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(CASE REPORT)

Neutrophilic dermatosis of the dorsum of the hands complicated by panniculitis revealing myelodysplastic syndrome

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Abstract

Neutrophilic dermatosis of the dorsum of the hands (NDDH) is a rare inflammatory condition that belongs to the spectrum of neutrophilic dermatoses and is frequently associated with systemic diseases, particularly hematologic malignancies. We report the case of an 80-year-old woman with NDDH revealing an underlying myelodysplastic syndrome, later complicated by neutrophilic panniculitis of the face. Cutaneous lesions of NDDH can often be misdiagnosed due to their variable and sometimes atypical presentation, leading to delays in appropriate management. The resemblance of NDDH lesions to infectious, inflammatory, or even vasculitic conditions can result in unnecessary treatments, including antibiotics or surgical interventions, without addressing the underlying cause. Given the frequent association between NDDH and hematologic disorders, clinicians should maintain a high level of suspicion and systematically perform appropriate biological investigations. Early recognition and accurate diagnosis are essential to avoid mismanagement and to ensure timely screening for potential systemic diseases, particularly myelodysplastic syndromes and other hematological malignancies.

Keywords: Hand; Neutrophilic dermatosis; Sweet syndrome; Neutrophilic panniculitis; Myelodysplastic syndrome

1. Introduction

Neutrophilic dermatoses encompass a spectrum of conditions characterized histologically by an aseptic neutrophilic infiltrate. Neutrophilic dermatosis of the dorsum of the hands (NDDH) is a poorly understood and often misdiagnosed entity. We report a case of neutrophilic dermatosis revealing an underlying myelodysplastic syndrome, complicated by neutrophilic panniculitis of the face after several weeks.

2. Case Report

We describe the case of an 80-year-old woman with a history of Hashimoto's thyroiditis who presented with post-traumatic bullous lesions (following infusion) on the dorsum of the hands. These lesions evolved over a month into non-healing areas of epidermal detachment, despite regular wound care.

Clinical examination revealed an infiltrated, edematous, and painful plaque on the dorsum of the hands, with a large hemorrhagic ulceration on the right hand (figure 1). Swab cultures were negative and a skin biopsy confirmed the diagnosis of neutrophilic dermatosis. Laboratory tests revealed pancytopenia, and a bone marrow biopsy diagnosed myelodysplastic syndrome.

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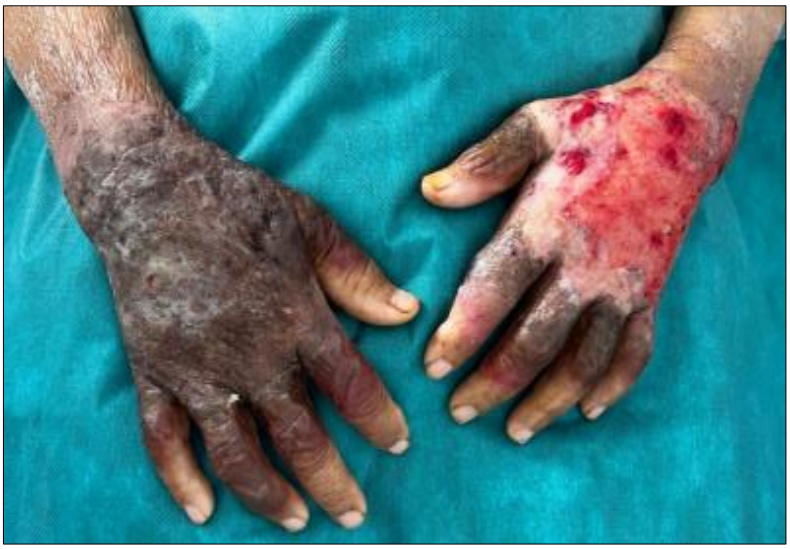

Figure 1 Clinical presentation of a cutaneous lesion on the dorsum of the hands persisting for over a month without healing despite regular wound care

The patient was treated with high-potency topical corticosteroids, resulting in significant improvement. However, two months later, she experienced a relapse of the hand lesions accompanied by erythematous, violaceous, infiltrated, and painful plaques on the face, predominantly on the left side. Infectious workup was negative. Neutrophilic panniculitis was suspected and oral corticosteroid therapy led to clinical resolution within three weeks. The patient was admitted to the hematology-oncology unit for the management of her hematologic disorder.

Figure 2 Facial infiltration emerging two months after the involvement of the dorsum of the hands

3. Discussion

Neutrophilic dermatosis of the hands was first described by Strutton et al in 1995 using the nomen clature “pustular vasculitis of the hands” [1]. Recent debate focuses on categorizing this disorder in the family of neutrophilic dermatoses (ND), as opposed to a primary vasculitis [2].

Neutrophilic dermatosis of the dorsal hands is a rare condition primarily affecting individuals around the age of 60, with a slight female predominance. It is characterized by erythematous, edematous, pustular, or ulcerative papules, nodules, or plaques, bilaterally localized on the dorsal hands. Palmar involvement is more likely to be with erythematous patches and bullae [3]. A pathergy phenomenon is often described following trauma.

Histological examination is essential in suspected cases and typically reveals an infiltrate of normal neutrophils. Epidermal necrosis, ulceration, pustulation and leukocytoclasis can also be observed. In some cases, vasculitis may be found on skin biopsy [3]. During disease progression, other tissues may also exhibit similar infiltrates, as observed in our case.

As with Sweet syndrome, there is a significant association between neutrophilic dermatosis of the dorsal hands and underlying disease such as haematological disorders, the commonest being myelodysplastic syndrome, monoclonal gammopathy, multiple myeloma, acute myeloid leukaemia. There were cases of active solid organ tumours, bowel diseases and autoimmune conditions [4].

Treatment depends on the severity and duration of the disease. Topical corticosteroids, colchicine, and dapsone are effective for mild forms, while systemic corticosteroids remain the first-line treatment for severe or disseminated cases.

Neutrophilic panniculitis is a rare subtype of neutrophilic dermatosis. It is characterized by the presence of a neutrophilic infiltrate in the fat lobules in the absence of either infection or vasculitis. The pathogenesis of NP associated with MDS remains unknown. It may precede or even appear during follow-up of MDS [5].

Abbreviation

- NDDH = Neutrophilic dermatosis of the dorsal hands
- ND = neutrophilic dermatoses
- NP = Neutrophilic panniculitis
- MDS = myelodysplastic syndrome

4. Conclusion

Recognition of localized presentations of neutrophilic dermatoses, although rare, is crucial for early and appropriate management, thereby preventing unnecessary and potentially disfiguring surgeries. Given that hematological disorders are often associated with NDDH, it is important to perform the necessary biological tests for proper screening.

Compliance with ethical standards

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Disclosure of conflict of interest

The authors declare no conflicts of interest related to this work.

Statement of informed consent

Informed consent for publication was obtained from the patient.

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