Case Report

Neutrophilic Dermatosis of Dorsal Hands and Legs

Elham Behrangi MD[1], Abbas Rasi MD[2], Bahamin Attar MD[3], Zahra Azizian MD•[4]

Abstract

We present a case of neutrophilic dermatosis of dorsal hands (NDDH), with lesions on the dorsal part of the hands with pustular features and histologic picture of vasculitis.This case highlights the notion that low dose oral steroid can offer a useful first therapeutic option for NDDH. Interstingly, in this case, a few additional lesions were located on the legs, making it different from some previously described cases of NDDH. As with other authors, we believe that this recently described neutrophilic dermatosis is a localized and acral variant of Sweet syndrome. To our knowledge, this is the first case report of NDDH from Iran.

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Introduction

Neutrophilic dermatosis (pustular vasculitis) of dorsal hands is a newly described disorder, the lesions of which may clinically resemble a localized varriant of the Sweet syndrome. Histologic evidence includes dense dermal infiltrate neutrophils. We describe the clinical and histological findings of a patient with neutrophilic dermatosis of dorsal hands.[1]

Case report

A 57-year-old Afghan woman was admitted to the dermatology department of the Hazrat-e-Rasul Hospital, Tehran, Iran, due to tender, deeply erythematous to violaceous plaques on the dorsal part of her hands, of 4 days duration. On physical examination, there were tender violaceous nodules and plaques over the dorsal surface of both hands (Figure 1A). Prominent minute pustules were also visible on the surface of lesions. The lesions rapidly increased in size and number so that 2 days after admision, the entire dorsal part of hands and fingers became edematous and erythematous, and finally large hemorrhagic flaccid bullae or superficial necrotic ulcers developed (Figure 1B). The palmar surface of hands were completely free of lesions (Figure 2). Five days later, a few cutaneous lesions appeared on the legs (Figure 3).

Incisional skin biopsy was taken from the lesions which demonstrated epidermal spongiosis and heavy neutrophilic inflammatory infiltrate in the superficial dermis, causing dermo-epidermal detachment. There was nuclear fragmentation (leucocytoclasia) in vascular walls with no evidence of fibrinoid necrosis (Figures 4A and 4B). The histologic features were similar to the Sweet syndrome.

Laboratory tests on admission showed leucocytosis and neutrophilia with a white blood cell count of 17000/μL, 85% neutrophils,and 12% lymphocytes. Serum electrolytes, rhematoid factor, ANA, and liver and renal function tests were normal. Fecal occult blood was positive on three occasions. Conventional chest X-ray was normal, but spiral chest CT showed two calcified nodules at the periphery of right lung, suspicious for tuberculosis or metastasis. Mamography showed an ovoid density about 10 mm in the left breast. The patient refused further investigations such as CT scan of chest, abdomen, and pelvis. Based on the patient's history, physical examination, and histopathologic and laboratory findings, a diagnosis of NDDH was made.

During her hospital admission, oral prednisolone 60 mg daily was started,and tapered to zero over 4–6 weeks. Five days after the introduction of steroid, no new lesions appeared. The systemic symptoms were rapidly relieved within one or two days, and complete clearing of skin lesions was observed in 6 weeks with no scarring. There was no recurrence during 6 months of follow-up.

Discussion

NDDH is an uncommon disease with less than 100 reported cases in the English literature.[2] The presentation of violaceous papulonodules on the dorsal aspects of the hands was first defined in 1995, by Strutton *et al*. as an acral variant of Sweet syndrome (SS).[3] In 1998, Curco *et al.* described two cases, one without fever and associated with metastatic renal adenocarcinoma.[4,5] The term neutrophilic dermatosis of the dorsal hands (NDDH) was suggested by Galaria *et al.* in 2000, who considered this condition to be a subset of SS. They described three cases with plaques, pustules, and hemorrhagic blisters located on the dorsal aspect of the hands. But these patients did not have any systemic symptoms or fever.[1] Our case, however, had high fever for 10 days before admission to the hospital. Weenig *et al*. in 2004 presented new cases with involvement of the dorsal and palmar surfaces of the hands.[6] Walling *et al.* in 2006 considered this disorder as an atypical and localized form of SS, starting on the hands and possibly spreading to other locations.[7] They proposed the term vasculitis of the dorsal hands. Recently, another author has presented a new case with lesions limitted to the palmar surface of hands.[8]

