The Relationship Between Neutrophilic Dermatosis of the Dorsal Hands and Sweet Syndrome

Report of 9 Cases and Comparison to Atypical Pyoderma Gangrenosum

Hobart W. Walling, MD, PhD; Clancy J. Snipes, MD; Pedram Gerami, MD; Warren W. Piette, MD

**Background:** Neutrophilic dermatoses are a collection of diseases with varying presentation unified by clinical and histologic features. Neutrophilic dermatosis of the dorsal hands is a recently described clinical entity and an evolving disease concept. Its relationship to acute febrile neutrophilic dermatosis (Sweet syndrome), pyoderma gangrenosum, and a primary vasculitis has been debated.

**Observations:** We present 9 cases (8 women and 1 man) of neutrophilic dermatosis of the dorsal hands, all with consistent histologic features. Two cases had histologic evidence of vasculitis, and 3 had clinical extension of lesions onto the forearms. Most showed fever, leukocytosis, and/or elevated erythrocyte sedimentation rate. Individual cases were associated with leukemia, lung carcinoma, and inflammatory bowel disease. All 9 patients responded to systemic corticosteroid therapy, with additional response to dapsone, methotrexate, and potassium iodide therapies in several cases. Of the 9 patients, 5 showed complete resolution of their skin disease, whereas 4 required ongoing therapy. We assessed the 43 cases previously reported in the literature.

**Conclusion:** The clinical presentation, laboratory data, histologic features, and response to corticosteroid therapy offer strong evidence that neutrophilic dermatosis of the dorsal hands is a localized variant of Sweet syndrome and is also identical to atypical pyoderma gangrenosum when that condition presents on the hands.

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**REPORT OF CASES**

**CASE 1**

A 61-year-old woman was seen for a painful ulcer on her right dorsal hand, which began as a pink papule 2 weeks prior. She...
had been treated with antibiotics (cephalexin hydrochloride and ampicillin sodium–sulbactam sodium) for the past 10 days. She denied having fever or prior trauma, though reported that similar lesions had developed intermittently over the joints of her dorsal hands for the past 10 years, typically lasting 1 to 2 weeks and treated with wound care and occasional oral antibiotics. Physical examination revealed a $6 \times 4$-cm violaceous plaque with central nonundermining superficial ulceration at the right dorsal hand over the second and third metacarpophalangeal joints extending onto the index finger (Figure 1 A). Lymphadenopathy was absent. Findings from laboratory studies showed moderate leukocytosis, neutrophilia, anemia, and elevated inflammatory indices (Table 1). Results of serum electrolyte assessment, renal and liver function panel, serum protein electrophoresis, and urinalysis, as well as hepatitis serologies and autoantibody profile (antinuclear antibody, SS-A, SS-B, double-stranded DNA, antineutrophil cytoplasmic antibody, and thyroid peroxidase), were normal. A cutaneous biopsy was performed (Table 1). Bacterial, fungal, and mycobacterial cultures were negative. She was initially treated with dapsone, 100 mg/d. Prednisone, 60 mg/d, was added 5 days later, with improvement over the next week (Figure 1B). The lesions resolved, and both medications were slowly tapered.

**CASE 2**

A 44-year-old woman with ulcerative colitis was seen for recurrent ulcerations on the dorsal hands. Lesions, associated with fever and chills, began as violaceous edematous plaques and would ulcerate within 2 days. A biopsy was performed (Table 1). Cultures for bacteria, fungi, and mycobacteria were negative. She responded rapidly to prednisone therapy, 1 mg/kg, tapered over 6 weeks as dapsone therapy was initiated.

**CASE 3**

A 48-year-old woman was seen for a painful pustular eruption on her dorsal fingers. The lesions started as small, edematous, pink papules evolving into pustules with subsequent ulceration. She did not improve with ceftriaxone sodium and azithromycin therapies. Complete blood cell count revealed leukocytosis with neutrophilia. A biopsy was performed (Table 1). Cultures for bacteria, fungi, and mycobacteria were negative. Her skin lesions, fever, and laboratory abnormalities resolved with prednisone tapered over 8 weeks, and she continues dapsone therapy without recurrence.

