A 78-year-old Taiwanese woman presented with fever and painful skin lesions over both palms for 2 weeks. Her medical history was unremarkable. On physical examination, infiltrating and juicy vesicular eruptions on violaceous bases were noted over the palms as well as on the ventral and lateral aspects of the fingers (Figure 1). Very few lesions extended to the dorsal aspect of the fingers. In addition, no mucosal lesions were found. Laboratory studies on admission revealed leukocytosis and neutrophilia with the white blood cell count at $12.54 \times 10^9/\mu L$ with 91.6% neutrophils. The C-reactive protein level was also elevated (3.18 mg/dL). The patient was treated under a provisional diagnosis of a cutaneous infection; however, the symptoms persisted despite a 5-day administration of systemic antibiotics. Further laboratory investigations including urinalysis, blood culture, and Tzanck smear—all demonstrated negative results. A skin biopsy specimen was obtained from a lesion on the patient’s left palm. Histopathologic evaluation revealed mild epidermal acanthosis with subepidermal edema with dense dermal neutrophilic infiltrate (Figure 2A). In addition, abundant leukocytoclastic debris, extravasated red blood cells, and fibrinoid necrosis of small vessels were evident (Figure 2B). Tissue culture and immunofluorescence revealed negative results. No associated underlying malignancy, infection, or inflammatory bowel disease was noted at the time of presentation. Based on the clinicopathologic correlations, the diagnosis of neutrophilic dermatosis of the hands was made, and the patient was treated with betamethasone 4 mg orally twice per day. The patient’s fever subsided in 3 days and cutaneous lesions resolved without scarring after 2 weeks of treatment.

In 1995, Strutton et al first described six patients displaying papules, hemorrhagic plaques, pustules, and bullous lesions over the dorsal surface of the hands in whom histopathologic features revealed dense dermal neutrophilic infiltration with leukocytoclastic vasculitis. The general clinical manifestations were also associated with fever, leukocytosis with neutrophil predominance, sterile culture result, and nonresponse to antibiotic therapy but showing rapid response to prednisone administration. This clinical presentation differed from, but was similar to, Sweet syndrome (SS) in terms of histologic features, particularly with respect to the presence of leukocytoclastic vasculitis. Hence, Strutton et al named this entity pustular vasculitis of the dorsal hands; however, these cases were still considered as localized variants of SS. In 2000, Galaria et al reported three cases with similar clinical findings albeit with no evidence of vasculitis on histopathology; therefore, this entity was renamed neutrophilic dermatosis of the dorsal hands instead of pustular vasculitis of the dorsal hands. In most cases reported under this diagnosis, the dorsal aspects of the hands comprised the most commonly affected sites; however, recently, a few similar cases involving only the palms and lateral aspects of the fingers have been reported. Hence, the term neutrophilic dermatosis of the dorsal hands was replaced by the term neutrophilic dermatosis of the hands (NDH). To date, although fewer than 100 cases of NDH have been reported, it is possible that the incidence of this disease might have been underestimated. For NDH, the average age at diagnosis was 60.5 years, with 58% of the patients being female. Clinically, the
lesions appeared as edematous, purpuric plaques with blisters and/or pustules on the dorsal and/or palmar aspects of the hands and the lateral aspects of the fingers. Most cases of NDH are associated with fever, peripheral leukocytosis with neutrophilia, and an elevated erythrocyte sedimentation rate or C-reactive protein level. Histopathologic features typically include subepidermal edema and dense dermal neutrophilic infiltrate with or without leukocytoclastic vasculitis. Patients in whom NDH is diagnosed should be assessed for comorbid medical conditions because previously reported cases have been associated with leukemia, myelodysplasia, other hematologic diseases, inflammatory bowel disease, or preceding vaccination.

