Acute Pustular Eruption on the Hands

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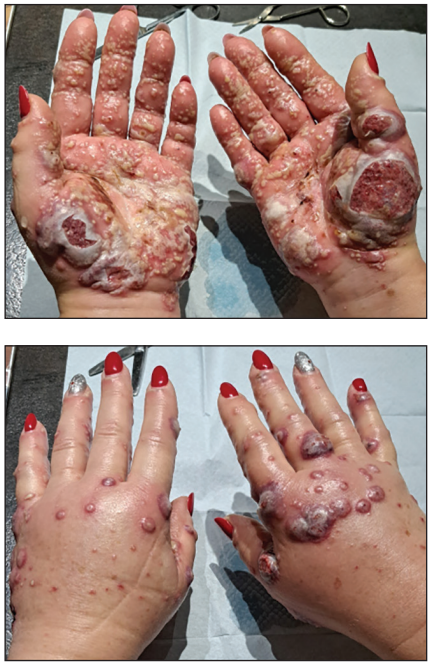

A 56-year-old woman was referred to the dermatology department for a painful acral pustular eruption of 6 days' duration. Her medical history was otherwise unremarkable. Physical examination revealed multiple pustules on the hands with large blisters on an erythematous base and painful surface ulceration (top). Papulonodular infiltrated lesions also were observed on the dorsal aspect of the hands (bottom). There were no additional systemic symptoms. Routine laboratory tests showed hyperleukocytosis at $17.9\times10^3/mm^3$ (reference range, $4\text{-}10\times10^3/mm^3$) with neutrophils at $12.3\times10^3/mm^3$ ($1.8\text{-}7.5\times10^3/mm^3$) and elevated C-reactive protein at 67 mg/L (<5 mg/L). Screening for hematologic neoplasms, solid tumors, and inflammatory bowel disease was negative. An incisional biopsy was performed on a pustule on the palm of the left hand.

WHAT'S YOUR **DIAGNOSIS?**

a. dyshidrosis
b. neutrophilic dermatosis of the dorsal hands
c. poststreptococcal pustulosis
d. pustular psoriasis
e. reactive arthritis (Reiter syndrome)

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THE **DIAGNOSIS:** Neutrophilic Dermatosis of the Dorsal Hands

Histopathology showed a unilocular pustule with a dense neutrophilic infiltrate of the superficial dermis. Minimal vascular alterations also were observed. These findings were consistent with a diagnosis of neutrophilic dermatosis of the dorsal hands (NDDH). Our patient was treated successfully with systemic corticosteroids (1 mg/kg/d) with rapid improvement after 10 days of treatment.

Neutrophilic dermatosis of the dorsal hands is an evolving disease concept that was first described as pustular vasculitis by Strutton et al[1] in 1995. Galaria et al[2] subsequently identified NDDH as a clinical entity associating tender erythematous plaques, pustules, bullae, and/or ulcers on the dorsal hands with histologic features of Sweet syndrome (SS). After reviewing 9 cases of NDDH—all of which demonstrated clinical, laboratory, and histologic characteristics of SS—Walling et al[3] concluded that NDDH was best understood as a distributional variant of SS.

Our patient presented with vascular alterations described as a reactive response to the neutrophilic infiltration. The presence of vasculitis in SS and NDDH biopsies is considered as an occasional epiphenomenon and should not rule out the diagnosis of NDDH.[3] A literature review of 123 cases of NDDH revealed the presence of vasculitis in 36 (29.5%) patients.[4] With regard to other clinical findings, it has been suggested that an increased white blood cell count and elevated C-reactive protein level, as was seen in our patient, may be observed in NDDH, albeit less frequently than in classical SS.[4]

While palmar involvement of NDDH is considered rare, the recent review of 123 cases of NDDH identified palmar lesions in 5 patients (4.1%).[4] Earlier reviews had identified 12 historical cases.[5] Palmar manifestations of NDDH have been shown to be associated with erythematous nonulcerated lesions (as opposed to the classical ulcerative or pustular plaques) and a lower association with hematologic malignancies.[5]

In our patient's case, dyshidrosis was excluded due to the presence of painful ulcerative plaques rather than pruritic, deep-seated vesicles. Pustular psoriasis typically manifests with sterile pustules on the palms and soles; however, the rapid onset of ulcerative, necrotic plaques and substantial edema are more specific to NDDH. Poststreptococcal pustulosis generally follows a streptococcal infection and lacks the violaceous undermined borders seen in NDDH. Reactive arthritis manifests with hyperkeratotic plaques and is associated with the clinical triad of urethritis, conjunctivitis, and arthritis, which were absent in our patient.

The histologic differential diagnosis of NDDH includes infection, pyoderma gangrenosum, bowel-associated dermatosis-arthritis syndrome, rheumatoid neutrophilic dermatitis, and erythema elevatum diutinum[3,4]; however, these conditions typically manifest with distinct clinical features that allow for differentiation, despite histologic similarities. The wide histologic spectrum of neutrophilic dermatosis may contribute to variable clinical manifestations and an evolving disease concept, as the classification of NDDH has changed from a primary vasculitis to a variant of SS. However, this evolution does not affect the appropriate management, as they all have shown good response to corticosteroid treatment.[4,6]

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