

LETTER – CLINICAL

Intravascular large B-cell lymphoma: presentation with generalized dendritic-type telangiectasias. A diagnostic challenge[☆]



Dear Editor,

Intravascular lymphoma is an extremely rare disease with diverse and atypical clinical manifestations, making early diagnosis challenging and impacting the prognosis.¹

We present the case of a 70-year-old female patient with a history of cervical cancer treated with surgery, brachytherapy, and radiotherapy, who presented with a two-year history of progressive edema in both lower extremities, associated with multiple generalized telangiectasias (Fig. 1), significant weight loss, episodes of fever up to 38 °C, and night sweats. Laboratory tests revealed hemoglobin of

8.7 g/dL, MCV 94 fL, leukocytes 3,240 μ L with neutrophil predominance, platelets 144,000 μ L, ESR 48 mm/h, uric acid 6.2 mg/dL, LDH 662 U/L (RI: 125–220), ALP 246 U/L (RI: 40–150), GGT 135 U/L (RI: 9–36), normal transaminases and bilirubin levels, and vitamin B12 at 1360 pg/mL, with normal renal function, calcium, and phosphorus levels. β 2-microglobulin was elevated (3.74 mg/L). Protein electrophoresis showed a monoclonal peak in the gamma region, with an IgM kappa monoclonal component and lambda light chain on immunofixation. Computed tomography (CT) of the chest, abdomen, and pelvis showed no significant pathological findings. Doppler ultrasound of the lower extremities revealed subcutaneous edema. Bone marrow biopsy was normal. A skin biopsy was performed, identifying large atypical lymphoid cells with prominent nucleoli within small vessels (Fig. 2). Immunohistochemistry was positive for CD20, PAX-5, MUM-1, and Bcl-2 and negative for CD3, Bcl-6, CD10, CD56, CD30, CD34, and cytokeratins (Fig. 3). A diagnosis of intravascular large B-cell lymphoma (IVLBCL)

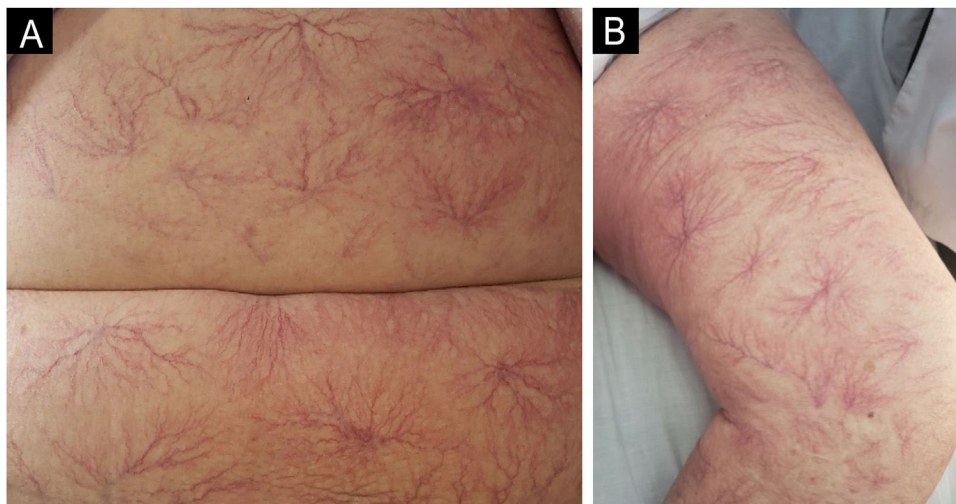


Figure 1 Clinical images. Multiple generalized dendritic type telangiectasias on the abdomen (A) and thigh (B).

[☆] Study conducted at the Hospital Dr. Sótero del Río, Puente Alto, Santiago, Chile.

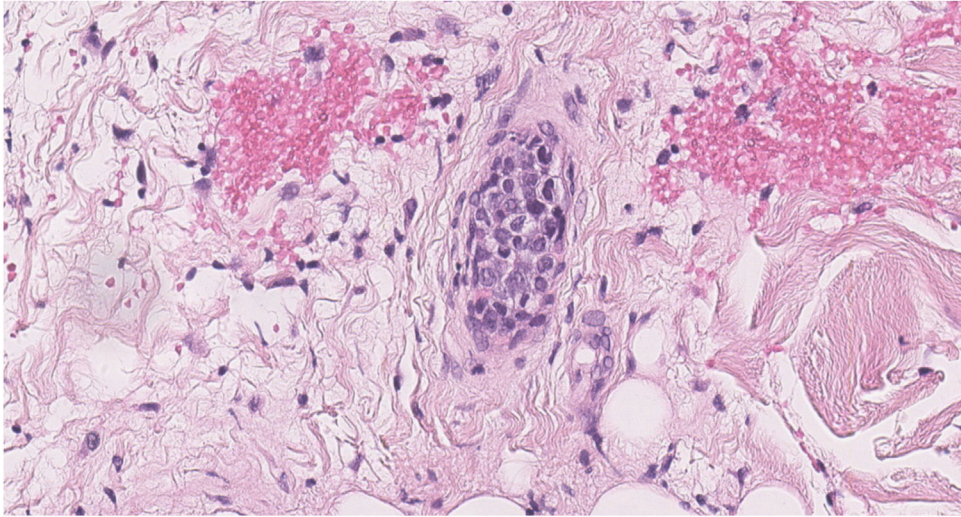


Figure 2 Atypical intravascular lymphoid population with monomorphic, uniformly enlarged cells with prominent nucleoli (Hematoxylin & eosin, $\times 400$).

