

Letter to Editor

## Poikilodermatous Mycosis Fungoides – A Rare and Challenging Diagnosis

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Dear Editor,

Mycosis fungoides (MF), a type of cutaneous T-cell lymphoma, classically presents as slow progressive patches, plaques or tumours. Various lesser-known variants of MF have also been described, such as polymorphic, verrucous, hyperkeratotic, lichenoid, hypo- and hyperpigmented, follicular, granulomatous, psoriasiform, vesicular, bullous, pustular, purpuric and poikilodermatous MF (PMF).<sup>[1,2]</sup> PMF constitutes 1–2% of all cases of MF and presents as localised or generalised patches with mottled hypo- and hyper-pigmentation, telangiectasias and atrophy.<sup>[3]</sup>

Here, we report a case of PMF, highlighting the importance of keeping PMF as a differential while dealing with poikilodermatous lesions in clinical practice.

A 41-year-old man presented with progressive generalised dryness, flaking and mottled discoloration of the skin for the past 5 years, with photosensitivity.

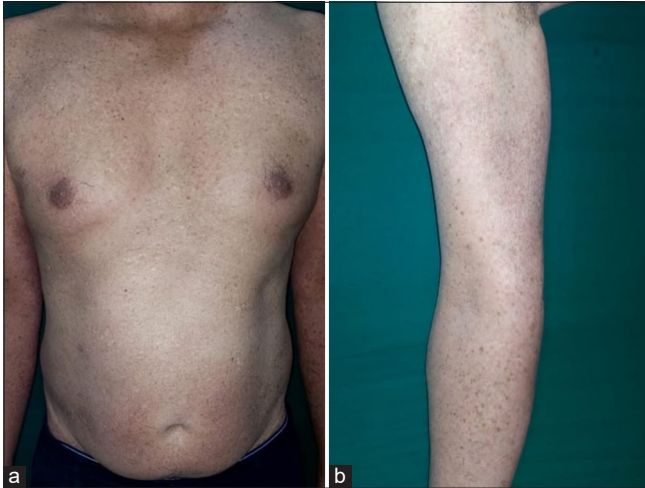
Dermatological examination revealed blanchable erythema, scaling and mottled hypo- and hyperpigmentation, along with a few depigmented macules showing atrophy and telangiectasia seen over the neck, chest, abdomen, back, upper limb and lower limbs [Figure 1]. Multiple, discrete, horizontal groups of inguinal lymph nodes were palpable bilaterally, the largest measuring 1.5 × 1.5 cm. All relevant biochemical and haematological tests showed no abnormality. Histopathological examination showed a thinned-out epidermis with effaced rete ridges; epidermotropism, with large epidermal lymphocytes having irregular nuclei with occasional Pautrier abscess. Nodular and interstitial infiltration by lymphocytes in the dermis and pigment incontinence was seen as shown in the Figure 1. Immunohistochemistry revealed lymphocytes showing co-expression of CD4 and CD8, with predominant CD4 cells [Figure 2]. CD3+ lymphoid cells in the dermis showed diffuse strong positivity. Excisional biopsy of the inguinal lymph node showed features of dermatopathic lymphadenopathy. Bone marrow aspirate and biopsy were well within the normal findings. F-18 fluorodeoxyglucose-positron emission tomography/computed tomography scan showed no visceral involvement.

A final diagnosis of PMF in T4N0M0B0–IIIA stage was made. The patient was initiated on topical high-potent corticosteroids, oral methotrexate (0.25 mg/kg/week) and psoralen ultraviolet A (PUVA). Since the patient developed a phototoxic reaction following PUVA, it was switched to narrow band ultraviolet B (NB UVB). The patient has been under regular follow-up for 1 year and reports considerable improvement and exhibits no signs of progression.

