

CASE REPORT

Purpuric mycosis fungoides: A rare presentation of a great masquerader

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ABSTRACT

Mycosis fungoides (MF) has many clinical presentations. It typically presents as indolent, progressive, and persistent erythematous patches or plaques with mild scaling and over time can evolve into tumor stage with tumor nodules. More than 50 clinical presentations of MF have been described. Here, we report a case of MF in a 49-year-old male presenting as pigmented purpuric dermatitis (PPD). This is a rare presentation of a condition known to mimic many clinical entities.

INTRODUCTION

Mycosis fungoides (MF), a low-grade lymphoproliferative disorder, is the most common type of cutaneous T-cell lymphoma. Patients usually present with patches, plaques, tumors and/or erythroderma.¹ Occasionally, less typical lesions are encountered which makes establishing the correct diagnosis difficult. What makes diagnosis even more difficult is that this diversity is observed not only in the clinical presentation but also in histopathologic findings. While almost any histological pattern can be seen on biopsy, the most common is a superficial perivascular lymphoid infiltrate in a somewhat lichenoid distribution accompanied by epidermotropic lymphoid cells.² We report a patient who presented with a clinical picture suggesting pigmented purpuric dermatitis but turned out to be one of the rare variants of mycosis fungoides.

CASE REPORT

A 49 year old Kuwaiti male presented with bi-

lateral non-itchy erythematous plaques of two months duration mainly over the buttocks, sacrum, thighs and feet. There was no history of drug intake, varicosities or family history of similar condition. The plaques were red-brown and some of them had annular configuration (Fig. 1). Complete blood count revealed leukocytosis. Bio-



Fig. 1 Red-brown plaques on the thigh. The one at the bottom has an annular configuration reminiscent of purpura annularis telangiectodes of Majocchi.

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chemical profile showed high cholesterol and uric acid levels. Histopathologic examination revealed psoriasiform hyperplasia with dense superficial prevascular and band-like lymphocytic infiltrate along with liquefaction degeneration of the basal layer. Abnormal single and grouped lymphocytes surrounded by a halo were seen in the epidermis (Fig. 2). The abnormal lymphocytes in the dermis had large irregular deeply stained basophilic nuclei. The cells were positive for CD3 and CD4. The patient was diagnosed as pigmented purpuric dermatitis-like mycosis fungoides and treated with PUVA with excellent outcome (Fig. 3 A,B).

DISCUSSION

Mycosis fungoides (MF) is a peripheral non-Hodgkin T-cell lymphoma initially presenting in the skin. In the majority of cases, the disease evolves clinically through three stages - the patch, plaque and tumour stage. Histologically, the disease is typified by the proliferation of small cerebriform lymphocytes showing epidermotropism. Such clinicopathological presentation and course are usually regarded as 'classical' for this lymphoma.³ In fact, however, MF has a plethora of clinicopathological manifestations, some of which differ substantially from the classical Alibert-Bazin disease and are therefore sometimes referred to as 'atypical' forms of MF.^{4,5} More than 50 forms of the so called 'atypical' forms of MF have been described in the literature. One of the rare forms of them is pigmented purpura-like MF. Pigmented purpuric dermatitis (PPD) associated with MF is a rare but documented phenomenon. In fact, the first instance of lichen aureus reported in the American literature was later recognized and reported to have evolved into MF.^{6,7} PPD may be related to MF in three ways: (i) MF can present clinically as

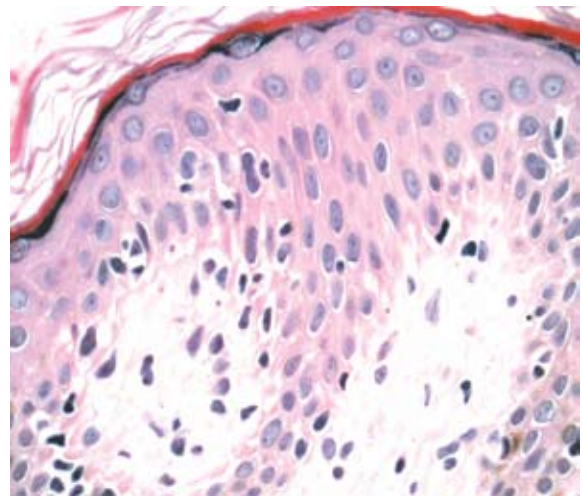


Fig. 2 Abnormal single and grouped lymphocytes surrounded by a halo in the epidermis.



Fig. 3 Red-brown plaques on the buttock before (A) and after (B) PUVA therapy.

PPD;⁸⁻¹¹ (ii) PPD can precede MF;^{6,7,12,13} and (iii) PPD can simulate MF histologically.^{14,15} Our case falls into the first category, namely MF presenting as PPD.

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