**CASE 4**

A 71-year-old man was referred for ulcers on his hands, which developed from spontaneous papules. Treatment with cephalixin hydrochloride and then amoxicillin sodium–clavulanic acid had been ineffective, as had acetic acid soaks and mupirocin cream. He reported chills but denied having arthralgias. He had a history of lung adenocarcinoma treated with lobectomy. Metastases to the mediastinum and left adrenal gland were discovered, and he had completed chemotherapy 8 months prior to presentation. Physical examination revealed superficial ulcerations with raised violaceous undermined borders on the dorsal aspect of both index fingers and the left fourth finger (Figure 2 A). Laboratory studies revealed leukocytosis, neutrophilia, and elevated erythrocyte sedimentation rate and C-reactive protein level. Radiography of the hands was unremarkable, and bacterial cultures were negative. A biopsy was obtained (Figure 2B, Table 1). He was treated with prednisone, 60 mg/d (0.8 mg/kg) for 4 days and then tapered over 3 weeks, and continued local wound care, with rapid improvement. He died of metastatic lung cancer 4 months later.
Table 1. Case Summaries of Neutrophilic Dermatosis of the Dorsal Hands

<table>
<thead>
<tr>
<th>Patient Age, y/</th>
<th>Site</th>
<th>Temperature</th>
<th>Laboratory Study Results</th>
<th>Histologic Features</th>
<th>Comorbidities</th>
<th>Treatment</th>
<th>Outcome</th>
<th>Follow-up, mo</th>
</tr>
</thead>
<tbody>
<tr>
<td>61/W/F</td>
<td>Right dorsal hand and index finger</td>
<td>(36°C)</td>
<td>WBC, 10.9 × 10^9/µL; PMN, 8.7 × 10^9/µL; ESR, 45 mm/h; CRP, 3.8 mg/dL; Hb, 11.9 mg/dL</td>
<td>Pseudoeppitheliomatous hyperplasia, dense PMN infiltrate epidermis to subcutis, vasculitis, gram-negative, and GMS negative</td>
<td>Hypothyroid, fibromyalgia, Raynaud phenomenon</td>
<td>Prednisone ×5 wk, dapsone</td>
<td>Resolved</td>
<td>31</td>
</tr>
<tr>
<td>44/W/F</td>
<td>Bilateral dorsal hands</td>
<td></td>
<td>WBC, 10.5 × 10^9/µL; PMN, 8.5 × 10^9/µL; ESR, 66 mm/h; CRP, 7.8 mg/dL; Hb, 11.3 mg/dL</td>
<td>Ulcerative colitis, dense dermal neutrophilic infiltrate with lymphocytes, no vasculitis, AFB, gram-negative, and GMS negative</td>
<td>Ulcerative colitis</td>
<td>Prednisone and dapsone ×13 wk</td>
<td>Resolved</td>
<td>6</td>
</tr>
<tr>
<td>48/W/F</td>
<td>Bilateral dorsal hands, arms, elbows</td>
<td>39°C</td>
<td>WBC, 14.8 × 10^9/µL; PMN, 12.3 × 10^9/µL; ESR, &gt;100 mm/h; CRP, 2.7 mg/dL; Hb, 10.0 mg/dL</td>
<td>Ulcerative, edema, sheets of PMNs in dermis, no vasculitis, PAS and GMS negative evolution to PNGD</td>
<td>Subepidermal blister, sheets of PMN in dermis, rare eosinophils, lymphocytes, no vasculitis, PAS and GMS negative</td>
<td>Goodpasture syndrome, ESRF on HD</td>
<td>Topical, intraliesional steroids, dapsone</td>
