Lymphomatoid papulosis

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Abstract

Lymphomatoid papulosis is often regarded as a low-grade variant of cutaneous T cell lymphoma (CTCL). Given the excellent long-term prognosis, recent consensus guidelines indicate that patients can be monitored off therapy. We report a case of a 67-year-old man who presented with lymphomatoid papulosis, with necrotic papules that have been intermittently present for over forty years.

Keywords: lymphomatoid papulosis, cutaneous T cell lymphoma

Introduction

Lymphomatoid papulosis is often regarded as a low-grade variant of cutaneous T cell lymphoma (CTCL), of which it accounts for approximately 15% of all cases. Lesions typically disappear without treatment within three to eight weeks, resulting in post-inflammatory pigment alteration, atrophic scars, or no sequelae. Disease duration ranges from months to more than four decades.

Case Synopsis

A 67-year-old man presented to the Manhattan Campus of the VA New York Harbor Healthcare System fifteen years ago for the evaluation of necrotic papules that had been intermittently present for the preceding thirty years. The patient reported the development of this eruption primarily on his trunk and extremities. At the time of presentation, the patient denied fevers, chills, arthritis, abdominal pain, or other systemic symptoms.

Exacerbations of the papules have been managed intermittently with clobetasol ointment. More recently, his course has been complicated by bacterial superinfection, for which he uses clindamycin pledgets. He also applies hydroquinone to post-inflammatory hyperpigmentation associated with resolved lesions. There were a few scattered hyperpigmented papules with central necrosis on his right forearm as well as some scattered hyperpigmented macules and patches on his trunk and extremities (Figures 1, 2). No inguinal, axillary, or cervical lymphadenopathy was appreciated. There was also no hepatosplenomegaly on palpation of the abdomen. A complete blood count at time of presentation was within normal limits. Lactate dehydrogenase at time of presentation was 171 U/L (reference range 91-180).

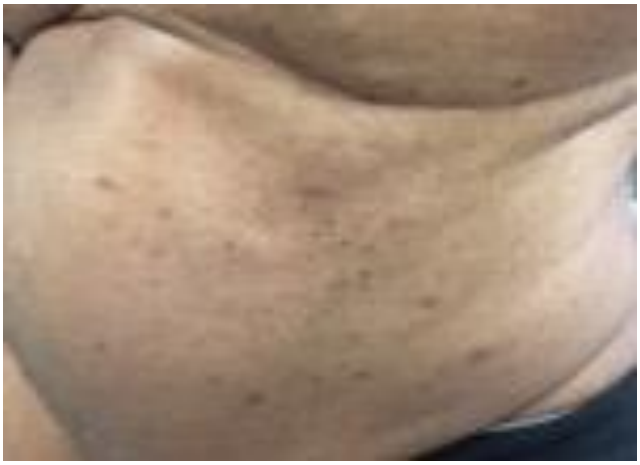

Figure 1. *There are few scattered hyperpigmented papules with central necrosis on his right forearm.*

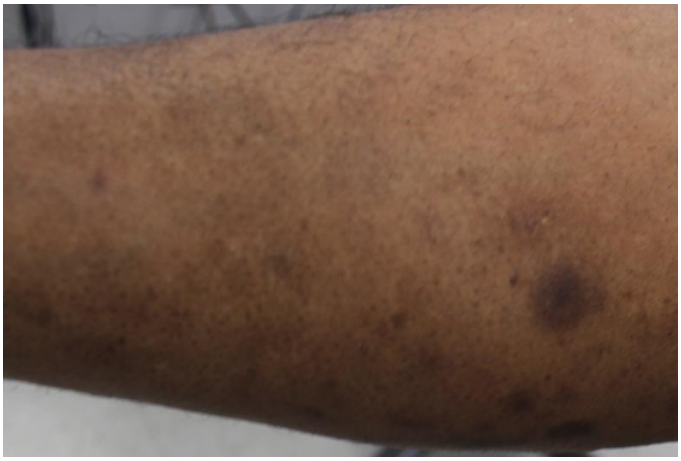

Figure 2. *There are some scattered hyperpigmented macules and patches on his trunk and extremities.*

A punch biopsy was performed of a lesion on the right forearm. This exhibited a superficial and deep, perivascular and interstitial infiltrate of lymphocytes, histiocytes, neutrophils, and numerous eosinophils (**Figure 3**). Scattered enlarged lymphocytes were present, highlighted by CD30. The epidermis was unremarkable. Bi-annual chest CT scans have all been unremarkable for the last 15 years.

Case Discussion

The term lymphomatoid papulosis (LyP) was first introduced in 1968 [1], and since that time, there has been controversy as to its malignant, pre-malignant, or benign nature. The World Health Organization and European Organization for the Research and Treatment of Cancer classify LyP as a subset of the CD30 positive lymphoproliferative disorders [2]. Most authors regard the entity as a low-grade variant of cutaneous T cell lymphoma (CTCL), of which it accounts for approximately 15% of all cases [3].

