"Missed" Diagnosis of Mycosis Fungoides: A case report

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Key Clinical Message

Mycosis fungoides (MF), a subtype of primary cutaneous T cell lymphoma**,** is often misdiagnosed at early stages due to non-specific clinical findings. Patients are at high risk for developing secondary malignancies.

MF should be suspected in patients with underlying malignancies and patients with MF should be screened for secondary malignancies.

Introduction

Mycosis fungoides (MF), although uncommon, is the most common subtype of primary cutaneous T cell lymphoma that occurs in middle aged and elderly adults.[1] MF has an indolent course, presents as erythematous scaly patches or plaques, and may progress to generalised erythroderma or cutaneous tumours. MF is often misdiagnosed at early stages due to non-specific clinical and pathological findings, and prognosis depends on the type and extent of skin involvement and extra-cutaneous invasion.[2] Patients with MF are at high risk for developing secondary malignancies, including hematological malignancies.[3,4] We, hereby report a case of Mycosis fungoides misdiagnosed and associated with underlying diffuse large B cell lymphoma.

Case Presentation

A 50 year old male presented with multiple itchy, ill-defined, scaly, erythematous-violaceous patches and plaques over generalized body surface for 7 years (Figure 1 a, b, c). Initially, skin lesions appeared bilaterally over the thighs and buttocks, for which he visited a medical centre and was prescribed anti-leprosy medication for 1 year. He was also treated with topical steroids on and off for chronic dermatitis with minimal improvement. He had an abdominal mass excised 2 years back and histology reports were suggestive of diffuse large B cell lymphoma, so he received R-CHOP regimen for 6 courses. Notably, the cutaneous lesions significantly improved while on chemotherapy. However, the skin lesions flared up after 6 months of stopping the chemotherapy.

Methods

Routine blood investigations revealed normal findings. Skin biopsy showed epidermotropism, Pautrier's microabscesses and perivascular lymphocytic infiltrates (Figure 2 a, b). Immunohistochemistry revealed intraepidermal and perivascular atypical cells which were immunopositive for CD2, CD3, CD4, CD5 with Ki-67 proliferation index of 20% and negative for CD20. Bone marrow aspiration and bone marrow biopsy performed were normal. CT scan reports of the neck, chest, abdomen and pelvis were normal. Based on clinical, histopathological and immunohistochemistry findings, the patient was diagnosed as case of Mycosis fungoides, a subtype of primary cutaneous T cell Non-Hodgkin Lymphoma.

Results

As he was diagnosed in the early stage (stage 1B) according to the modified tumour-node-metastasis-blood (TNMB) classification, was started on low dose methotrexate and is under regular follow-up.

Discussion

MF is characterized by malignant proliferation of T cells with epidermotropism in the skin.[1] The diagnosis of MF is often delayed by many years from the initial appearance of skin lesions due to its indolent course and diagnostic difficulties as in our patient. In the early stages of disease, it can mimic common skin conditions like chronic eczema, psoriasis and atopic dermatitis.[1,2]

The factors predisposing for higher rates of certain malignancy in MF is not known, however an inherent potential of disease itself or therapy utilized for treatment could be the potential cause.[3]Viral infections, genetic factors, and environmental exposures implicated in carcinogenesis might be the common mechanism linking the onset of MF with increased risk for a second malignancy.[4] Furthermore, altered T-cell activation in MF might contribute to develop secondary lymphoid malignancy, as it is evident that a subset of highly immunosuppressive regulatory T cells emerging from the infiltrating malignant T cells increases the risk of severe infections and malignancy in MF as in our case.[3,4]

Diagnosis is difficult especially in the early stages, but it is made through clinical examination and is confirmed by a skin biopsy, immunohistochemistry and staged appropriately. Histologically, early MF are characterized by epidermotropism, atypical lymphocytes with cerebriform nuclei, Pautrier's microabscesses

and basal alignment of neoplastic lymphocytes as seen in our case. In immunohistochemistry, MF cells are positive for CD2, CD3, CD4, CD5 and negative for CD20 as reported in our case.[3,5] Management is based on disease stage and includes skin-directed therapies, systemic therapies and allogenic stem cell transplantation.[1,2]

MF is a clinical diagnosis; however, is easily misdiagnosed at an early stage due to its nonspecific clinical features and lack of awareness, warranting a high index of clinical suspicion. MF should be suspected in patients with underlying malignancies and patients with MF are at high risk for developing secondary malignancies which should be screened for and the other way around.

