

Serpiginous mycosis fungoides in a 21-year-old man

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INTRODUCTION

Mycosis fungoides (MF) is a common type of cutaneous non-Hodgkin mature T cell lymphoma (CTCL). The median age of diagnosis is in the mid-50s with an increased incidence seen in patients older than 70.¹ Before the diagnosis of MF, patients may experience a premycotic phase, during which nonspecific scaly skin lesions are present. These may fluctuate and resemble more common skin conditions, such as eczema and psoriasis, rendering a definitive diagnosis difficult to establish. Further complicating matters, MF may present with a variety of morphologically distinct cutaneous lesions. In addition to the classic form of MF, there are 3 other common variants: folliculotropic, pagetoid reticuloid, and granulomatous cutis laxa. More rare subtypes include, among others, hypopigmented/hyperpigmented, erythrodermic, poikilodermic, pigmented purpurallike, bullous/dyshidrotic, papular, and the invisible subtypes.² These cutaneous lesions may be pruritic and have a profound impact on a patient's quality of life.³ Because of the nonspecific and variable course of presentation, the diagnosis of MF usually takes many years.¹

We present a case of MF in a young adult Asian male with an atypical presentation.

CASE REPORT

A previously healthy 21-year-old Asian man presented with a 4-year history of nonpruritic, hyperpigmented lesions that initially started as small, eczema-like, erythematous papules on his legs. After clinical and histologic evaluations by numerous dermatologists, these lesions were initially diagnosed as postinflammatory hyperpigmentation and

Abbreviations used:

CTCL: cutaneous T cell lymphoma
 EAC: erythema annulare centrifugum
 MF: mycosis fungoides

nonspecific dermatitis. His condition continued to progress despite topical corticosteroids, so his family physician referred him to our clinic for evaluation and treatment. During his initial consultation with us, he reported no systemic symptoms, joint pain, or shortness of breath and appeared well and healthy. Family history was significant for leukemia in his grandfather. There was no personal or family history of eczema or atopy. Interestingly, he was assessed for possible Lisch nodules and café au lait macules in 2000 but did not meet criteria for a diagnosis of neurofibromatosis type I. Other than a remote history of a slight partial thromboplastin time elevation, which was deemed nonsignificant by his hematologist, his medical history was unremarkable. He was not on any medications nor did he have any allergies.

Examination found large annular to serpiginous, polycyclic, brown, nonscaly patches with mild atrophy and a rim of hyperpigmentation on his limbs (Fig 1). Numerous ecchymoses and red/brown 1- to 2-mm papules and macules on his legs were noted. Punch biopsies of 3 separate areas taken from both legs showed a perivascular and bandlike infiltrate of lymphocytes, histiocytes, and melanophages within a papillary dermis that was fibrotically thickened. There was an increased number of atypical lymphocytes with irregularly shaped hyperchromatic nuclei, arranged singly, within the basal cell region of an epidermis in which there was no

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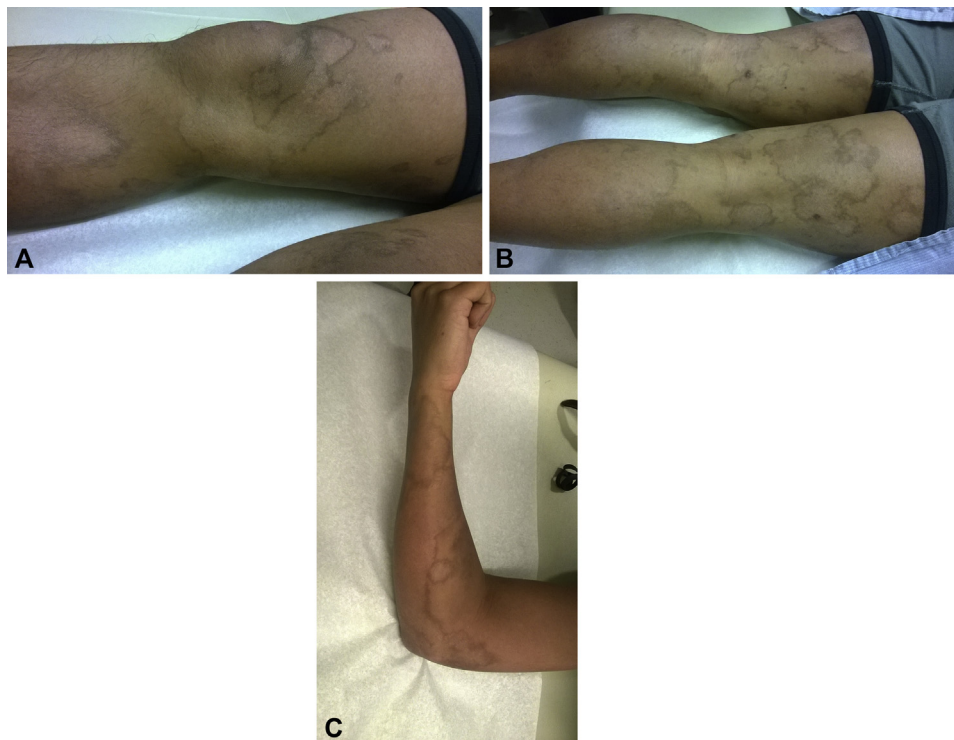


