Long-term Follow-up of a Case of Lymphomatoid Papulosis with a Benign Course

Dear Editor,

We present the case of a 40-year old male patient with lymphomatoid papulosis of a waxing and waning course on whom three biopsies were performed during a 14-year period with no change in histopathological or immunophenotypical characteristics.

Lymphomatoid papulosis (LP) is a chronic, recurrent, self-healing papulonodular skin eruption with the histopathologic features of a cutaneous T-cell lymphoma but an often benign and indolent clinical course (1). It is designated as a primary, cutaneous, CD30+ lymphoproliferative disorder. The histopathologic features of LP are variable, with five main types (A-E) and several other variants (2). In most cases, LP presents with a generalized eruption of reddish-brown papules or nodules usually smaller than one cm on the trunk and limbs. Rarely, large, rapidly growing nodules may be the first manifestation of the disease (3). Patients with LP have an excellent prognosis even though they are at increased risk of developing secondary cutaneous or nodal lymphomas such as mycosis fungoides, primary cutaneous anaplastic large cell lymphoma (PC-ALCL), or Hodgkin lymphoma (4). LP-associated lymphomas develop in between 10% and, as recently reported, 52% percent of patients and may occur before, concurrent with, or after the onset of LP (4,5).

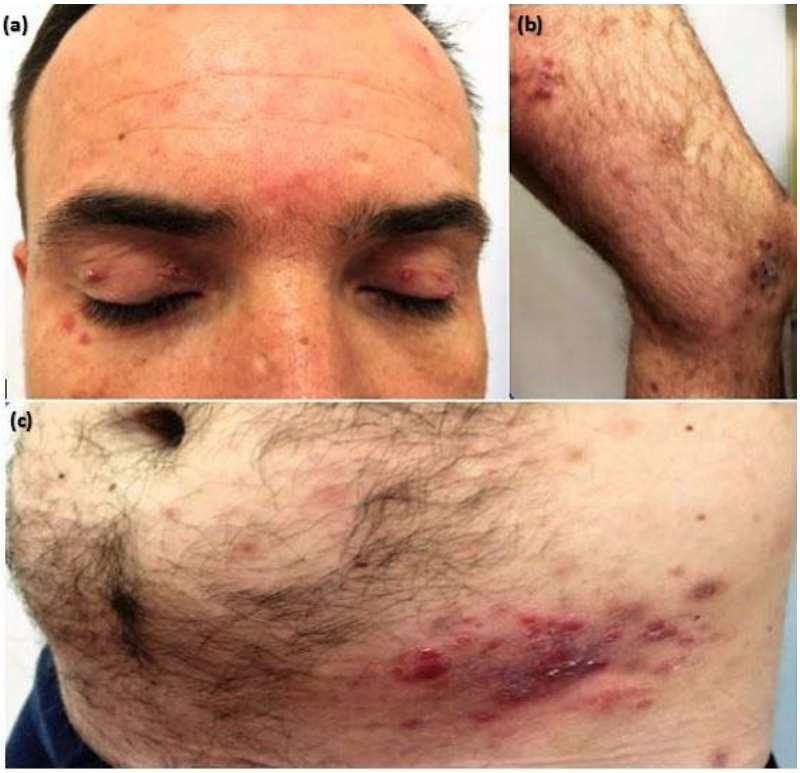

Figure 1. Recent exacerbation of the disease. (a) Multiple papules and small nodules on the eyelids and face. (b) Clustered erythematosus papules and nodules, partially necrotic and ulcerated with scarring and post-inflammatory hyperpigmentation. (c) Tumor on the abdomen with smaller satellite papules, mimicking the clinical picture of primary cutaneous anaplastic large cell lymphoma (PC-ALCL).

