Is Mycosis Fungoides Truly an Indolent Pathology?

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Abstract

The course of Mycosis Fungoides (MF) is indolent except when the transformation to a large T-cell lymphoma occurs. The diagnosis of transformed MF relies on the presence of more than 25% of large cells on biopsy of an MF lesion. Treatment of transformed MF is a major challenge as these patients have poor outcome. Here, we report a 40-year-old man presenting in an advanced stage of disease with nodules all over the body diagnosed as cutaneous CD30(−) large cell transformed MF of fatal evolution.

Keywords: Cutaneous T-cell lymphomas; Transformed mycosis fungoides; Tumor stage; CD30-

Introduction

Mycosis Fungoides (MF) is the most common type of T-cell lymphoma. Its development is generally slow and indolent, however it can transform and acquire a poor prognosis [1,2]. We describe an original and impressive case of transformed MF.

Case History

Patient 40-year-old male consults the emergency room for generalized erythematous nodules that appeared suddenly 20 days earlier, associated with profuse sweating, fever and weight loss. The patient had pruritic erythematous plaques of the trunk for 10 years treated with corticosteroids by self-medication. On examination, there were about a hundred erythemato-violins tumors, rapidly extensive, that became confluent in large poly-lobed placards affecting almost the entire skin surface (Figure 1). The skin biopsy showed a diffuse infiltration of atypical lymphocytes with vesicular nuclei involving the full thickness of dermis and extending to the superficial part of the hypoderm. Of all infiltrating atypical lymphocytes, about 60% were large cells. On immunohistopathologic examination, infiltration was found to be CD3 (+), CD4 (+), CD8 (-) and CD30 (-), Ki67 more than 90% (Figure 2-4). There were no palpable lymph nodes or hepatomegaly. Anti-HIV and...
Anti-HTLV serologies were negative. Results of a complete blood count, liver function, and lactate dehydrogenase were within normal limits. Peripheral smear showed no atypical cells. There was no involvement of the bone marrow and CT scans of the chest, abdomen and pelvis showed no systemic involvement. Based on the clinical manifestation and histopathological findings, the patient was diagnosed with transformed MF. The staging of disease was T3N0M0, Stage IIB. A chemotherapy, CHOP protocol (cyclophosphamide, vincristine, doxorubicin, prednisone), was established, allowing thus a rapid resolution of the tumors at the first cure, then a complete bleaching after the third (Figure A). After the 5th cure of CHOP, the disease flared with infiltrated erythematous plaques as well as generalized papulo-nodules. Intensification of chemotherapy with Isosfosphamide-Carboplatin-Etoposide, allowed a spectacular but transient control of the disease. An allograft was then proposed, but the patient died due to septic shock.

**Discussion**

Mycosis fungoides is usually indolent cutaneous T-cell lymphomas, which can evolve as macules or infiltrated plaques over many years [1]. Rarely, transformation to large cell lymphoma may be seen and in this case, clinically it may be more aggressive [2]. Transformation to a large cell in MF is defined as showing large cells (>4 times the size of a small lymphocyte) in 25% or more of the dermal infiltrate or forming microscopic nodules in which large cells are CD30 (+) or CD30 (-) [3,4]. A recent article from the Dutch Cutaneous Lymphoma Group suggests that CD30 (+) large-cell tumors in patients with MF carry a better prognosis than CD30 (-) large-cell tumors [4]. This transformation, which is possible at any stage of the disease, is more frequent as the stage of the disease is advanced [2]. It has a pejorative use a better word prognostic value, especially in case of nodal or visceral involvement. In 6% and 23% of cases, the disease will develop into a highly malignant lymphoma with extensive spread to various organs [2]. The extent of skin involvement, high lactate dehydrogenase levels, transformation at the time of initial diagnosis of MF, and especially the occurrence at an old age beyond 60 years, as well as extracutaneous involvement, are factors of poor prognosis. They shorten the evolution of MF, which is usually slow and insidious [5].

**References**