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INTERESTING MEDICAL IMAGE

Primary Cutaneous Gamma/Delta T-cell Lymphoma Simulating Lupus Erythematosus Panniculitis

What do we know about this aggressive mimicker?

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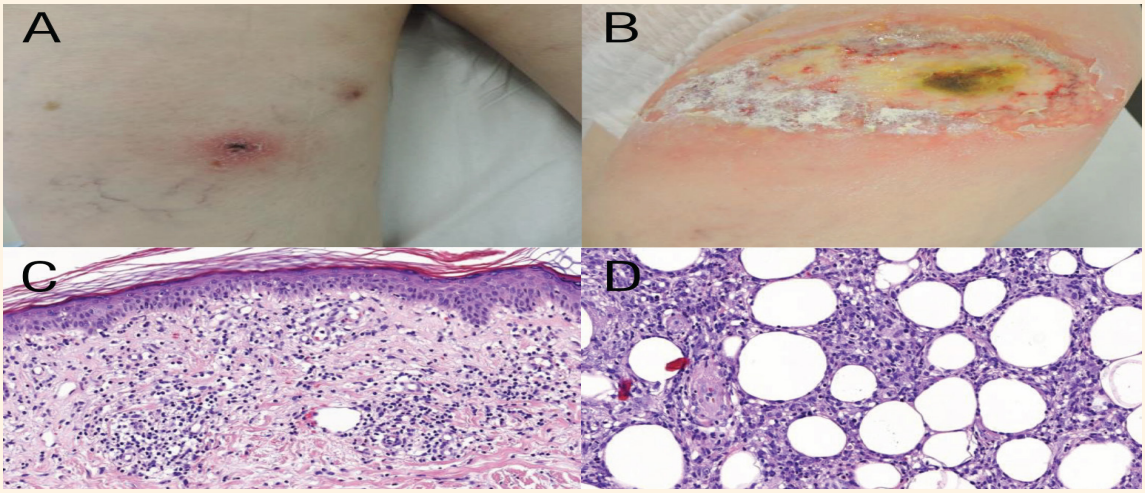

Figure 1: A: Well-delimited erythematous nodule located on the front thigh. **B:** Ulcerated plaque with an erythematous border located on the left thigh. **C:** Haematoxylin-eosin (H&E) staining ×20 magnification showing epidermal and dermal histological appearance, with a perivascular inflammatory lymphocytic infiltrate and vacuolisation of the basal layer. **D:** H&E staining ×40 magnification showing inflammatory infiltrate, mainly lymphocytes, surrounding the subcutaneous fat, phenomenon known as rimming.

A 62-year-old female patient with a 4-month history of asymptomatic erythematous nodules located on both thighs, hips and abdomen [Figures 1A and B]. Some lesions were ulcerated. Histopathological examination revealed an interface dermatitis, dermal mucin deposits, as well as a lobar panniculitis, with focal rimming of adipocytes by lymphocytes [Figures 1C and D]. This was suggestive of lupus erythematosus panniculitis (LEP). Complete blood count (CBC) and antinuclear antibodies were normal and the patient refused treatment. Over a nine-month period new ulceronecrotic plaques developed on the left thigh [Figure 2A]. She became febrile and her overall status worsened.

CBC revealed pancytopenia—haemoglobin 8.2 g/dL (12.0–16.0 g/dL), white blood cells 1,900/mm^3 (4,500–11,000/mm^3) and platelets 90,000/mm^3 (150,000–400,000/mm^3). Abdominal CT-scan revealed splenomegaly but no nodal involvement. Bone marrow biopsy was unremarkable.

Histopathollogy of the left thigh ulcer revealed an atypical T-cell lymphoid proliferation [Figure 2B]. CD2, CD3, CD56, granzyme B and gamma TCR were positive; CD4, CD20, CD8, TCRBF1, EBV-LMP1 and EBER were not detected [Figure 2C]. Patient consent has been obtained for publication purposes.

Comment

Primary cutaneous gamma-delta T-cell lymphoma (PCGD-TCL) was diagnosed. Cyclophosphamide, doxorubicin, vincristine, prednisolone and etoposide (CHOEP) were administered. The patient died 10 days later.