Fig 1. MF: the patient presented with these annular, serpiginous, hyperpigmented lesions on his limbs. **A**, Right knee. **B**, Posterior aspect of lower limbs. **C**, Left forearm.

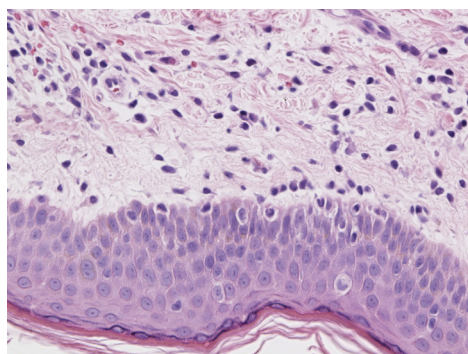


Fig 2. Hematoxylin and eosin stain shows lymphocytic infiltration and an increased number of atypical lymphocytes, with irregularly-shaped hyperchromatic nuclei within the basal cell region of the epidermis.

significant spongiosis (Fig 2). Periodic acid-Schiff stain was negative for fungi.

These findings were consistent with that of cutaneous T-cell lymphoma.

The patient responded well to treatment with narrow band ultraviolet B therapy 3 times weekly, showing partial resolution of his lesions after 8 weeks of treatment.

DISCUSSION

The etiology of MF is poorly understood; however, associations with human T-lymphocyte virus I

have been made.¹ It is thought that continuous activation of cutaneous T-helper lymphocytes leads to malignant transformation of a specific clone.⁴ A recent meta-analysis found an increased risk of MF associated with body mass index equal to or larger than 30 kg/m², cigarette smoking for 40 years or more, eczema, and family history of multiple myeloma.⁵ Other possible risk factors that have been implicated include drugs, infections, radiation, and chemical exposure, especially aromatic halogenated hydrocarbons and pesticides.⁴ MF can be diagnosed using a combination of clinical impression and histopathology, but the diagnosis often is not made until a few years after the onset after numerous skin biopsies. Immunophenotyping and polymerase chain reaction can also be used to aid in diagnosis.¹

MF is more common among older individuals, with a male preponderance. MF in patients younger than 35 years is rare and even more unusual in patients younger than 25 years, although cases even in infants have been reported.⁶ Nevertheless, the young age at onset seen in our patient is somewhat uncommon. Our initial differential diagnosis of his cutaneous eruption included, among others, granuloma annulare, sarcoidosis, annular lichen planus, annular psoriasis, and partially treated atopic dermatitis.

MF presenting as serpiginous or ecchymotic lesions is rare. A literature search found only 1 case of MF in a 61-year-old man with a 30-year history of serpiginous eruption on his limbs.⁷ We came across 1 published case of MF presenting as ecchymosislike, hyperpigmented mycosis fungoides.⁸ We were able to find about 20 cases of purpuric MF, with most cases reported in patients in their teens or early 20s.⁹⁻¹² Similarly, searching for arcuate or polycyclic MF/CTCL yielded several reports. However, having purpuric/ecchymotic, arcuate/polycyclic, and serpiginous features in a single patient was rare.

At first glance, the lesions in our patient may resemble erythema annulare centrifugum (EAC). We found 9 previous reports of EAC-like MF, with most cases arising in patients older than 40.¹³⁻²¹ Only a few cases of EAC-like MF were found in individuals younger than 30 years.^{13,14,17} The youngest reported MF resembling EAC was in a 12-year-old girl.¹⁷ The constellation of features—hyperpigmentation, EAC-like figurate/serpiginous, and ecchymotic/purpuric lesions in a patient with darker complexion—makes this case unique.

The prognosis for MF is generally good, especially in patients with patch/plaque stage, but median survival rates decrease in patients with more advanced disease. Furthermore, the frequency of large cell transformation to a more aggressive clinical form of CTCL increases with advancing clinical stage of MF. It is, therefore, important to identify MF early so that appropriate therapies could be instituted to reduce potential progression and subsequent complications.

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