

Cutaneous Necrosis Associated with Cold Agglutinins



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A 31-YEAR-OLD WOMAN PRESENTED WITH MULTIPLE NECROTIC SKIN LESIONS ON HER NOSE, EARS, AND LEGS that had developed over a period of a few days. The lesions had begun as erythematous-bluish macules that soon became bullous and finally, after approximately 10 days, developed into thick necrotic plaques. She had no history of underlying diseases or infections, but she described several days of exposure to cold after the heating in her home failed and outside temperatures dropped to 50°F (10°C). Histologic examination revealed thrombosis of all dermal and subcutaneous vessels but almost no inflammatory infiltration and no signs of vasculitis. A test for cold agglutinins was positive at a 1:64 dilution, and there were signs of a mild hemolytic anemia. Cold agglutinins (immunoglobulins, predominantly IgM) typically lead to agglutination of erythrocytes below certain temperatures, causing impaired microcirculation and hemolysis. Screening for hypercoagulability, cryoglobulins, and paraproteins was negative, and there were no signs of cancer. The patient was treated with heparin, iloprost, plasmapheresis, and surgical débridement of necrotic skin and was given strict instructions to protect herself against cold. After initial improvement, she was lost to follow-up.

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