

Primary cutaneous large B-cell lymphoma of atypical presentation - Case report

Linfoma cutâneo primário de grandes células B de apresentação atípica -
Relato de caso

Antônio René Diógenes de Sousa¹
Edmar Fernandes de Araujo Filho³
Weline Lucena Landim Miranda³

Igor Santos Costa²
Natália Braga Hortêncio Jucá³

Abstract: Primary cutaneous lymphomas are defined as lymphocytic neoplasias that present themselves clinically in the skin without extracutaneous disease at diagnosis and up to 6 months after it. The authors report the case of an elderly male patient, with a three-month history of papules in the axilla which evolved into painful ulceration. Examination found deep ulcer with irregular borders, infiltrates, in the right axilla. Physical and additional examinations did not evidence disease at distance. Histopathology revealed dense and diffuse dermic sample infiltrate of atypical lymphocytes. Immunohistochemistry shows expression of CD20 and bcl-2 antigens, with negative CD10, configuring diagnosis of cutaneous large B-cell lymphoma. In this type of cutaneous lymphoma, primary cutaneous manifestation is rare, the incidence in men is lower and it is most commonly located in the lower limbs.

Keywords: Immunohistochemistry; Lymphoma, Large B-Cell, Diffuse; Skin ulcer

Resumo: Linfomas cutâneos primários são definidos como neoplasias linfocíticas que se apresentam clinicamente na pele sem doença extracutânea no momento do diagnóstico e até por 6 meses após. Os autores relatam o caso de um paciente masculino, idoso, com história de pápulas em axila, há 3 meses, que evoluíram para ulceração. Ao exame, úlcera profunda de bordos irregulares, infiltrados, em axila direita. Exames físico e complementares não evidenciaram doença à distância. O histopatológico mostra infiltrado dérmico denso e difuso de linfócitos atípicos. A imuno-histoquímica evidencia expressão de antígenos CD20 e bcl-2, com CD10 negativo, configurando diagnóstico de linfoma cutâneo difuso de grandes células B. Neste tipo de linfoma, é rara a manifestação cutânea primária, assim como a incidência é menor em homens e a localização, mais comum em membros inferiores.

Palavras-chave: Imunoistoquímica; Linfoma difuso de grandes células B; Úlcera cutânea

INTRODUCTION

Lymphomas are neoplasms of the lymphoreticular system and may originate from three lineages of lymphoid cells: B, T and NK (natural killer). They may originate from lymph nodes or from extranodal sites and include Hodgkin and non Hodgkin lymphomas being the last one the most common¹

Primary cutaneous lymphomas are defined as lymphoid neoplasms that present themselves clinically on the skin and that do not have extra

cutaneous disease when the diagnosis is made or even after 6 months of it. They show considerable variation in its clinical, histological, immunophenotypic presentation and prognosis. Annual incidence of primary cutaneous lymphomas in the USA is 1/100.00 inhabitants, being 75% of T-cells, with predominance of MF and its variants. They are more common in men, after 45 years of age.

Primary cutaneous lymphomas of B cells (CLBC) are less frequent than lymphomas of T cells.

Received on 03.12.2009.

Approved by the Advisory Board and accepted for publication on 11.05.10.

* Work carried out at the Cancer Institute of Ceará (ICC) – Fortaleza (CE), Brazil.

Conflict of interest: None / *Conflito de interesse: Nenhum*

Financial funding: None / *Suporte financeiro: Nenhum*

¹ Dermatology Residency – Coordinator of the Dermatology Residency at Dona Libânia Dermatology Center (CDERM) – Dermatology Professor at the State University of Ceará (UECE) – Dermatology Professor at the University of Fortaleza (UNIFOR) – Fortaleza (CE), Brazil.

² Medical doctor. Residency in Pathology at the University of São Paulo (USP) – Pathologist at Dona Libânia Patology Center (CDERM) – Fortaleza (CE), Brazil.

