Mycosis fungoides presenting as symmetric concentric patches mimicking figurate erythema

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CASE REPORT

A 52-year-old white male presented to outpatient dermatology clinic for evaluation of a 4-year history of a recurrent, intermittent rash that appeared as symmetrically distributed, red annular patches on non−sun-exposed areas of his trunk and extremities. The lesions gradually expanded in concentric rings with scale over several months. The affected areas were mildly pruritic, and he experienced arthralgias with each flare. His medical history was noncontributory, and his only medication was a proton pump inhibitor, which he had been taking for many years.

At an outside facility, Lyme disease testing was negative, and he was given the initial clinical diagnosis of erythema annulare centrifugum (EAC) and was treated with topical and systemic corticosteroids, which provided only temporary improvement.

On examination, the bilateral upper and lower extremities had pink-red, symmetric, concentric annular patches with trailing scale covering 10% to 15% of his body surface area (Fig 1). There was no palpable cervical or axillary lymphadenopathy. Clinically, the differential diagnosis included EAC, erythema gyratum repens, tinea corporis, and mycosis fungoides (MF).

Results of laboratory investigations were within normal limits. A punch biopsy and a broad shave skin biopsy from the leading edge of a lesion on the left lower extremity found a superficial perivascular and lichenoid lymphocytic infiltrate with exocytosis of small lymphocytes with mild nuclear contour irregularities, accompanied by minimal spongiosis and vacuolar alteration at the dermoepidermal junction as well as papillary dermal fibrosis (Fig 2). Immunohistochemical staining found a mostly $CD3^+$ lymphocytic infiltrate with approximately an equal ratio of $CD4^+$ to $CD8^+$ cells (Fig 3). Periodic acid−Schiff diastase staining was negative for fungi. No perivascular cuffing, interstitial lymphocytic infiltrate, or significant red blood cell extravasation was evident. Loss of CD7 on lymphocytes was estimated to be approximately 40%. The morphologic and histologic features raised concern for evolving MF, prompting further molecular studies. T-cell receptor (TCR) gene rearrangement studies of the TCRγ gene locus using polymerase chain reaction and capillary electrophoresis[1] were positive in 2 concurrently submitted specimens for oligoclonal peaks using

Abbreviations used:

EAC: erythema annulare centrifugum
MF: mycosis fungoides
TCR: T-cell receptor

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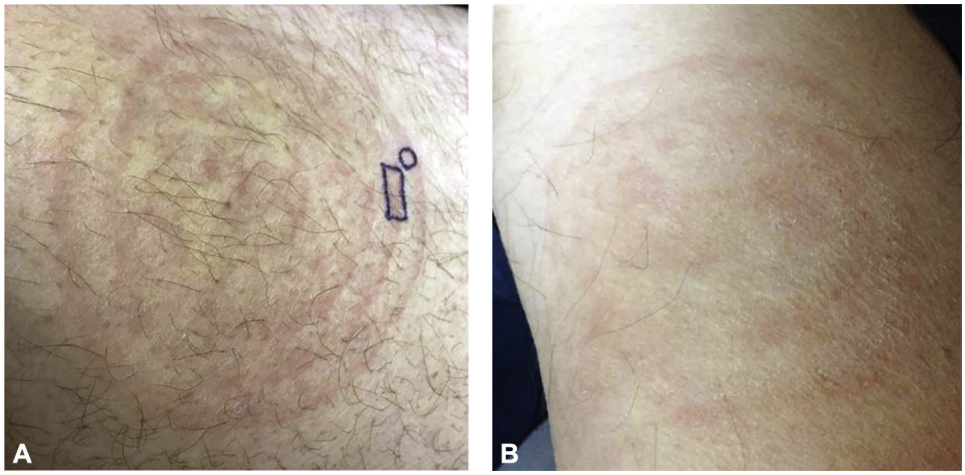

Fig 1. Concentric lesions present on the (**A**) left thigh and (**B**) left upper arm. Skin markings correspond to biopsied sites.

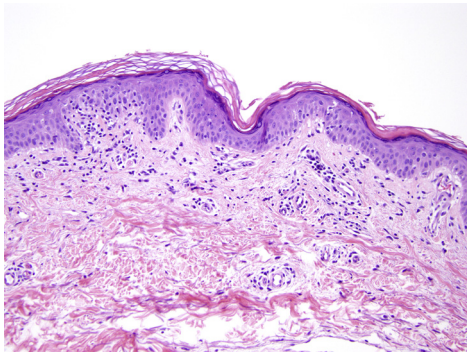

Fig 2. Histology shows a superficial perivascular and lichenoid lymphocytic infiltrate with exocytosis of predominantly small lymphocytes. (Hematoxylin-eosin stain; original magnification: ×20.)