<td>Improved</td>
</tr>
<tr>
<td>71/W/M</td>
<td>Bilateral dorsal fingers</td>
<td>(35.8°C)</td>
<td>WBC, 20.1 × 10^9/µL; PMN, 18 × 10^9/µL; ESR &gt;100 mm/h; CRP, 12.7 mg/dL; Hb, 9.5 mg/dL</td>
<td>Subepidermal blister, sheets of PMNs in dermis, florid, folliculocentric neutrophilic infiltrate in papillary/reticular dermis, fibrinoid necrosis</td>
<td>Metastatic lung cancer</td>
<td>Prednisone ×3 wk</td>
<td>Resolved but died of cancer</td>
<td>4</td>
</tr>
<tr>
<td>32/W/F</td>
<td>Left dorsal hand</td>
<td></td>
<td>WBC, 9.8 × 10^9/µL; ESR, 34 mm/h; CRP, 3.4 mg/dL; Hb, 12.9 mg/dL</td>
<td>Ulcerative, sheets of PMNs in dermis, no vasculitis, PAS and GMS negative</td>
<td>None</td>
<td>Prednisone ×3 wk</td>
<td>Resolved</td>
<td>1</td>
</tr>
<tr>
<td>69/W/F</td>
<td>Right dorsal hand</td>
<td>38.5°C</td>
<td>WBC, 10.3 × 10^9/µL; PMN, 8.9 × 10^9/µL; ESR, 43 mm/h; CRP, 18.1 mg/dL; Hb, 11.9 mg/dL</td>
<td>Ulcerative, sheets of PMNs in dermis, no vasculitis, PAS and GMS negative</td>
<td>B cell lymphoma, HTN, COPD</td>
<td>Prednisone tapered over 4 mo</td>
<td>Resolved</td>
<td>7</td>
</tr>
<tr>
<td>71/W/F</td>
<td>Bilateral dorsal hands, forearms, buttocks, thighs</td>
<td></td>
<td>WBC, 15.8 × 10^9/µL; PMN, 11.9 × 10^9/µL; ESR, 23 mm/h; Hb, 14.7 mg/dL</td>
<td>Subepidermal edema and microabscess, dense interstitial and perivascular neutrophilic infiltrate, no vasculitis</td>
<td>HTN, GERD</td>
<td>Prednisone then dapsone, tacrolimus ointment</td>
<td>Resolved</td>
<td>9</td>
</tr>
<tr>
<td>32/W/M</td>
<td>Bilateral dorsal hands, fingers</td>
<td></td>
<td>WBC, 12.1 × 10^9/µL; PMN, 9.5 × 10^9/µL; CRP, 18.5 mg/dL; Hb, 10.6 mg/dL</td>
<td>Florid, folliculocentric neutrophilic infiltrate in papillary/reticular dermis, fibrinoid necrosis</td>
<td>GERD</td>
<td>Prednisone, methotrexate, cyclophosphamide; refractory to colchicine, dapsone, SSKI, cyclosporin, thalidomide, minocycline</td>
<td>Improved</td>
<td>54</td>
</tr>
<tr>
<td>51/W/F</td>
<td>Bilateral dorsal hands, fingers, arms</td>
<td>37.6°C</td>
<td>WBC, 12.1 × 10^9/µL; PMN, 9.5 × 10^9/µL; CRP, 18.5 mg/dL; Hb, 10.6 mg/dL</td>
<td>Dense neutrophilic infiltrate subepidermal to subcutis, no vasculitis</td>
<td>Seizure disorder</td>
<td>Prednisone and SSKI; could not tolerate dapsone or colchicine</td>
<td>Improved</td>
<td>17</td>
</tr>
<tr>
<td>Summary: 8 F and 1 M</td>
<td>3 extending onto arms; 1 to thigh, 1 to hip</td>
<td>7/9 Afebrile</td>
<td>4/8 WBC; 7/7 TPMN; 7/7 TESR; 7/7 TCRP; 6/9 Anemic</td>
<td>1/9 Lymphoma, 1/9 solid malignancy, 1/9 IBD</td>
<td>3 Prednisone monotherapy, 4 prednisone + dapsone, 1 prednisone + SSKI, 1 prednisone + methotrexate</td>
<td>9/9 Improved, 5/9 resolved off therapy</td>
<td>Average follow-up, 15.4</td>
<td></td>
</tr>
</tbody>
</table>

