CASE REPORT

A SEVERE CASE OF LYMPHOMATOID PAPULOSIS TYPE B SUCCESSFULLY TREATED WITH LOW-DOSE METHOTREXATE

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ABSTRACT

Lymphomatoid papulosis type B, also defined as "Mycosis fungoides – like lymphomatoid papulosis", is a rare type of cutaneous lymphoproliferative disorder, characterized by chronic, recurrent, self-limiting papulonodular rash.
The diagnosis is established through the correlation of clinical, histopathological, immunohistochemical and molecular genetic evaluation. The disease has a benign course but further investigations are necessary to exclude a systemic lymphoid malignancy.
The available treatments are not curative but may dramatically improve the severity and the frequency of the episodic flares.
Here we present the case of a 68-year-old female patient with a disseminated lymphomatoid papulosis type B evolving for 10 years. She had only been kept under observation, with no systemic treatment during the last decade. We rendered treatment with low-dose methotrexate, as recommended by current guidelines. The course of the disease was satisfactory, with a complete regression of the skin lesions and no development of new lesions. We report this case to evidentiate the role of immunosuppressive therapy in the stabilization of this unpredictable, extremely rare condition.

Keywords: Lymphomatoid papulosis, case report, cutaneous lymphoproliferative disorder.

INTRODUCTION

Lymphomatoid papulosis type B is a rare disease, often misdiagnosed, characterized by a chronic, recurrent, self-limiting papulonodular rash (1). Although the disease is generally associated with a good prognosis, there is a risk of malignancy and patients may develop secondary lymphoma (5). No clear predictive markers for disease progression are currently available. Thus, lifelong monitoring is required (9).

The characteristic feature of this condition is its benign clinical evolution combined with aggressive cytological and morphological characteristics closely mimicking lymphoma.

The disease was first described by Macaulay in 1968 and since then there are different opinions as to whether lmphomatoid papulosis is a benign, premalignant, or malignant condition (1,2).

Currently, lymphomatoid papulosis is considered by the World Health Organization (WHO) (3) as a type of primary cutaneous CD30+ T-cell lymphoproliferative disorders.

The condition is extremely rare, with an overall prevalence rate of 1.2 to 1.9 cases per 1 million population (4).

The available treatments are not curative but may dramatically improve the severity and the frequency of the episodic flares (9).

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CASE PRESENTATION

A 68-year-old female patient presented to the dermatology department at Elias University Emergency Hospital, with a history of recurring skin lesions of 10 years duration. On presentation, she had a severe, disseminated, itchy eruption, consisting of waxing and waning papules and erythematous nodules, in different stages of evolution, worsening dramatically in the last 3 years (Figure 1). The physical examination also showed some residual hyperpigmented macules from previous lesions, with scarring. The lesions were not accompanied by fever, palpable peripheral lymph nodes or systemic ill-health. She was otherwise healthy and her family history was unremarkable.

Laboratory investigations, abdominal ultrasound and chest radiography were normal. There were no atypical cells in the peripheral blood smear. The correlation of histopathological and immunohistochemical examinations confirmed the diagnosis of lymphomatoid papulosis type B, CD3 positive, CD4 positive, CD8 negative, CD30 negative (Figures 2 and 3). Due to technical inadequacies and high costs, we were unable to perform a molecular genetic evaluation of our patient.

UVB 311 nm phototherapy was initiated and systemic therapy with methotrexate 15 mg weekly was instituted, which resulted in the improvement of pruritus and remission of the disease. Currently, the patient is still under regular follow up at our department with a stable clinical picture and no evidence of malignant transformation after 6 months from the initial presentation.

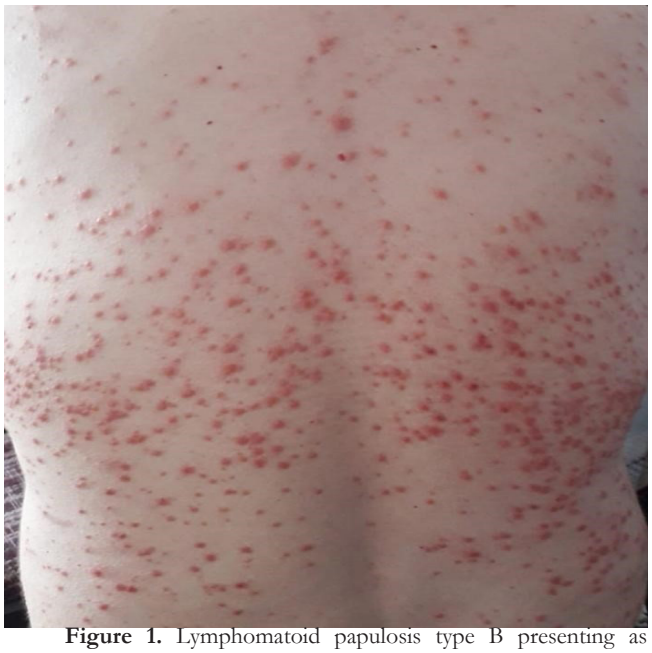

Figure 1. Lymphomatoid papulosis type B presenting as multiple erythematous papules and nodules on the back

