Neutrophilic Dermatosis of the Hands: A Case Report

Neutrophilic dermatosis of the hands (NDDH) is a localized variant of Sweet's syndrome which has been recently introduced. Strutton *et al.*in 1996 and then in Galaria *et al.* in 2000 reported cases with violaceous papulonodules on the dorsal surfaces of the hands with histopathological findings of a neutrophilic dermatosis in association with leukocytoclasia, but clinically and histologically without true vasculitis findings. Eventually, they proposed the term NDDH for these lesions (1,2).

A 46-year-old man was referred to our outpatient dermatology clinic with a painful ulcerative lesion on the dorsal side of the left hand that had been present for one year. Initially, the lesion had appeared as a small purulent papule, which gradually extended to a large ulcer. The patient underwent frequent referrals to several physicians and had taken a variety of topical and systemic antibiotics, meglumine antimoniate (Glucantime), and amphotericin with the clinical diagnosis of cutaneous bacterial or fungal infections, or leishmaniasis. All of these therapeutic regimes were ineffective in eradicating the lesion. Given the history, he denied any trauma to the site of lesion; he also did not report any similar lesions in his family.

The physical examination revealed an extensive tender ulcer of 4×7 cm^2 in size, with a shallow violaceous border superimposed on an edematous region on the dorsal side of the left hand. Atrophic scars resulting from old similar lesions were visible on the dorsal aspects of the 3rd and 4th proximal and middle metacarpal joints (Figure 1). The examination of the other parts of the body was unremarkable.

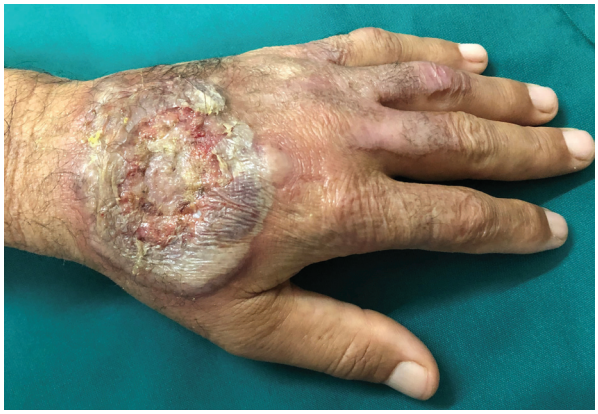

Figure 1. An extensive shallow ulcer with a raised border involving the dorsal aspect of the left hand along with two atrophic scars seen on the third and fourth fingers.

Laboratory tests showed an impaired white blood cell count and their differentiation, including leukocy-

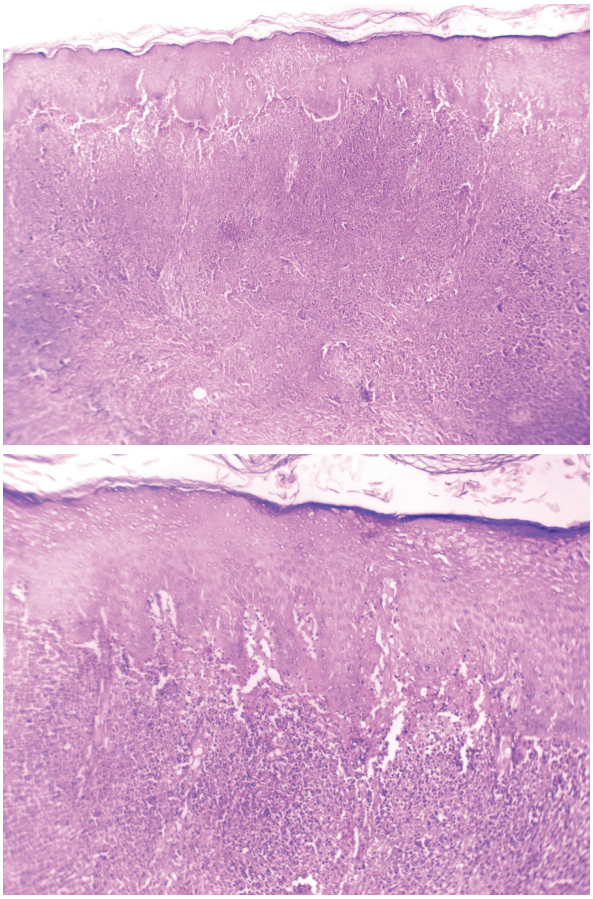

Figure 2. Histologic features of skin lesion shown in **Figure 1**, showing: (A) mild hyperkeratosis, spongiosis, irregular acanthosis, basal layer degeneration, and a dense infiltrate of neutrophils in the dermis (hematoxylin and eosin, ×40); (B) dermal edema and a dense massive neutrophilic infiltration in the dermis (hematoxylin and eosin, ×100); and (C) a dense neutrophilic infiltrate, slight vascular damage, and erythrocyte extravasation in the dermis (hematoxylin and eosin, ×100).

