Chilblain Lupus Erythematosus Lesions Precipitated by the Cold

Thomas N. Helm, MD; Cornelie M. Jones, MD

Lupus erythematosus (LE) may exhibit a broad array of clinical presentations. Cutaneous manifestations include malar rash, discoid lesions, alopecia, and panniculitis. Cold-induced lesions are uncommon. To illustrate this unusual presentation, we describe a case of typical chilblain LE.

Lupus erythematosus (LE) may present with a characteristic malar butterfly rash or a discoid lesion. Occasionally, less common presentations—for example, red lunula—may be noted. Chilblain LE is an uncommon but characteristic presentation in which histologically typical cutaneous lupus lesions are precipitated by the cold. The following report illustrates a typical case.

Case Report

A 56-year-old farmer presented with complaints of recurrent lesions upon exposure to the cold. Sun exposure had no similar effect. Asymptomatic lesions developed episodically on the cheeks, nose, and abdomen and on acral sites. No residual scars or pigmentary changes were noted. Persistent erythema was noted on the abdominal lesions after resolution of induration. Results of a physical examination revealed 1- to 2-cm erythematous, indurated plaques on the cheeks (Figure 1) and on the abdomen above the belt line (Figure 2), as well as a few lesions on the nose. No scaling or epidermal changes were noted.

Figure not available online

Figure 1. Erythematous lesions on the cheek precipitated by the cold.

Initial biopsy results revealed a perivascular lymphocytic infiltrate (Figure 3). Basal layer vacuolopathy was not identified, but lymphocytes and histiocytes extended into the panniculus. Increased mucin was noted in the dermis. Another biopsy was performed 2 weeks later, and results revealed new erythematous lesions after cold exposure. In addition, a biopsy of the right cheek was performed, and those results revealed parakeratosis and serous crust. Basal layer vacuolopathy was noted, and dyskeratotic keratinocytes were evident (Figure 4). A superficial and deep perivascular and periappendageal lymphocytic infiltrate was noted. Histologic findings were consistent with a diagnosis of cutaneous LE, and the history of cold exposure confirmed the clinical suspicion of chilblain LE.

Comment

Chilblain (perniosis) is characterized by localized redness and swelling, precipitated by the cold.[1,2] The face, ears, hands, and feet are most commonly involved. Onset of lesions may be insidious, but

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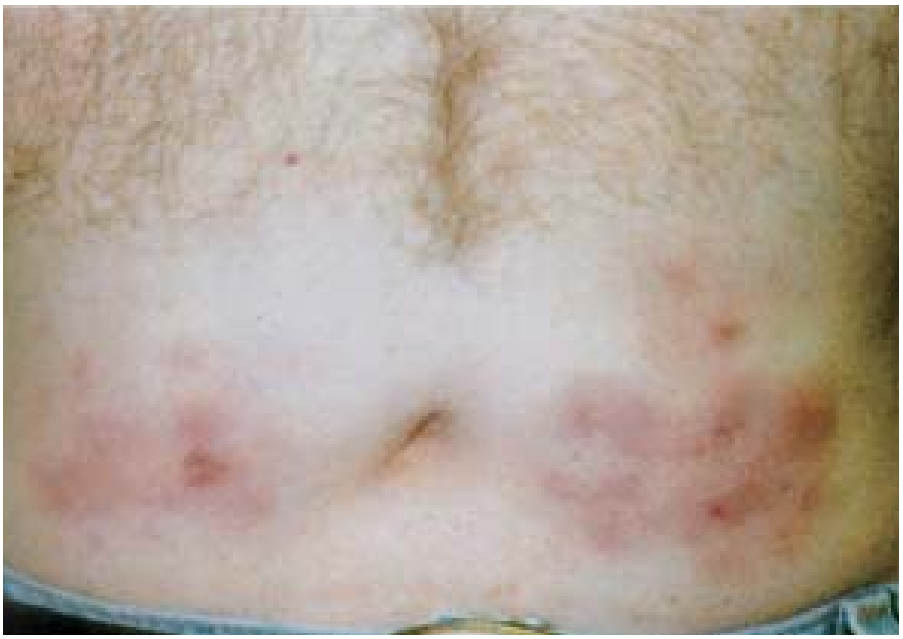

Figure 2. Erythematous and indurated lesions on the abdomen.

