# Tiger-like mycosis fungoides: an unusual clinical presentation of a rare variant of mycosis fungoides.

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## Abstract

Mycosis fungoides (MF) is the most common type of cutaneous T-cell lymphoma. Mycosis fungoides classically presents in the skin as patches, plaques, tumors, or erythroderma, progressing to involve the lymph nodes and peripheral blood. The many clinical variants, with different histologic patterns, and the subtle early clinical and histologic changes may delay early diagnosis and present a diagnostic challenge for clinicians. The greatest challenge in diagnosis is the pre-mycotic stage, which may closely resemble eczematous or psoriasiform dermatitis clinically and histologically. The persistence of lesions and inadequate response to treatment are the first warning signs. Later stages of MF have a poor prognosis with poor therapeutic response and fatal outcome. We describe a 72-year-old man, who presented with a two-year history of an unusual eruption, which started on the abdomen, around the waistline, and gradually spread to involve his back, trunk, and buttocks. Clinically, the skin eruption presented as tiger-like stripes. The diagnosis was confirmed after histopathologic examination. The patient was treated with NB-UVB phototherapy with marked improvement.

*Keywords: mycosis fungoides, poikiloderma, tiger-like*

## Introduction

Mycosis fungoides (MF) is the most common type of primary cutaneous T-cell lymphoma. It usually shows skin predilection with characteristic clinical and histopathologic features and can progress to involve the lymph nodes and peripheral blood [1]. There are multiple unusual clinical variants of mycosis fungoides, such as the pigmented purpuric type, the poikilodermatous type, and the Ofugi type/papuloerythroderma; the early features may be very subtle, making it difficult to diagnose [2].

Early diagnosis may be especially difficult in the pre-mycotic stage of mycosis fungoides that closely resembles eczema. The late stages of mycosis fungoides have a poorer prognosis, with an unsatisfactory therapeutic response, and occasionally fatal outcome [3]. The median survival following diagnosis varies according to stage. Patients with an early stage IA disease have a survival identical to age and sex matched controls. In contrast, more than 50% of patients with stage III through stage IV disease have a fatal outcome, with a median survival of less than five years [4].

## Case Synopsis

We present a 72-year-old man, with a two-year history of an occasionally pruritic rash, which started on the abdomen around the waistline. He was treated with various topical emollient treatments without improvement. He was treated with topical corticosteroids but he discontinued after two weeks owing to no apparent benefit. His past medical history revealed epilepsy, which had been treated with sodium valproate for 10 years. Clinical examination revealed poikilodermatous, subtle, but

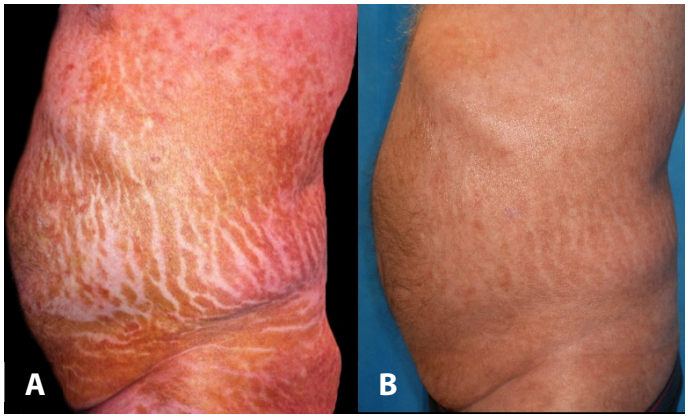


**Figure 1**. *'Tiger-dermatitis' at presentation **(A)** and status-post phototherapy **(B)**.*

demarcated, vertically-orientated pink-to-orange elongated plaques with fine scaling and epidermal atrophy, mainly at the waistline. The rash extended onto his trunk, back, and buttocks. The orange-red color of the eruption and the characteristic distribution imparted a tiger stripe-like appearance to the rash (**Figure 1A)**. The main clinical differential diagnosis included a chronic eczematous dermattits, an early stage of mycosis fungoides, and a drug-induced dermatosis.

