

## CASE REPORT

# A Case of Neutrophilic Dermatositis of the Dorsal Hands with Concomitant Involvement of the Lips

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Neutrophilic dermatosis of the dorsal hands (NDDH) is a localized, pustular variant of acute febrile neutrophilic dermatosis (Sweet syndrome). The lesions of NDDH clinically resemble those of Sweet syndrome (SS), but they differ from classic SS according to their locations (NDDH is predominantly restricted to the dorsal hands) and the smaller number of accompanying systemic symptoms. The histology of the NDDH lesion shows a dense dermal neutrophilic infiltration. The lesions rapidly resolve after systemic corticosteroid and/or dapsone therapy. We herein report on a case of neutrophilic dermatosis of the dorsal hands in a 34-year-old woman. The patient also had skin changes with erythematous plaque on the right lips. (*Ann Dermatol* 22(1) 106~109, 2010)

**-Keywords-**

Dorsal hand, Neutrophilic dermatosis, Sweet syndrome

## INTRODUCTION

Sweet syndrome (SS) is an acute febrile neutrophilic dermatosis that is clinically characterized by fever, neutrophilia and painful erythematous skin lesions, and it is histologically characterized by a predominantly mature neutrophilic dermal infiltrate<sup>1</sup>. Neutrophilic dermatosis of the dorsal hands (NDDH) is considered to be a rare localized variant of SS with less frequent systemic symptoms. NDDH begins on the hands and it can spread to

other locations<sup>2</sup>. We present here the case of a 34-year-old woman with recurrent solitary tender lesions on the right hand and lips, but she had no associated systemic symptoms.

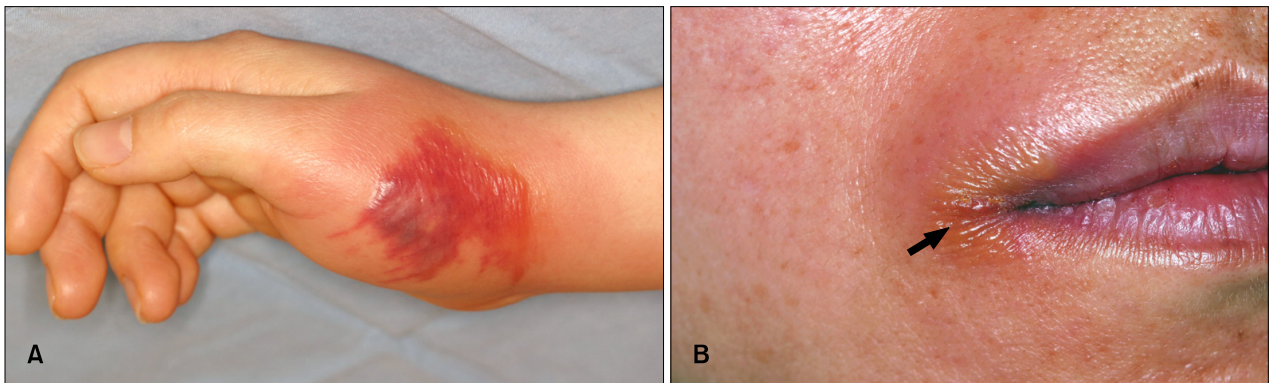
## CASE REPORT

A 34-year-old woman presented with a one-week history of erythematous skin rash on the hand. This lesion had recurred each spring for the last four years. She had another cutaneous lesion on the right lips that had first developed one day previously. Her medical and family histories were noncontributory. She was receiving no medications. She had no underlying diseases such as hematologic malignancy or inflammatory bowel disease. Physical examination revealed a solitary, tender, erythematous, well-demarcated plaque on the dorsum of the right thumb (Fig. 1A) and an erythematous plaque with small vesicles on her right lips (Fig. 1B). She had a febrile and chilling sensation one week previously. She had no fever, arthralgia or generalized malaise at the time of diagnosis. Biopsy specimens from both the lesions on the dorsal hand and lips revealed edema of the papillary dermis and a dense perivascular infiltrate of neutrophils with leukocytoclasia throughout the upper to mid dermis, but there was no evidence of true vasculitis (Fig. 2, 3). Gram staining of the tissue was negative. The laboratory studies revealed a normal erythrocyte sedimentation rate (ESR). Her white blood cell count was 9,800 cells  $\mu\text{l}^{-1}$  with 68.8% neutrophils, the haemoglobin level was 13.5 g/dl and the platelet count was 330,000/ $\mu\text{l}$ . The result of the serum electrolyte assessment, the renal and liver function panel and the urinalysis were normal. The serum albumin and alkaline phosphatase were normal.

