



## Cutaneous diffuse large B-cell lymphoma, leg type - a typical case

*Linfoma cutâneo difuso de grandes células B, tipo perna: sobre um caso típico*

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### ABSTRACT

Cutaneous diffuse large B-cell lymphoma, leg type, is a rare, aggressive, and poorly prognostic neoplasm. It corresponds to 10-20% of cutaneous B-cell lymphomas and mainly affects the lower limbs of older women. We report the case of an 81-year-old woman with painful, fast-growing nodules and tumors in her left leg. Histopathological and immunohistochemical findings, associated with the absence of extra-cutaneous involvement during staging, concluded the diagnosis of cutaneous diffuse large B-cell lymphoma, leg type. The rarity of this lymphoma, its typical clinic and epidemiology, and the excellent response to treatment motivated this report.

**Keywords:** Immunohistochemistry; Diffuse; Lymphoma non Hodgkin; Lymphoma large B-cell, Neoplasms; Therapeutics

### RESUMO

*O linfoma cutâneo difuso de grandes células B, tipo perna, compreende uma neoplasia rara, agressiva e de mau prognóstico. Corresponde a 10-20% dos linfomas cutâneos de células B e afeta principalmente membros inferiores de mulheres idosas. Relatamos o caso de mulher de 81 anos, com nódulos e tumorações dolorosos, de crescimento rápido em perna esquerda. Os achados histopatológicos e a imuno-histoquímica, associados à ausência de comprometimento extracutâneo no estadiamento, concluíram o diagnóstico de linfoma cutâneo difuso de grandes células B, tipo perna. A raridade, a clínica e a epidemiologia típicas e a excelente resposta ao tratamento motivaram este relato.*

**Palavras-chave:** Imuno-histoquímica; Linfoma difuso de grandes células B; Linfoma não Hodgkin; Neoplasias; Terapêutica

## Case report

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**INTRODUCTION**

Primary cutaneous lymphomas are defined as a heterogeneous group of lymphoproliferative malignancies that affect the skin, with no evidence of extracutaneous involvement at the time of diagnosis.<sup>1</sup>

Cutaneous B-cell lymphomas (CBCL) account for 20% to 25% of all primary cutaneous lymphomas and are more common in women.<sup>1,2</sup>

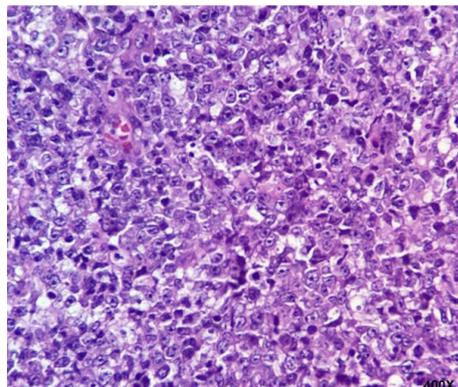
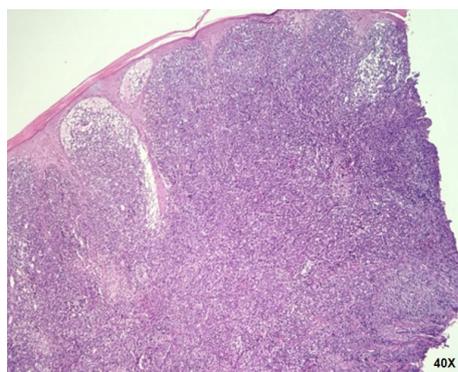
According to clinical, histopathological, immunohistochemical, and molecular criteria, CBCLs are classified by the World Health Organization–European Organization for Research and Treatment of Cancer (WHO-EORTC) – 2018, as centrofollicular and diffuse large B cells, leg type; the latter represents only 4% of all cutaneous lymphomas.<sup>1</sup>

The rarity of this type of lymphoma, its typical clinic and epidemiology, and the excellent response obtained to treatment motivated this report.

