A rare case of mycosis fungoides with rapid progression, ulceration and a fatal outcome

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Introduction

Primary cutaneous T cell lymphomas (CTCL) are a heterogeneous group of clonal T cell lympho-proliferative disorders. Mycosis fungoides (MF) is the most common form of primary cutaneous lymphoma accounting for 55-60% of cases[1]. It most commonly occurs in adults with a median age at diagnosis between 55-60 years[2]. MF is twice as common in males than in females.

Case presentation

A 67-year-old man with multiple cardiovascular risk factors presented with asymptomatic plaques over his face, scalp, chest, abdomen and back for 3 months. Lesions initially started on the face and involved the other areas subsequently. It was not associated with mucosal involvement, photo-exacerbation, connective tissue disease features, constitutional symptoms, or fever. He denied dysphagia, altered bowel habits, and respiratory or urinary symptoms. Examination revealed multiple, well-defined, erythematous to skin-coloured plaques. Some were annular with central depression. The center of some plaques had a violaceous hue with few lesions showing fine scales (Figure 1). There was no evidence of sensory impairment over the plaques, peripheral nerve thickening, madarosis or earlobe infiltration. The differential diagnoses considered were CTCL, borderline lepromatous (BL) leprosy with type II reaction and sarcoidosis.

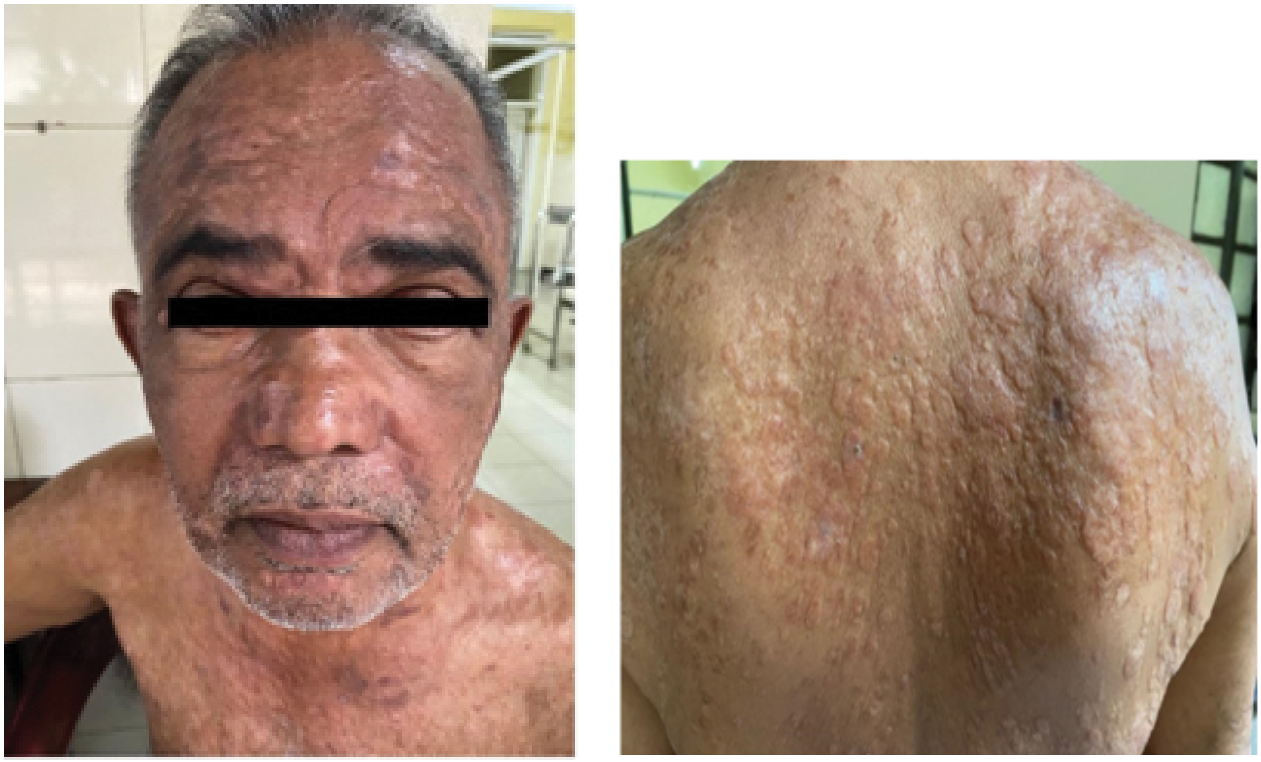

Figure 1.