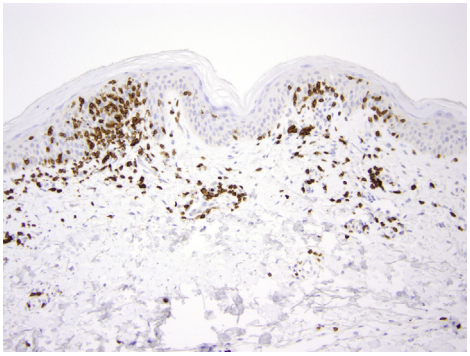

Fig 3. Immunohistochemistry with an anti-CD3 stain shows a $CD3^+$ infiltrate. (Original magnification: ×20.)

the V9_J13n23 and the V1to8_J13n23 primers. Based on the sum of his clinical, molecular, and histologic findings, our patient had patch stage MF diagnosed (T2 stage 1b).[2] He responded well to high-potency topical corticosteroids and natural sunlight exposure (in lieu of narrowband ultraviolet B phototherapy) with near resolution of all the patches and continues to be clinically monitored for disease progression.

DISCUSSION

The annual incidence of cutaneous T cell lymphoma in the United States is 6.4 to 9.4 cases per million[3] with MF, the most common variant, representing approximately 50% of new cases.[4] The diagnosis of MF can be challenging because of the diversity of clinical presentation and the array of histologic/molecular findings that commonly, but not always, occur in this disease.[3]

Several case reports of MF presenting similarly to EAC as annular or polycyclic plaques are present in the literature.[5] Characteristic lesions of EAC present as erythematous rings, which expand outward with central clearing.[6] In the case presented here, however, lesions were striking, symmetrically distributed red patches with multiple concentric, almost targetoid, rings with trailing scale that radiated from the center presenting similarly to that of gyratum repens. Unlike EAC, there was no obvious central clearing, as the entire patch was composed of these striking bandlike rings.

There is variation in the histologic findings in MF depending on the stage at which the lesions are biopsied.[2] The histologic features of early patch stage MF are often subtle, and it is not uncommon that multiple biopsies are necessary to reach the correct diagnosis. The characteristic histology in MF shows epidermotropic and lichenoid (palisading) infiltrates of small- to medium-sized T lymphocytes with cerebriform nuclei. Other reported useful features include disproportionate epidermotropism and wiry bundles of papillary dermal collagen

associated with lichenoid infiltrates.[7] Notably, we observed some of these features, including epidermotropism, in our patient that raised the suspicion for MF despite clinical features that were unusual for this diagnosis. Interestingly, immunophenotyping of the T-cell infiltrate did not show an elevated CD4:CD8 ratio that has been reported in many patients with MF. However, a normal CD4:CD8 ratio does not necessarily exclude MF. Furthermore, although rare, MF with $CD8^+$ phenotype could also be considered, given the near equal ratio of CD4:CD8 observed in our case.[3] Our patient did show partial loss of CD7 surface antigen, which is a feature of MF that can also occur in benign inflammatory conditions.[7]

Because of the suggestive, but not diagnostic, histologic and immunologic features shown by our patient, we performed molecular T-cell gene rearrangement analysis. Again, although detection of TCR clonality is not entirely specific for MF, clonal peaks have been shown to be present in 50% of those with patch stage disease and to increase to nearly 100% of those with tumor stage disease (3). Our polymerase chain reaction–based methodology did detect clear oligoclonal peaks, which prompted us to give a diagnosis of MF to this patient. In the future, advanced next-generation sequencing techniques may offer greater sensitivity and specificity for the detection of T-cell clonality.[8] The presence of T-cell clonality in 2 different anatomic sites has been reported to increase specificity for a diagnosis of MF.[9]

Although MF can present with myriad clinical findings, the current case presentation of concentric erythematous rings with trailing scale was unusual and mimicked figurate erythema. The initial diagnosis of EAC was understandable given the presence of trailing scale, but typically EAC does not show central clearing. Indeed, an EAC-like presentation of MF has been reported.[10] A morphology, however, manifested by concentric rings of affected skin, is rare in dermatology and is observed in a few conditions such as erythema gyratum repens and tinea imbricata. To the best of our knowledge, the complex concentric/annular morphology exhibited by our MF patient has not been reported elsewhere. Accordingly, we advise that skin biopsy, histology, and immunohistology as well as T-cell gene rearrangement studies may need to be performed in patients with chronic dermatoses with gyrate (figurate) morphologies to rule out T-cell malignancy.

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