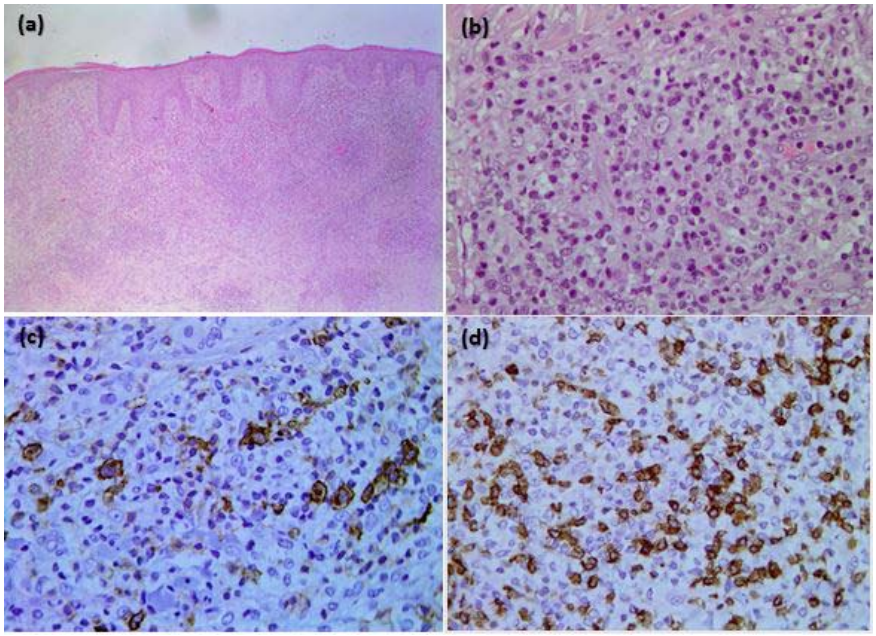

Figure 2. Lymphomatoid papulosis, type A. (a) Wedge-shaped infiltrate of predominantly clustered, large, atypical lymphocytes admixed with small lymphocytes, eosinophils, histiocytes, and neutrophil in the dermis (hematoxylin and eosin ×4). (b) Large, atypical cells arranged within collagen bundles (hematoxylin and eosin ×40). (c) Immunohistochemical staining for CD30 shows positive atypical cells in small clusters. (d) Staining for CD3 highlights the positive cells.

Our patient was diagnosed with "conventional" type An LP 14 years earlier based on the clinicopathologic correlation. The diagnosis was confirmed a year later after excision of a rapid growing ulcerated nodule on the forearm measuring 17 mm in diameter, which was clinically suspected to be anaplastic large cell lymphoma. During these 14 years, there were only a few worrisome recurrences of the disease, which resolved spontaneously or were successfully controlled with local steroids. During a recent exacerbation, when the third biopsy was performed, the patient presented with a large number of generalized reddish-brown pruritic papules and nodules on the trunk, extremities, neck, and face, predominantly up to one cm, some among which were necrotic and excoriated (Figure 1). There were three sites of clustered papules on the trunk, groin, and neck that resembled large, infiltrated plaques larger than two cm, at a glance mimicking cutaneous lymphoma (Figure 1, b, c). There were also older residual hyper- and hypopigmentations on the skin with prominent scarring. Excisional skin biopsy of one larger papule from the abdominal plaque was performed and was not morphologically or immunophenotypically different from the previous ones (Figure 2). Immunohistochemistry showed expression of CD 30 and the phenotypic markers of T-helper lymphocytes (CD 3+/-, CD4+) by neoplastic cells (Figure 2, c, d). Associated systemic malignant lymphoma was excluded based on examination findings, normal laboratory tests, the absence of palpably enlarged lymph nodes, hepatosplenomegaly, and systemic symptoms followed with MSCT. Serology for HIV and EBV was performed and was positive for EBV EBNA, VCA IgG, and IgM, which could be associated with the exacerbation of LP. Topical corticosteroids and phototherapy were administered when needed in this 14-year period, and methotrexate in a lower dose was prescribed during the extensive generalized eruptions. All of the applied therapeutic modalities led to a partial response.

LP is a self-limiting disease for which many patients do not require specific treatment. Therapy should be directed at controlling symptoms in generalized eruptions or minimizing the frequency of recurrences, but none of the available treatment options disrupt the natural history of LP or reduce the risk of developing an associated lymphoma (6). Low-dose methotrexate is the initial therapy of choice in patients with the extensive or symptomatic disease or disease involving cosmetically sensitive areas like the face or hands, which were the affected areas in our patient (6,7). There are no markers that can help predict the course of the disease in a given patient, although some indicators have been suggested (8,9). Because of this lack of markers that can help predict the course of the disease and occurrence of malignant lymphoma, patients should remain in monitoring for the rest of their life.

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