Associations have been reported with a variety of diseases. A

Authors' affiliations: [1]Assosciate Professor of Dermatology, Rasoul-e-Akram Hospital, Iran University of Medical Sciences, Tehran, Iran. [2]Professor of Dermatology, Rasoul-e Akram Hospital, Iran University of Medical Sciences, Tehran, Iran. [3]Orthopaedic Surgery Resident, Shohada Hospital, Tabriz University of Medical Sciences, Tabriz, Iran. [4]Dermatology Resident, Skin and Stem Cell Research Center, Tehran University of Medical Sciences, Tehran, Iran.
•**Corresponding author and reprints:** Zahra Azizian MD, Dermatology Resident, Skin and Stem Cell Research Center, Tehran University of Medical Sciences, Tehran, Iran. Tel: 9122788611, E-mail:azizian_z@yahoo.com.
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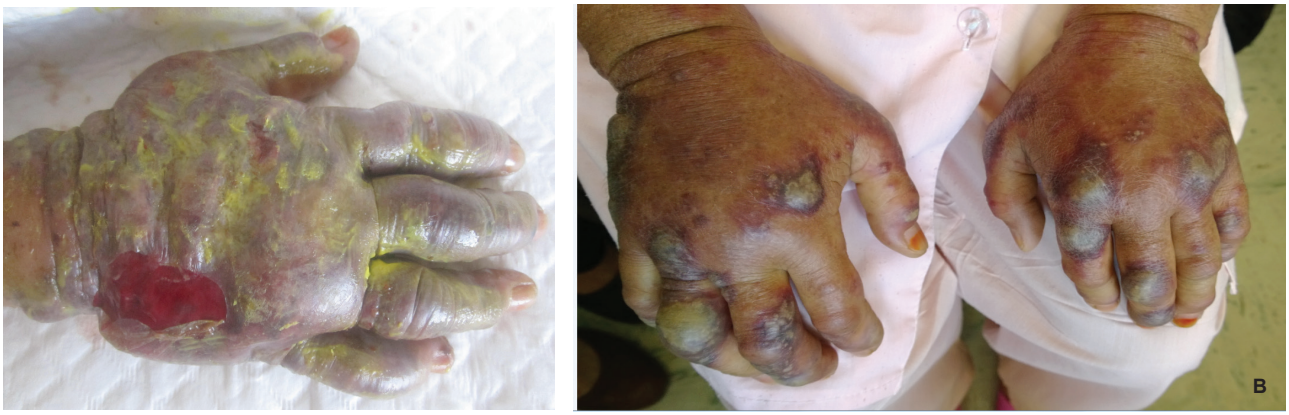

Figure 1 (A and B). Violaceous nodules and plaques over the dorsal surface of both hands.

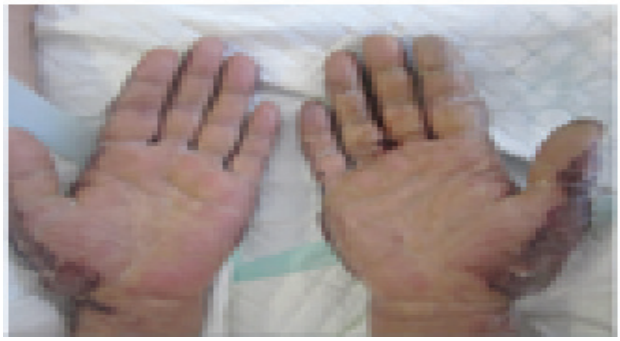

Figure 2. Palmar aspect of hands without lesions.

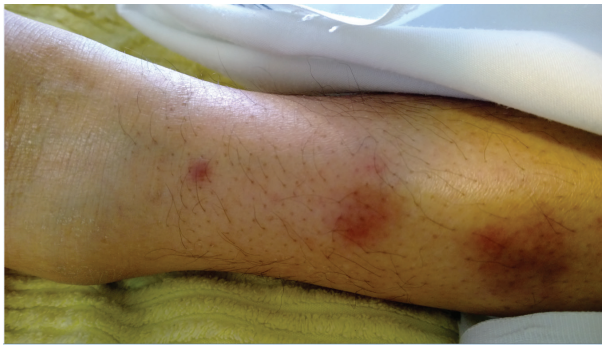

Figure 3. Few lesions on the legs.

