Case for diagnosis*

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CASE REPORT

A 81-year-old female patient had a 2-year history of painful lesions on both legs. She complained of asthenia, decreased appetite and changes in the sleep-wake cycle. Physical examination evidenced multiple brown-violet nodular lesions on both legs. Some of the lesions were ulcerated, with drainage of purulent and serosanguineous secretion (Figure 1). There was non-pitting edema extending throughout the length of the legs. During hospitalization, atrial flutter and decompensated heart failure were diagnosed. Histopathological examination showed extensive dermal infiltration by malignant neoplasm of large cells with voluminous, irregular nuclei, prominent nucleoli and scant cytoplasm (Figure 2). Immunohistochemistry showed strong expression of CD20, negativity for CD3, and high levels (>80%) of Ki67 index of cell proliferation (Figure 3). Computed tomography of the abdomen and thorax showed no abnormalities.

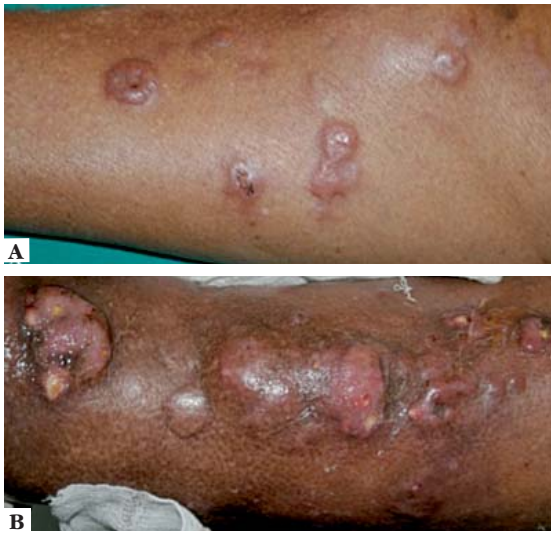

FIGURE 1: A. Violet-colored erythematous nodular lesions with central ulceration on the anteromedial region of the right leg; **B.** More abundant, similar lesions on the anterior region of the left leg

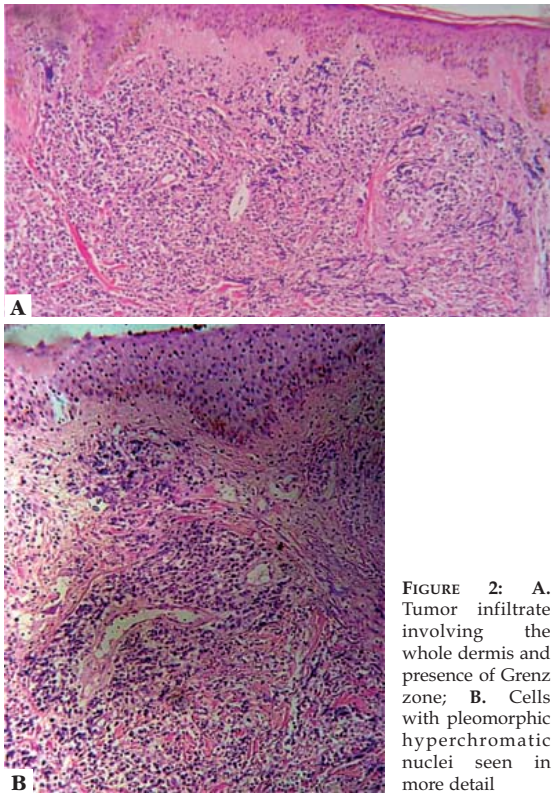

FIGURE 2: A. Tumor infiltrate involving the whole dermis and presence of Grenz zone; **B.** Cells with pleomorphic hyperchromatic nuclei seen in more detail

DISCUSSION

The diagnosis of primary cutaneous large B-cell lymphoma, leg type (PCLBCL-LT) was confirmed by clinical, histological and immunohistochemistry findings. These lymphomas are malignancies of the lymphoreticular system that originate from three lymphoid cells lines: B, T and NK (natural killer). Primary cutaneous lymphomas are neoplasias restricted to the skin. According to the WHO-EORTC classification (2008), cutaneous B-cell lymphomas are classified into three types: marginal zone primary cutaneous B-cell lymphoma; centrofolicular primary cutaneous lym-

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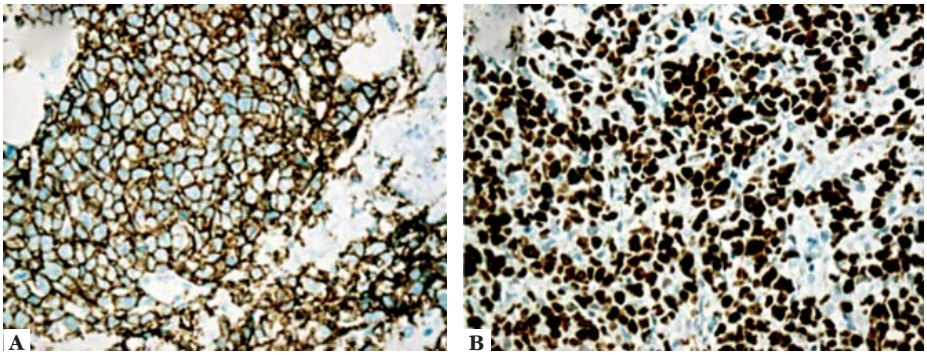

