A rare presentation of cutaneous T-cell lymphoma mimicking morphea

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INTRODUCTION

Cutaneous T-cell lymphoma (CTCL) is a non-Hodgkin lymphoma of skin-homing malignant T cells, with mycosis fungoides (MF) being the most common subtype.[1] MF can mimic benign inflammatory skin disorders and can be diagnostically challenging in some cases.[2] In such cases, identification of T cells with aberrant immunophenotypes and T-cell receptor gene rearrangement assays for clonality can inform the diagnosis. Morphea is an inflammatory fibrosing disorder of the skin that typically presents as pink to violaceous patches that may progress to more indurated and sclerotic plaques.[3] Early forms of morphea can pose a diagnostic challenge and, to date, 3 cases of morphea with clinical and histologic features of patch MF have been reported in the literature.[3,4] In all cases, initial histopathology showed acanthosis and lymphocytic epidermotropism consistent with MF, but subsequent histopathology revealed dermal fibrosis with loss of adnexal structures compatible with morphea. Early extragenital lichen sclerosus can also constitute a mimicker of MF and can be seen in conjunction with morphea, as may have been present in reported cases of morphea mimicking CTCL.[3,5] Conversely, 5 patients with interstitial MF mimicking morphea histologically have been reported.[6] However, the authors noted that none of the lesions were violaceous or indurated as expected in morphea. Cases of CTCL mimicking morphea either clinically or histologically may thus be underappreciated. Herein, we describe a rare presentation of interstitial CTCL as violaceous indurated plaques.

Abbbreviations used:

CD:	cluster of differentiation
CTCL:	cutaneous T cell lymphoma
MF:	mycosis fungoides

CASE REPORT

A 50-year-old woman presented with a 4-year history of pruritic cutaneous lesions on her abdomen and breast. Physical examination revealed a round to oval pink patch on the left breast and thin pink plaque on the lower abdomen. Over 3 months, both lesions progressed to violaceous indurated plaques without scale, with the abdominal plaque manifesting peau d'orange changes (Fig 1). Given the progression from pink patch to indurated plaques, a clinical diagnosis of morphea was initially favored. Biopsies were obtained from both lesions, and histopathology revealed a dense band-like dermal lymphocytic infiltrate without epidermotropism (Fig 2, *A* and *B*). As the histopathology was not diagnostic, T-cell receptor gene rearrangement testing was performed via polymerase chain reaction and a dominant T-cell clone was identified in the abdominal lesion, but not the breast lesion. Immunohistochemical staining revealed that a majority of the lymphocytes were cluster of

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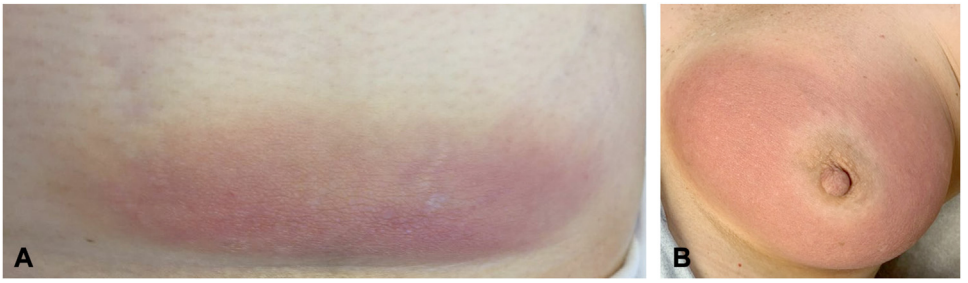

Fig 1. Cutaneous T-cell lymphoma presenting as indurated plaques. **A,** *Pink* indurated plaque on the *lower* abdomen with peau d'orange changes. **B,** Violaceous indurated patch without scale on the left breast with sparing of the nipple.