Abbreviations and Acronyms

CD: Cluster of Differentiation

MF: Mycosis fungoides

R-CHOP: Rituximab- Cyclophosphamide, Doxorubicin, Vincristine, Prednisone

TNMB: Tumour-Node-Metastasis-Blood

Authors Contribution Statement

Prof. Dr. Mahesh Mathur: Conceptualization; Formal analysis; Resources; Supervision; Validation; Visualization; Writing-original draft

Dr. Neha Thakur: Conceptualization; Formal analysis; Resources; Supervision; Validation; Visualization; Writing-original draft.

Dr. Sandhya Regmi: Conceptualization; Formal analysis; Resources; Supervision; Validation; Visualization; Writing-original draft.

Dr. Supriya Paudel: Formal analysis; Resources; Supervision; Visualization; Writing-original draft; Writing- review and editing

Dr. Sambidha Karki: Data curation; Investigation; Resources; Visualization; Writing- review and editing.

Dr. Nabita Bhattarai : Data curation; Investigation; Resources; Visualization; Writing - review and editing.

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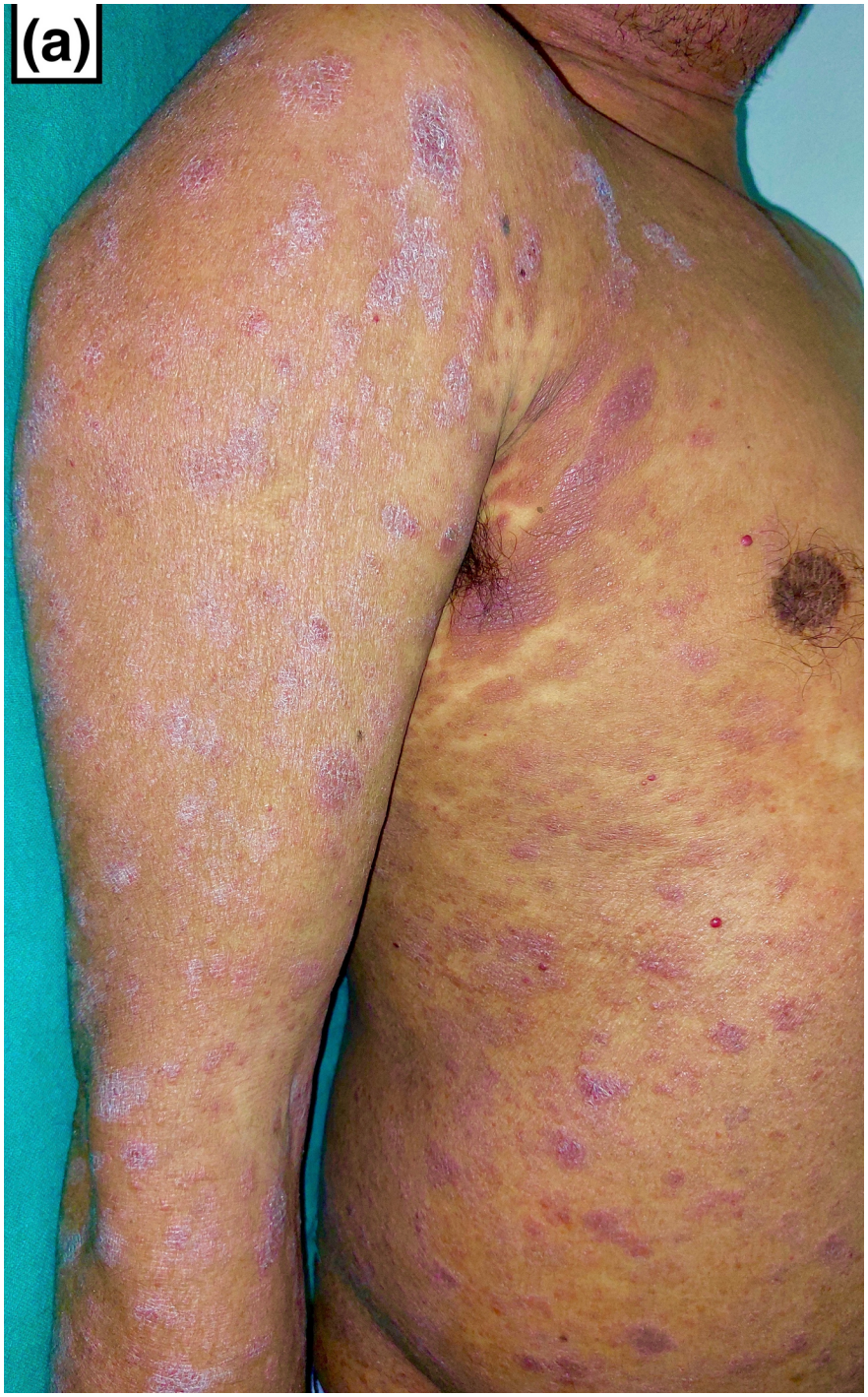

