Angiodestructive lymphomatoid papulosis lasting more than 45 years

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INTRODUCTION

Lymphomatoid papulosis (LyP) is a benign lymphoproliferative disorder characterized by recurrent crops of multiple erythematous papules on the trunk and extremities that spontaneously resolve within 1 to 4 months.[1] LyP belongs to a spectrum of $CD30^+$ lymphoproliferative disorders, which also includes primary cutaneous anaplastic large cell lymphoma. Although it is associated with mycosis fungoides, primary cutaneous anaplastic large cell lymphoma, systemic anaplastic large cell lymphoma, Hodgkin disease,[2] chronic lymphocytic leukemia,[3] multiple myeloma, and myelodysplastic syndrome,[4] the disease itself is considered benign.

Several histologic subtypes of LyP have been described, including A, B, C, D, E, and $Dusp22^+$.[5] Type E is characterized by angiocentric and angiodestructive infiltrates of small- to medium-sized $CD30^+$ and $CD8^+$ lymphocytes. Clinically, it presents as papular lesions that rapidly develop into necrotic eschar-like ulcers and plaques that spontaneously resolve.[6]

Despite the indolent clinical course of LyP type E, its histology can mimic that of aggressive angiocentric cutaneous T-cell lymphoma. Therefore, clinicopathologic correlation is essential to diagnosing the disease.[7] Here, we present a patient with LyP type E with a regressing papular eruption lasting for more than 45 years.

Abbreviation used:

LyP: lymphomatoid papulosis

CASE REPORT

A healthy 58-year-old man presented to the dermatology clinic with a 45-year history of recurrent, asymptomatic eruptions of red papules. The lesions predominantly appeared on his arms, thighs, and trunk. They healed spontaneously in 2 to 4 weeks, with minimal scarring. The patient denied weight loss, night sweats, or fevers. Biopsy of a characteristic lesion from 1973 showed "vasculitis of allergic granulomatous character" with "features of lymphomatoid papulosis."

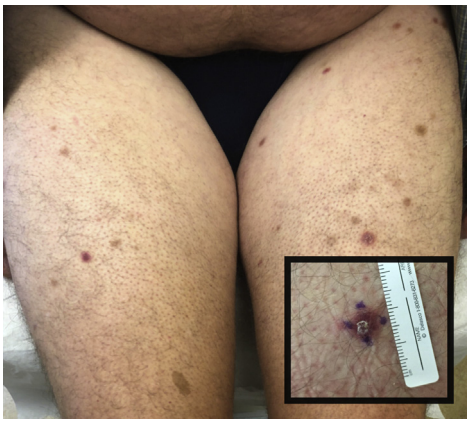

Fig 1. LyP type E clinical presentation. Erythematous to violaceous papules that emerge in crops on the dorsal hands, thighs, and dorsal feet. The inset shows the collarette of scale seen in some of the lesions.

On examination, there were scattered erythematous to violaceous edematous papules, some with overlying collarette of scale and crust on his dorsal hands, arms, thighs, and trunk (Fig 1). There was no lymphadenopathy or hepatosplenomegaly on

