SCIENTIFIC ARTICLE

Neutrophilic Dermatosis of the Hands: A Review of 17 Cases

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Purpose Neutrophilic dermatosis of the hands is an inflammatory skin condition related to Sweet syndrome that responds to corticosteroids. It commonly affects the dorsum of the hand and often mimics infection, with violaceous inflammatory papules and plaques that may ulcerate. The aim of this study was to review the clinical presentation of neutrophilic dermatosis of the hands.

Methods A retrospective review was undertaken of all cases of neutrophilic dermatosis of the hands seen at a tertiary hospital in New South Wales, Australia, over a 5-year period.

Results Seventeen cases were identified. The mean time to diagnosis was 9 days after lesion onset. Most cases were older adults (mean age, 71 years). The most common referral diagnoses were infection or a nonhealing wound and 65% of cases reported a history of trauma. The dorsal index finger was the site of involvement in 41% of cases. One case involved the palm. Histopathology reports were available for skin punch biopsy for 14 of 17 cases, which showed dermal neutrophilic infiltrate (93%) and epidermal involvement with necrosis, ulceration, or pustulation (64%). Six cases were treated surgically prior to the correct diagnosis and management being introduced.

Conclusions Neutrophilic dermatosis of the hands was often misdiagnosed as infection. A history of trauma is common and may be misleading. Dermatological consultation and skin punch biopsy are useful in confirming the diagnosis, ideally prior to surgical management. *(J Hand Surg Am. 2017;■(■):1.e1-e5. Copyright © 2017 by the American Society for Surgery of the Hand. All rights reserved.)*

Type of study/level of evidence Diagnostic IV.

Key words Hand, neutrophilic dermatosis, Sweet syndrome.

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Ethics: ■ ■ ■

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Neutrophilic dermatoses are skin disorders characterized by dense dermal or subcutaneous neutrophilic infiltrate. The spectrum includes acute febrile neutrophilic dermatosis (ie, Sweet syndrome), neutrophilic eccrine hidradenitis, erythema elevatum diutinum, Behçet disease, and pyoderma gangrenosum (PG). These conditions may occur in association with recognized predisposing factors including rheumatoid arthritis, inflammatory bowel disease, and hematological malignancy.[1]

Neutrophilic dermatosis of the hands (NDH) represents a dilemma in diagnosis and in classification. It presents with violaceous papules, nodules, and plaques

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FIGURE 1: Clinical presentation of NDH with epidermal necrosis and violaceous plaques distributed over the dorsal hand. The violaceous hue is best appreciated at the periphery of the lesions in this case, which is also a typical feature of PG.

on the hands that may be accompanied by epidermal changes such as necrosis and ulceration (Fig. 1). It typically involves the dorsum of the hands, although palmar cases are reported.[2] As such, NDH shares features with the other neutrophilic dermatoses. Indeed, it has been suggested that these conditions form a spectrum, varying in the intensity and extent of the inflammatory response.[3] Where these conditions occur in the hand, all could be classified as variations of the distinct disease entity of NDH.[4,5]

Histopathologically, NDH typically has a dermal neutrophilic infiltrate. The presence of vasculitis has traditionally been suggested to not be a feature of febrile neutrophilic dermatosis (Sweet syndrome and, therefore, NDH when lesions occur in the hands); however, it may be that histological evidence of vasculitis is secondary to the inflammatory dermatosis and not a primary feature that could help with diagnosis.[4] The differentiation of NDH from typical PG is both clinical and histological. Neutrophilic dermatosis of the hands tends to have blisters and bullae, whereas PG will exhibit pus and early ulceration. Pyoderma gangrenosum is more often associated with inflammatory bowel disease, and NDH is more commonly associated with malignancy. Histologically, there is a greater neutrophilic infiltrate in NDH.

This condition may closely mimic an infective process.[4] Systemic involvement, including fever, neutrophil leukocytosis, and raised inflammatory markers can occur and further implicate an infective cause. This condition does not respond to antibiotics; however, improvement with corticosteroids is usually apparent within a few days, and this can help to confirm the diagnosis.

The aim of this study was to review cases of NDH seen at a single tertiary referral hospital, document the patient journey to diagnosis, and describe the clinicopathological features of this condition.

TABLE 1. Comorbidities of Patients Diagnosed With NDH (n = 17)

Comorbidity	n (%)
Diabetes mellitus	3 (17.6)
Myelodysplastic syndrome	3 (17.6)
Multiple myeloma	3 (17.6)
Inflammatory bowel disease	1 (5.9)
Rheumatoid arthritis	1 (5.9)
Prostate cancer	1 (5.9)
Pseudogout of ipsilateral wrist	1 (5.9)

MATERIALS AND METHODS

A retrospective review was undertaken following approval from the hospital's human research ethics committee. This study complied with the ethical requirements of this institution—Northern Sydney Local Health District Human Research Ethics Committee reference RESP/16/146.

A data search was conducted for presentations of neutrophilic dermatosis to this metropolitan hospital's tertiary hand surgery service over a 5-year period.

Cases were identified via their International Classification of Diseases (ICD) code, with a search undertaken to capture presentations with codes 695.89 or L98.2 (ICD 9 and ICD 10, respectively, for febrile neutrophilic dermatosis [Sweet]), or 696.01 and L88 (ICD 9 and ICD 10, respectively, for PG). Pyoderma gangrenosum was included as a search term to avoid missing cases coded under this related condition. Medical records were then reviewed to ascertain cases in which the most appropriate diagnosis was NDH (based on specialist dermatologist diagnosis and/or histopathology).

Data recorded included patient age, sex, occupation, ethnicity, clinical presentation, initial diagnosis and management, presence of comorbidities, results of investigations, final diagnosis, and treatment. Follow-up information and clinical photographs were included if available.

RESULTS

The ICD coding search identified 50 admissions (37 patients) from 2011 to 2016. Following review of the individual medical records to identify cases involving the hands and to confirm the diagnosis of NDH, 17 cases were included. Of these, 9 were initially coded as febrile neutrophilic dermatosis (Sweet), 7 as PG,

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