Figures:

Figure 1 Multiple ill-defined, scaly, erythematous-violaceous patches and plaques over right arm and anterior trunk (a), left arm and posterior trunk (b) and bilateral thighs and legs (c).

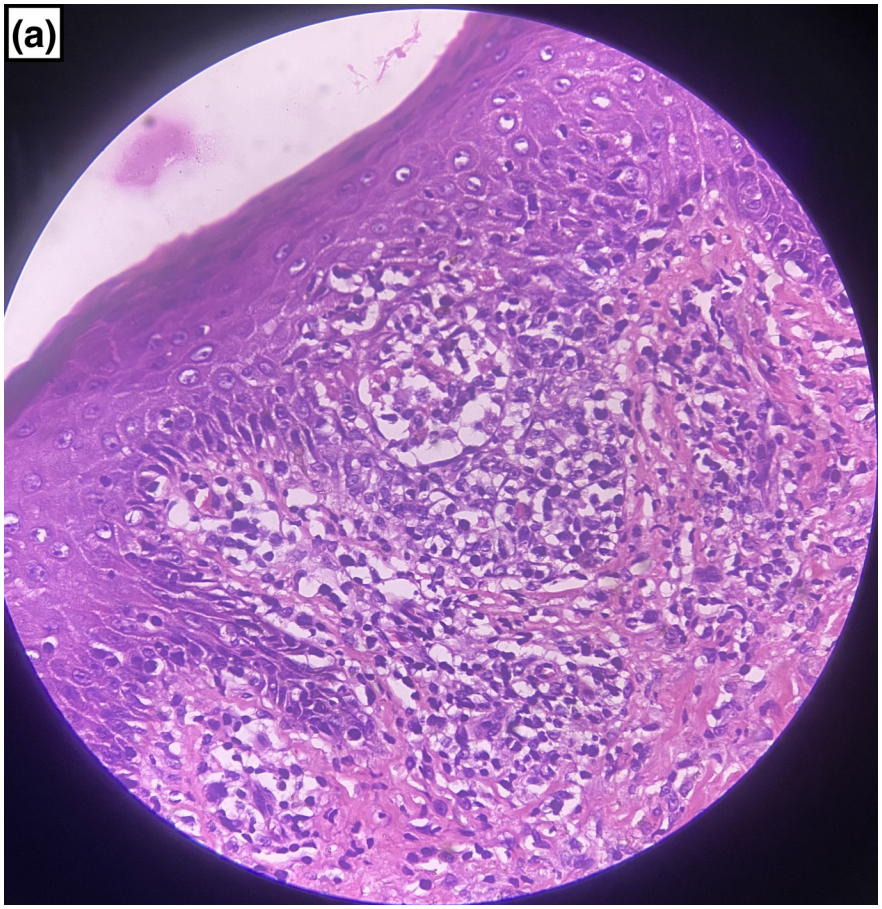

Figure 2 (a, b) Haematoxylin and eosin staining (40x) showed epidermotropism, Pautriner's microabcesses and perivascular lymphocytic infiltrates.