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Case Report

An Aggressive Epidermotropic Type of Cutaneous T-Cell Lymphoma: A Case Report

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Abstract

Mycosis Fungoides (MF) is the most common primary cutaneous lymphoma. Co-expression of CD4 and CD8 in a subset of atypical T-lymphocytes of MF is rare. Herein, we reported a case of CD4/CD8 dual-positive MF in a 70-year-old man. The biopsy demonstrated significant epidermotropism of atypical, hyperchromatic, cerebriform of lymphocytes. Immunohistochemical staining revealed that the lymphoid infiltrate is CD4/CD8 double positive with CD5 and CD7-positive.

Introduction

Mycosis Fungoides (MF) is the most common type of Cutaneous T-Cell Lymphoma (CTCL) and represents nearly 50% of all primary cutaneous lymphomas [1]. MF is characterized by an epidermotropic skin infiltrate of atypical CD4+ phenotype lymphocytes. In a minority of MF cases, the atypical infiltrate is CD8 positive. The later form of CTCL generally follows a rapid, aggressive clinical course [2]. Herein, we reported a case of CD4/CD8 dual-positive MF, which is rarely described in the literature.

Case Report

A 70-year-old man presented to Dermatology Department at Southwest Hospital for evaluation of symptomless erythematous plaques and nodules for just one year. Physical examination revealed disseminated well-defined eruptive papules, nodules and mushroom-shaped tumorous masses with central ulceration and necrosis (Figure 1). He had received several treatments including NB-UVB (Narrow Band Ultraviolet B) and topical corticosteroid, but failed to prevent the development of the disease. Histopathologic examination showed significant epidermotropism of atypical, hyperchromatic, cerebriform of lymphocytes colonizing the epidermic basal layer in a linear or Pautrier pattern (Figure 2). In addition, we found that atypical lymphocytes infiltrated deeper with the development of MF from the papules, to nodules and tumorous masses (Figure 2). Immunohistochemical analysis revealed that the infiltrating neoplastic cells were CD4+, CD5+, CD7+, CD8+, Ki67+, and CD20- (Figure 3). The human immunodeficiency virus and Human

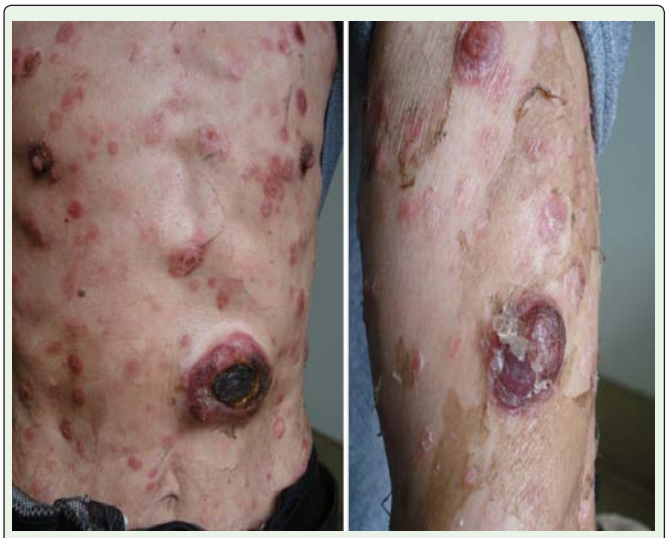

Figure 1: The clinical manifestation of this patient. Physical examination revealed disseminated well-defined eruptive papules, nodules and mushroom-shaped tumorous masses with central ulceration and necrosis.