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tosis (white blood cell count of 16.12/mm^3) with neutrophilia (neutrophil percent at 65.9%). Additionally, altered liver function tests were remarkable for high serum levels of AST (SGOT) (105 IU/L) and ALT(SGPT) (355 IU/L), while the total bilirubin and alkaline phosphatase were within normal limits. Hemoglobin levels (13.90 g/dL) and platelet count (272/mm^3) were within normal range. The other laboratory tests, including serological tests for fasting blood sugar, hemoglobin A1c, creatinine, BUN, and an immunoassay for ruling out vasculitis lesions (anti-MPO (P-ANCA) and anti-PR3 (C-ANCA)) revealed no remarkable results. An erythrocyte sedimentation rate of 16 mm/h was reported.

A biopsy was performed. Histologic features demonstrated a dense, diffuse dermal infiltrate comprised almost entirely of neutrophils. The epidermis was slightly acanthotic and showed small foci of spongiosis, but the inflammatory infiltrate remained largely in the dermis. Sheets of neutrophils were present, admixed with karyorrhectic debris. The infiltrate did not appear to be peri-vascular, and most vessels that could be observed clearly appeared to be undamaged. However, some vessels appeared to show some neutrophils infiltrating vessel walls (Figures 2a, b). Based upon histopathologic examination, the diagnosis of neutrophilic dermatosis of the hands (NDDH) was suggested. The work-up findings for ruling out neoplastic diseases were unremarkable.

Clinically, patients with NDDH show various morphologic patterns of the lesions on the dorsal aspect of the hands, including violaceous edematous plaques or ulcers with undermined borders, hemorrhagic bullae, necrotic pyoderma-like lesions with pseudovesiculation, and atypical pyoderma gangrenosum-like lesions (1). This disease is more common in women (70%) than in men (3). NDDH has been reported in association with malignancies (such as leukemia and lymphoma), myelodysplasia, inflammatory bowel diseases, seropositive arthritis, sarcoidosis, HCV infection, and medications (such as lenalidomide, thalidomide, vaccinations, fertilizer, etc.) (1). Among them, neoplastic diseases are the most common association, which has been reported in 27% of the cases. It may thus represent a paraneoplastic phenomenon (3).

Histopathological study is mandatory for achieving a definite diagnosis of NDDH. Its pathological findings include subepidermal edema, a dense and diffuse dermal infiltration of neutrophils along with leukocytoclastic debris, and extravasated erythrocytes, which are not associated with true vasculitis (1,3). However, the presence or absence of some vasculitic features as a histopathological finding depends on the time of biopsy with regard to the evolutionary phases of the lesion (3). In our case, the diffuse nature of the infiltrate was somewhat indicative against the diagnosis of leukocytoclastic vasculitis. Additionally, the possibility of infection was excluded empirically (due to the ineffectiveness of previous therapies without doing cultures or PCR), and indirectly through biopsy.

Cohen (4) and Cohen and Kurzrok (5) explained the presence of vasculitis in Sweet's syndrome and NDDH as an epiphenomenon in which the damaged vessel is as an "innocent bystander" in the background of an inflammatory dermatosis. Eventually, they concluded that the presence or absence of vasculitis has a secondary importance in the diagnosis of NDDH.

The following entities should be considered in the differential diagnoses of NDDH: cutaneous infections, vesiculobullous pyoderma gangrenosum (atypical), bullous erythema multiforme, pustular drug reactions, rheumatoid neutrophilic dermatosis, bowel-associated dermatosis-arthritis syndrome, and erythema elevatum diutinum (1-3). In our case, based on the pathological examination, the differential diagnosis included neutrophilic dermatosis such as Sweet's syndrome or neutrophilic dermatosis of the dorsal hands. It is essential to exclude an infectious etiology that might include a bacterial infection, or less likely a fungal or atypical mycobacterial infection, given the lack of any granulomatous component. However, some atypical mycobacterial infections can demonstrate a brisk neutrophilic infiltrate and relatively sparse granulomatous responses (6). For the same reason (lack of significant histiocytes), we thought that palisaded neutrophilic and granulomatous dermatosis associated with connective tissue disease was less likely. The relationship between this disease entity and a superficial variant of pyoderma gangrenosum remains unclear.

The treatment of NDDH includes systemic corticosteroids, dapsone, methotrexate, potassium iodide, colchicine, and minocycline (2). NDDH is often misdiagnosed as an infectious condition, which can result in inappropriate antibiotic therapy, surgical debridement, and even amputation (7). Therefore, early diagnosis and initiation of appropriate treatment should be mainstay of its treatment.

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