The similar entities of SS, vesiculobullous pyoderma gangrenosum (PG), and NDH present a difficult diagnostic dilemma (Table 1). NDH is fairly similar to SS in terms of clinical presentation except that the lesions are limited to the hands. Basically, the main histopathologic feature of NDH is similar to that of SS, which shows papillary dermal edema with dense neutrophilic infiltrate without vasculitis. However, vasculitis might be shown in some cases of NDH; therefore, it has long been debated whether NDH comprises a true variant of SS or a separate clinical entity. Some authors have observed that vasculitis is correlated with long-standing lesions in SS and NDH, where the damaged vessels could be considered as the so-called innocent bystander. In other words, vasculitis is considered as a time-dependent change rather than a fundamental one for the diagnosis of SS and NDH. Therefore, NDH continues to be regarded as a localized variant of SS. Vesiculobullous PG is another disease entity resembling NDH. Clinically, vesiculobullous PG presents as hemorrhagic bullae predominantly located on the dorsal hands. Histologically, the condition is characterized by dense neutrophilic infiltrate with a prominent vascular reaction, but not true vacuities. Thus, both its clinical and histologic features are considerably similar to those of NDH. Because of this overlap in clinical and microscopic features, some authors consider that NDH, SS, and vesiculobullous PG possibly represent different aspects of the same disease spectrum.

Many of the reported cases of NDH—including the current case—were initially diagnosed as localized cutaneous infections, such as cellulitis, and treated with systemic antibiotics. However, antimicrobial therapy achieves no response in these patients. Hence, differentiating this clinical entity from cellulitis is important for accurate and timely therapy. In general, cellulitis clinically presents as an ill-defined erythematous patch, in contrast to NDH, which presents as well-demarcated violaceous vesicles on erythematous bases. Although cellulitis requires systemic antibiotic therapy, NDH shows good response to oral corticosteroids, potassium iodide, minocycline, or dapsone. The skin lesions show gradual resolution without scarring in variable periods (from 3 weeks to 3 months) and the overall outcome is good. However, in patients with underlying disease including inflammatory bowel disease or malignancy, the clinical outcome is correlated with the associated disease.

In conclusion, for patients with tender and/or hemorrhagic palmar vesiculobullous rashes accompanied by fever and leukocytosis, NDH should be included in the differential diagnosis, with skin

Table 1 Common and distinct features among different neutrophilic dermatoses.

<table>
<thead>
<tr>
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<th>SS</th>
<th>NDH</th>
<th>Vesiculobullous PG</th>
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<tbody>
<tr>
<td>Clinical manifestation</td>
<td>Asymmetrically distributed on the face, trunk, and limbs</td>
<td>Limited on the palmar surface and/or dorsal aspects of hands</td>
<td>Usually limited on the extremities, especially dorsal aspects of hands</td>
</tr>
<tr>
<td>Histopathologic finding</td>
<td>Dermal edema with dense infiltration by neutrophils; vasculitis may be found in long-standing lesions</td>
<td>Dermal edema with dense infiltration by neutrophils; with/without vasculitis</td>
<td>Dermal edema with dense infiltration by neutrophils; with/without vasculitis</td>
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<tr>
<td>Possible comorbidity</td>
<td>Internal malignancy, infection, pregnancy, vaccination, inflammatory bowel disease</td>
<td>Internal malignancy, infection, vaccination, inflammatory bowel disease</td>
<td>Internal malignancy, rarely inflammatory bowel disease</td>
</tr>
<tr>
<td>Prognosis</td>
<td>Good response to systemic steroid administration. Overall outcome depends on underlying disease.</td>
<td>Good response to systemic steroid administration. Overall outcome depends on underlying disease.</td>
<td>Moderate response to systemic steroid administration. Overall outcome depends on underlying disease.</td>
</tr>
</tbody>
</table>

NDH = neutrophilic dermatosis of the hands; PG = pyoderma gangrenosum; SS = Sweet syndrome.
biopsy being the recommended examination to reach an accurate
diagnosis. Based on the resemblance of the clinical and histopath-
ologic features of NDH, SS, and vesiculobullous PG, we consider
neutrophilic dermatosis of the hands to be the most appropriate
designation for such acral neutrophilic eruptions.

Ching-Fu Huang
Department of Dermatology, Tri-Service General Hospital, National Defense Medical
Center, Taipei, Taiwan

Wei-Ming Wang*
Department of Dermatology, Tri-Service General Hospital, National Defense Medical
Center, Taipei, Taiwan

Department of Biochemistry, National Defense Medical Center, Taipei, Taiwan

* Corresponding author. Department of Biochemistry, National Defense Medical
Center, No. 325, Sec. 2, Chenggong Rd., Taipei 114, Taiwan. Tel.: +886 87923311.
E-mail address: adest0431@ms38.hinet.net

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Received: May 30, 2012
Revised: Dec 5, 2012
Accepted: Dec 17, 2012