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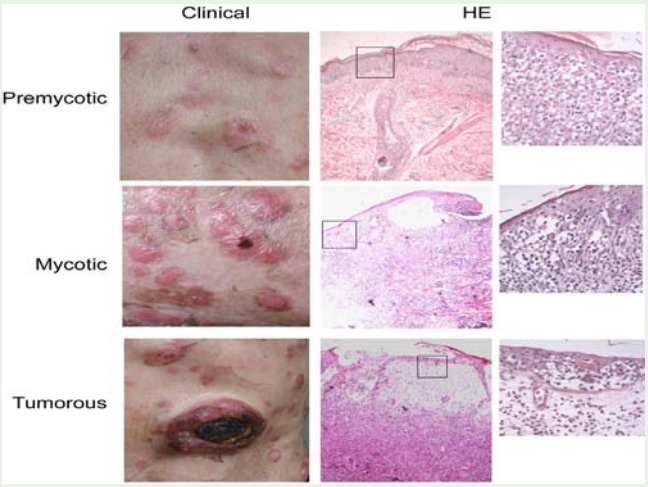

Figure 2: Histopathological examination of this patient (HE). Histopathologic examination showed significant epidermotropism of atypical, hyperchromatic, cerebriform of lymphocytes colonizing the epidermic basal layer in a linear or Pautrier pattern. Atypical lymphocytes infiltrated deeper with the development of MF from the papules, to nodules and tumorous masses.

T-Lymphotropic Virus (HTLV)-1 serologic status were negative. The clinical manifestations, histopathological and immunohistochemical results supported the diagnosis of CD4/CD8 double positive MF.

The patient was given Acitretin (30mg/d) and electron beam radiotherapy. Unfortunately, he died from severe lung infection four months after the first visit.

Discussion

The classical CD4+ Alibert-Bazin type of MF and its clinical and/or histologic variants have an indolent clinical course with slow progression over years or sometimes decades, from patches to more infiltrated plaques and eventually to tumors [3-5]. For CD8+ MF, several reports showed that CD8+ MF had more aggressive clinical behavior and progressed in a short time from scaly macules to plaques

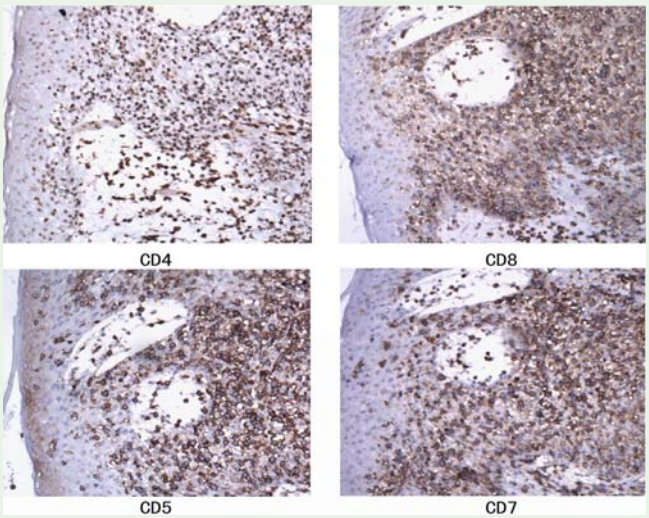

Figure 3: The results of the immunohistochemical stainings. Immunohistochemical studies revealed that the infiltrating neoplastic cells were CD3+, CD4+, CD5+, CD7+, CD8+.

to ulcerated lesions, and led to infiltrate into other organs and death [6]. However, others reported that CD8+ MF was considered to have the same clinical behavior and prognosis as CD4+MF [7]. Recently, there have been only two reports of CD4/CD8 dual positive MF [8,9]. It has been demonstrated a switch between CD4 and CD8 phenotype during the course of CTCL, and the ratio of CD4 to CD8 has been shown to change during the course of the disease [10]. We suppose that this phenotype switch may be associated with the immune microenvironment, especially certain infection triggers. There should be loss of pan-T-cell antigens including CD5 and CD7 in MF, but CD5 and CD7 were positive in our case. The above mechanism of immune microenvironment disorder may also play certain roles in CD5+CD7+ phenotype. Moreover, the immunophenotypical shift between CD4 and CD8 in CTCL may be associated with the aggressive disease progression and/or prognosis. All these hypotheses need further investigations to confirm.

In brief, this case we reported presented a spectrum ranging from the papules to the tumor formation within a short time, suggesting an aggressive behavior and leading to the patient's death. Although we presented just one case and clinically meaningful conclusion is difficult to establish, this case report provides an important resource for disease classification and disease staging, meanwhile provides a key basis for the diagnosis and treatment of the disease.

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