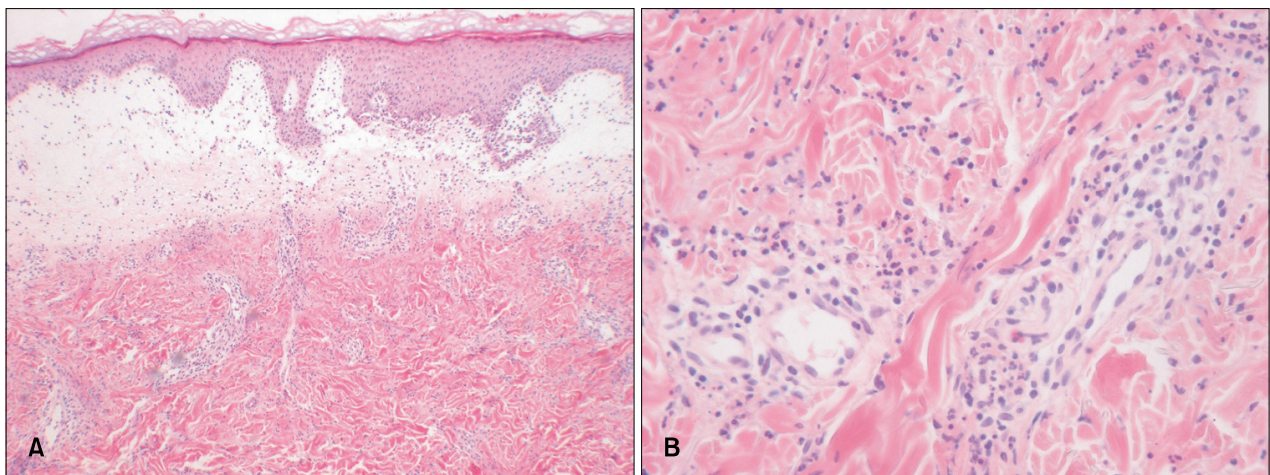
A diagnosis of NDDH was made, and the patient was treated with prednisone 20 mg daily for one week and

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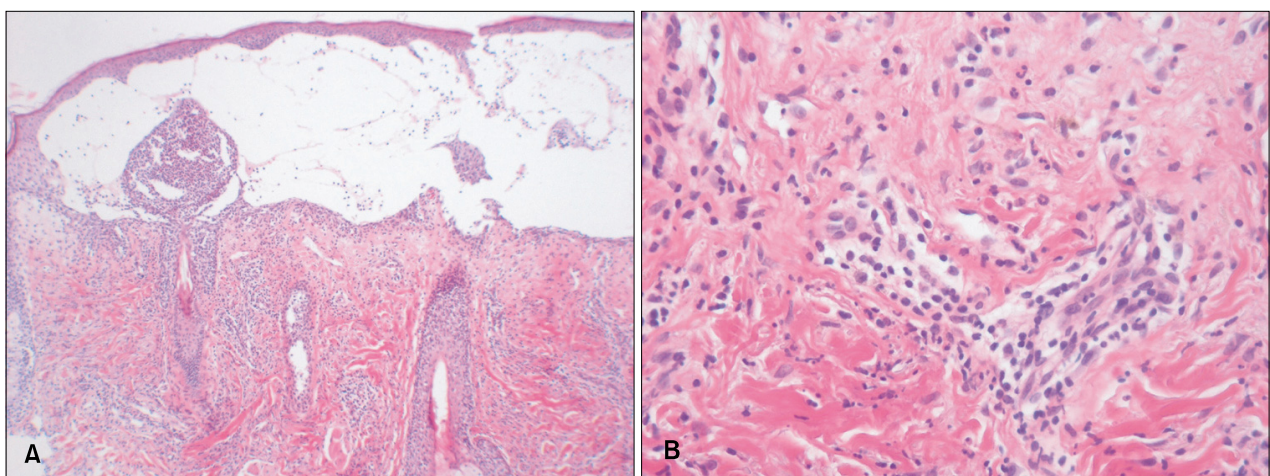
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**Fig. 1.** (A) Tender, erythematous, well-demarcated plaques on the dorsum of the right hand. (B) Erythematous plaque with small vesicles (arrow) on the right lips.