Abbreviations: AFB, acid-fast bacilli stain; COPD, chronic obstructive pulmonary disease; CRP, C-reactive protein; ESR, erythrocyte sedimentation rate; ESRF, end-stage renal failure; GERD, gastroesophageal reflux disease; GMS, Gomori methenamine silver; Hb, hemoglobin; HD, hemodialysis; HTN, hypertension; IBD, inflammatory bowel disease; PAS, periodic acid–Schiff stain; PMN, polymorphonuclear cell; PNGD, palisaded neutrophilic granulomatous dermatitis; SSKI, saturated solution of potassium iodide; W, white; WBC, white blood cell count; ↑, increased.
CASE 5

A 30-year-old woman was seen for an enlarging ulcer on her left dorsal hand for 4 days. She denied having fever, prior trauma, or a history of similar lesions. Physical examination revealed a 4 × 4-cm violaceous, firm, centrally necrotic plaque on the left dorsal hand with 2 erythematosus 4-mm papules on the forearm. Laboratory studies revealed elevated inflammatory indexes and leukocytosis. A cutaneous biopsy was performed (Table 1). She was treated with prednisone, 60 mg/d (tapered over 2 weeks), and topically with a combination of bacitracin zinc and polymyxin B sulfate (Polysporin; Glaxo Wellcome, Research Triangle Park, NC) and adaptic gauze, with resolution.

CASE 6

A 69-year-old woman was seen for a tender ulcer on her right thumb for 2 weeks, which was treated with oral antibiotic without improvement. She then sustained cat bites on her right index and middle fingers with resulting pathergic ulcerations, which was unresponsive to treatments with intravenous (IV) levofoxacin and metronidazole hydrochloride and then ampicillin-sulbactam. A biopsy was performed (Table 1). On transfer to our institution, physical examination revealed a 2-cm ulceration with a violaceous border on her right dorsal thumb with smaller ulcers on bilateral second and third dorsal fingers. Laboratory studies revealed elevated inflammatory indexes and a monoclonal gamma globulinemia. Antibiotic treatments were discontinued, and prednisone therapy was started at 60 mg/d. The lesions slowly resolved, and prednisone therapy was tapered over 4 months. Subsequent oncologic evaluation disclosed a low-grade B-cell lymphoma, which was monitored clinically.

CASE 7

A 71-year-old woman was seen for a pruritic eruption initially appearing on her right wrist and hand spreading to the forearms and trunk. The eruption had begun 2 weeks after right knee arthroplasty. She denied having fever, local trauma, or a history of similar lesions. Physical examination revealed violaceous 3- to 4-cm plaques on the right dorsal wrist and hand, with smaller (1- to 1.5-cm) erythematous, succulent plaques on the thighs. A cutaneous biopsy was performed (Table 1). Treatment with prednisone, 30 mg/d, and 0.1% triamcinolone acetonide ointment resulted in improvement after 1 week. After 3 weeks of tapering prednisone therapy, her lesions flared, prompting a 4-week tapering of prednisone therapy, while dapsone therapy was started at 50 mg/d. She continues to receive treatment with dapsone and 0.1% tacrolimus ointment without recurrence.

