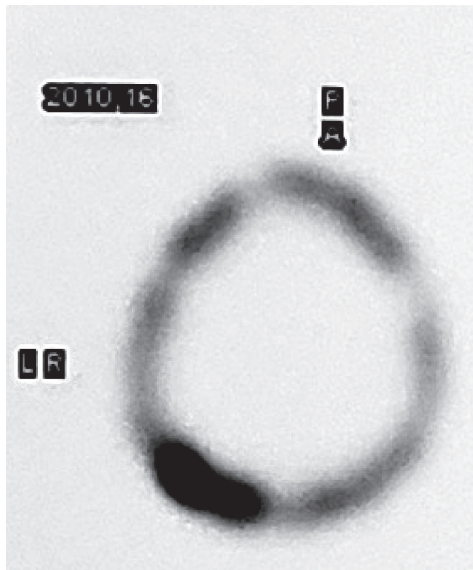


FIGURE 4 – Bone scintigraphy revealing focal radiopharmaceutical uptake of moderate degree in the right parietal region of the calvaria

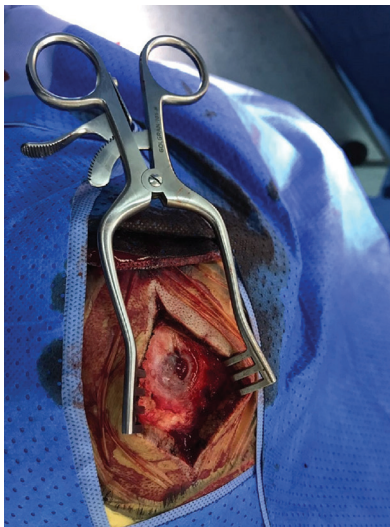


FIGURE 5 – Craniotomy with surgical margin and resection of the lytic bone lesion

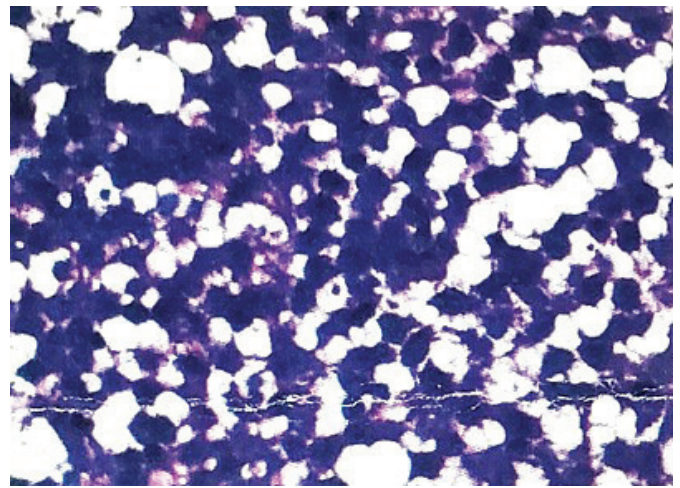


FIGURE 6 – Photomicrograph (HE stain; 200× magnification) of the anatomopathological examination, demonstrating histiocytic proliferation of large cells, with wide cytoplasm and presence of some cells with reniform nuclei and mixed B-cell and T-cell infiltrate of small lymphocytes, without atypias and co-expression of CD68, S100 protein and CD1. The constellation of findings is compatible with Langerhans histiocytosis
HE: hematoxylin and eosin.

DISCUSSION

The osseous location of EG generally exhibits few symptoms and runs a benign course⁽⁶⁾. Therefore, the therapeutic approach is, as a rule, conservative⁽⁶⁾. In fact, after establishing the correct diagnosis, corticosteroid injections or the simple surveillance seem good conducts⁽⁶⁾.

EG is a disease described as the most benign form of histiocytosis X, characterized by infiltration of histiocytes, eosinophils, macrophages, and lymphocytes in the medullary cavity of the bone, forming osteolytic lesions that can be localized or multifocal; mainly affecting skull bones, ribs, pelvis, mandible, femur and spine. It is more frequent in children and teenagers⁽¹⁾. It represents 50%-60% of the cases of Langerhans cell histiocytosis and affects more commonly patients aged 5 to 15 years, but can occur at any age⁽⁷⁾.

The skull is the most common site of EG⁽⁸⁾. The calvaria is more affected than the base of the skull⁽⁹⁾. The parietal bone is the most affected, followed by the frontal bone⁽⁸⁾. Medical literature shows involvement of the temporal bone in 18%-61% of the cases⁽⁵⁾.

Symptoms of the cranial EG vary depending on the evolution of the disease⁽¹⁰⁾. Initially, when the tumor develops intradiploically, it is asymptomatic⁽¹⁰⁾. Pain is general the presenting symptom, beginning as the tumor destroys the outer table, the diploe and the inner table of the calvaria, when it finally invades the dura mater⁽¹⁰⁾. In some patients, pain and localized swelling are the

most common complaints, what is in accordance with reports by other authors^(10, 11). Imaging methods are very important to help establish diagnosis and prescribe treatment^(9, 10). At a plain skull radiograph, EG presents as a well-defined punched out osteolytic lesion^(9, 10).

CT shows an osteolytic lesion, with intracranial extension and involvement⁽¹²⁾. MR defines the extension of bone involvement and of the adjacent soft tissues⁽¹³⁾. The lesions display low signal on T1-weighted image, high signal on T2-weighted image and show enhancement after administration of gadolinium⁽¹³⁾.

Differential diagnosis must be made with osteomyelitis, tuberculosis, syphilis, bone cyst, giant-cell tumor, multiple myeloma, metastatic neuroblastoma or lymphoma, and cystic fibrous osteitis^(12, 14). When located in the temporal bone, the differential diagnosis includes cholesteatoma, chronic mastoiditis, rhabdomyosarcoma, and metastases⁽¹¹⁾.

The treatment of EG depends on factors such as location, number of lesions, and patient's age^(9, 14). In case of a single circumscribed lesion, curettage with complete excision has been the most adequate procedure^(9, 14). This method is quite efficient in small children, because it avoids the use of complementary radiation therapy, with its known complications⁽¹⁰⁾. In cases of multiple or recurrent lesions, chemotherapy is indicated, as well as systemic or intralesional corticosteroids⁽¹⁰⁾.

CONCLUSION

This case illustrates a typical presentation of an uncommon disease in adults. It must, therefore, be included in the differential diagnosis of lytic lesions of the skull. Besides, it highlights the correct methodology of diagnostic and therapeutic procedures.

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CORRESPONDING AUTHOR

Bruno Fernandes Barros Brehme de Abreu  0000-0003-4882-8299
e-mail: bruno_brehme@hotmail.com



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