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Initial skin biopsy for histopathological examination revealed nonspecific features and he was treated as leprosy with reaction with steroids at a local hospital without any improvement. Basic investigations were normal including blood picture without evidence of atypical lymphocytes. Bilateral axially and left submandibular lymphadenopathy was noted in the ultrasound scan. Hepatosplenomegaly or paraaortic lymphadenopathy was absent. Slit skin smear for leprosy and mantoux test were negative. Multiple skin biopsies were obtained. After one week of hospital stay, the patient became acutely unwell with painful ulceration of skin lesions associated with facial swelling (Figure 2). The results of repeated basic investigations were unremarkable. Histology showed nests of epidermotrophic lymphoid cells in the epidermis and at the dermo-epidermal junction. Dermis contained a moderate perivascular and periadnexal lymphocytic infiltration including atypical cells with enlarged hyperchromatic nuclei. Pautrier microabscesses and spongiosis were absent (Figure 3). Immunohistochemistry showed CD3, CD4, CD8, CD30 and MUM1 positivity with a Ki67 index of 70% indicating high cell proliferation. (Figure 4) However, CD7 was negative. Lymph node architecture was partially effaced with malignant cell infiltration. A diagnosis of MF with TNMB stage IVA2 was made. He was referred for oncological management and was started on gemcitabine. During chemotherapy, he succumbed due to sepsis.

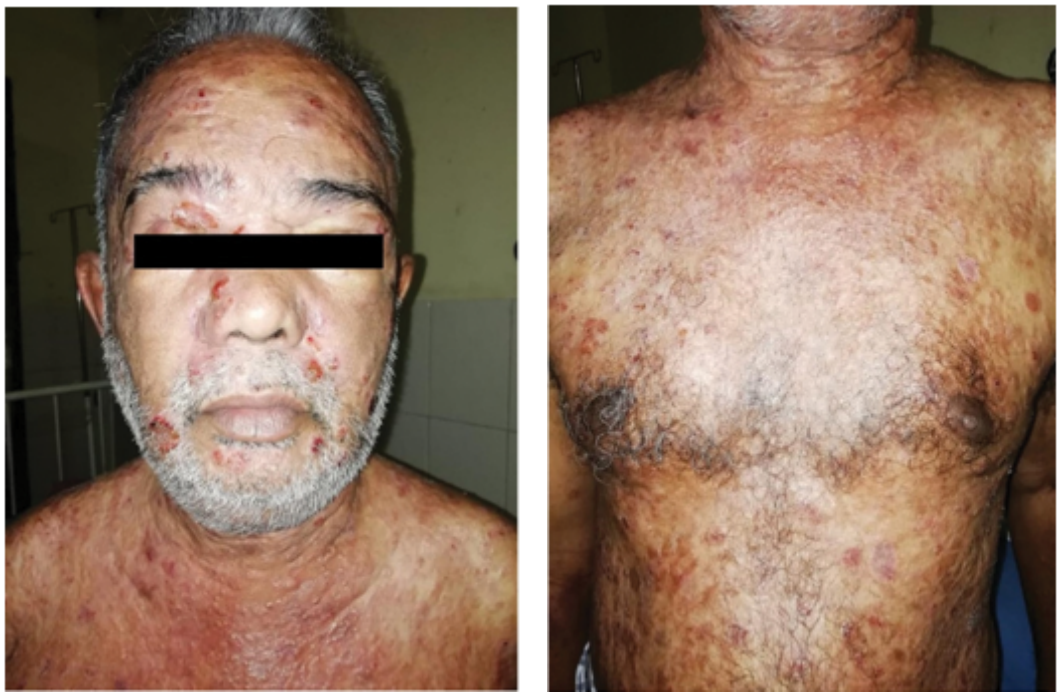

Figure 2.

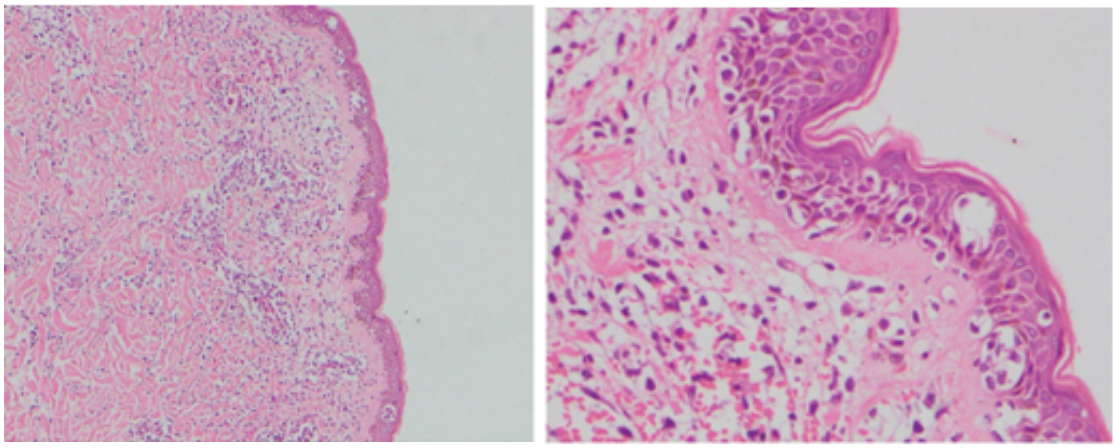

Figure 3.