CASE 8

A 31-year-old man presented in November 1998 with spontaneously arising bullae on his dorsal hands and fingers, which expanded into tender ulcers. A biopsy revealed a dense neutrophilic infiltrate with papillary dermal edema without vasculitis. Laboratory studies revealed leukocytosis, neutrophilia, and elevated liver function test results. Findings from serum protein electrophoresis, urinalysis, and urine porphyrins were normal. Bullous Sweet syndrome was diagnosed. He began treatment with prednisone, 60 mg/d, and topical Polysporin, with initial improvement. His disease flared over the next 6 months during any attempt to taper prednisone therapy, and he did not tolerate treatments with colchicine or dapsone. Flares responded briefly to treatment with IV methylprednisolone sodium phosphate. Minocycline hydrochloride therapy...
was added in August 1999 without significant benefit. A reevaluation in October 1999 revealed violaceous ulcers on the right first and left fourth fingers (2 cm) and the left dorsal hand (5 cm). A second biopsy (Table 1) revealed vasculitis, and his condition was now interpreted as atypical PG. Findings from colonoscopy and bone marrow biopsies were normal. Treatment with topical tacrolimus was not helpful, and 3-month trials of cyclosporine and thalidomide failed. By May 2000, lesions had developed on his ears and shoulders, which were poorly controlled with prednisone therapy, 40 mg/d, with methylprednisolone pulses every 3 to 4 weeks. Cyclophosphamide, 500 mg, was administered IV 4 times over 2 months with improvement, allowing for tapering of prednisone therapy to 25 mg/d. Therapy with saturated solution of potassium iodide was added in October 2000. He developed indurated papules on his forearms, and a biopsy confirmed lymphocytic infiltrate of Jessner. Therapy with hydroxychloroquine sulfate, 200 mg/d, was added. His disease flared despite increased use of corticosteroid, and in December 2001, cyclophosphamide was resumed monthly for 5 months, allowing for tapering of prednisone therapy to 20 mg/d. Cyclophosphamide therapy was subsequently discontinued because of concern for toxic effects, and he began methotrexate therapy in July 2002, at a weekly dose of 17.5 mg, allowing for tapering of prednisone therapy to 10 mg/d. His disease was well controlled in May 2003, at the time of his accidental death.

**CASE 9**

A 51-year-old woman developed expanding ulcers on her hands and arms for 2 weeks. The onset was associated with scratches from her domestic cat. A local physician began therapy with antibiotics (amoxicillin–clavulanic acid and then IV ampicillin-sulbactam) without improvement. Bacterial and fungal cultures were negative, and she denied having chills or sweats. Her medical history included depression, migraines, and syncopal episodes during the past year, which were diagnosed as transient ischemic attacks. On physical examination, superficially ulcerated plaques with violaceous undermined borders were present on the dorsal hands, left fifth finger, and left dorsal forearm, without purulent discharge (Figure 3A). Laboratory studies revealed an elevated C-reactive protein level, leukocytosis, neutrophilia, and anemia. Therapy with oral antibiotics was discontinued, and her wounds were treated with bacitracin zinc and nonadherent gauze. Prednisone therapy was begun at 60 mg/d (1.25 mg/kg), and she was hospitalized. A skin biopsy was performed (Figure 3B; Table 1). The ulcerations were improving 3 days later, and she was discharged on therapy with tapering doses of pred-
nisone and fluocinonide ointment. New plaques on the hands developed as the prednisone dose was decreased to 15 mg/d, and dapsone therapy was added (50 mg/d), with doses changed to 10 mg/d and 100 mg/d, respectively, after 4 weeks. Two months later, dapsone therapy was discontinued owing to hemolytic anemia, and the prednisone dose was increased to 20 mg/d. Shortly thereafter she was hospitalized for confusion and new-onset seizures; encephalopathic Sweet syndrome was considered. Magnetic resonance imaging disclosed chronic ischemia of the right frontal lobe; findings from electroencephalography and lumbar puncture were normal. A bone marrow biopsy revealed mildly hypercellular marrow. She began gabapentin therapy for her seizure disorder. During this time, she developed new ulcers on her hands and arms at venipuncture sites, prompting an increase in the prednisone dose to 30 mg/d. Colchicine therapy was not tolerated. Therapy with saturated solution of potassium iodide was begun (15 drops 3 times daily). Her cutaneous lesions gradually regressed (Figure 3C). Prednisone and topical corticosteroid treatments were withdrawn over 2 months, and therapy with saturated solution of potassium iodide was tapered over the following 2 months.