³ Dermatology residency at Dona Libânia Dermatology Center (CDERM) – Fortaleza (CE), Brazil

They represent approximately 20-25% of all primary cutaneous lymphomas. From a dermatological point of view they are characterized by few lesions, in general showing nodules or infiltrations of relatively fast growth, that are different from the T lymphomas and have no itching. From a histopathological view they are in general monomorphic (large or small cell), and the infiltrate is separated from the epidermis by a collagen band (Grenz zone). Rare cases show epidermotropism and are confused with Mycoises fungoides³. Primary B-cells lymphomas have a better prognosis than the secondary ones. Neoplastic B cells express markers CD19, CD20 and CD79a. In the immunocytomas and plasmocytomas the neoplastic infiltrate and the cells are CD20 negative.⁴

Primary B-cell cutaneous lymphomas are classified in 5 types, according to the WHO-EORTC classification: marginal zone primary cutaneous B-cell lymphoma; centrollicular primary cutaneous lymphoma, diffuse large B-cell primary cutaneous lymphoma, leg type, large B-cell primary cutaneous lymphoma, others that include the intravascular large B-cell primary cutaneous lymphoma.⁵ The diagnosis is made by the clinical-pathologic correlation, immunohistochemical findings and molecular biology that complement the investigation.

The Diffuse large B-cell primary cutaneous lymphoma corresponds to approximately 5-10% of the B-cell cutaneous lymphomas, affecting more frequently the lower limbs although they may also affect other areas. It is predominant in the elderly people, mainly females. Lesions can be single or multiple and even grouped. It has been observed a five-year survival rate in 36 to 100% of the cases. The expression of bcl-2, presence of multiple lesions and involvement of the two upper limbs lead to a worse prognosis.⁶

CASE REPORT

Male patient, aged 80, mullato, farmer, coming from Quixeramobim-CE, sought medical assistance in December, 2008 complaining of skin tumor. The patient reported first the appearance of small papules in the right axilla three months before that evolved into tumor and that a month before had evolved into painful ulceration. He also reported loss of 9 kg, hyporexia and adynamia. He denied rash or fever.

Clinical examination showed ulcers of irregular edges, reddish-violet infiltrates reaching cutaneous and sub-cutaneous plans of the right axilla and measuring 9,2cm x 7,7cm (Figure 1). Neither lymphadenopathy nor hepatosplenomegaly were detected.

Exams were held: hemoglobin=10,3; platelets=163.000; VHS=78; Urea=24;



FIGURE 1: Image of deep ulcer in the right axilla, with irregular erythematous infiltrate borders

Creatinine=0,7; TGO=26; TGP=32; abdominal ultrasound and chest CT without alterations.

The histopathological exam of the edge of the lesion showed dense diffuse dermal infiltrate of monotonous cells medium sized, scant cytoplasm and large and hyperchromatic nuclei, with distortions of the natural architecture of the skin and destruction of the cutaneous appendages and suggesting lymphoproliferative disease. (Figure 2). Complementary immunohistochemistry revealed diffuse expression of CD20 antigens (Figure 3) and bcl-2, with negativity for CD10, configuring diagnosis of diffuse large B-cell primary cutaneous lymphoma.

DISCUSSION

In the last decade, studies have proved an

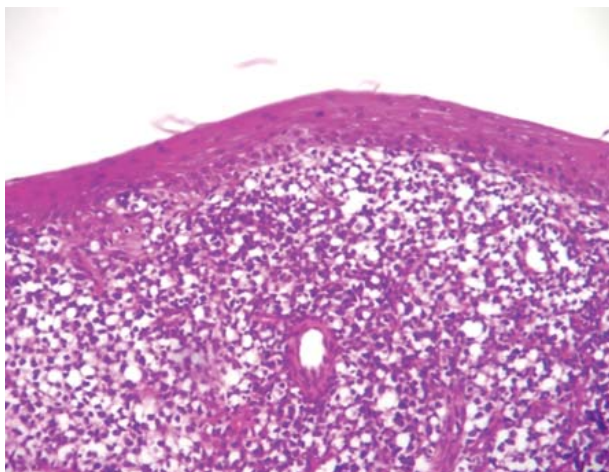


FIGURE 2: Image of the histopathological, HE enlarged 100x, showing dense and diffuse dermal infiltrate of monomorphic cells of average size

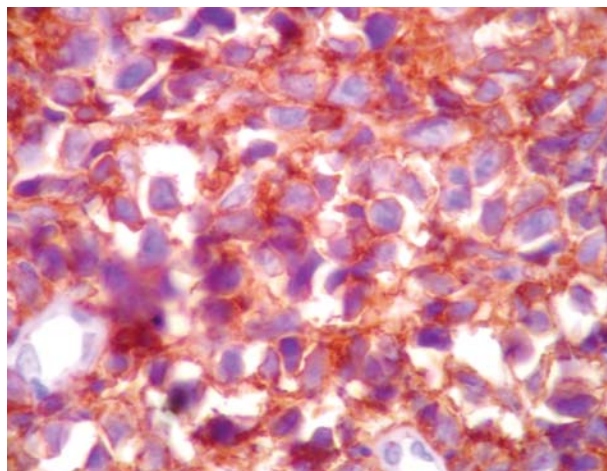


FIGURE 3: Image of immunohistochemistry, showing positivity for CD20 antigens, enlarged 400x for antos eritaumento 400x

increase in the incidence of non-Hodgkin lymphomas in relation to the other subtypes of blood-dermal neoplasms.