**CASE REPORT**

A Caucasian, 81-year-old woman came to the Dermatology Outpatient Clinic complaining of fast-growing “lumps” on her left leg, with one-month evolution. Dermatological examination revealed vegetating nodules and tumors, with an erythematous–yellow fibrinoid surface and the presence of blackened crusts (Figure 1). There was no lymph node enlargement or visceromegaly. The lesions were painful, and the patient denied any systemic symptoms. She mentioned previous trauma as a triggering factor. Also, she had peripheral venous insufficiency and type II *diabetes mellitus* as comorbidities. The hypotheses put

forward were squamous cell carcinoma, amelanotic melanoma, cutaneous metastases, and cutaneous lymphoma. Incisional biopsies were performed in two points. The histopathological study showed ulcerated epidermis with areas of necrosis and the presence of dense and diffuse lymphocytic infiltrate consisting of large cells in the superficial and deep dermis, sometimes with nuclei with irregular contours and abundant mitotic figures (Figure 2). The immunohistochemical panel was positive for Bcl-2, CD20, and MUM1 (Figure 3) and negative for CD3, CD10, and cyclin D1 (Figure 4), concluding a B-cell non-



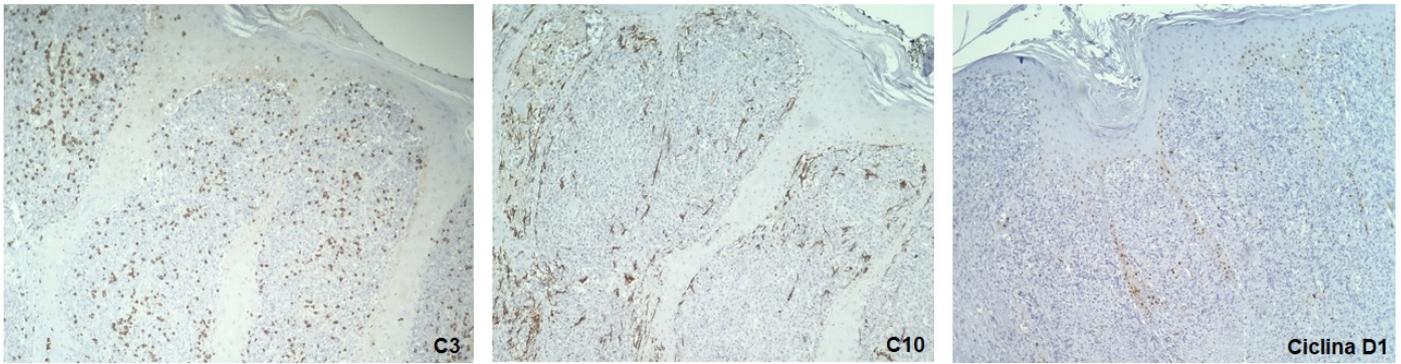
**FIGURE 2:** Superficial and deep dermis with presence of dense diffuse lymphocytic infiltrate of large cells, sometimes with irregularly contoured nuclei, and abundant mitotic figures (Hematoxylin & Eosin 40x, 400x)



**FIGURE 3:** Immunohistochemistry: MUM-1 positive



**FIGURE 1:** A: Left leg: vegetating nodules and tumors, sometimes covered by a blackened crust. B: Greater detail: medial face

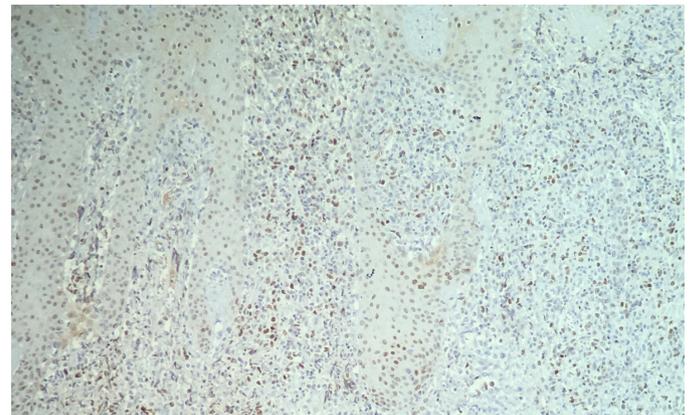


**FIGURE 4:** Immunohistochemistry: CD3, CD10, and cyclin D1 negative

Hodgkin lymphoma diagnosis. Bcl-6 was not performed. The proliferative index by Ki-67 was 90%, and C-MYC was positive in about 30% of the cells (Figure 5).

