

Itchy Papules And Plaques on the Dorsal Hands



A 69-year-old man presented with tender, itchy papules and plaques on the bilateral dorsal hands of 2 months' duration. The plaques had started as small papules that gradually enlarged and then became ulcerated. The patient denied prior trauma or constitutional symptoms. Laboratory testing revealed macrocytic anemia, thrombocytosis, and hypoalbuminemia. A complete blood count and complete metabolic panel were otherwise unremarkable.

A recent bone marrow biopsy for macrocytic anemia performed prior to the current presentation suggested early myelodysplastic syndrome, and endoscopic retrograde cholangiopancreatography revealed a large mass in the common bile duct that was suspicious for malignancy. Two punch biopsies were performed and were negative for fungus and acid-fast bacteria and positive for methicillin-sensitive *Staphylococcus aureus*. Treatment with topical clobetasol 0.05% twice daily was initiated with complete healing of the plaques on the hands after 2 weeks of use; however, the patient continued to develop new ulcerated papulonodules distally.

What's the diagnosis?

- atypical mycobacterial infection
- blastomycosis
- neutrophilic dermatosis of the dorsal hands
- phototoxic drug eruption
- sporotrichosis

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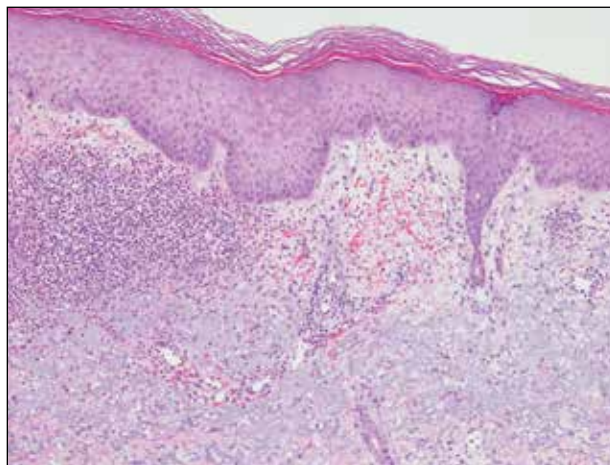
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The Diagnosis: Neutrophilic Dermatitis of the Dorsal Hands

Neutrophilic dermatitis of the dorsal hands (NDDH) is considered to be an uncommon localized variant of Sweet syndrome (SS). The term *pustular vasculitis* originally was used to describe this condition by Strutton et al¹ in 1995 due to the presence of leukocytoclastic vasculitis on histology. In 2000, Galaria et al² suggested this eruption was a localized variant of SS based on clinical presentations that demonstrated associated fever and lack of necrotizing vasculitis and proposed the term *neutrophilic dermatitis of the dorsal hands* to describe the condition. Cases of similar cutaneous eruptions on the hands associated with fever, leukocytosis, elevated erythrocyte sedimentation rate, and leukocytoclasia have since been reported.³⁻⁵ Some authors have concluded that these eruptions, previously termed *atypical pyoderma gangrenosum* and *pustular vasculitis of the hands*, represent a single disease entity and should be designated as NDDH.^{3,4}

Neutrophilic dermatitis of the dorsal hands characteristically presents with hemorrhagic pustular ulcerations limited to or predominantly located on the dorsal hands, as seen in our patient. Histopathologically, NDDH demonstrates a neutrophil-predominant infiltrate of the upper dermis and marked papillary dermal edema; a punch biopsy specimen from our patient was consistent with these features (Figure). Two punch biopsies were performed and were negative for fungus and acid-fast bacteria and positive for methicillin-sensitive *Staphylococcus aureus*. Vasculitis, if present, is more commonly seen in eruptions of longer duration (ie, months to years) and is thought to be secondary to the dense neutrophilic infiltrate and not a primary vasculitis.^{3,6,7} Similar to classic SS, NDDH is inherently responsive to corticosteroid therapy. Successful treatment also has been reported with dapsons, colchicine, sulfapyridine, potassium iodide, intralesional and topical corticosteroids, and topical tacrolimus.²⁻⁸ Oral minocycline has shown variable results.^{3,4}

Numerous case series have demonstrated that a majority of cases of NDDH are associated with hematologic or solid organ malignancies, myelodysplastic syndrome (MDS), inflammatory bowel disease, or other underlying systemic diseases.^{3,5,9} It is important for dermatologists to recognize NDDH, distinguish it from localized infection, and perform the appropriate workup (eg, basic laboratory tests [complete blood



Biopsy from the edge of an ulcerated lesion on the left hand revealed papillary dermal edema with a dense, bandlike neutrophilic infiltrate and hemorrhage (H&E, original magnification $\times 10$).

count, complete metabolic panel], age-appropriate malignancy screening, colonoscopy, bone marrow biopsy) to exclude associated systemic diseases.

Our patient demonstrated characteristic clinical and histopathologic findings of NDDH in association with early MDS and possible common bile duct (CBD) malignancy. The lesions showed a rapid response to topical corticosteroid therapy. The initial differential diagnoses included NDDH or other neutrophilic dermatosis, phototoxic drug eruption, and atypical mycobacterial or fungal infection (cultures were negative in our patient). Physical examination and histopathologic findings along with the patient's clinical course and rapid response to topical corticosteroid therapy supported the diagnosis of NDDH. Our patient's multiple comorbidities, including macrocytic anemia, MDS, and potential CBD malignancy, presented a therapeutic challenge. Oral dapsons, an ideal steroid-sparing agent for neutrophilic dermatoses including NDDH, was avoided given its associated hematologic side effects including hemolysis, methemoglobinemia, and possible agranulocytosis. To date, the patient has not received any further treatment for MDS or the CBD mass and continues regular follow-up with hematology, gastroenterology, and dermatology.

This case highlights the importance of including NDDH in the differential diagnosis of papules and plaques on the hands, especially in patients with known malignancies, and emphasizes the association of neutrophilic dermatoses with malignancy and systemic disease.

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