Dr. Julia S. Lehman

WALDENSTROM'S MACROGLOBULINEMIA AND THE SKIN

by Julia S. Lehman

Waldenstrom's macroglobulinemia (WM) involves excessive production of a particular protein called immunoglobulin M (IgM). Although the most common symptoms of WM include tiredness, weakness, weight loss, and bleeding of the nose or gums, some patients may develop skin rashes.

Such rashes may develop from direct infiltration of the skin by cancerous blood cells (lymphoplasmacytoid B-cells). In these cases, patients can develop non- specific reddish-brown to purple patches or plaques (flat-topped, raised patches) on their skin. A very rare phenomenon related to WM that may affect the skin is a condition called "Schnitzler Syndrome." Patients with this condition develop hives (urticaria) in association with fever and bone pain.

More commonly, patients with WM will develop skin lesions as a consequence of excess blood proteins sludging in the blood vessels. Because skin blood vessels near the toes, fingers, and ears are among the smallest vessels, and therefore the most likely to get plugged, these areas are most frequently affected. At these sites, patients may get purplish change of the skin (purpura), which may be painful if the associated small blood vessel occlusion reduces blood supply to these locations. Sometimes, patients will develop net-like purple skin lesions (livedo reticularis), which represents low blood flow to the areas between larger blood vessels in the skin. Uncommonly, the skin can break down over these skin changes, leading to skin erosions and ulcers.

Because people with WM have impaired immunity, they are at higher risk for certain types of infections. A common infection associated with rash that may affect patients with WM is shingles (caused by reactivation of the chicken pox virus). Shingles, or herpes zoster, is characterized by the appearance of clustered blisters, usually confined to a single area of the body, which can be itchy or painful. The best treatment for shingles, antiviral medications, is most effective if it is started early in the disease course. Unfortunately, shingles may leave patients with long-lasting discomfort at the site of involvement, even if the original rash has subsided. Some medications, both topical and oral, may be helpful in alleviating this discomfort.

Another non-specific infection that may occur in patients with WM is cellulitis. When bacteria enter through breaks in the skin and cause cellulitis, patients may experience fever and pain, swelling, and expanding redness at the site of involvement.

Finally, the chemotherapy medications used to treat WM can cause a variety of rashes. The most common chemotherapy- related rashes include painful red bumps on the palms (neutrophilic eccrine hidradenitis), which is not usually intrinsically serious but may be bothersome to patients. Loss of hair is another common and expected cutaneous side-effect of certain chemotherapeutic agents.

If you or someone you know with WM develops a rash that is painful, purple, or associated with fevers, chills, or feeling ill, a doctor should be consulted immediately.

REFERENCES:

Chan I, Calonje E, Whittaker SJ. Cutaneous Waldenstrom's macroglobulinemia. Clin Exp Dermatol 2003; 28(5): 491-2.

Daoud MS, Lust JA, Kyle RA, Pittelkow MR. Monoclonal gammopathies and associated skin disorders. J Am Acad Dermatol 1999; 40(4): 507-35.

Libow LF, Mawhinney JP, Bessinger GT. Cutaneous Waldenstrom's macroglobulinemia: report of a case and overview of the spectrum of cutaneous disease. J Am Acad Dermatol 2001; 45(6 Suppl): S202-6.

Dr. Julia S. Lehman is the Chief Resident of Dermatology at Mayo Clinic in Rochester, Minnesota. Before starting her dermatology training at Mayo Clinic in 2007, she completed medical school at the University of Wisconsin School of Medicine and Public Health, followed by an internship at Gundersen Lutheran Medical Center in La Crosse, Wisconsin. Her special interests within dermatology include the cutaneous manifestations of systemic disease, immune- mediated skin disease, and dermatopathology.

This article was published in the IWMF *Torch*, volume 11.1 (January 2010) page 14.