Case Report
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Mycosis Fungoides: A Case Report

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Objectives: Mycosis fungoides (MF) is the most common group of cutaneous T-cell lymphomas. We reported this rare diagnosed case of MF in National Dermatology Center of Mongolia. **Methods:** Diagnosis was based on physical examination and laboratory testing. Bioethical permission for this research was given by the Biomedical Ethics Committee of Ministry of Health in Mongolia, May 23, 2017. **Results:** A 78-year-old woman presented for evaluation of a 7-year history of recurrent pruritic lesions that appeared as red patches and plaques on non-sun exposed areas. She was given the initial clinical diagnosis of psoriasis. Upon examination, the morphology, histopathology and immunohistochemistry indicated MF, IB stage. **Conclusions:** A reliable diagnosis of mycosis fungoides might be feasible only in conjunction with clinical, histopathological, and molecular criteria.

Keywords: Mycosis Fungoides, Lymphoma T-Cell Cutaneous, Histopathology, Immunohistochemistry

Introduction

Mycosis fungoides (MF) is the most common type of cutaneous T-cell lymphoma (CTCL) and affect most commonly middle-aged and elderly adults of all races [1, 2]. The male-to-female ratio is about 2:1[3]. Morphologically, the neoplastic lymphoid infiltrate is epidermotropic and composed predominantly of small to intermediate-sized atypical lymphocytes with enlarged hyperchromatic, cerebriform nuclei and clear cytoplasm (haloed cells). These atypical lymphocytes often colonize the basal layer of the epidermis singly or in a linear fashion, forming a "string of pearls." Pautrier micro abscesses, which consist of small aggregates of atypical lymphocytes are found often in association with Langerhans cases [4, 5]. MF in its early stage may mimic psoriasiform and lichenoid inflammatory dermatoses both clinically and histologically; therefore, the diagnosis remains a major challenge for dermatologists and dermatopathologists. Investigators have proposed histologic criteria distinguishing MF from non-MF [6, 7]. Herein, we report the patient with MF who was diagnosed by different diagnosis.

Case report

A 78-year-old woman presented with generalized hyper pigmented skin, scaly patches and plaques associated with itching from 2010. On examination, the trunk and abdomen and bilateral upper and lower extremities had pink-red, symmetric, concentric annular patches and flat plaques with trailing scale covering 60% to 70% of her body surface area (Figure 1).

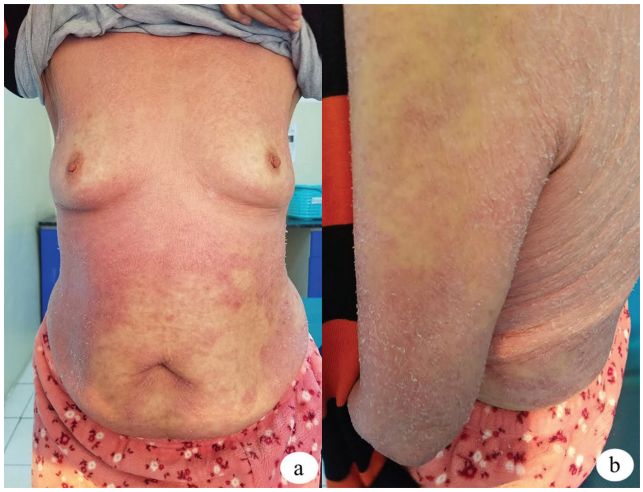

Figure 1. Plaques of Mycosis Fungoides. Erythematous scaly patches (a) and flat plaques (b) on the trunk and upper extremities (on sun-protected skin). No lymphadenopathy or organomegaly was observed.

Histologic examination of the skin punch biopsy showed skin with exocytosis of small abnormal mononuclear cells with irregular nuclei, Pautrier micro abscess, basal alignment of neoplastic lymphocytes (Figure 2), and grandiosity sign. Based on the peripheral blood smear, no Sezary cells were identified.

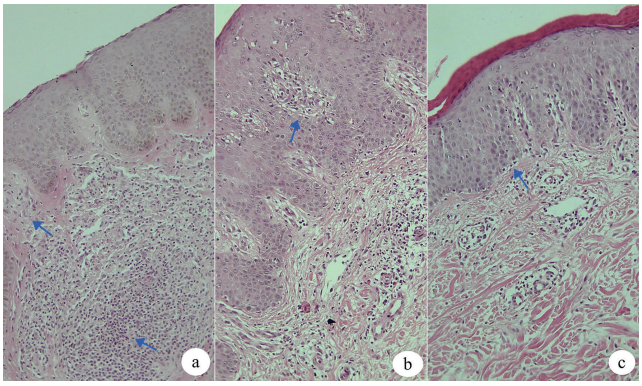

Figure 2. Histopathology images by hematoxylin and eosin at 10x. Papillary dermal neoplastic lymphoid infiltrates with exocytosis (a), Pautrier micro abscess (b) and basal alignment of neoplastic lymphocytes (c).

On immunohistochemistry, the tumor cells were positive for CD2, CD3, CD4, CD5, CD7 and negative for CD8 (Figure 3).