The patient was referred to the Hebe Camargo Cancer-Fighting Network, where she was staged and diagnosed with diffuse cutaneous large B-cell lymphoma, leg type, T2bN0M0 (Table 1).

We instituted therapy with R-CHOP regimen (rituximab, cyclophosphamide, doxorubicin, vincristine, and prednisone) and scheduled surgical intervention, which was not performed due to the excellent evolution of the patient (Figure 6). Currently, the patient has only dyschromic cicatricial areas and is under joint follow-up with Dermatology and Oncology.



**FIGURE 5:** Immunohistochemistry: C-MYC positive in 30% of cells

**DISCUSSION**

Diffuse cutaneous large B-cell lymphoma, leg type, comprises a rare, aggressive neoplasm with a poor prognosis.<sup>1,3</sup>

It corresponds to 10-20% of CBCLs and has a 5-year survival rate of 50%.<sup>1,3,4</sup> It mainly affects women of advanced age (70-82 years).<sup>1,3,5</sup> Clinically, it manifests as nodular, infiltrative, single, or multiple lesions, which may be located in different sites (lower limbs are the most common). It also presents a rapid pro-

gression, agreeing with what was observed in this case. Phlogistic signs and pruritus may be present.<sup>1,3</sup>

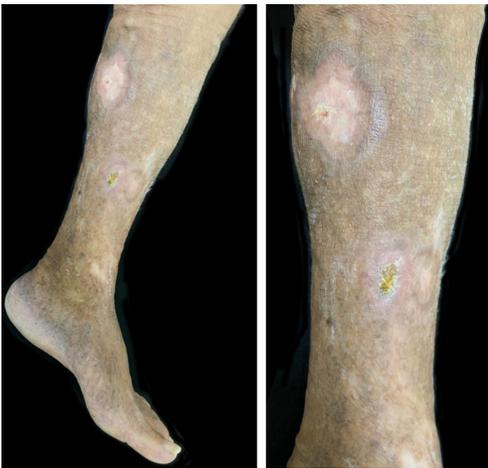
The development of extracutaneous lesions is common, especially among patients who have involvement in the lower limbs, with bone marrow, lymph nodes, and central nervous system is the most affected sites.<sup>1,5,6</sup>

**TABLE 1: Tumor classification according to the EORTC/ISCL for primary cutaneous lymphomas other than mycosis fungoides and Sézary syndrome**

Tumor	
<b>T1</b>	Only a skin lesion:
	T1a. The lesion size is up to 5 cm in diameter T1b. The lesion size is greater than 5 cm in diameter
<b>T2</b>	Two or more skin lesions. They can be in a single region of the body or nearby region:
	T2a. All skin lesions can be placed within a 15 cm diameter circle
	T2b. The circle needed to round all lesions is between 15 and 30 cm in diameter T2c. The circle needed to round all skin lesions is greater than 30 cm in diameter
<b>T3</b>	Skin lesions in different regions of the body or at least three different regions:
	T3a. Many injuries involving two regions of the body, distant from each other T3b. Many injuries involving three or more regions of the body.

Source: American Cancer Society

EORTC/ISCL: European Organization for Research and Treatment of Cancer/ International Society for Cutaneous Lymphomas



**FIGURE 6:**  
Scar dyschromic areas after chemotherapy with R-CHOP regimen

The diagnosis is made by clinical, histopathological, and immunohistochemical findings.<sup>1,2</sup>

In histopathology, it is characterized by a dense infiltrate of large cells in the dermis and subcutaneous tissue, separated from the epidermis by a narrow band of collagen called the grenz zone (not observed in the present case).<sup>2,5</sup> Often, this infiltrate tends to be more intense in the deep dermis, called a “bottom-heavy pattern”.<sup>2</sup>

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