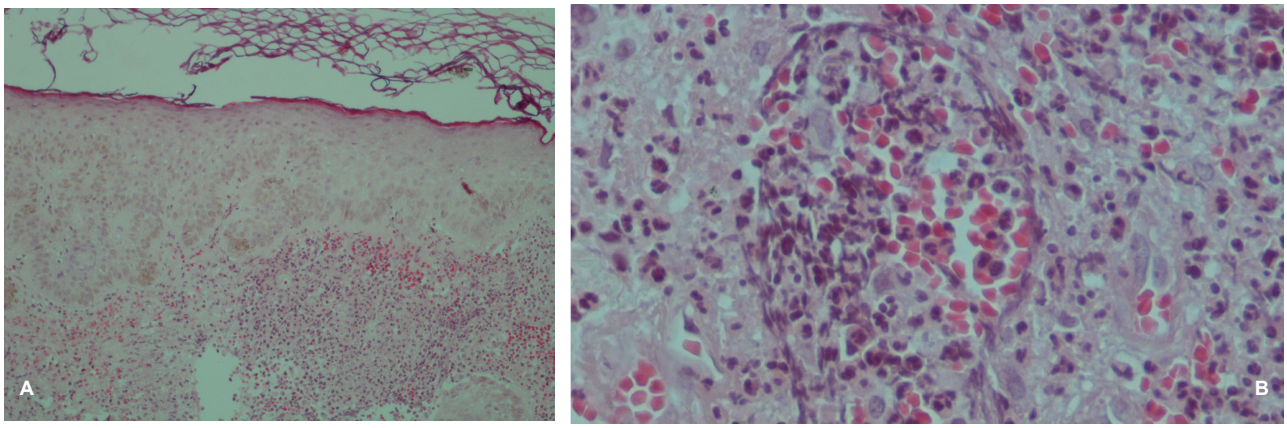

Figure 4. A) Histopathology micrograph of hand lesions skin biopsy reveals neutrophilic inflammatory infiltrate in superficial dermis with perivascular inflammation (Hematoxylin and Eosin; x40) **B)** Nuclear fragmentation (leucocytoclasia).

controlled study of 55 NDDH patients revealed that hematologic diseases (18%) were most commonly associated with NDDH, followed by ulcerative colitis (11%), and solid tumors (9%).[9] Recently an association with HLA B54 (similar to SS) has been reported.[10]

The differential diagnoses of NDDH include SS, fungal, bacterial, and myco-bacterial infections, pustular drug reaction, pyoderma gangrenosum and erythema elevatum diutinum. Although the clinicopathologic features of this condition are similar to those of SS, the lesions are mainly distributed on the dorsal aspects of the hands. SS is usually seen in the third to fifth decades of life. However, the age at onset in our patient was 57 years, which is similar to the mean age reported in previous studies.[6,11]

The clues to diagnosis are: (I) abrupt onset of painful erythematous plaques or nodules, and (II) histologic evidence of a dense neutrophilic infiltrate without evidence of leukocytoclastic vasculitis.[12]

Multiple therapies have been used for NDDH with different rates of success and relapse. The treatment options for NDDH are similar to those for the Sweet´s syndrome. Corticosteroids are commonly used as first line therapy for treating this condition. The majority of cases usually respond well to moderate doses

of systemic corticosteroids. Dapsone has also been used either alone or in combination with systemic corticosteroids.[1,6,7] Other treatment options that have been used as adjuncts to steroids include cyclosporine, methotrexate, supersaturated potassium iodide and salazopyrine. Successful treatment with indomethacin, IVIG colchicine, minocycline, azathiopurine, intralesional triamcinolone, topical steroids, and topical tacrolimus have also been reported.[6,13,14]

The prognosis of NDDH is generally good, as the disease is usually readily treatable. Galaria *et al*. reported the recurrence of hand lesions in their cases as frequent, but this did not happen with our case.[5]

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