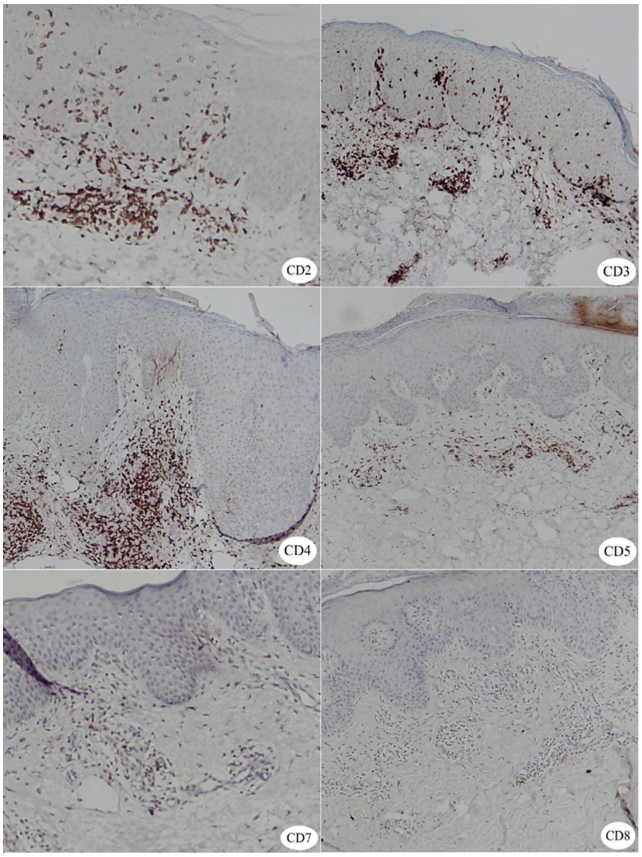

Figure 3. Immunohistochemistry showing positivity for CD2, CD3, CD4, CD5, CD7 and negativity for CD8 in skin biopsy (each 10x)

Based on the clinical features, histology and immunohistochemistry results, the diagnosis was CTCL and its type was the MF with IB stage (T2b N0 M0 B0) according to the TNMB classification.

Discussion

MF is the most common cutaneous lymphoma. It is a relatively rare, extra nodal, non-Hodgkin's lymphoma with a stable incidence of approximately 0.36 per 100,000 person years [8]. It most often presents in those aged 45 to 60 years but has been diagnosed in children and adolescents [9]. MF is classically divided according to its clinical presentation as patches, plaques, or tumors [10]. The disease progresses slowly for years, evolving from erythematous patches on sun-protected skin to plaques and then to tumors and erythroderma [10].

Diagnosis Mycosis fungoides in earliest stages may be prognostically or therapeutically important in patients [2]. However, accurate diagnosis of early MF remains elusive because of clinical and histological features which overlap with those of benign conditions [11, 12]. The histopathologic features of MF in an early stage vary from person to person, over time, and even between multiple sites in a single patient [13, 14]. MF should also be distinguished from several benign inflammatory conditions, including lichenoid keratosis, lymphomatoid eczematous dermatitis, lymphoid drug eruptions, and the inflammatory stage of lichen sclerosis, among many other [15]. On the other hand, in many cases of early MF, "nonspecific" histopathologic features may be observed in a given specimen, often due to long periods of improper "therapy resistant" dermatitis [15-18] treatment before the correct diagnosis is made by biopsy

The diagnosis is difficult especially in the early stages, but it is made through a combination of the clinical picture and examination, and is confirmed by a skin biopsy (two or more times) and staged appropriately based on the results. Others studies focused on the histopathologic features of this correlation and confirming the characteristic histologic features of MF in early stage include enlarged epidermal lymphocytes with cerebriform nuclei within the epidermis and epidermotropism, bandlike infiltrate in a thickened papillary dermis with coarse collagen bundles, lymphocytes with perinuclear clearing, and a linear arrangement of boiler epidermal lymphocytes along the dermal-epidermal junction [19].

In our case the histology report revealed prominent exocytosis of the lymphocytes with cytological atypia. The cells are irregular and convolute infiltrating mainly the basal cell layer of the epidermis. In the superficial to mid-dermis, there were mild to moderately prominent perivascular and interstitial cell infiltrates consisting of round lymphocytes, eosinophils, and atypical lymphoid cells. These findings are similar to standard histologic criteria. The presence of Pautrier's Microabscess is considered more specific for MF but is observed in only few cases in an early stage [14, 20].

The MF tumor cells are characterized by epidermotropic peripheral T lymphocytes whose phenotype is CD2+, CD3+, CD4+, CD5+, and, in some cases, T lymphocytes may present CD4-, CD8+ and others [11]. The case described here presented CD2+, CD3+, CD4+, CD5+, just a few CD7+ and CD8- cells phenotypes.

A reliable diagnosis of MF might be feasible only in strong correlation with clinical, histopathological and sometimes molecular findings to exclude benign inflammatory diseases, more aggressive primary cutaneous lymphomas and other.

Conflict of Interest

The authors declare that they have no competing interests.

Acknowledgments

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