Cutaneous Polyarteritis Nodosa as the Inaugural Manifestation of Hairy Cell Leukemia

Poliarterite Nodosa Cutânea como Manifestação Inaugural de Tricoleucemia

Keywords: Leukemia, Hairy Cell; Panniculitis; Polyarteritis Nodosa; Skin Diseases, Vascular
Palavras-chave: Dermatopatias Vasculares; Paniculite; Poliarterite Nodosa; Tricoleucemia

Panniculitides are inflammatory dermatoses of the subcutaneous tissue resulting from multiple etiologies, including infections, autoimmune diseases, drugs, and, less commonly, hematologic malignancies, particularly lymphoproliferative disorders.[1] Among these, hairy cell leukemia (HCL) is a rare B-cell lymphoproliferative disorder, typically presenting with splenomegaly and cytopenias.[2] While rare, cutaneous manifestations can occur at any stage of the disease.[3]

We report the case of a 45-year-old previously healthy man who presented with painful erythematous nodules on the lower limbs (Fig. 1A), fever (38.2°C), fatigue, and myalgias for four days. He also reported an unintentional weight loss of 5 kg over the previous four months. Physical examination revealed hepatosplenomegaly.

Laboratory tests showed neutropenia (0.99 × 10^9/L), monocytopenia (0.00 × 10^9/L), thrombocytopenia (50 × 10^9/L), and elevated C-reactive protein (8.11 mg/dL; normal < 0.5 mg/dL). The peripheral blood smear revealed atypical lymphoid cells with abundant pale cytoplasm and irregular circumferential cytoplasmic projections, suggestive of hairy cells. Computed tomography demonstrated homogeneous splenomegaly (20 cm). The skin biopsy showed a medium-caliber arteriole in the deep dermis with fibrinoid necrosis, luminal occlusion, and neutrophilic infiltrate with leukocytoclasia and erythrocyte extravasation, associated with adjacent neutrophilic panniculitis (Fig. 1B). No atypical hematolymphoid cells or microorganisms were identified, which could support cutaneous polyarteritis nodosa. Septic vasculitis was considered but excluded based on negative microbiological studies and absence of organisms on histochemical stains. Bone marrow biopsy demonstrated extensive infiltration by monoclonal B lymphocytes (CD5-, CD10-, CD19+, CD20+, CD25+, CD103+, CD123+, CD200+) with the BRAF V600E mutation, which confirmed the diagnosis of HCL. Treatment with cladribine and adjunctive oral prednisolone for the vasculitic cutaneous manifestations resulted in progressive resolution of the skin lesions, cytopenias, and splenomegaly.

Although polyarteritis nodosa has been described in association with HCL, it remains rare and is usually reported after the hematologic diagnosis, especially after splenectomy or infection. Although the underlying mechanism remains unclear, immune dysregulation and cytokine-mediated vascular inflammation may contribute to the association between HCL and vasculitis.[3]

In our patient, vasculitic panniculitis represented the inaugural presentation. Importantly, the coexistence of constitutional symptoms, including fever and weight loss, together with splenomegaly and skin lesions, provided key clinical clues suggestive of an underlying systemic disorder.[2,4] This case highlights the importance of recognizing such warning signs when evaluating panniculitis or vasculitis of unclear etiology. Early skin biopsy and multidisciplinary investigation may allow prompt identification of occult hematologic malignancies, reinforcing the role of the skin as a potential window to systemic disease.

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The authors declare that no AI tools were used during the preparation of this work.

AUTHOR CONTRIBUTIONS

FM, SF: Study design, data acquisition and analysis, manuscript writing and critical review.
GAS, PV: Data acquisition, manuscript writing and critical review.
JA: Study design, data acquisition and analysis, manuscript critical review.
All authors approved the final version to be published.

PROTECTION OF HUMANS AND ANIMALS

The authors declare that the procedures were followed according to the regulations established by the Clinical Research and Ethics Committee and to the Helsinki Declaration of the World Medical Association updated in October 2024.

DATA CONFIDENTIALITY

The authors declare having followed the protocols in use at their working center regarding patients' data publication.

PATIENT CONSENT

Obtained.

CONFLICTS OF INTEREST

The authors have no conflicts of interest to declare.

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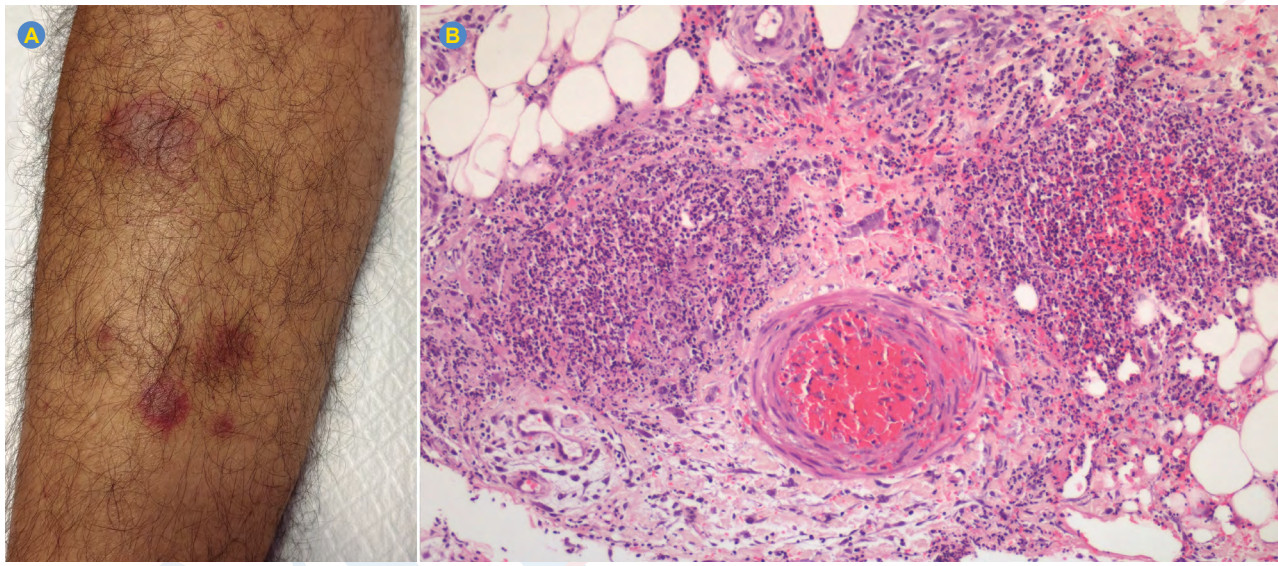

Figure 1 – (A) Physical examination revealing erythematous subcutaneous nodules on the anterior surface of the lower limbs. (B) Haematoxylin & eosin, 100×. Deep dermis with a medium-calibre arteriole showing luminal occlusion with fibrin deposition in the vessel wall, surrounded by a dense neutrophilic infiltrate with leukocytoclasia and marked erythrocyte extravasation. Adjacent mixed panniculitis with neutrophils.

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