The work up included routine complete blood count and differential, which revealed normal lymphocyte count; blood morphology showed lymphocytosis. Flow cytometry including immunophenotyping detected an excess of CD4 cells, raised CD4/CD8 ratio of 16.22 (normal range is 1-3.6), and a T-cell clone in the position V-beta, suggestive of T-cell lymphoma. Liver function test, urea and electrolytes, and serum lactate dehydrogenase were all within normal range. HTLV-1/2 serology was negative. A full body staging CT scan excluded systemic lymphoma and a bone

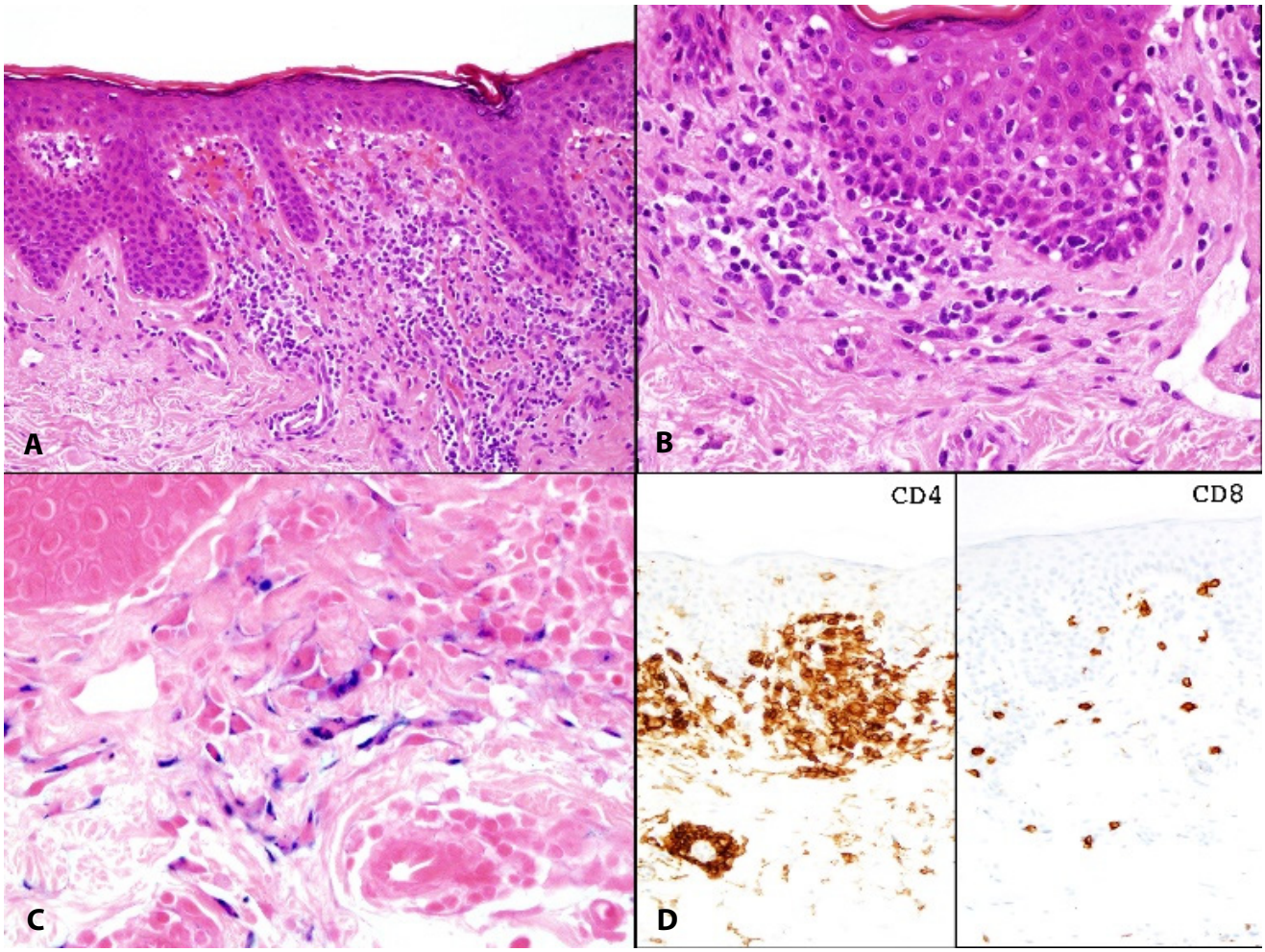


**Figure 2**. ***A)*** *Atypical lymphoid cell infiltrate on a background of papillary dermal fibrosis with extravasated erythrocytes. H&E, 200×.* ***B)*** *Detail of the papillary dermal atypical lymphocytes with 'tagging' along the basal cell layer and basilar epidermotropism. H&E, 600×.* ***C)*** *Perls stain highlights the hemosiderin deposits, 600×.* ***D)*** *Immunoprofile of the lymphoid cell infiltrate documents that it is CD4 predominant with almost complete loss of CD8 expression, 400×.*

marrow biopsy came back as negative. Four diagnostic incisional biopsies were carried out from the affected areas.