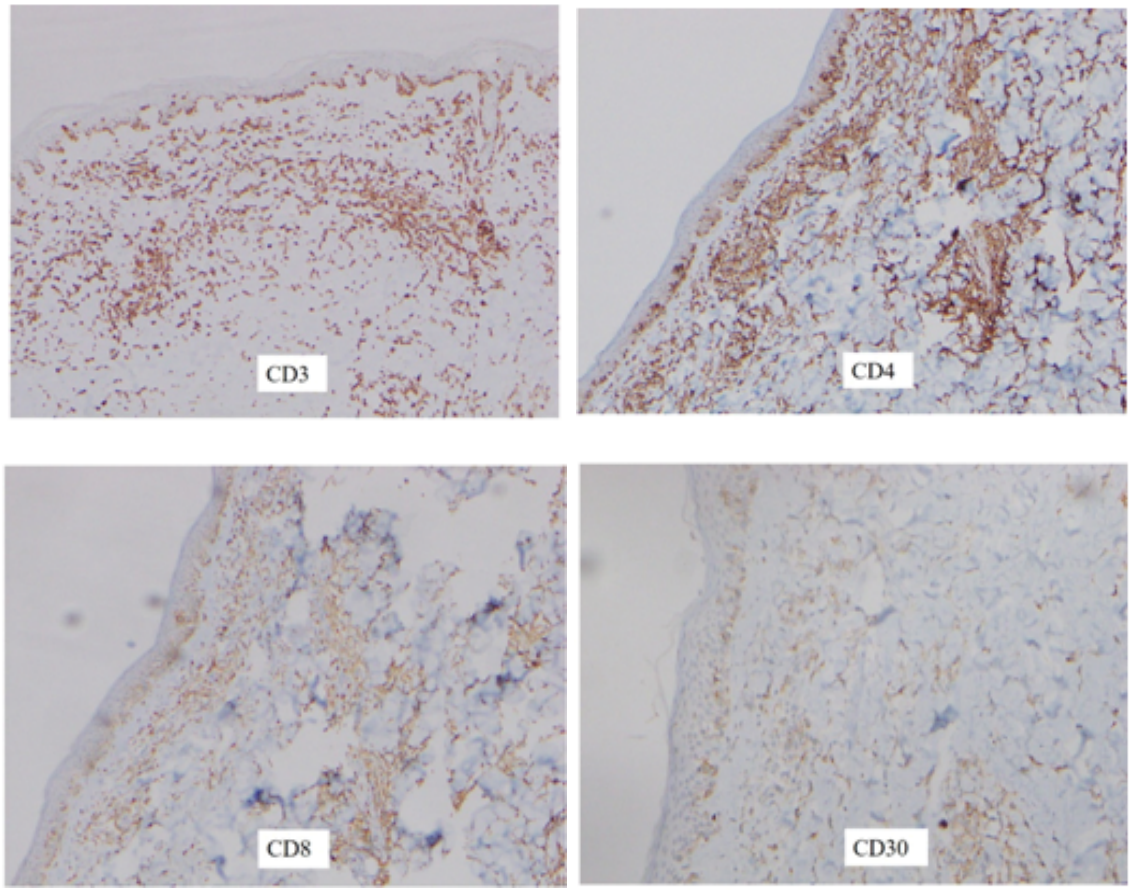

Figure 4. *Immunohistochemistry showing CD3, CD4, CD8 and CD30 positivity.*

Discussion

Patients with a classical type of MF progress from the patch stage to plaque stage and finally to tumour stage. Usually, MF has a protracted clinical course over the years or even decades[3]. Ulceration is rare in patches and plaques of MF, but is common in the tumour stage due to fast tumour growth and is associated with poor prognosis and high morbidity and mortality[4]. In our patient, plaques were ulcerated acutely and his disease rapidly progressed over a few months.

Conclusion

Clinicopathologically, the initial stages of MF mimic various benign skin diseases like leprosy, sarcoidosis, and psoriasis leading to delayed diagnosis and treatment[5]. Therefore, clinical suspicion, periodic skin biopsies for histopathological evaluation, and regular follow-up are of paramount importance. The commonest forms of CTCL have an indolent course, but some forms can be aggressive with the development of ulcerations even in the plaque stage of MF[6].

References

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