

Discordant mycosis fungoides and cutaneous B-cell lymphoma: A case report and review of the literature

Cory Maughan, DO,^a Lauren Boudreaux, DO,^b William Lear, MD,^a and Angela Bohlke, MD^a
Salem and Corvallis, Oregon

Key words: B-cell lymphoma; mycosis fungoides; secondary lymphoma.

INTRODUCTION

An elderly woman was seen for evaluation of a pruritic rash on her trunk and proximal extremities. Clinically and histologically the lesions resembled mycosis fungoides (MF). Two additional lesions on her back, with differing morphology, were proven to be cutaneous B-cell lymphoma on pathology and gene rearrangement. Several case reports have shown that MF is a risk factor for the development of a secondary B-cell lymphoma. However, it is extremely rare for the two discordant lymphomas to present simultaneously in the same patient.

CASE REPORT

An 81-year-old immunocompetent woman presented to the dermatology department on referral from her primary care physician for treatment of a pruritic rash. The rash had been present on her trunk for approximately 18 months, and to the knowledge of the patient, the lesions all presented at the same time. She otherwise felt well with no complaints of fevers, chills, night sweats, or weight loss. Upon examination, the patient was found to have multiple pink, scaly patches and plaques clinically suggestive of MF on the trunk and bilateral axillae (Fig 1) in addition to 2 erythematous, indurated plaques on the mid back (Fig 2). There was no palpable lymphadenopathy or hepatosplenomegaly.

A biopsy of the plaque at the right portion of the upper chest, stained with hematoxylin-eosin, found enlarged lymphocytes with hyperchromatic irregular nuclei. Epidermotropism and focal Pautrier microabscesses were present. The papillary dermis was hyalinized. The lymphocytes were highlighted on CD3 staining, and most cells expressed CD4 with little CD8 expression. No significant CD20 staining was present. Periodic acidised CD4 staining was

Abbreviation used:

MF: mycosis fungoides

negative for fungal organisms. The histologic findings were consistent with MF.

Biopsies of the right and left mid back found a dense, diffuse infiltrate of predominantly lymphocytes with histiocytes and occasional plasma cells. The lymphocytes were not significantly enlarged and had angulated to round nuclei and inconspicuous nucleoli. CD20 and CD79a highlighted most of the lymphocytes and a smaller population of CD3⁺ lymphocytes was seen at the base of the lesion. Kappa and Lambda staining highlighted plasma cells, but no significant clonal shift was identified. B- and T-cell gene rearrangement was performed, and a B-cell (IgH) clone was identified. Additional immunostaining was performed and the morphologic features, together with the immunophenotype (CD20⁺, CD79a⁺, bcl6⁺, bcl2⁺partial, CD10⁻) and identification of a B-cell clone, was most suggestive of a low-grade B-cell lymphoma (Figs 3 and 4). In situ hybridization stain for Epstein Barr virus was also performed, and results were negative.

On referral to the oncology department, laboratory results showed a white blood cell count of 6,760, hemoglobin of 12.5, platelet count of 127,000, and basophilia with a basophil count of 3.8%. Sezary cell count showed 5% lymphocytes with features consistent with Sezary cells. Computed tomography scan of the chest found a 3.3-cm enlarged node in the preaortic space, small bilateral axillary adenopathy, and small bilateral pulmonary nodules worrisome for metastatic disease. Bone marrow biopsy found increased numbers of T cells both in small lymphoid

From Silver Falls Dermatology/Western University of Health Sciences^a and Good Samaritan Regional Medical Center.^b

Funding sources: None.

Conflicts of interest: None declared.

Correspondence to: Cory Maughan, DO, 1430 Commercial St. SE, Salem, OR 97306. E-mail: cory.maughan@gmail.com.

JAAD Case Reports 2015;1:219-21.

2352-5126

© 2015 by the American Academy of Dermatology, Inc. Published by Elsevier, Inc. This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>).

<http://dx.doi.org/10.1016/j.jidcr.2015.04.015>



Fig 1. Erythematous scaling plaque clinically suggestive of MF on the right upper portion of the chest.

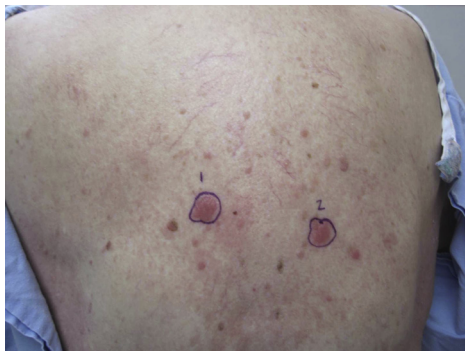


Fig 2. Erythematous, indurated plaques on the midback demonstrating B-cell lymphoma.

aggregates and scattered diffusely through the marrow. Flow cytometry found gating on 19% of cells, 87% CD3, 73% CD4/CD3, and 10% CD8/CD3. Left axillary lymph node histology found small to medium lymphoid cells with some nuclear irregularity and inconspicuous nucleoli. The cells showed lack of expression of CD30 on immunohistochemical staining. Cytometry found an abundance of T lymphocytes, of which most expressed CD4 and were negative for CD8. The cells also expressed CD2, CD3, CD5, and CD7. The staging for her T-cell lymphoma was stage IVb.

The patient was initially treated with pazopanib and experienced no improvement after 2 months of 200-mg daily therapy. Cyclosporine and cyclophosphamide were also considered as treatment options. Because of persistent and significant pruritus, the patient was started on systemic psoralen and ultraviolet A therapy which provided symptomatic improvement. Further systemic chemotherapy was offered and declined by the patient.