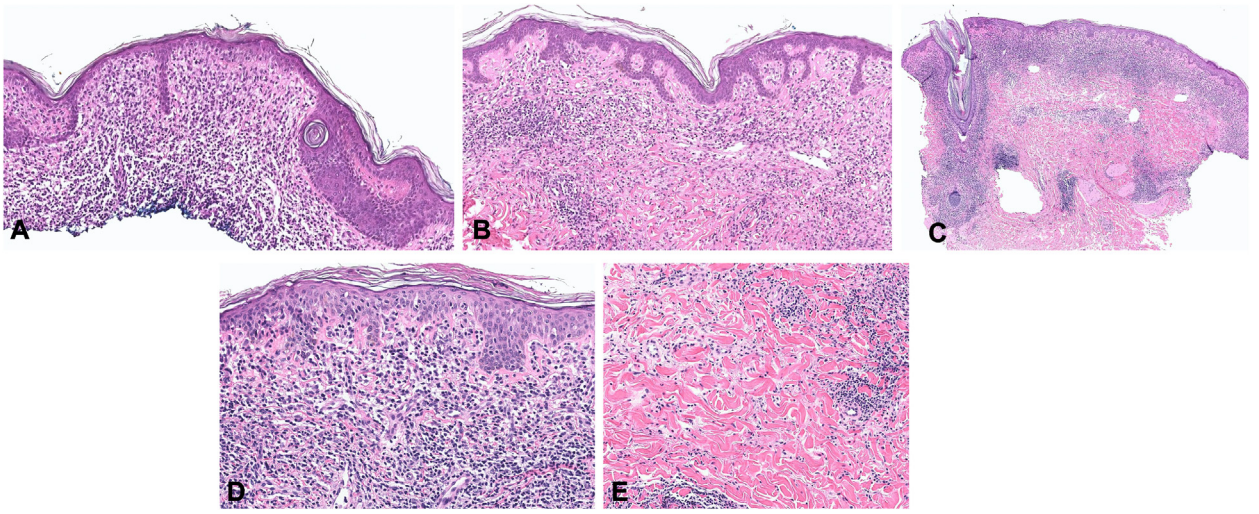

Fig 2. An initial biopsy specimen from the *lower* abdomen revealed a dense band-like dermal lymphocytic infiltrate without epidermotropism (**A**, hematoxylin and eosin [H&E], original magnification × 100). A biopsy from the left breast demonstrated nearly identical histopathologic features (**B**, H&E, original magnification × 100) with a slightly atrophic epidermis and no epidermotropism. The third biopsy specimen obtained 3 years subsequently from a plaque on the left breast showed features consistent with cutaneous T-cell lymphoma (**C**, H&E, original magnification ×20). A dense lymphocytic infiltrate was present in the superficial and deep dermis. Folliculotropism was demonstrated (**C**) along with characteristic epidermal extension of monotonous moderately enlarged lymphocytes (**D**, H&E, original magnification × 100). The reticular dermis showed myxoid alteration of collagen with an interstitial increase in lymphocytes and eosinophils (**E**, H&E, original magnification × 100).

differentiation (CD)4 positive and had diminished CD5 expression, but without loss of CD7 typical of MF. Flow cytometry of peripheral blood revealed no evidence of blood involvement. Despite the lack of epidermotropism or lymphocyte atypia histologically, and weighing the distribution and evidence of T-cell clonality, the diagnosis of MF stage 1A was made.

The patient was sequentially treated with narrow band UV-B for 5 weeks, topical imiquimod 3.75% for 10 months, and oral bexarotene 225 mg daily for 5 months, but without significant improvement. Bexarotene was discontinued due to leukopenia. The patient subsequently underwent spot electron beam radiation therapy to the abdominal lesion that resulted in complete resolution. The indurated plaque on the left breast persisted despite the aforementioned treatments and another biopsy specimen was obtained. Histopathology revealed a band-like lymphocytic infiltrate in the superficial and deep dermis. Folliculotropism and epidermotropism were noted (Fig 2, *C-E*). The lymphocytes were monotonous and moderately enlarged. The reticular dermis showed myxoid alteration of non-thickened collagen bundles with an interstitial increase in lymphocytes and eosinophils (Fig 2, *E*). The

histopathology was most compatible with an interstitial pattern of MF, although the interstitial pattern was reminiscent of early morphea. The patient was treated with acitretin 10 mg daily and systemic interferon-α 180 mcg weekly by subcutaneous injection for 4 years with near resolution of the lesion and improvement in induration. No new lesions or progression have been noted for the past 3 years.

DISCUSSION

Stage IA lesions of MF classically present as pink patches or thin plaques with fine scale, and favor the sun-protected areas of the body.[1] They can frequently mimic benign inflammatory skin disorders such as psoriasis and chronic eczematous dermatitis, leading to delays in diagnosis.[2] Our patient presented with pink patches that later progressed to violaceous indurated plaques, seemingly following the clinical course of early morphea. Initial histopathology was nondiagnostic but the identification of T-cell clonality was instrumental in the diagnosis of MF. Subsequent biopsy revealed epidermotropism with monotonous, moderately enlarged lymphocytes, providing further support for the diagnosis of MF. The dermal alterations such as the myxoid dermal changes and interstitial increase in lymphocytes and eosinophils were suggestive of interstitial MF and could explain the indurated clinical aspect of the lesions. However, it is important to note that identification of a clonal T-cell population is not specific for MF and immunohistochemistry can provide another tool toward the correct diagnosis.[7,8] For instance, the loss of CD7 expression by lymphocytes is more suggestive of MF than benign inflammatory disorders such as morphea.[9] In fact, 1 of the 3 reported cases of morphea mimicking CTCL showed a monoclonal amplification of T cell receptorγ on 1 of 2 specimens; however, there was a mixed population of $CD4^+$ and $CD8^+$ cells without significant deletion of CD5 or CD7 expression.[3] Additionally, a subsequent biopsy showed dermal fibrosis with entrapment of eccrine glands and loss of hair follicles, features consistent with morphea.[3]

In summary, we report herein a rare case of CTCL with clinical features mimicking morphea to raise awareness of this potential presentation. While molecular studies can be used to aid with the diagnosis, the clinical course of the disease should be followed with a low threshold to repeat biopsy in diagnostically challenging cases.

Conflicts of interest

None disclosed.

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