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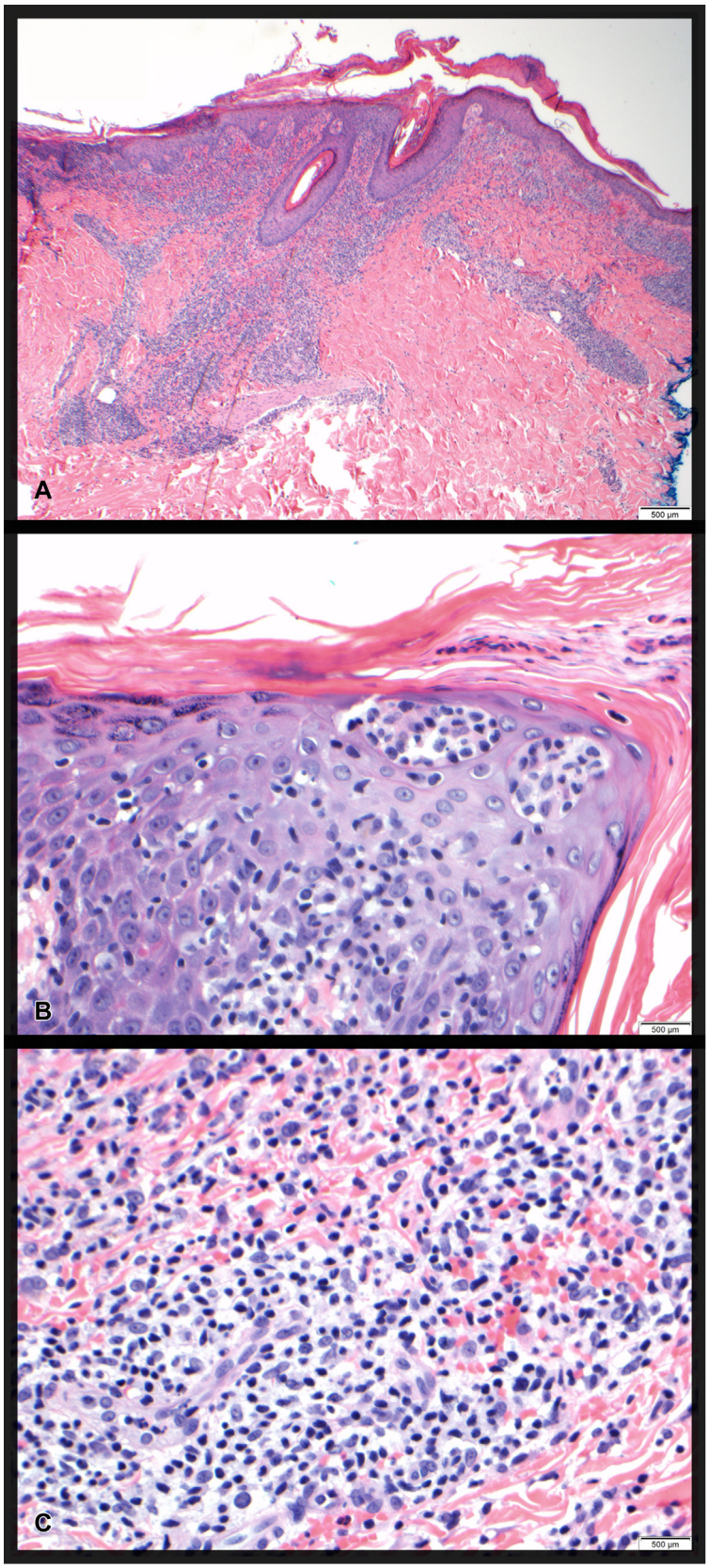

Fig 2. LyP type E histopathology. **A**, Wedge-shaped infiltrate extending from the papillary dermis to the deep reticular dermis. **B**, High-power image of the infiltrate shows small- and medium-sized lymphocytes with occasional scattered large lymphocytes. **C**, High-power image of small- and large-vessel destruction in the setting of this infiltrate. (Hematoxylin-eosin stain; original magnifications: **A**, ×2; **B** and **C**, ×20).

examination. Punch biopsy of a lesion showed a wedge-shaped perivascular and interstitial infiltrate primarily composed of small and medium-sized lymphocytes with occasional scattered large lymphocytes, causing some small and large vessel destruction (Fig 2, *A*–*C*). $CD3^+$ lymphocytes extended from the epidermis to the deep reticular dermis, with loss of CD7 expression. There was a 10:1 predominance of CD4 to CD8 lymphocytes, and 75% of the lymphocytes were $CD30^+$. Test results for T-cell receptor β were positive. Staining results for Epstein-Barr virus and anaplastic lymphoma kinase 1 were negative.

The histologic differential diagnosis included LyP, $CD30^+$ mycosis fungoides, anaplastic large cell lymphoma (primary cutaneous or systemic with secondary cutaneous involvement), and angiodestructive peripheral T-cell lymphoma. When the history and examination were taken into account, these findings supported a diagnosis of angiodestructive LyP. Methotrexate was discussed as a therapy. However, because the lesions remained asymptomatic, the patient declined treatment.

DISCUSSION

LyP has an excellent prognosis.[1] One study of 118 patients with LyP reported only a 4% absolute risk for systemic lymphoma within 10 years of diagnosis and a 5-year disease-related survival rate of 100%.[8] Additionally, the lesions of LyP type E resolve spontaneously, as do those of other types of LyP. In a case series of 16 patients with type E LyP, 56% experienced complete remission of symptoms in a mean follow-up of 37.5 months.[6]

Consensus recommendations for the treatment of LyP endorse noninterventional monitoring as a first-line approach because no therapy has been shown to achieve sustained remission or to prevent secondary lymphomas.[9] However, patients with extensive and/or symptomatic lesions may benefit from therapies that have been shown to reduce the number and duration of the lesions, including light therapy and low-dose methotrexate.[9] Bexarotene and topical mechlorethamine, both of which are approved by the US Food and Drug Administration for mycosis fungoides/Sézary syndrome, can also be used in cases of extensive LyP.[10] Case studies have also shown successful management of LyP using interferon alfa-2a.[2] For cases refractory to methotrexate, national guidelines suggest low-dose brentuximab vedotin.[10]

To our knowledge, our case shows the longest course of LyP type E reported to date.[2,6] The disease course is benign and indolent despite its aggressive, malignant appearance on histology, which highlights the importance of clinicopathologic correlation for diagnosis of angiodestructive LyP.

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