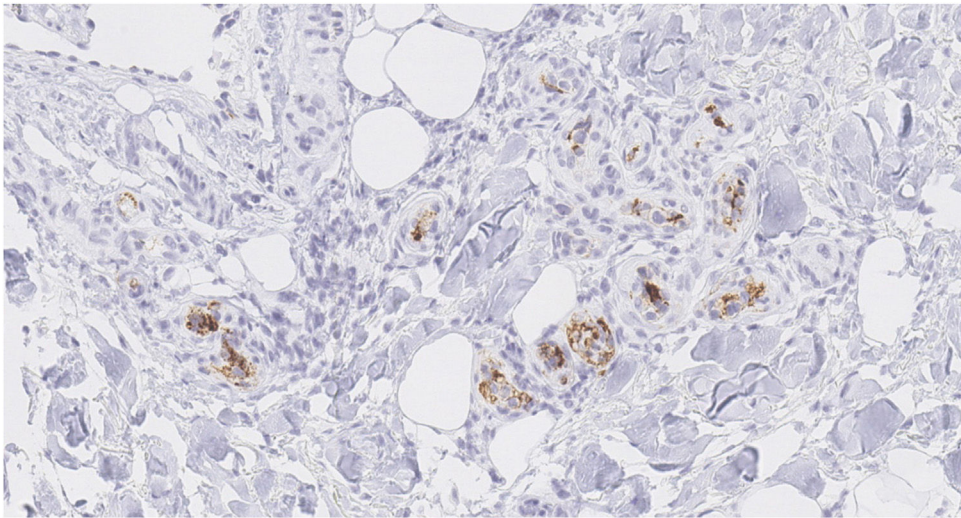


Figure 3 Immunohistochemistry showing hyperchromatic cells showing positive immunohistochemical staining with the CD20 B-cell marker ($\times 400$).

was established. The patient was initiated on the R-CHOP chemotherapy regimen (rituximab, cyclophosphamide, doxorubicin, vincristine, and prednisone). A marked reduction in cutaneous telangiectasias – estimated at approximately 60% – was noted following the first cycle. Intrathecal methotrexate was also administered due to the presence of pathological mononuclear cells in moderate quantity in the cerebrospinal fluid, which subsequently cleared. The patient remains under multidisciplinary follow-up by hematology and dermatology, exhibiting a favorable clinical response after six cycles of R-CHOP, with only sparse residual telangiectasias noted on the chest.

Intravascular lymphoma is a rare type of extranodal non-Hodgkin lymphoma (<1% of cutaneous lymphomas), characterized by the proliferation of neoplastic lymphocytes in the lumen of small vessels, described in 1959 as “endotheliomatosis and reticuloendotheliosis”.² An inci-

dence of 0.09 per 1 million inhabitants was reported in the USA between 2000–2013.³ The average age of presentation is 70-years, with no gender differences.^{2,3} 85% originate from B-cells (IVLBCL) and 15% from NK or T-cells.⁴ Two clinical variants are described: the “Asian” variant, associated with multi-organ involvement and hemophagocytosis, and the “Western” variant, associated with skin lesions and central nervous system involvement.^{5,6} Clinical presentation is variable, including the presence of B-symptoms (55%–85%) without associated lymphadenopathy, neurological symptoms (39%–76%), and skin lesions (17%–39%) with erythematous-violet indurated plaques and nodules that simulate panniculitis, associated with edema. Its presentation as dendritic-type telangiectasias is very rare, with only 24 reported cases.^{1–6}

Due to its variable and nonspecific symptoms, the diagnosis tends to be delayed, leading to a poor prognosis,

with a median survival of 46.1% at 5-years.³ In cases of timely treatment with R-CHOP, the survival rate is 66% at 24-months, making it a potentially curable disease with chemotherapy.^{3,5} If there is CNS involvement, methotrexate or cytarabine is recommended.⁵ In certain cases, an autologous stem cell transplant should be considered.^{2,5}

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Authors' contributions

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Research data availability

Does not apply.

Conflicts of interest

None declared.

Editor


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Appendix A. Supplementary data

Supplementary material related to this article can be found, in the online version, at doi:<https://doi.org/10.1016/j.abd.2025.501221>.

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