DISCUSSION

Lymphomas coexisting in the same patient are classified by the Working Formulation of

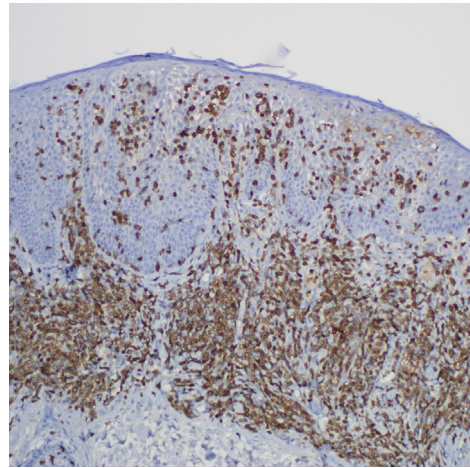


Fig 3. Biopsy specimen of chest shows CD3 positivity of lymphocytic infiltrate with moderate cytologic atypia, hyperchromatism, and scattered mitotic figures consistent with MF. (Hematoxylin-eosin stain.)

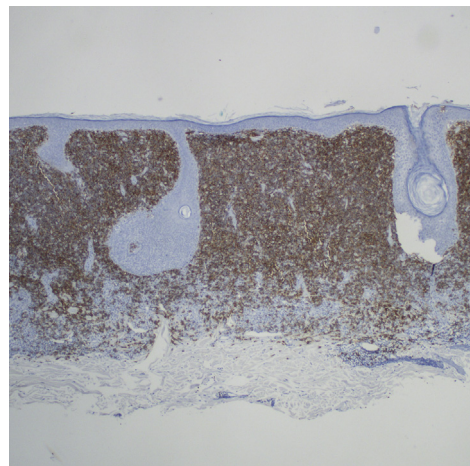


Fig 4. Biopsy specimen of back shows CD20 positivity of a dense, lymphoid infiltrate consistent with B-cell lymphoma. (Hematoxylin-eosin stain.)

non-Hodgkins lymphomas into three categories.¹ Discordant lymphomas are two histologically distinct lymphomas at two different anatomic sites. Composite lymphomas have two types of lymphoma within the same anatomic lesion. Lymphomas may also occur sequentially in which the second is known as *secondary lymphoma*.

Several cases in the literature report MF as a risk factor for secondary non-Hodgkins lymphomas, and, rarely, Hodgkins lymphomas.²⁻⁸ Explanations have been proposed for this association; the use of immunosuppressants in the treatment of the primary lesion, a genetic predisposition to malignancy, and the monoclonal proliferation of T cells in MF modulating the B-cell system, have all been

implicated in causing secondary malignancies.⁹ The Epstein-Barr virus has also been implicated in driving a B-cell lymphoproliferative process in patients with MF; the Epstein Barr virus studies in our patient were negative.¹⁰

Although it is a well known, yet rare, occurrence for MF to present with a secondary B-cell malignancy, it is exceedingly rare to find the two presenting simultaneously. Our patient presented with discordant cutaneous lymphomas in two anatomically distinct locations occurring roughly within the same timeframe. There are no documented cases of simultaneous presentation of discordant cutaneous lymphomas, to our knowledge, as most occur as a secondary lymphoma.

Our case further supports the need to perform multiple biopsies when patients present with morphologically different cutaneous findings.

REFERENCES

1. A clinical evaluation of the International Lymphoma Study Group Classification of Non-Hodgkin's Lymphoma. The Non-Hodgkin's Lymphoma Classification Project. *Blood*. 1997; 89:3909-3918.
2. Barzilai A, Trau H, David M, et al. Mycosis fungoides associated with B-cell malignancies. *Br J Dermatol*. 2006;155:379-386.
3. Sidwell RU, McLaughlin JE, Jones A, Whittaker SJ. Hodgkin Lymphoma in a patient with mycosis fungoides: molecular evidence for separate cellular origins. *Br J Dermatol*. 2003; 148(4):810-812.
4. Yang Q, Pei X, Tian X, Li Y, Li Z. Secondary cutaneous Epstein-Barr virus-associated diffuse large B-cell lymphoma in a patient with angioimmunoblastic T-cell lymphoma: a case report and review of literature. *Diagn Pathol*. 2012;7:7.
5. Metzman MS, Stevens SR, Griffiths CE, Ross CW, Barnett JM, Cooper KD. A clinical and histologic mycosis fungoides simulant occurring as a T-cell infiltrate coexisting with B-cell leukemia cutis. *J Am Acad Dermatol*. 1995;33(2):341-345.
6. Marschalk M, Csomor J, Eros N, et al. Coexistence of primary cutaneous anaplastic large cell lymphoma and mycosis fungoides in a patient with B-cell chronic lymphocytic leukaemia. *Br J Dermatol*. 2007;157(6): 1291-1293.
7. Rausch T, Cairoli A, Benhattar J, Spring P, Hohl D, de Leval L. EBV cutaneous B-cell lymphoproliferation of the leg in an elderly patient with mycosis fungoides and methotrexate treatment. *APMIS*. 2013;121(1):79-84.
8. Au WY, Lie AK, Lam WW, Chan JK. Refractory, Concomitant, Cutaneous, and Systemic Lymphomas of Discordant B-Cell and T-Cell Lineages. *Am J Dermatopathol*. 2010;32(1): 102-104.
9. Au WY, Lie AK, Lam WW, Chan JK. Refractory, Concomitant, Cutaneous, and Systemic Lymphomas of Discordant B-Cell and T-Cell Lineageshoproliferative diseases. A report of two cases. *Am J Dermatol*. 1990;23(4):677-681.
10. Novelli M, Merlino C, Ponti R, et al. Epstein-Barr virus in cutaneous T-cell lymphomas: evaluation of the viral presence and significance in skin and peripheral blood. *J Invest Dermatol*. 2009;129(6):1556-1561.