

Mycosis Fungoides Associated with Kaposi's Sarcoma, T-cell Rich B-cell Lymphoma, and T-cell Lymphoma with Angioimmunoblastic Features

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ABSTRACT A patient with mycosis fungoides (MF), Kaposi's sarcoma, T-cell rich B-cell lymphoma, and T-cell lymphoma with angioimmunoblastic features is described. The appearance of multiple malignancies in this patient may have been caused by previous exposure to radiation in the Chernobyl accident and/or systemic chemotherapy for the initial T-cell rich B-cell lymphoma which he underwent.

KEY WORDS: mycosis fungoides; Kaposi's sarcoma; cutaneous lymphoma

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INTRODUCTION

Mycosis fungoides (MF) is the most common type of primary cutaneous T-cell lymphoma. The disease is initially characterized by slightly scaly erythematous patches or plaques, which later may evolve into tumors and/or involve the entire surface of the body, finally culminating in blood and visceral dissemination (1). Many clinicopathological variants of MF have been described in the literature (2). Atypical presentations often cause a delay in the diagnosis because these forms tend to imitate other inflammatory conditions.

In the present case report, we describe an unusual type of MF associated with other hematological malignancies in the same patient.

CASE REPORT

A 55-year-old man, born in Ukraine and a survivor of the Chernobyl disaster in 1986, was hospitalized to investigate a disseminated skin eruption. His past medical history included ischemic heart disease, hyperlipidemia, hypothyroidism, diabetes mellitus, hypertension, and rheumatoid arthritis. Two years prior to his admission he underwent treatment with CHOP (cyclophosphamide, hydroxydaunorubicin, oncovine, and prednisolone) for a systemic T-cell rich B-cell lymphoma, leading to a complete remission. In addition, Kaposi's sarcoma (KS) of the lower extremities with diffuse lymph node involvement had been diagnosed one year prior to his hospitalization and was treated with vincristine and radiation.



Figure 1. Brown, poorly demarcated, firm, non-tender plaques over the lower back.

The patient presented with a mildly pruritic skin eruption associated with bilateral inguinal lymph node enlargement. He was previously treated with UVA1 phototherapy at another medical center where a skin biopsy had been interpreted as suggestive of morphea.

The clinical examination revealed a generalized symmetric rash over the trunk, abdomen, thighs, and buttocks, composed of brown, poorly demarcated, firm, and non-tender plaques, which were considered to be morphea (Figure 1). In addition, Kaposi's sarcoma nodules were observed over the lower extremities (Figure 2), and bilateral inguinal lymph node enlargement was detected.

The routine blood tests were normal, except elevated serum levels of C-reactive protein, lactic dehydrogenase (LDH) and $\beta 2$ microglobulin.

The skin biopsy obtained from one of the plaques revealed a subepidermal, perivascular, and interstitial dense infiltrate of small to medium pleomorphic lymphocytes in the upper dermis. In addition, there was exocytosis of lymphocytes into the epidermis as



Figure 2. Pinkish nodules of Kaposi's sarcoma over the lower extremities.

simple units, pairs, and triplets without parakeratosis and with only slight spongiosis (Figure 3). Immunohistochemical staining of the skin biopsies revealed a predominance of CD4⁺ CD3⁺ cells with a small number of CD8⁺ cells. CD30⁺ cells were not identified. The histopathologic findings were compatible with MF.

Inguinal lymph node biopsy showed complete effacement of the lymph node architecture due to diffuse infiltration of small to medium sized lymphocytes characterized by a pale cytoplasm. There was also marked proliferation of arborizing vessels (Figure 4). Immunohistochemical studies found positive staining for CD3, CD4, CD5, CD10, and bcl6 of the neoplastic cells. Positive staining for CD21 highlighted an expanded dendritic framework. Stains for CD31, CD34, and factor VIII highlighted the expanded vascular proliferation, and 80% of the cells also stained positive for Ki67. Staining for HHV8 (LANA) was negative.

There was no evidence of Sézary cells in the blood smear. Polymerase chain reaction (PCR) analysis of DNA extracted from a lesional skin biopsy showed monoclonality of the T-cell infiltrate. Involvement of the bone marrow was ruled out by biopsy. Positron emission tomography-CT (PET-CT) demonstrated increased fluorinated deoxy glucose (FDG) uptake in the axillary, retroperitoneal, and inguinal lymph nodes.

The clinical, histopathologic and immunohistochemical findings were compatible with a diagnosis of morphea-like mycosis fungoides (MF) in the skin and peripheral T-cell lymphoma with angioimmunoblastic features in the lymph nodes.