Since the original description of acute febrile neutrophilic dermatosis by Sweet in 1964, diagnostic criteria have been variably proposed and generally include the abrupt onset of typical skin lesions with characteristic histopathologic features as major criteria and elevation of serum inflammatory markers, presence of constitutional symptoms or associated infection, inflammatory, or malignant disease, and responsiveness to corticosteroid therapy as minor criteria. In 1995, Strutton et al reported 6 patients with violaceous plaques on the radial aspect of the dorsal hands, with histologic features of papillary dermal edema, brisk neutrophilic infiltrate, and leukocytoclastic vasculitis. The clinicopathologic features and responsiveness to corticosteroid therapy were suggestive of acute febrile neutrophilic dermatosis (Sweet syndrome), though the authors termed the presentation *pustular vasculitis of the hands* to reflect the vascular damage. Pustular vasculitis had been initially applied to purpuric lesions of Behçet syndrome and bowel-associated dermatosis-arthrosis syndrome. In 2000, Galaria et al presented 3 similar cases that lacked leukocytoclastic vasculitis and proposed the term *neutrophilic dermatosis of the dorsal hands* as a localized variant of Sweet syndrome. In 2002, DiCaudo and Connolly reported 7 additional cases, all with vasculitis and unassociated with systemic illness. In 2004, Weenig et al presented 4 new cases and reviewed the other 32 cases then in the literature, concluding that the presentation warranted evaluation for occult malignancy or other systemic disease. Three additional cases have since been reported. Many of these authors have commented on the morphologic similarity of NDDH to Sweet syndrome. Clinical features of our 9 patients are summarized in Table 1.

We agree with Galaria et al that NDDH is a subset of Sweet syndrome. Assimilation of reported cases supports this idea. Of 52 patients, 36 (69%) were female, in accordance with the female preponderance for Sweet syndrome. Of 52 cases of NDDH, 11 (21%) (1 in our series and 10 in the review by Weenig et al) had associated leukemia, myelodysplasia, or other hematologic disease. This is in agreement with the incidence of hematopoietic disease (16%-54%) in 3 large reviews of Sweet syndrome. Of 52 cases of NDDH, 8 (15%) (1 in our series and 7 in the review by Weenig et al) had associated inflammatory bowel disease, similar to the association (up to 19%) with Sweet syndrome. Most of our 9 patients showed fever, peripheral neutrophilia, leukocytosis, and/or elevated erythrocyte sedimentation rate or C-reactive protein level. Of the 52 assimilated cases, 17 (33%) mentioned fever and 17 mentioned leukocytosis or peripheral neutrophilia, and 37 (71%) responded to systemic corticosteroid therapy.

While an ulcerative pattern is uncommon in Sweet syndrome (and when present may suggest hematologic malignancy), ulceration is not unusual in cases of NDDH (27 of 52 cases) and is the clinical sine qua non of PG. While NDDH is clearly distinct from classic PG, it bears resemblance to vesiculobullous PG, also called atypical PG, which is also similar and may be identical to bullous or atypical Sweet syndrome. Atypical PG characteristically presents as hemorrhagic bullous lesions, which superficially ulcerate and is most commonly found on the dorsal hands. Characteristically, PG involves the lower extremities as deeper ulcers with overhanging borders and often excruciating pain. Atypical PG is distinguished from typical PG histologically by the frequent presence of a diffuse neutrophilic infiltrate with associated vascular reaction and therapeutically by a much prompter remission. Each of these features of atypical PG is similar to NDDH.

It is also likely that many of the cases termed *atypical Sweet syndrome or PG-Sweet overlap* belong to the spectrum of neutrophilic dermatoses now increasingly recognized as NDDH when present in this distribution. Atypical Sweet syndrome has been reported to occur frequently in patients with hematologic malignancy, and many of the published photographs of cancer-associated Sweet syndrome closely resemble lesions of NDDH seen in the presence and absence of malignancy. Callen has also noted the similarity between bullous Sweet syndrome and atypical PG and opined that pustular vasculitis of the hands is a variant of atypical PG. At present, as more cases of NDDH have been recognized and reported, the association has crystallized and can be stated more definitively; these 3 entities in fact represent the same disease. When features of NDDH, atypical PG, and Sweet syndrome are numerically compared (Table 2), the similarity is striking.

