Neutrophilic Dermatosis of the Dorsal Hand

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Abstract: Neutrophilic dermatosis of the dorsal hands (NDDH) is a rare condition often misdiagnosed as a localized cutaneous infection. A type of neutrophilic dermatosis, NDDH is considered to be a subset of Sweet’s syndrome and is similar to pyoderma gangrenosum. The authors report 2 cases of male patients who presented with persistent ulcerative skin lesions. In both cases, biopsy revealed neutrophilic infiltration consistent with NDDH. Patient 1 had been unsuccessfully treated with antibiotics. Lesion biopsy showed epidermal hyperplasia with spongiosis and overlying scale crust, as well as dense neutrophilic infiltration of the underlying dermis. Periodic acid-Schiff (PAS) stain was negative for fungal and yeast organisms. A biopsy of a lesion from patient 2 indicated focal parakeratosis, perivascular inflammatory cell infiltrate, and scarring. Both patients were prescribed oral and/or topical corticosteroids and the condition resolved in 1-2 months. The clinical presentation, histological features, and excellent response to corticosteroid treatment are consistent with diagnosis of NDDH. Delayed recognition of neutrophilic dermatosis and treatment of secondary infection resulted in delayed treatment. Improved recognition of neutrophilic dermatoses is important for optimal management of NDDH.

Key words: acute febrile neutrophilic dermatosis, pustular vasculitis, Sweet’s syndrome, wounds, ulcers

Neutrophilic dermatosis of the dorsal hand (NDDH) is a rare condition characterized by painful pustule ulcerations usually localized to the dorsal and lateral aspect of the hand, however additional lesions may also present elsewhere.1,2 Lesions are characterized by heavy neutrophilic infiltration of the dermis, which may be accompanied by vasculitis in some cases.3,4 Neutrophilic dermatosis of the dorsal hand is more common in women and is morphologically similar to Sweet’s syndrome (Table 1).5,6 Sweet’s syndrome, also called acute febrile neutrophilic dermatosis, is a condition characterized by painful erythematous cutaneous lesions with neutrophilic infiltration. These are classical, malignancy-associated or drug-induced variants, which improve rapidly with corticosteroid therapy.5,6 Like Sweet’s syndrome, the pathogenesis of NDDH is generally unknown, but may be precipitated by an immunologic or drug reaction.1,2 The hypothesis of altered immunological activity is
consistent with neutrophilic infiltration and response to topical and/or systemic glucocorticoid treatment.

Though NDDH is not thought to originate from a bacterial infection, infection of the lesion is not uncommon and can often lead to misdiagnosis, delaying appropriate treatment.7

Case Reports

**Patient 1**. A 64-year-old male presented with a lesion on the left index finger and complained of increasing redness, tenderness, and swelling, which the patient thought was due to a spider bite. Minimal violaceous erythema was also evident on the dorsum of the right index finger. No other symptoms were present. Patient had a history of type 2 diabetes, hypertension, and arthritis, as well as a remote history of seizures, and was taking glipizide, losartan, atenolol, diltiazem, aspirin, and hydrocodone. Cutaneous infection was suspected and had been unsuccessfully treated with antibiotics (levofloxacin) over the previous month. Multiple lesions on both hands coalesced and formed blistered, bullous, and ulcerative lesions, with a secondary finding of infection. Cultures of the lesions on the left index finger were positive for yeast. The patient was placed on doxycycline 100 mg twice a day, and fluconazole 100 mg daily. Cellulitis and necrosis of the left index finger improved spontaneously, although erythema on the right hand index finger continued.

