Atypical Cutaneous Presentation of Waldenström Macroglobulinemia: An Extensive Erythematous Patch Mimicking an Angiosarcoma

Erin L. Spillane, BA; CPT Yang Xia, MC, USA; COL George W. Turiansky, MC, USA

Waldenström macroglobulinemia (WM) is an immunoglobulin M–producing lymphoproliferative disorder in elderly individuals. Cutaneous manifestations of WM are rare and typically consist of plaques or nodules. We describe a case of a man with WM who presented with an extensive erythematous patch on the scalp that clinically mimicked an angiosarcoma.

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Case Report

As described by Libow et al,[1] in 1991, an otherwise healthy 46-year-old man presented to a hematology clinic for evaluation of anemia. Laboratory results revealed low levels of hemoglobin and hematocrit, an elevated erythrocyte sedimentation rate, free κ light chains in the urine, and periaortic lymphadenopathy. Serum protien electrophoresis showed an immunoglobulin M (IgM) monoclonal gammopathy. Results of a bone marrow biopsy demonstrated diffuse infiltration of plasmacytoid lymphocytes. A diagnosis of Waldenström macroglobulinemia (WM) was made on the basis of these findings. Results of a physical examination 6 years later revealed asymptomatic, red-brown to violaceous plaques distributed symmetrically over the anterior chest and scalp that were present for 3 years. Subsequent histologic examination, immunohistochemistry, and polymerase chain reaction–based IgM heavy chain gene rearrangement analysis of the chest lesions confirmed that the plaques were the result of cutaneous infiltration by a monoclonal population of lymphoplasmacytoid B cells.[1]

The patient's WM remained stable for 10 years before he became severely anemic at 57 years of age.[2] At that time, he was noted to have hepatosplenomegaly, retroperitoneal adenopathy, and an IgM monoclonal gammopathy in his serum and urine. He was successfully treated with rituximab, a chimeric anti-CD20 monoclonal antibody, previously described in the literature as being useful in the treatment of B-cell malignancies.[2,3]

In 2005, the patient again presented to Walter Reed Army Medical Center, Washington, DC, with an unusual new scalp lesion. The findings from the physical examination revealed a large, well-demarcated erythematous patch encompassing the majority of the patient's scalp (Figure). The differential diagnosis at the time included angiosarcoma, but lymphoma, contact dermatitis, and dermatomyositis also were considered as possible diagnoses. Surprisingly, however, both histologic examination of the lesions and immunohistochemistry revealed findings consistent with the patient's previously diagnosed lymphoproliferative disorder.

Comment

The extensive erythematous patch on the patient's scalp is an atypical cutaneous manifestation of WM and reveals the heterogenous nature with which WM can clinically present. WM, an indolent B-cell lymphoproliferative disorder characterized by an elevated circulating level of monoclonal IgM and bone marrow, spleen, and lymph node infiltration with lymphoplasmacytoid cells, is uncommonly

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Ms. Spillane is from the University of Maryland, Baltimore.
Drs. Xia and Turiansky are from Walter Reed Army Medical Center, Washington, DC.

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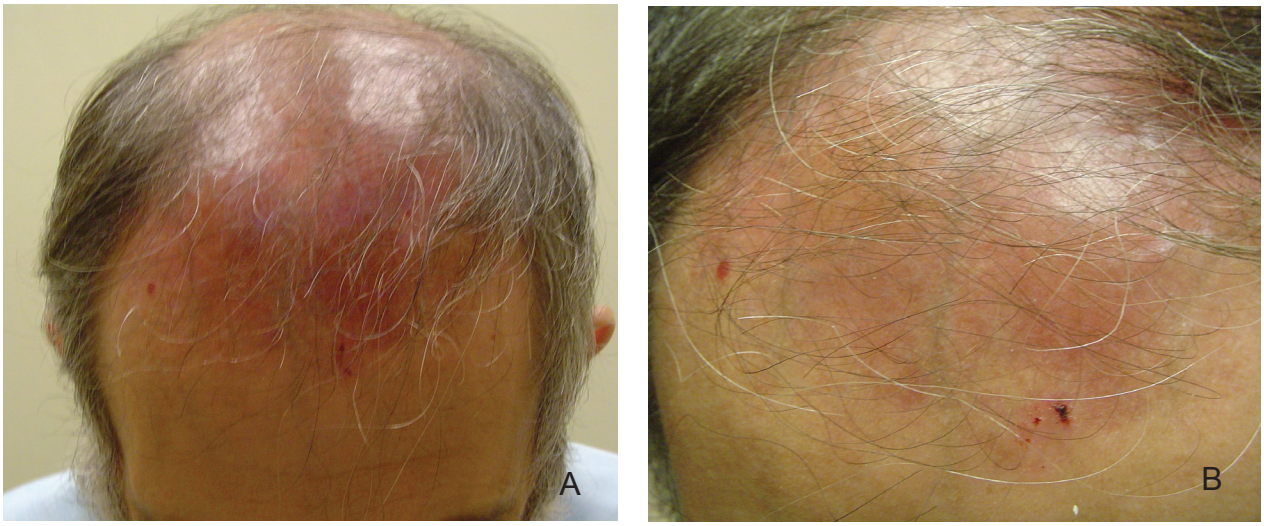
Correspondence: CPT Yang Xia, MC, USA, Dermatology Service, Walter Reed Army Medical Center, 6900 Georgia Ave NW, Washington, DC 20307 (yang.xia@na.amedd.army.mil).



An extensive erythematous patch on the scalp (A and B).

associated with cutaneous manifestations.[4] Libow et al[1] noted that the cutaneous manifestations of WM may be divided into 3 general categories: (1) secondary to paraproteinemia; (2) secondary to paraprotein-specific autoimmune processes; and (3) rarely caused by specific infiltration of the skin by neoplastic cells.

Specific cutaneous lesions in patients with WM typically present as either plaques or nodules. Nonneoplastic cutaneous lesions of WM may be associated with hyperviscosity syndrome, cryoglobulinemia, deposition of immunoglobulin, and paraprotein-specific autoimmune phenomenon, and may manifest as purpura, cold urticaria, acral cyanosis, Raynaud phenomenon, cold hypersensitivity, ulcers, livedo reticularis, leukocytoclastic vasculitis, storage disease,[1] vesicles, and bullae.[5] Because WM rarely presents in an atypical manner, such as an extensive erythematous patch on the scalp mimicking an angiosarcoma, this case report further expands the clinical spectrum of skin lesion morphology in WM.

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