PCGD-TCL is an extremely rare and aggressive entity that accounts for less than 1% of cutaneous lymphomas.[1] Clinical manifestations of PCGD-TCL are variable. The condition most commonly presents as subcutaneous nodules and plaques often involving lower extremities, which tend to ulcerate early in the

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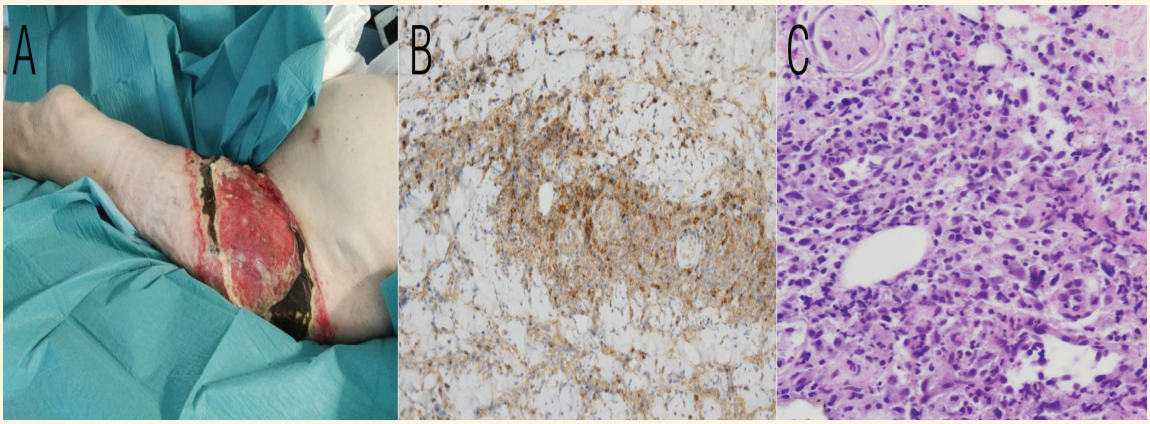

Figure 2: A: Ulcerated and necrotic erythematous plaque located on the left thigh. **B:** Hematoxylin-eosin staining ×40 magnification showing atypical heterogeneous lymphoid infiltrate. **C:** Cytoplasmic staining of TCR gamma ×10 magnification.

disease.[1] Hemophagocytic lymphohistiocytosis may be observed, but the involvement of lymph nodes, spleen or bone marrow is uncommon.[2] Elevated liver enzymes levels and leukopenia are associated with adverse prognosis.[3]

Histologically, PCGD-TCL may be heterogeneous with epidermotropic, dermal and/or subcutaneous infiltrates or combinations of these patterns. Other features such as lichenoid or vacuolar interface reaction, scattered necrotic keratinocytes or dermal mucin deposits have been infrequently described. Histopathological differentiation from LEP may be challenging. Histologic features of LEP such as the presence of lymphoid follicles conforming germinal centers, septal fibrosis, plasma cells and clusters of CD123+ plasmacytoid dendritic cells are rarely observed in PCGD-TCL. PCGD-TCL is characterised by the presence of atypical lymphocytes, angioinvasion and adipocyte rimming by neoplastic cells. However, these findings are not specific enough to diagnose PCGD-TCL.[2,4]

On immunohistochemistry, a gamma-delta T-cell immunophenotype with loss of CD4 and CD8, expression of CD56 and at least one cytotoxic marker supports the diagnosis of PCGD-TCL. TCR gamma clonality is frequently detected.[1]

Furthermore, the differential diagnosis of PCGD-TCL includes entities such as subcutaneous panniculitis-like T-cell lymphoma or mycosis fungoides, as well as erythema nodosum or pancreatic panniculitis.

Patients are usually treated with multiagent chemotherapy (eg. CHOEP) or allogeneic stem cell transplantation. Therapeutic alternatives include bexarotene, phototherapy, radiotherapy, denileukin diftitox and bendamustine.[5] PCGD-TCL responds poorly to conventional treatments, having an average survival of 31 months.[3]

PCGD-TCL needs to be carefully excluded in patients with panniculitis-like lesions unresponsive to therapy. Multiple biopsies may be required to correctly diagnose this disorder. This case raises the question of whether premalignant or early gamma-delta T-cell proliferations exist and can be identified.

AUTHORS' CONTRIBUTION

FJ-TG, J-GC, A-SL and I-AM contributed equally to the design, drafting and critical review of the manuscript. All authors approve the final version of the manuscript.

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