The fact that the eruptions of NDDH share features of both Sweet syndrome and atypical PG underscores the utility of the designation. Weenig et al proposed shortening the designation of NDDH to *neutrophilic dermatosis of the hands*, though we prefer to retain the term dorsal to reflect the primary distribution in most cases. Many of the reported cases of NDDH discuss suspected infection and failure of antibiotic therapy prior to dermatologic consultation. Of our 9 patients, 6 had been treated as such prior NDDH diagnosis. While most cases respond to treatment with prednisone or steroid-sparing immunosuppressants, at least 10 of 52 reported cases responded to
Table 2. Comparison of Features of NDDH, Sweet Syndrome, and Atypical Pustular Vasculitis (PG)

<table>
<thead>
<tr>
<th>Feature</th>
<th>NDDH (n = 52)</th>
<th>Sweet Syndrome (n = 22)</th>
<th>Atypical PG</th>
</tr>
</thead>
<tbody>
<tr>
<td>Female-male ratio</td>
<td>2.2</td>
<td>3.7*</td>
<td>1.44</td>
</tr>
<tr>
<td>Mean age at onset, y</td>
<td>60</td>
<td>53*</td>
<td>52</td>
</tr>
<tr>
<td>Malignancy, %</td>
<td>27</td>
<td>21*</td>
<td>27</td>
</tr>
<tr>
<td>Inflammatory bowel disease, %</td>
<td>15</td>
<td>16*</td>
<td>5</td>
</tr>
<tr>
<td>Total with systemic disease, %</td>
<td>42</td>
<td>27*</td>
<td>45</td>
</tr>
<tr>
<td>Neutrophil infiltrate on histologic examination, %</td>
<td>95</td>
<td>100</td>
<td>83</td>
</tr>
<tr>
<td>Vasculitis, %</td>
<td>25</td>
<td>18*</td>
<td>0</td>
</tr>
<tr>
<td>Response to steroid, %</td>
<td>71</td>
<td>97*</td>
<td>23</td>
</tr>
</tbody>
</table>

Abbreviation: NDDH, neutrophilic dermatosis of the dorsal hands.
*Modified from Bennett et al.†
†4:1 for classic Sweet syndrome; 1:1 for malignancy-associated Sweet syndrome.

The nosologic classification of this disease as a limited distribution variant of Sweet syndrome is supported by the clinical presentation, laboratory and histologic findings, presence of comorbid medical conditions, and response to treatment. When present on the dorsal hands, atypical PG is clinically indistinguishable and represents the same disease. We view NDDH as a distribution variant of Sweet syndrome, which is identical to atypical PG and pustular vasculitis of the hands, and propose that the term neutrophilic dermatosis of the dorsal hands is the most appropriate designation for such eruptions.

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Correspondence: Hobart W. Walling, MD, PhD, Dermatology, P.C., 6000 University Ave, Suite 450, West Des Moines, IA 50266 (hobartwalling@yahoo.com).

Author Contributions: Study concept and design: Walling and Piette. Acquisition of data: Walling, Snipes, and Gerami. Analysis and interpretation of data: Walling. Drafting of the manuscript: Walling, Snipes, and Gerami. Critical revision of the manuscript for important intellectual content: Walling and Piette. Statistical analysis: Walling. Study supervision: Walling and Piette.

Financial Disclosure: None.

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18. Bennett ML, Jackson JM, Jorizzo JL, Fleisher AB, White WL, Callen JP. Pyoderma gangrenosum and Sweet’s syndrome, which is identical to atypical PG and pustular vasculitis of the hands, and propose that the term neutrophilic dermatosis of the dorsal hands is the most appropriate designation for such eruptions.