The patient presented 7 months later with painful blisters, erythematous, violaceous purple areas on the right and left index fingers (Figure 1). Shave biopsies of the left hand revealed subepidermal bulla with neutrophils. The patient was placed on co-trimoxazole 1 tablet twice a day, and betamethasone ointment 0.05% to be applied twice daily. An additional culture was positive for pseudomonas and the patient was placed on ciprofloxacin 500 mg twice a day for 10 days. Due to an excellent response to the betamethasone, the patient was prescribed oral methyl prednisolone to supplement the topical betamethasone and significant

<table>
<thead>
<tr>
<th>Types of neutrophilic dermatoses</th>
<th>Pyoderma gangrenosum</th>
<th>Sweet’s syndrome (acute febrile neutrophilic dermatosis)</th>
<th>NDDH (subset of Sweet’s syndrome)</th>
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<tbody>
<tr>
<td><strong>Clinical features</strong></td>
<td>Painful enlarging cutaneous ulcerations with a purple undermined border, most common in the lower legs but may present on the hands (atypical) or elsewhere.</td>
<td>Tender erythematous nonulcerated papules and nodules that coalesce to form plaques. Most occur on the face, arms, and neck.</td>
<td>Similar to Sweet’s syndrome, but lesions are located on the dorsal aspect of the hand (may also occur on the palmar surface); subsequent lesions may present, usually on the arms and back.</td>
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<td><strong>Presentation</strong></td>
<td>More common in individuals &gt; 50 years old; often associated with systemic disease, or may be drug-induced.</td>
<td>Often associated with systemic disease, especially fever, carcinoma, or inflammatory disease; may be drug-induced.</td>
<td>Similar to Sweet’s syndrome.</td>
</tr>
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<td><strong>Diagnosis</strong></td>
<td>Clinical features and exclusion of other causes; neutrophilic infiltration of dermis may not always be present.</td>
<td>Clinical features and skin biopsy showing neutrophilic infiltration of the dermis, and vasculitis may also be present.</td>
<td>Similar to Sweet’s syndrome.</td>
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<td><strong>Treatment</strong></td>
<td>Removal of necrotic tissue; immunosuppressive treatment consisting of systemic or topical corticosteroids, dapsone, or both; potassium iodide.</td>
<td>Immunosuppressive treatment consisting of systemic or topical corticosteroids, dapsone, or both; potassium iodide, or colchicine is effective in individuals where corticosteroid therapy is contraindicated; may also resolve spontaneously.</td>
<td>Similar to Sweet’s syndrome.</td>
</tr>
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Table 1. Overview of pyoderma gangrenosum, Sweet’s syndrome, and neutrophilic dermatosis of the dorsal hands (NDDH).
improvement was noted within 1 week. The lesions resolved within 3 weeks after starting betamethasone (Figure 1). Length of follow-up was 2 months.

Patient 2. A 54-year-old male presented due to an intermittent, ongoing problem for which he had received treatment from multiple providers for 2 years. The patient had an ulcer on the fourth finger of the right hand, and serous weeping cellulitis was evident (Figure 2). The patient had been prescribed a lidocaine patch (for pain), as well as minocycline and ciprofloxacin. There was a history of hypertension and hypercholesterolemia, for which the patient was receiving lisinopril and simvastatin. No systemic symptoms were evident. Aerobic cultures were positive for *Enterococcus* and skin flora. A biopsy revealed focal parakeratosis, perivascular neutrophilic infiltration, scarring, and prominent solar elastosis. An x-ray was negative for any erosive changes, foreign bodies, or fracture, and metabolic and lipid panels were normal. Because of neutrophils present in the biopsy, the patient was started on prednisone 20 mg daily. Significant improvement was evident as early as 4 days after beginning prednisone. The condition resolved within 2 months (Figure 2).

Discussion

Neutrophilic dermatoses include Sweet’s syndrome and pyoderma gangrenosum (Table 1). The etiology of pyoderma gangrenosum and Sweet’s syndrome is similar, and both conditions respond to immunosuppressive treatment; however, Sweet’s syndrome is characterized by tender erythematous nonulcerated plaques, whereas pyoderma gangrenosum consists of cutaneous ulceration with a purple undermined border. Neutrophilic dermatosis of the dorsal hand and Sweet’s syndrome lesions are similar, although Sweet’s syndrome lesions appear mainly on the face, arms, and neck. In NDDH, lesions present on the dorsal aspect of the hand but may also occur on the palmar surface of the hand. Subsequent lesions may present in other locations including the arm and back. Lesions are characterized by dense dermal infiltration consisting of neutrophils; the extent of vascular endothelial damage varies and may range from endothelial swelling to ulceration, vasculitis, and fibrinoid necrosis. The exact cause of NDDH is unknown. Similar to Sweet’s syndrome, NDDH often presents with concurrent disorders, including fever, carcinoma, hepatitis C, bowel disorders, urinary tract infection, drug reaction, arthritis, and thermal injury.