The patient was treated with several courses of chemotherapy, including fludarabine, cytoxan, and

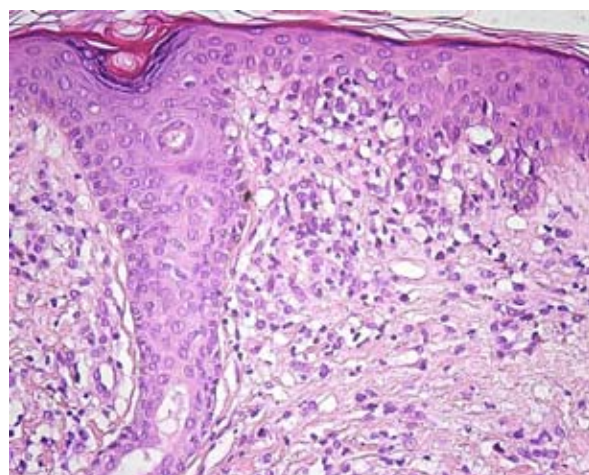


Figure 3. A skin biopsy from a lesion on the trunk showing perivascular and interstitial pleomorphic lymphocytic infiltrate in the papillary dermis with focal exocytosis into the epidermis (hematoxylin-eosin $\times 200$).

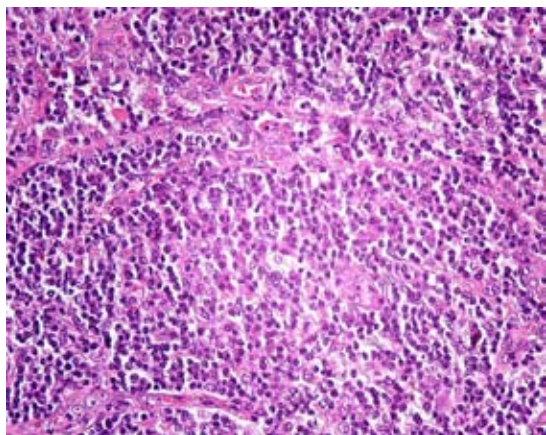


Figure 4. Inguinal lymph node biopsy showing small to medium sized lymphoid cells and marked proliferation of arborizing vessels (hematoxylin-eosin $\times 200$).

methotrexate, with only partial and transient response; consequently he underwent successful allogeneic bone marrow transplantation with resolution of the skin and lymph node involvement over a two-year follow-up period.

DISCUSSION

We described three major hematological malignancies in a single patient: systemic T-cell rich B-cell lymphoma, MF, and T-cell lymphoma with angioimmunoblastic features. Additionally, the patient had KS. Several etiologic factors may underlie the unusual coexistence of these multiple and infrequent malignancies in a relatively young individual: previous exposure to massive amounts of ionizing radiation (the patient is a survivor of the Chernobyl disaster), chemotherapy in the past due to T-cell rich B-cell lymphoma, and the fact he was affected by an autoimmune disease (rheumatoid arthritis).

The appearance of lymphoid infiltrate in the skin lesion raised the question whether the cutaneous lesions were an expression of skin involvement by peripheral T-cell lymphoma with angioimmunoblastic features or MF. Cutaneous involvement of the former may be present in 50% of cases, most often as macules and papules over the trunk and extremities. According to the literature, there are 4 histologic patterns of skin involvement by peripheral T cell lymphoma with angioimmunoblastic features consisting of (a) a superficial perivascular infiltrate with or without atypical lymphocytes; (b) a dense superficial and deep perivascular infiltrate of atypical lymphocytes; (c) a leukocytoclastic vasculitis-like infiltrate; and (c) necrotizing granulomas (3). None of those histologic patterns were recognizable in the skin of our patient, indicating there was no secondary involvement of the skin with peripheral T-cell lymphoma with angio-

immunoblastic features. In addition, the fact that the lymph node enlargement occurred prior to appearance of the skin lesions and the absence of histologic findings compatible with MF in the lymph nodes ruled out the possibility of lymph node involvement with MF and led to the diagnosis of two concomitant malignancies of different origin. Unfortunately, no material from the lymph node biopsy was available to molecularly compare the clones detected in the skin and in the nodal tissues.

MF typically presents with slightly scaly erythematous or light brown macular patches, which later evolve into plaques or tumors. Several clinicopathologic variants have been described in the literature over the past several decades. These variants may complicate the diagnosis of the disease because the classical features of the disease may be absent or masked (4). To our knowledge, only one study previously described a morpheiform type of MF resembling our case (5).

CONCLUSION

We have described a patient with MF presenting with morphea-like clinical characteristics associated with two other hematological malignancies and KS which may have been caused by ionizing radiation and/or previous chemotherapy.

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