Erythroderma and panniculitis as protean manifestations of nodal T-cell lymphomas

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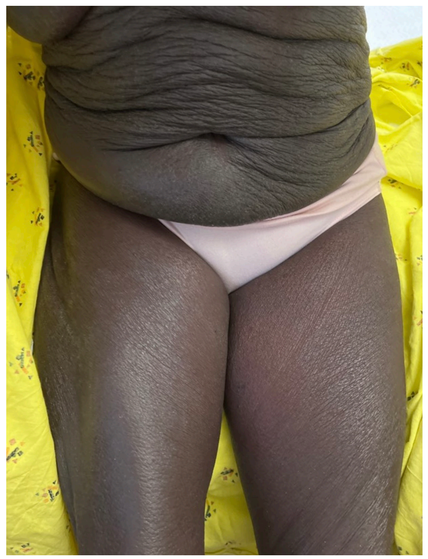
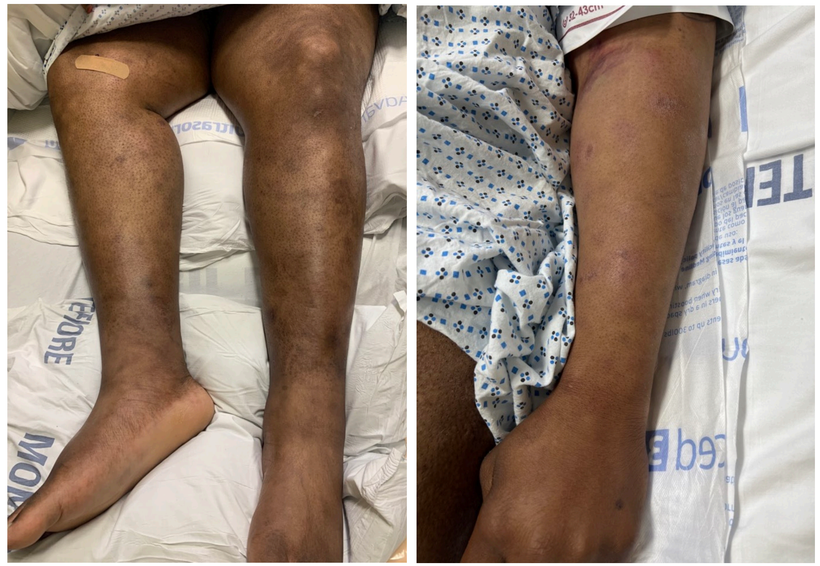
Introduction

- Lymphoproliferative disorders constitute a group of malignant lymphocytic clonal proliferations whose diagnosis remains challenging given their varied clinical presentation and immunophenotype[1]
- Mature T-cell lymphomas (MTCLs) comprise mature neoplastic T-cells that can remain absent in the blood and skin but have significant skin manifestations[2]
- The average age of diagnosis is 65, and men are affected at higher rates than women[3]
- MTCLs are generally aggressive diseases, except for cutaneous T cell lymphomas (CTCLs) such as mycosis fungoides, which are considered more indolent
- MTCLs are a diverse group of cancers due to the proliferation of different subsets of T cells, which secrete distinct cytokines[3]
- Skin manifestations may include ulcers, panniculitis, plaques, nodules, erythroderma, and pruritus[3]
- Herein, we introduce two cases of nodal MTCLs presenting as erythroderma and panniculitis to stress their clinical heterogenicity and the critical role of nodal biopsies for accurate diagnosis

Clinical Images



Case 1:
Widespread erythroderma with prominent xerosis of the back, abdomen, and lower extremities



Case 2:
Ovoid red to violaceous papulonodules, plaques, and hypopigmented patches on the bilateral upper and lower extremities

Clinical Cases

Case 1:

- A 71-year-old woman presented with erythroderma and severe pruritus
- Multiple punch biopsies spanning several months reported purpuric spongiotic dermatitis without atypical lymphocytes
- Laboratory workup was negative for HIV, RPR, and HTLV-1 & 2 antibodies
- Peripheral blood flow cytometry (PBFC) yielded nonspecific findings of heterogeneous T-cells with an increased CD4/CD8 ratio (25:1)
- The axillary and inguinal nodes were easily palpable; an inguinal node excisional biopsy confirmed MTCL

Case 2:

- A 35-year-old man presented with minimally scaly ovoid red to violaceous papulonodules, plaques, and hypopigmented patches on the bilateral upper and lower extremities
- Hospital course complicated by acute-onset multiorgan failure and hemophagocytic lymphohistiocytosis (HLH)
- PBFC was negative
- Incisional and punch biopsies revealed atypical lobular panniculitis consistent with MTCL involving the fat
- Numerous enlarged lymph nodes were palpable; an inguinal node excisional biopsy confirmed MTCL

Conclusions

- Unlike lymphoid B-cell disorders, there are no immunophenotypic signatures for lymphoid T-cell disorders to determine clonality[1]
- This renders flow cytometric analysis challenging and oftentimes insufficient[4]
- Histological changes in the skin can be nonspecific, delaying accurate diagnosis[5]
- A comprehensive evaluation and workup should always include a lymph node exam, with nodal biopsy if pathologically enlarged, irrespective of negative skin biopsies or PBFC results

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