Cutaneous lymphomas may be primary or secondary. Primary lymphoma is the one which is exclusively cutaneous when diagnosed or even up to six months after it had been diagnosed, as evidenced in the case through physical and complementary imaging exams. Despite being identical in morphological appearance they have different clinical

behaviors. The primary one presents a more indolent history when compared to the secondary one, with good prognosis.⁷ It presents local recurrence in up to 68% of the cases and rare extracutaneous dissemination, with an average rate of five-year survival varying from 89 to 96 %.⁸

The presentation of the variant Diffuse large B-cell non-Hodgkin lymphoma is rare as a primary cutaneous manifestation. In this case, epidemiological factors of interest were added to this fact, to quote: sex (lower incidence in men), atypical location (more commonly found in the lower limbs) and local aggressiveness.

Histopathological diagnosis is essential for the correct characterization of the tumor and adequate treatment. The immuno-histochemical exam is fundamental and serves, initially, to differentiate between cutaneous neoplasms that are morphologically similar to lymphoma, such as the Merkel cell carcinoma and the small cells melanoma⁹, and, later, to define the subtype of lymphoma.. It also serves as an important tool to exclude systemic lymphomas with secondary skin involvement.

The suggested therapeutic proposal was chemotherapy using cycles of CHOP (cyclophosphamide, hidroxidoxorubicina, Oncovin and prednisone) every three weeks, adding a total of 8 cycles. □

REFERENCES

1. Fletcher CDM. Diagnostic Histopathology of Tumors. Vol. 2. Churchill Livingstone, 2007. p.1140-1142.
2. Moricz CZM, Sanches Jr JA. Processos linfoproliferativos da pele. Parte: 1 - Linfomas cutâneos de células B. An Bras Dermatol. 2005;80:461-71.
3. Chiang S, DiCauda DJ, Valdez R, Swanson DL. Cutaneous B-cell lymphoma with histologic features of mycosis fungoides. J Am Acad Dermatol. 2010;62:320-3.
4. Azulay RD, Azulay DR, Azulay-Abulafia L. Dermatologia. 5. ed. Rio de Janeiro: Guanabara koogan; 2008. p.604, 611.
5. Willemze R, Jaffe ES, Burg G, Cerroni L, Berti E, Swerdlow SH, et al. WHO-EORTC classification for cutaneous lymphomas. Blood. 2005;105:3768-85.
6. Sampaio SAP, Rivitti EA. Dermatologia. 3. ed. São Paulo: Artes Médicas; 2007. p. 1268.
7. Nunes MG, Pierro APSM, Coutinho MFV, Morais JCO, Carneiro SCS, Azulay DR. Linfoma cutâneo de células B: relato de caso. An Bras Dermatol. 2004;79:715-20.
8. Sterry W, Jahn S. Other systemic lymphomas with skin infiltrations. In: Fitzpatrick T, Eisen AZ, Wolff K, Freedberg IM, Austin KF, editors. Fitzpatrick's: Dermatology in General Medicine. 5th ed. New York: McGraw-Hill; 1999. p.1250-8.
9. Bahrami A, Truong LD, Ro JY. Undifferentiated tumor: true identity by immunohistochemistry. Arch Pathol Lab Med. 2008;132:326-48.

MAILING ADDRESS / ENDEREÇO PARA CORRESPONDÊNCIA:

Antônio René Diógenes de Sousa
Rua Pedro I, 1033, Centro
60035-101 Fortaleza (CE) - Brasil
E-mail: renediogenes@terra.com.br

How to cite this article/Como citar este artigo: Sousa ARD, Costa IS, Araújo Filho EF, Jucá NBH, Miranda WLL. Primary cutaneous large B-cell lymphoma of atypical presentation: a case report. 2011;86(3):549-51.