Clinically, lymphomatoid papulosis is characterized by chronic, recurrent, and self-healing red-brown papules and nodules that may develop central necrosis, hemorrhage, and crusting. LyP can occur at any age, with the average onset between 35 and 45 years [3]. There is a 1.5:1 male to female ratio. Lesions in different stages of evolution are typically noted concurrently, and lesion counts range from several to more than 100. Distribution of lesions can be focal, clustered, or generalized, and the eruption is usually asymptomatic, typically involving the trunk and limbs. Lesions typically disappear without treatment within three to eight weeks, resulting in post-inflammatory pigment alteration, atrophic scars, or no sequelae. Disease duration ranges from months to more than four decades [4]. Patients are followed closely given LyP's association (in 5-25% of patients) with another cutaneous or systemic lymphoma (often mycosis fungoides, cutaneous anaplastic large cell lymphoma (C-ALCL), or Hodgkin lymphoma), [5]. There also may be a small associated risk of development of a non-hematologic malignancy [6]. Patients with LyP have an excellent prognosis, with only a minority of patients developing a systemic lymphoma. A recent study of over 100 patients with LyP reported only two patient deaths over a median follow-up period of 77 months [7].

The pathophysiology of LyP is not well understood. Although a viral etiology has been suggested as an

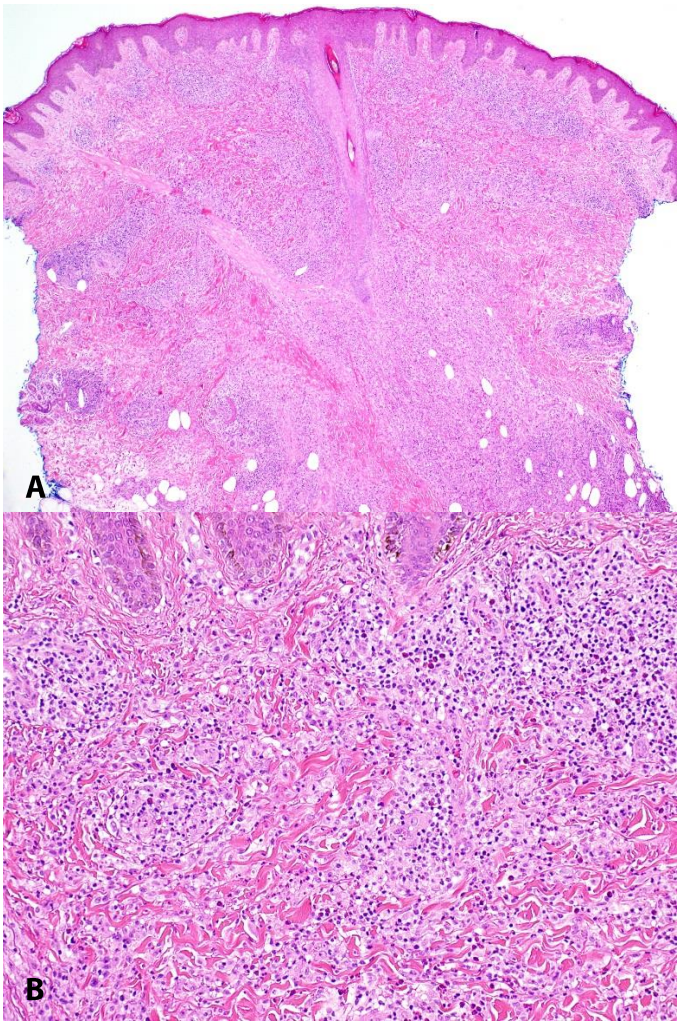

Figure 3. *There is a superficial and deep, perivascular and interstitial infiltrate of lymphocytes, histiocytes, neutrophils, and numerous eosinophils. H&E, A) 4×, B) 20×.*

initiating event, none has been elucidated. Studies searching for involvement of viruses, including HTLV-1, EBV, HSV-1, HSV-2, and HHV-6, have been negative [8]. Similarly, the reasons why most patients experience spontaneous lesion resolution whereas others experience tumor progression are poorly understood. Interactions between CD30 and its ligand may contribute to apoptosis of neoplastic T cells and regression of skin lesions [9].

Several histologic subtypes of LyP have been described, all of which contain a variable infiltrate of CD30+ lymphocytes [10]. Because the histologic subtypes do not differ clinically, histologic subtyping of lesions is not thought to be necessary [3, 11].

The differential diagnosis for LyP includes folliculitis, arthropod assault, pityriasis lichenoides et varioliformis acuta, and pityriasis lichenoides chronica. Since histologic features are consistent with C-ALCL, LyP can inappropriately be treated with chemotherapy by those unfamiliar with this condition. C-ALCL and LyP fall along a disease spectrum and have overlapping clinical, immunophenotypical, and histologic features. Disease course and clinical appearance are typically used as decisive criteria for definitive diagnosis. Longitudinal evaluation typically discriminates between the two entities, although the term "borderline case" is used for patients in whom a clear distinction cannot be made [3, 7].

Given the excellent long-term prognosis, recent consensus guidelines indicate that patients can be monitored off therapy [12]. Methotrexate is considered a first-line treatment for symptomatic disease although recurrence is reported with the discontinuation of therapy [13]. Other reported treatments include topical corticosteroids, mechlorethamine, radiotherapy, imiquimod, carmustine, excimer laser, PUVA, interferon-alpha, and brentuximab [8].

Conclusion

We report a case of a 67-year-old man who presented with lymphomatoid papulosis, with necrotic papules that have been intermittently present for over forty years.

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