**Figure 1.** Lesions on the right and left index finger of Patient 1 (left); and after treatment (right).

**Keypoints**

- Neutrophilic dermatosis of the dorsal hand is a relatively new and rare condition.
- To date, NDDH reports include approximately 60 individuals, the majority occurring in females.
- NDDH is often misdiagnosed as a cutaneous infection and unsuccessfully treated with topical or systemic antibiotics.
- Prompt diagnosis may prevent unnecessary medical or surgical therapy.

Neutrophilic dermatosis of the dorsal hand is a relatively new and rare condition. In 1995, Strutton et al described 6 cases in adult women where NDDH lesions were noted for their resemblance to those observed in Sweet’s syndrome, except that they were localized to...
the dorsal and radial aspect of the hand and the first 3 fingers. Lesions consisted of a symmetric eruption of papules and plaques, were associated with fever, sterile culture, neutrophils, nonresponse to antibiotic therapy, and rapid response to corticosteroid treatment. Due to the presence of leukocytoclastic vasculitis, the condition was termed pustular vasculitis of the hands. Subsequent publication of similar cases also included 2 men.9,13 In 2000, Galaria et al11 described 3 additional patients with hand lesions and introduced the term NDDH because vasculitis was not always present. Neutrophilic dermatosis of the dorsal hand was also introduced as a subset of Sweet’s syndrome.4,11 A genetic similarity between Sweet’s syndrome and NDDH has been suggested.18 To date, NDDH reports include approximately 60 individuals, the majority of whom are females.1,4,7,10,14-24

Neutrophilic dermatosis of the dorsal hand presentation is distinctive but rare. Thus, NDDH is often misdiagnosed as a cutaneous infection and unsuccessfully treated with topical or systemic antibiotics.7,17 Prompt diagnosis may prevent unnecessary medical or surgical therapy.7 In NDDH and Sweet’s syndrome, lesions rapidly respond to treatment with systemic or topical corticosteroids, dapsone, or both; the use of topical corticosteroids is effective in minor lesions.1,2,7 It has been suggested that other treatments for Sweet’s syndrome, potassium iodide or colchicine, may also be effective for NDDH in patients whom corticosteroid therapy is contraindicated.1,2 Cases of spontaneous healing have also been reported.16,24,25 The incidence of reoccurrence varies from frequent11 to rare.1

**Conclusion**

This report looked at the treatment of 2 male patients who presented with dorsal hand ulcerative lesions. The primary diagnosis was NDDH, though the purple violaceous border noted in Patient 1 is similar to the wound margins commonly seen in ulcerative pyoderma gangrenosum.8 Bacterial and fungal infections were presumed to be secondary to NDDH and were treated with antibiotics and antifungals. Lesions improved spontaneously in Patient 1 and subsequently reappeared. Presence of neutrophils on biopsy precipitated treatment with corticosteroids (prednisone, betamethasone, or methylprednisolone, systemic and/or topical), which resolved the condition in both cases.

Neutrophilic dermatosis of the dorsal hand is a seldom-seen condition that responds well to treatment with corticosteroids. Because of the rarity of the condition and concurrent presentation of inflammation, ulceration, and infection, treatment may be delayed due to incorrect or belated diagnosis. Often, patients are treated for secondary infection while the primary cause of the lesions and inflammation is overlooked. Greater awareness of NDDH and similar conditions among primary care physicians would speed appropriate treatment and resolution of symptoms.

**References**


