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Focus area: Challenging Cases of Cutaneous Lymphomas
Abstract n 144

If purpuric lesions, think of mycosis fungoides

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INTRODUCTION

Pigmented purpuric dermatoses (PPD) are a group of capillaropathy of undetermined etiology. Its relationship with mycosis fungoides (MF) is still poorly understood. It is known that MF may initially present with purpuric lesions, PPD may precede the diagnosis of MF, or both dermatoses may coexist.

CASE REPORT

A 48-year-old male presented in 2017 a red-brown spot on the right calf treated as a drug eruption, but had progression. He was admitted in Jan 2022 with erythematous brown patches and plaques with desquamation on the legs, buttocks, arms, and palmoplantar region and petechiae on the lower limbs (Fig 1). Tourniquet test was positive. Systemic investigation was negative.

A biopsy was performed with the hypotheses of chronic purpura pigmentosa, purpuric contact dermatitis, and purpuric lymphoproliferative disease. The histopathological examination demonstrated a superficial lymphocytic infiltrate forming a subepidermal band, expanding the papillary dermis, associated with fibroplasia, plasma cells, eosinophils, foci of extravasation of red blood cells and hemosiderosis. Epidermis exhibited focal areas of spongiosis and scattered isolated lymphocytes. Lymphocytes showed expression of CD3, CD5, and predominantly CD4, with partial loss of CD7 (Fig 2).

Purpuric MF was considered the most likely diagnosis. Phototherapy with PUVA was started with a satisfactory response (Fig 3).

Figure 1: Patient presenting red-brown plaques with desquamation on the surface and patechiae.

Figure 2: Superficial lymphocytic band (left image, hematoxylin and eosin), associated with evident dermal hemosiderosis (right image, Perls).

Figure 3: Superficial lymphocytic infiltrate, forming a subepidermal band associated with fibroplasia of the papillary dermis; foci of spongiosis and sparse lymphocytes in the epidermis (left image, hematoxylin and eosin). Immunohistochemical study showing CD3 expression by frequent lymphocytes, located in the dermis, epidermis and infundibulo-isthmic follicular epithelium, outlining a linear arrangement in the latter location (central image), and hypoimmunoexpression of CD7 (right image).

DISCUSSION

Atypical purpura pigmentosa is defined as cases that manifest clinically as PPDs, but present morphological aspects typically associated with MF. The presence of Pautrier's microabscesses, cerebriform lymphocytes, and atypical intraepidermal lymphocytosis suggest the diagnosis of MF. However, PPDs can also present similar findings. Monoclonality in T cell gene rearrangement studies favors MF, but is not enough for the diagnosis.

In this case report, widespread lesions affecting areas typically spared in PPD, like palmoplantar areas, and pruritus for more than 1 year, associated with atypical T cell lymphocytic infiltrate with partial CD7 loss, favors the diagnosis of the purpuric variant of MF. The treatment chosen is considered first-line for cases of MF without systemic involvement.

Figure 4: Patient presenting partial regression of the lesions after 10 sessions of PUVA.

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