Figure 3: A. Positive immunohistochemistry for CD20; **B.** High levels (>80%) of Ki67 index of cell proliferation

phoma and PCLBCL-LT. The PCLBCL-LT corresponds to 4% of all cutaneous lymphomas and to 20% of all primary B-cell lymphomas.[1,2] Clinically, it presents itself as single or multiple violet-colored erythematous tumors on one or both legs. Affection of the lymph node, bone marrow and central nervous system may rarely occur in PCLBCL-LT. The five-year survival is 50%.[3-5] Histological examination of tumor lesions shows a diffuse, monotonous cellular infiltrate of immunoblasts, without epidermotropism. The presence of the normal collagen band in the superficial dermis, separating the epidermis from the dermal lymphocytic infiltrate (Grenz zone) is a common finding in cutaneous B-cell lymphomas. Neoplastic cells are positive for CD20, CD79a and negative for CD3. There is strong expression of BCL-2, BCL-6 and MUM-1. Anti-Ki67 antibodies are also an important aid in the diagnosis of B-cell lymphoproliferative processes.[1,6,7] Currently, the R-CHOP chemotherapy regimen (rituximab, cyclophosphamide, doxorubicin, vincristine and prednisone) is the most widely accepted treatment for PCLBCL-LT.[2,8,9] The alternative COP chemotherapy regimen (cyclophosphamide, vincristine and prednisone) was chosen due to the presence of heart failure (which contraindicates the use of doxorubicin), the delay in the release of the immunohistochemistry examination results (required for the release of rituximab), and, especially, the rapid progression of the disease. There was a partial regression of cutaneous lesions and reduction of cutaneous infiltration after the first cycle of chemotherapy. However, the patient acquired nosocomial pneumonia, progressed to septic shock and died four months after the diagnosis of PCLBCL-LT. ❑

Abstract: We report the case of a 81-year-old female patient who had a two-year history of violet-colored erythematous tumors on both legs. Histopathological and immunohistochemical examinations confirmed the diagnosis of primary cutaneous large B-cell lymphoma, leg type. This rare, cutaneous lymphoma affects predominantly elderly females. Clinically, patients present with tumoral lesions on one or both legs (worst prognosis). Diagnosis is based on clinical, histopathological and immunohistochemical findings. The strong expression of BCL2, BCL6, MUM-1 and CD20, and the positivity for Ki67 antigen confirm the diagnosis. R-CHOP chemotherapy regimen (rituximab, cyclophosphamide, doxorubicin, vincristine and prednisone) is the most widely accepted treatment.
Keywords: B-cell lymphomas; Legs; Combination chemotherapy

REFERENCES

1. 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.
2. Willemze R. Primary cutaneous lymphomas. Ann Oncol. 2011;22:72-5.
3. Hristov AC. Primay cutaneous diffuse large B-cell lymphoma, leg type: diagnostic considerations. Arch Pathol Lab Med. 2012;136:876-81.
4. Bessell EM, Humber CE, O'Connor S, English JS, Perkins W, Dickinson PD, et al. Primary cutaneous B-cell lymphoma in Nottinghamshire U.K.: prognosis of subtypes defined in the WHO-EORTC classification. Br J Dermatol. 2012;167:1118-23.
5. Cwynarski K, Gldstone A. Non-Hodgkin lymphoma. In: Hoffbrand AV, Catovsky D, Tuddenham, Edward GD, editors. Postgraduate Haematology. 6th ed. Oxford, UK.: Willey-Blackwell; 2011. p.655-85.
6. Moricz CZM, Sanches Jr JA. Part 1 - Primary cutaneous B-cell lymphomas. An Bras Dermatol. 2005;80:461-71.
7. Grange F, Beylot-Barry M, Courville P, Maubec E, Bagot M, Vergier B, et al. Primary Cutaneous Diffuse Large B-Cell Lymphoma, Leg Type. Clinicopathologic Features and Prognostic Analysis in 60 Cases. Arch Dermatol. 2007 ;143:1144-50.
8. Fisher RI, Gaynor ER, Dahlberg S, Oken MM, Grogan TM, Mize EM, et al. Comparison of a standard regimen (CHOP) with three intensive chemotherapy regimens for advanced non-Hodgkin's lymphoma. N Engl J Med. 1993;328:1002-6.
9. Sehn LH, Donaldson J, Chhanabhai M, Fitzgerald C, Gill K, Klasa R, et al. Introduction of combined CHOP plus rituximab therapy dramatically improved outcome of diffuse large B-cell lymphoma in British Columbia. J Clin Oncol. 2005;23:5027-33.

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