**Fig. 2.** The skin biopsy from the right hand shows prominent edema in the papillary dermis and a perivascular cellular infiltration that is mainly composed of neutrophils with no features of true vasculitis (A: H&E, ×40; B: H&E, ×200).



**Fig. 3.** The skin biopsy from the lips shows subepidermal separation and a perivascular and interstitial mixed-cell infiltrate composed of neutrophils and lymphocytes (A: H&E, ×40; B: H&E, ×200).

then 5 mg daily for the following one week. The lesions resolved without scarring or recurring during the 6 months of follow-up.

## DISCUSSION

NDDH is a rare, localized variant of SS, and this was first described by Galaria et al.<sup>3</sup> in 2000. The pathogenesis of SS may be multifactorial and it remains to be definitively determined<sup>4</sup>. A septic process, a hypersensitivity reaction, leukotactic mechanisms and cytokines have all been postulated to contribute to the pathogenesis of SS<sup>4</sup>. Clinically, NDDH is characterized by tender, erythematous plaques, pustules and bullae that are generally limited to the dorsal hands and fingers<sup>3</sup>. This is usually limited to the dorsal hands, and often with a predilection for the lateral aspect of the hand between the thumb and index finger<sup>5</sup>. However, only several cases have been reported in which there were concurrent lesions located on either the arm, leg, back and/or face<sup>1</sup>.

The histology of NDDH shows prominent papillary dermal edema and a dense diffuse infiltration of mature neutrophils throughout the upper dermis<sup>3</sup>. Swollen endothelial cells, dilated small blood vessels and fragmented neutrophil nuclei can also be present<sup>4</sup>. Fibrin deposition or neutrophils within the vessel walls (the changes of "primary" leukocytoclastic vasculitis) is usually absent<sup>4</sup>. In 1995, Strutton et al.<sup>5</sup> described six patients with hand lesions that clinically resembled NDDH, but the lesions had the histological features of leukocytoclastic vasculitis, and so he proposed a new entity called "pustular vasculitis of the hands". However, Gilaberte et al.<sup>6</sup> suggested that the vasculitis might be a secondary event related to the intensity of the neutrophilic infiltrate and the time of evolution of the lesions.

The clinical presentation of NDDH differs from that of classic SS. In classic SS, fever, leukocytosis and an increased ESR are observed in 80~90% of the cases; inflammatory bowel disease (16%) and hematologic disease (54%) may be present as the accompanying diseases<sup>4,7</sup>. In NDDH, the lesions are clinically restricted to the hands; fever, leukocytosis and an increased ESR are observed in 33% of the cases and hematologic malignancy is present as an accompanying disease in 21% of the cases<sup>7</sup>.

The differential diagnosis of NDDH includes allergic contact dermatitis, cellulitis and pyoderma gangrenosum (PG). Allergic contact dermatitis is an eczematous dermatitis that presents as severe pruritus that develops in regions exposed to allergen, and allergic contact dermatitis shows spongiotic dermatitis histologically<sup>8</sup>. Cellulitis or erysipelas is excluded according to the negative

tissue cultures and stains and a lack of response to antibiotics<sup>3</sup>. Atypical pyoderma gangrenosum, also known as vesiculobullous PG, presents with chronic hemorrhagic bullous lesions that ulcerate superficially and are usually seen in patients with leukemia or polycythemia vera<sup>7</sup>. However, pyoderma gangrenosum may be histologically indistinguishable from NDDH, although the first usually shows true vasculitis<sup>7</sup>. Thus, atypical PG and pustular vasculitis of the dorsal hands are arguably within the spectrum of a single disease entity, which is most appropriately termed NDDH<sup>9</sup>.

The treatment for NDDH is believed to be same as that for SS. Many different treatments have been used for SS with different rates of success and relapse<sup>10</sup>. These include corticosteroids, dapsone, potassium iodide, colchicine, clofazimine, azathioprine, danazol, tetracyclines and cyclosporine. Systemic corticosteroids are the most common first-line therapy<sup>10</sup>. Walling et al.<sup>7</sup> showed that treatment with systemic corticosteroids was successful in 71% of the cases.

In summary, we have presented a case of recurrent NDDH with no associated systemic signs and symptoms and the patient was successfully managed with low dose systemic corticosteroids. There has been only one case of NDDH with bacterial endocarditis in the dermatological literature<sup>11</sup>. The remarkable characteristic of our patient was the concomitant involvement of the lips. We suggest that dermatologists consider this disease when they encounter a patient with a tender erythematous bullous lesion occurred on the dorsal hands.

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