Mycosis fungoides with large cell transformation (CD30+) and B-cell chronic lymphocytic leukemia

Mycosis fugnoides (MF) is an indolent cutaneous T-cell lymphoma (CTLC) and is the most common of all cutaneous lymphomas. An increased risk for developing a second primary malignancy in patients with CTCL has been described in several studies, with a range from 1.04 to 2.4 (1-4). Caucasian males are at higher risk for MF development. MF is often diagnosed at ages between 55 and 67 years, and second malignancy usually occurs 5 or 6 years after the diagnosis of MF was established (5). The most common second primary malignancies include non-Hodgkin lymphoma (NHL), Hodgkin lymphoma (HL), lung carcinoma, bladder carcinoma, and melanoma. Even though a higher incidence rate of all NHL was described in patients with MF (15/1000) in comparison with the general population (0.32/1000), there are still only a few cases of B-cell NHL following MF described in the literature (6,7). We describe a rare case of a patient with MF and simultaneous large cell transformation (LCT) and a small B-cell lymphocytic lymphoma/chronic lymphocytic leukemia (B-CLL).

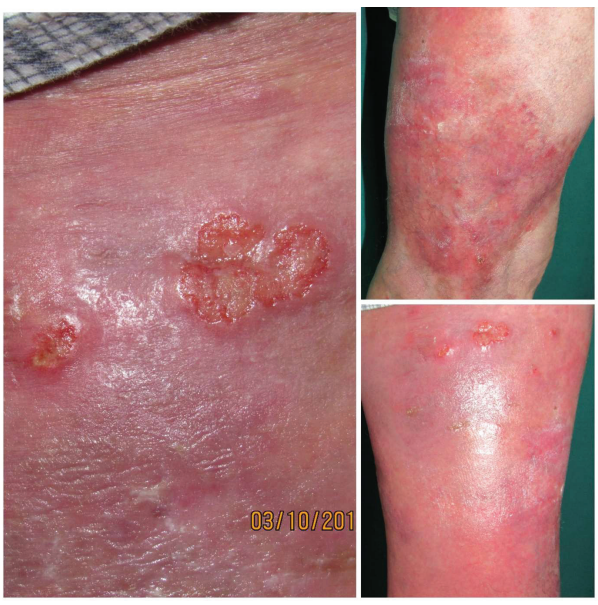

Figure 1. Clinical presentation of an ulcerated tumorous lesion on an erythematous plaque on the right tight.

In 2017, an 82-year-old man previously treated for MF presented with two fast growing tumorous lesions with ulceration on the right tight (Figure 1). A biopsy was performed, and a diagnosis of MF with LCT was established (Figure 2). During hospitalization, mild leukocytosis (12.2 x10^9 L^-1), lymphocytosis (64%, total count of 7.81 x10^9 L^{-1}), and anemia were found. Bone marrow biopsy was not performed due to low pain threshold. Bone marrow aspirate showed 70% of atypical lymphocytes and few "smudged" cells. Immunophenotyping by flow cytometry detected 49% monoclonal kappa+ B-cells with phenotypic features typical for B-CLL (CD5+, CD23+, kappa +). Of overall bone marrow cells, the ratio of monoclonal kappa + B-cells with the B-CLL phenotype was 21%.

Immunophenotyping of peripheral blood showed up to 50% monoclonal kappa+ B-cells with phenotypic features typical for B-CLL (CD5+, CD23+, kappa +). Of overall peripheral blood cells, the ratio of monoclonal kappa+ B-cells with the B-CLL phenotype was 28%. Multi-sliced computed tomography was within normal ranges. A flow cytometry showed lymphocytes with phenotypic findings for CD20+ B-CLL. A diagnosis of MF with LCT (CD30+) clinical

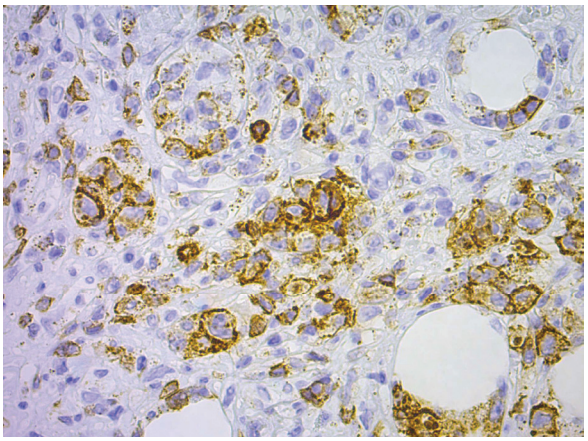

Figure 2. There was diffuse infiltrate of mostly small and medium size atypical lymphocytes with a large number of CD30-positive large cells in the dermis (immunohistochemistry for CD30, original magnification x400).

grade IIB (T3, N0, M0) and B-CLL was established. The patient was treated with fractionated superficial irradiation that resulted in applanation and regression of the tumorous lesions. No hematologic treatment was indicated other than regular follow-up. On dermatologic follow up for 2 years, the patient was stable, with no active skin lesions and no progression of MF. The patient was subsequently lost to follow-up.

This is a rare case of MF with LCT and B-CLL occurring simultaneously. Large cell transformation in patients with MF can occur in 20-55% of advanced MF, as in our case, and this something physicians must be aware of, so repeated biopsies are advised (8). We also should keep in mind that patients with MF are at higher risk of developing a second malignancy. Of those second malignancies, a coexistence of lymphoproliferative disorders in two lineages, T-cell and B-cell, such as CTCL and B-CLL, is very uncommon, and only a few cases have been published (6,7,10). In most of these cases, CTCL preceded B-CLL, and with the only established explanation being increased risk of second malignancy in patients with CTCL (3,5,10). Other explanatory hypotheses include neoplastic stem cells, a genetic predisposition to malignancy, the use of immunosuppressive agents for the treatment for a first neoplasm, viral agents, and modulation of the B-cell system by monoclonal T-cell proliferation (1,5,6,9,10). Regular follow-up is mandatory for all patients with CTCL as well as MF, in order to identify the disease progression but for the timely detection of second malignancies.

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Received: May 9, 2022

Accepted: December 1, 2023