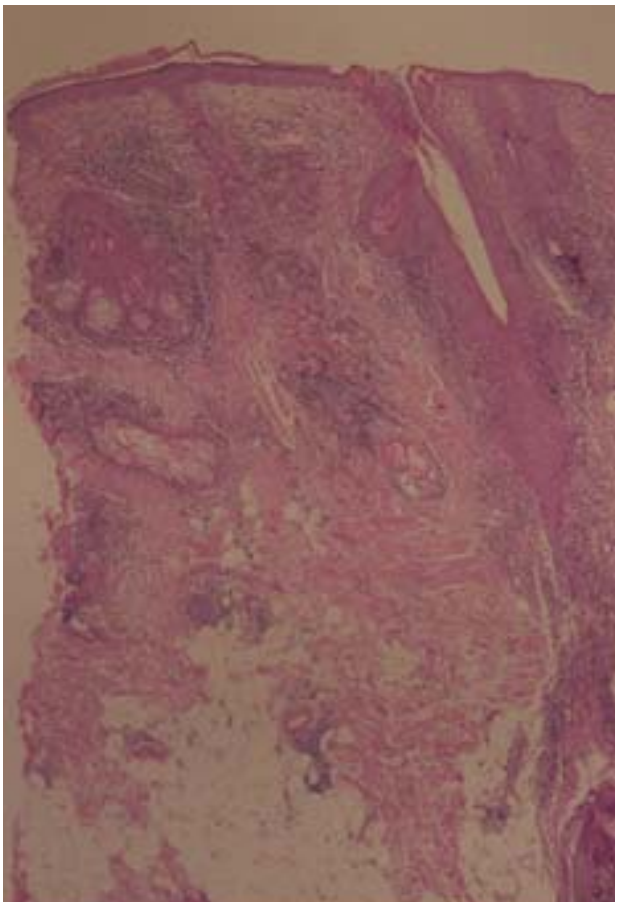

Figure 3. Interface changes and a perivascular and periappendageal lymphocytic infiltrate (H&E, original magnification ×40).

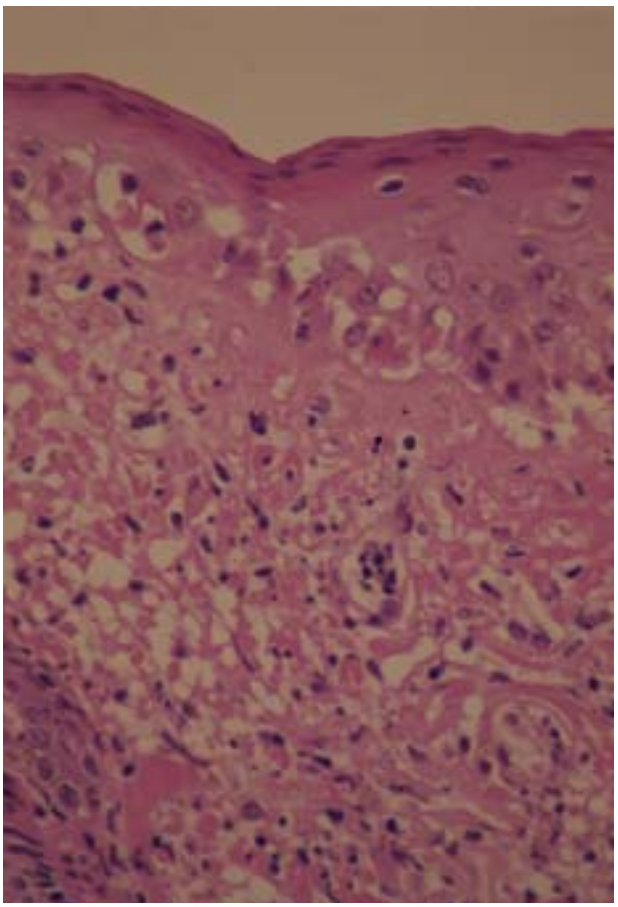

Figure 4. Dyskeratotic keratinocytes and basal layer vacuolopathy (H&E, original magnification ×100).

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pain and itching soon become bothersome. Hyperhidrosis may be noted in affected areas. At times, LE may be associated with identical lesions. Biopsy results help differentiate chilblain from chilblain LE. In chilblain LE, the characteristic interface changes of epidermal atrophy, basal layer vacuolopathy, and dyskeratotic keratinocytes are noted. Chilblain is characterized by lymphocytic infiltrate, especially around the eccrine coils.

Recent studies suggest that Ro/SSA antibodies may be associated with chilblain LE.[3-5] Cryoglobulin levels also should be ascertained.[6] Vasculitic lesions, such as palpable purpura and cutaneous infarction, rather than the blanching cutaneous lesions of chilblain and chilblain LE characterize cryoglobulinemia.

Treatment includes avoidance of the cold, rapid warming of affected areas, and use of agents such as dipyridamole, pentoxifylline, or calcium channel blockers. Chilblain LE also may be helped by hydroxychloroquine or other agents directed at underlying LE.

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