Erythematous Edematous Plaques on the Dorsal Aspects of the Hands

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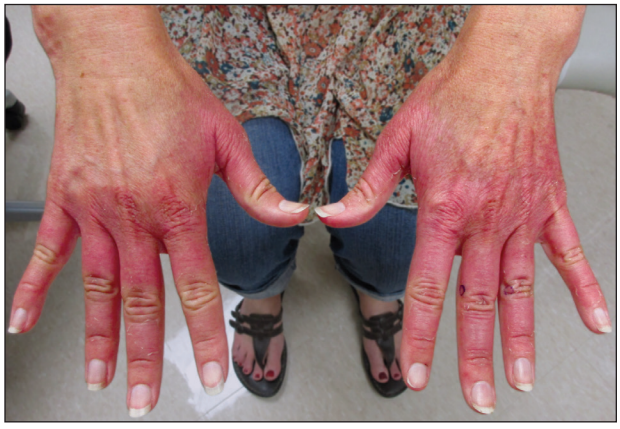

A 48-year-old woman presented with erythematous swelling of the dorsal aspects of the bilateral hands followed by desquamation and pruritus of 2 weeks' duration. She denied any recent contact with plants, chemicals, or topical products or use of over-the-counter medications. A 6-day course of prednisone provided by her primary care physician relieved the swelling and pruritus; however, the erythema persisted. Physical examination revealed clearly demarcated, erythematous to violaceous, edematous plaques with peripheral scaling that involved all digits. There was notable sparing of the proximal interphalangeal joints and volar aspects of the hands extending proximally to the metacarpophalangeal joints.

WHAT'S THE **DIAGNOSIS?**

a. CREST syndrome
b. dermatomyositis
c. erythromelalgia
d. lupus erythematosus
e. phytophotodermatitis

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The authors report no conflict of interest.
The views expressed in this article are those of the authors and do not reflect the official policy of the Department of Defense or US Government.
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THE **DIAGNOSIS:** Phytophotodermatitis

Initially, there was concern for autoimmune or connective tissue disease because of the edematous plaques localized over sun-exposed regions of the hands with marked sparing of the knuckles. Lupus erythematosus (LE), mixed connective tissue disease, CREST (calcinosis, Raynaud phenomenon, esophageal motility disorders, sclerodactyly, telangiectasia) syndrome, dermatomyositis (DM), and erythromelalgia all were considered. Common disorders such as contact dermatitis and phytophotodermatitis remained in the differential diagnosis, though the patient adamantly denied any recent exposures. As part of the initial workup, laboratory studies including a complete blood cell count, comprehensive metabolic panel, serum lactate dehydrogenase, serum creatinine kinase, erythrocyte sedimentation rate, and an antinuclear antibody panel were performed. Additionally, a punch biopsy at the border of the lesion was performed.

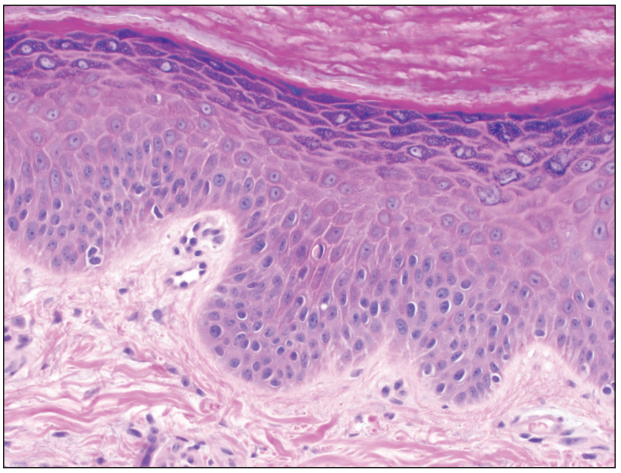
Lupus erythematosus was considered given the patient's age and sex and the photoexposed location of the plaques. The photosensitive rash of LE classically affects the dorsal aspects of the hands while sparing the interphalangeal joints.[1,2] However, the patient had no nail fold findings consistent with systemic LE with no evidence of erythema or dilated tortuous vessels.[3] Furthermore, there were no other cutaneous symptoms, and there was a negative review of systems, including malar/discoid rash, oral ulcers, photosensitivity, history of hematologic abnormalities, and end organ damage.[4,5] A negative antinuclear antibody serologic panel combined with a negative review of systems made the diagnosis of LE less likely.