Histopathology revealed a papillary dermal mild-to-moderately dense superficial perivascular and band-like lymphoid cell infiltrate with cytological atypia on a background of papillary dermal fibrosis. Lymphocytic epidermotropism as well as 'tagging' along the dermo-epidermal junction was present (**Figure 2A**, **B)**. Multifocal papillary dermal areas of extravasated erythrocytes with hemosiderin deposition highlighted by Perl stain were identified, accounting for the tiger- like orange-red color of the clinical presentation (**Figure 2C)**. The striped appearance could have been related to corticosteroid-related striae distensae. However, the patient was adamant that he had not used topical corticosteroids for more than two weeks. No skin atrophy was identified clinically or histologically.

The lymphocytic infiltrate exhibited a T-helper CD4-positive predominant immunoprofile with loss of expression for CD7 with only a few reactive CD8-positive cells (**Figure 2D)**. T-cell clonality was negative. However, the overall features were in keeping with the rare purpuric variant of mycosis fungoides, but with a clinical tiger-like appearance, stage 1B, and without B symptoms. The patient was started on a twice-weekly regime of narrow band UVB phototherapy (NB-UVB), which resulted in marked improvement (**Figure 1B)**. Our patient has not relapsed to date and has continued to take his anti-epileptic medication, valproic acid.

## Case Discussion

Differentiating early mycosis fungoides from other benign dermatoses is difficult and diagnosis of early stage MF is often elusive, hence an algorithm for the diagnosis of early-stage mycosis fungoides has been suggested by the International Society of Cutaneous Lymphoma (ISCL), [5]. The algorithm includes clinico-pathological correlation and immunophenotypic and clonal T-cell receptor gene rearrangement studies, providing minimal criteria for diagnosis. In early mycosis fungoides, detection of a clone may vary between 45% to 71% of cases and does not seem to have a prognostic significance [6].

Lesion distribution is important in correctly diagnosing mycosis fungoides. Atypical presentations have also been reported [7]. Our patient had a rare purpuric variant [8], with an atypical distribution resembling tiger stripes extending from his waistline in a linear and vertical fashion to his chest and buttocks, reminiscent of a vertically-oriented deckchair sign. Of note, the deckchair sign can also be seen in papuloerythroderma of Ofuji, an inflammatory disorder characterized by coalescence of solid papules/plaques that typically spare the skin folds on the abdomen, antecubital, and axillary areas [9]. It would have been interesting to carry out a diagnostic biopsy on the normal appearing skin between the affected areas but regrettably this was not done. The possibilities would include either sparing of skin in a unique tiger-like pattern versus sparing of pre-existing striae distensea if we assume the skin was not affected by MF. However, clinical and histological atrophy was not appreciated. Both the clinical history of short-term administration of topical corticosteroids and the marked improvement of the tiger-stripes following phototherapy would point more towards these skin changes being caused by MF.

The pigmented purpura-like variant of mycosis fungoides may initially have the histologic appearance of classical pigmented purpura. The association of mycosis fungoides with pigmented purpuric dermatosis (PPD) is a rare, but a recognised phenomenon [8]. Pigmented purpuric dermatoses (PPD), a group of vascular disorders with variable clinical picture is reported in all races and age groups with a male predilection. There are reports of mycosis fungoides manifesting as pigmented purpura as well as progression of pigmented purpuric dermatoses to cutaneous T-cell lymphoma [10].

Anticonvulsants are known to cause pseudolymphoma syndrome. Clinically, pseudolymphoma syndrome is characterised by either an erythematous patch or a maculopapular eruption or nodules developing within 2–3 weeks after administering the causative agents. Phenytoin, carbamazepine, and sodium valproate are known to induce pseudolymphoma

syndrome. The syndrome is associated with fever, lymphadenopathy, arthralgia, hepatosplenomegaly, severe hepatitis, and eosinophilia [11]. Histopathologically, pseudolymphoma syndrome is distinguished from mycosis fungoides by the presence of spongiosis, necrotic keratinocytes, a mixed inflammatory cell infiltrate with eosinophils and neutrophils, and papillary dermal edema, features consistent with a drug eruption.

Clinical symptoms of pseudolymphoma syndrome will improve in two-to-nine weeks after the cessation of the causative agent [11]. In our case, the patient had been on sodium valproate for over 10 years prior to developing his rash and did not suffer with any of the associated systemic symptoms. Typically, the patients with drug-induced pseudomycosis fungoides do not have symptoms that differentiate them from true mycosis fungoides, unless they have drug reaction with eosinophilia and systemic symptoms (DRESS syndrome). However, there are non-DRESS cases of mycosis fungoides-like rashes that occur due to anti-seizure medications. In addition, our patient has continued to take his anti-epileptic medication, valproic acid, without recurrence.

## Conclusion

Our case raises awareness of the multifaceted presentations of mycosis fungoides and its pitfalls, particularly in absence of T-cell clonality.

## Potential conflicts of interest

The authors declare no conflicts of interests.

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