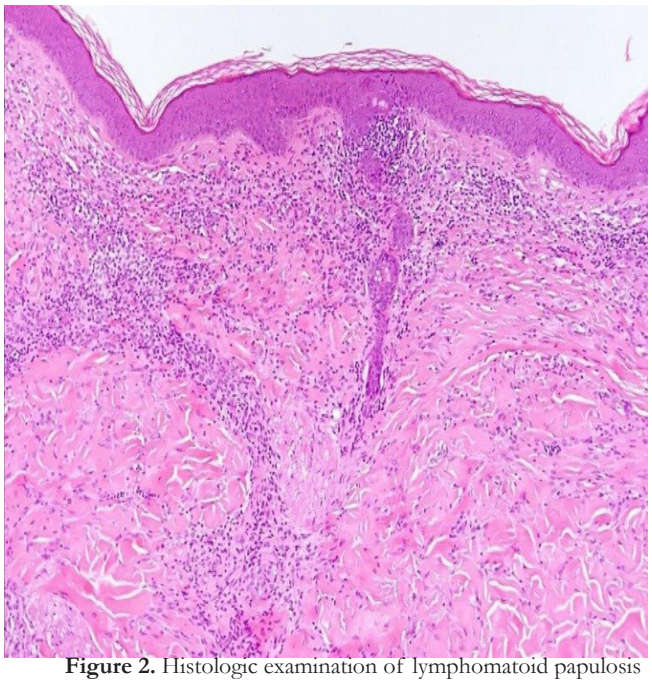

Figure 2. Histologic examination of lymphomatoid papulosis

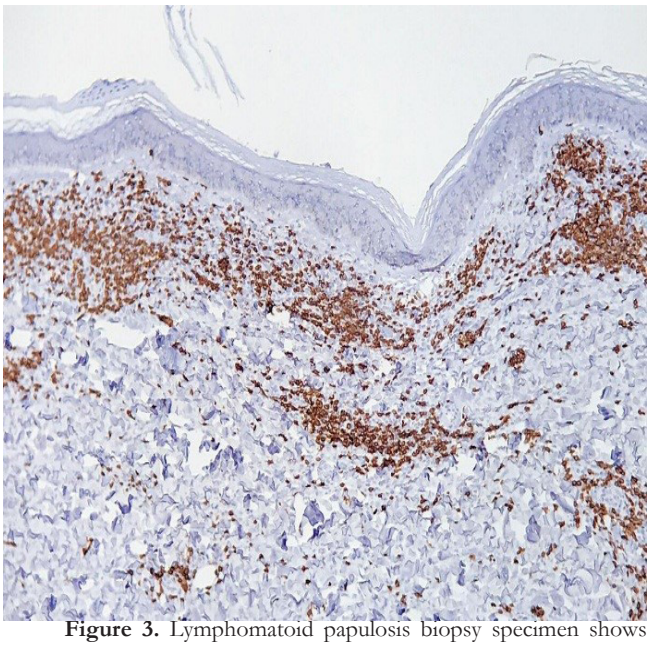

Figure 3. Lymphomatoid papulosis biopsy specimen shows cytoplasmic membrane

DISCUSSIONS

Lymphomatoid papulosis generally occurs in adults. Lesions are mostly red-brown asymptomatic papules and nodules, which usually slowly involute within a period of 3-8 weeks, leaving hypopigmentation, hyperpigmentation or scarring (2,3).

Clinically, the disease resembles many conditions and the biopsy is needed to make a correct diagnosis.

Lymphomatoid papulosis has been histopathologically classified into 3 subtypes: A, B, and C. Type B, the least common subtype of lymphomatoid papulosis, resembles Mycosis fungoides. Lymphocytes of this subtype may not express the CD30 antigen, as in our case, but are generally CD3 positive, CD4 positive, and CD8 negative(2). The characteristic histopathological and immunopathological findings are essential to establish the correct diagnosis.

The disease is often misdiagnosed, because it can mimic or even coexist with other conditions including PLEVA, mycosis fungoides, or other cutaneous lymphomas including large cell anaplastic lymphoma (5). In our case, the lack of type B symptoms and signs of systemic involvement supported the diagnosis of a benign process. Lymphomatoid papulosis type B, CD 30 negative, with epidermotropsim, Pautrier microabscesses, may be difficult to differentiate from Mycosis fungoides. However, in the context of the clinical picture and due to the long and benign course of the disease, the diagnosis is suggestive for lymphomatoid papulosis.

Treatment of LyP is generally unsatisfactory (5). Available options include topical corticosteroids, narrowband UVB therapy, low-dose methotrexate or PUVA therapy (5). In severe cases, with scarring or numerous papulo-nodular lesions, low-dose methotrexate (5-20 mg/week) has shown to be effective (6,7).

Overall, the duration of the disease might range from a few months to more than 40 years. Many studies estimate that in 10-20 % of cases, the disease may be preceded, accompanied or followed by the development of lymphoma, generally Mycosis fungoides, CD30+ large T-cell lymphoma or systemic Hodgkin's disease (2,5,6,8,9). However, the prognosis is usually excellent. In a study of 118 patients, only five patients (4%) developed a systemic lymphoma, and only two patients (2%) died of systemic disease over a median follow up period of 77 months (10).

In conclusion, this case report points out the satisfying course of the disease, treated with a low-dose of methotrexate and the importance of correctly

diagnosing patients with lymphomatoid papulosis due to the possible correlation with other cutaneous and systemic lymphoproliferative disorders.

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