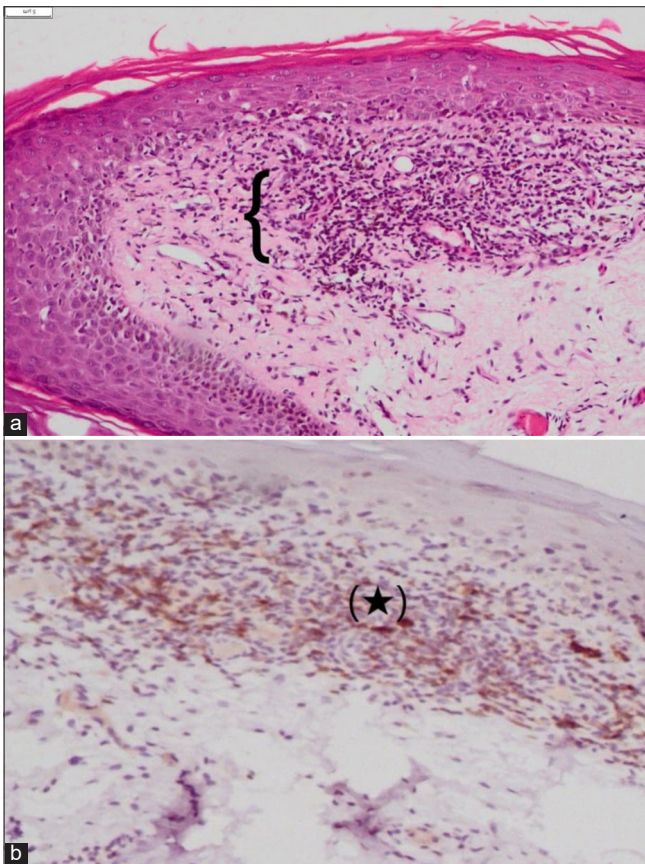
PMF presents as localised or generalised patches with erythema, scaling, hyper- and hypopigmentation with telangiectasia and atrophy with a predilection for trunk and body folds.

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**Figure 1:** Poikiloderma over the (a) Trunk and (b) Upper limb.



**Figure 2:** (a) Haematoxylin and eosin-stained section showing band-like lymphocytic infiltrate (curly bracket) with epidermotropism under x100 magnification, (b) CD4-positive neoplastic T lymphocytes (starred) on immunohistochemistry.

A male preponderance, a propensity to affect younger age and a long duration of symptoms before presentation have been documented in PMF compared to classical MF. This

can be attributed to the atypical clinical presentation, which delays the diagnosis.<sup>[4,5]</sup>

The histological features of PMF may resemble those of classical patch/plaque stage MF, by showing features such as epidermotropism and Pautrier microabscess. In addition, these cases may also show epidermal atrophy, keratinocyte apoptosis, basal hydropic degeneration, interface dermatitis, pigment incontinence, telangiectatic vessels and rarely granulomas.

The therapeutic options are skin-directed (SDT) and systemic therapy, the choice of which depends largely on the stage of the disease. Various skin-directed therapies are topical corticosteroids, PUVA, NB-UVB, extracorporeal photopheresis, bexarotene, carmustine, mechlorethamine, UVA1, radiation and excimer laser. Topical corticosteroids were found to be ineffective, while phototherapy produced a favourable result in PMF. For SDT-resistant cases, systemic retinoids (acitretin, isotretinoin and bexarotene) can be used. Denileukin difitox, alemtuzumab (anti-CD52 antibodies), interferon- $\gamma$  and histone deacetylase inhibitors have been tried for visceral/lymph node involvement. Brentuximab vedotin has been successfully used for PMF with CD30 large-cell transformation.

The overall prognosis and treatment response of PMF are better than classical MF due to the sparse atypical lymphocytic infiltrate in PMF. The diagnosis of PMF may be delayed due to its atypical presentation and histopathological resemblance to several other benign dermatological conditions. In such cases, careful correlation between clinical symptoms and pathological findings, along with regular follow-up and repeat histopathology, is crucial.

**Ethical approval:** Institutional Review Board approval is not required.

**Declaration of patient consent:** The authors certify that they have obtained all appropriate patient consent forms. In the form, the patients have given their consent for their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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