Given the presenting clinical appearance, DM also was considered. Dermatomyositis traditionally displays ragged cuticular dystrophy with nail fold telangiectasia, mechanic hands, and involvement of the dorsal aspects of the hands with violaceous accentuation of the knuckles.[6] The patient reported pruritus, which is common among DM patients; however, the nail folds were unaffected.[7] Finally, she demonstrated sparing rather than involvement of the knuckles, which would be an unlikely presentation for DM.[6]

CREST syndrome, systemic sclerosis, and syndromes with overlapping features such as mixed connective tissue disease also were considered. The cutaneous features of CREST syndrome are characterized by initial edema of the digits with a subsequent taut and shiny indurated phase. Flexion contractures, ulceration, tapering of the digits, and loss of cutaneous fat pads can progressively occur.[8,9] Raynaud phenomenon is a common early finding in CREST syndrome or systemic sclerosis, and patients may develop ice pick digital infarcts and calcinosis in progressed disease.[8] Common nail fold findings include periungual telangiectasia with dropout areas.[10,11] The marked edema and white discoloration of the knuckles in this patient could be mistaken for Raynaud phenomenon; however, she lacked pain or cold sensitivity and her discoloration was static.[12] Without sclerodermoid changes, nail fold findings, matted telangiectasia, taut skin, or systemic findings, a diagnosis of CREST syndrome, scleroderma, or other mixed connective tissue disease would be unlikely.[8]

Erythromelalgia is a clinical syndrome characterized by burning pain, erythema, and increased skin temperature that intermittently affects both the arms and legs. This rare disorder can be further classified into type 1 (associated with thrombocytopenia), type 2 (primary or idiopathic), and type 3 (associated with other medical cause excluding thrombocytopenia).[1,13] The patient endorsed some discomfort from the lesions but denied any subjective feeling of burning pain or increased skin temperature. Additionally, she had no family history of inheritable skin disorders and no personal history of polycythemia. Consequently, erythromelalgia remained less likely on the differential diagnosis.

The histology of the acral skin revealed mild focal spongiosis with no increase in dermal mucin on colloidal iron or mucopolysaccharide stains (Figure). After receiving the biopsy results and additional questioning of the patient, it was discovered that 2 days prior to her initial presentation she had juiced numerous limes by hand and subsequently spent a long period of time outside with sunlight exposure. Upon discovery of this additional historical information, the diagnosis of phytophotodermatitis was made.



A punch biopsy of the left hand showed epidermal changes on colloidal iron or mucopolysaccharide stains (original magnification ×200).

Phytophotodermatitis is an erythematous inflammatory reaction that occurs on the skin after exposure to a plant-derived photosensitizer followed by UVA light radiation.[14] This phenomenon was first described by the ancient Egyptians as a treatment for vitiligo.[1] The most common plant families that can cause this nonimmune cutaneous reaction include Apiaceae (eg, hogweed, celery, dill, fennel) and Rutaceae (eg, citrus plants, rue).[14] The psoralens or furocoumarins found in these plants bind loosely to DNA at their ground state but covalently bond to pyrimidine bases during photoexcitation with UVA, resulting in DNA damage and subsequent local inflammation.[14] Given the patient's clinical examination, pathology findings, and history, phytophotodermatitis secondary to lime juice exposure was confirmed. Two weeks after applying clobetasol ointment twice daily, the patient's hands had returned to baseline with complete resolution of the erythematous lesions.

Although lime phytophotodermatitis is a routine diagnosis, this clinical case stands as an important reminder to demonstrate how common diseases can masquerade as more exotic cutaneous disorders. There often is a clinical desire to seek out more complicated diagnoses, particularly during residency training; however, this case reinforces the invaluable importance of collecting a thorough patient history, as it can ultimately minimize excessive testing and in some cases prevent unnecessary therapy.

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