Archives of Clinical & Experimental Dermatology

Yumed Text
OPEN ACCESS JOURNALS

Case Report | Vol 6 Iss 2
ISSN: 2583-6374
https://dx.doi.org/10.46527/2583-6374.158

Neutrophilic Dermatosis of the Hand Associated to Community-Acquired Pneumonia: A Case Report and Literature Review

Nava-Martínez JF[1*], Varela-Coronado LF[1], Vazquez-Lara SE[1], Román-Barba, MR[1], Hernández-Torres MM[2] and Murillo-Tuercas XI[1]

[1]Dermatology Department, Instituto Dermatológico de Jalisco "Dr. José Barba Rubio", Zapopan, Jalisco, Mexico
[2]Pathology Department, Instituto Dermatológico de Jalisco "Dr. José Barba Rubio", Zapopan, Jalisco, Mexico

[*]**Corresponding author:** Nava-Martínez JF, Dermatology Department, Instituto Dermatológico de Jalisco "Dr. José Barba Rubio", Av. Federalismo nte 3201, Col. Atemajac del Valle, C.P. 45190, Zapopan, Jalisco, Mexico, Tel: 3330304536; E-mail: judith.navamrt@unal.edu.mx

Received: June 20, 2024; **Accepted:** July 01, 2024; **Published:** July 11, 2024

Abstract

Acral neutrophilic dermatosis is a localized variant of Sweet syndrome in which cutaneous manifestations are predominantly restricted to the dorsal area of the hands. It is often misdiagnosed as an infectious process in most cases, and treatment is often delayed. We present the case of a 75-year-old Mexican woman with clinical characteristics of a respiratory infection and the development of acral dermatosis, which represents a diagnostic challenge.

Keywords: ***Neutrophilic dermatosis; Sweet syndrome; Cutaneous reactions; Infectious diseases***

1. Introduction

Neutrophilic dermatoses are a heterogeneous group of inflammatory skin disorders characterized histopathologically by a sterile infiltrate with a predominance of neutrophils [1]. Acral neutrophilic dermatosis (DNA), was first described 20 years ago by Strutton et al., [2] as a rare localized variant of Sweet syndrome (also known as acute febrile neutrophilic dermatosis) [3]. These entities are part of a spectrum of benign dermatoses that share some clinical and histological characteristics, with a predominance in women [4,5].

Citation: Nava-Martínez JF, Varela-Coronado LF, Vazquez-Lara SE, et al. Neutrophilic Dermatosis of the Hand Associated to Community-Acquired Pneumonia: A Case Report and Literature Review. Arc Clin Exp Dermatol. 2024;6(2):158.

DNA is a rare entity with approximately 120 current reports in the medical literature; although the understanding of its etiopathogenesis remains unknown, it has been related to infectious, inflammatory, and neoplastic diseases, predominantly of a hematological nature [6]. DNA, similar to Sweet's syndrome, can present with leukocytosis with neutrophilia, fever, and the presence of infiltrated and painful erythematous papules and plaques [1,5]. A difference that contrasts with Sweet's syndrome is that acral neutrophilic dermatosis is distinguished by presenting with blisters and ulceration. Although acral lesions were initially described on the dorsum of the hand, in 2004 Weenig et al., excluded the term “back” from the name since they observed that many patients also present lesions in the palms and lateral regions of the hands. However, the use of this topographical name remains prevalent in the literature [5,7].

The recognition of DNA is important because an accurate diagnosis should promote the search for underlying diseases, such as neoplasms. Although neutrophilic dermatoses can resolve spontaneously, most cases require treatment. Therapeutic modalities range from systemic corticosteroids to steroid-sparing agents, such as dapsone, colchicine, and tetracyclines, which are almost always curative [6,8].

This article reports the dermatological description of a case of acral neutrophilic dermatosis, possibly triggered by community-acquired pneumonia.

2. Case Presentation

This was a 75-year-old female patient, with no history of known comorbidities. She presented with general malaise, asthenia, adynamia, and anorexia accompanied by a predominantly nocturnal cough, in addition to painful lesions and edema of both hands. Physical examination revealed a fever and right consolidation pulmonary syndrome. Dermatological examination revealed dermatosis that mainly affected the back of both hands, localized and tended toward symmetry, consisting of multiple well-defined erythematous-violaceous plaques, pustules of variable size, edema, pain on palpation, and increased local temperature. (FIG. 1 & 2), respectively.

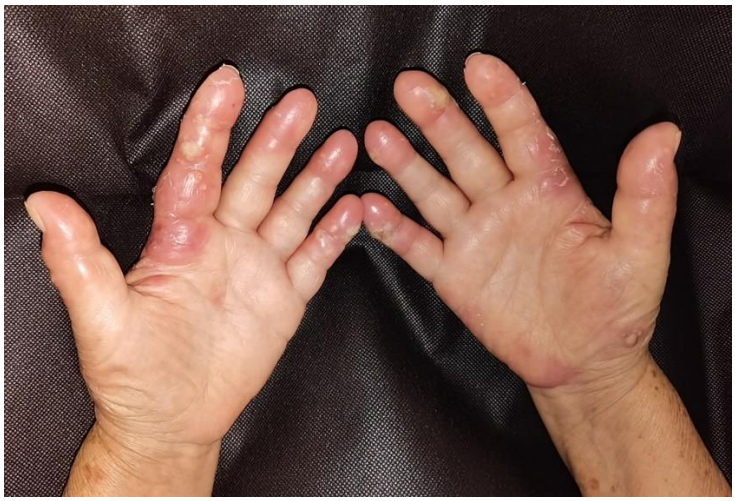

FIG. 1. **Both hands with edema and pustules with an erythematous base.**

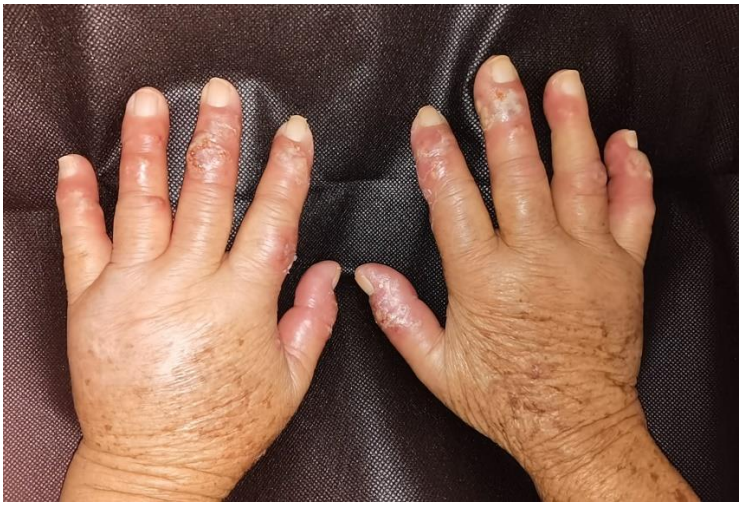

FIG. 2. **Dorsum of the hands with edematous plaques and targetoid erythematous nodules.**

Extension studies were requested to identify anemia, lymphopenia, leukocytes with neutrophilia, elevated acute-phase reactants, and an X-ray that corroborates a consolidation. Histopathological examination of the lesions revealed acanthosis, spongiosis, and neutrophilic infiltrate in the superficial and deep dermis, an image compatible with acral neutrophilic dermatitis. (FIG. 3 & 4)

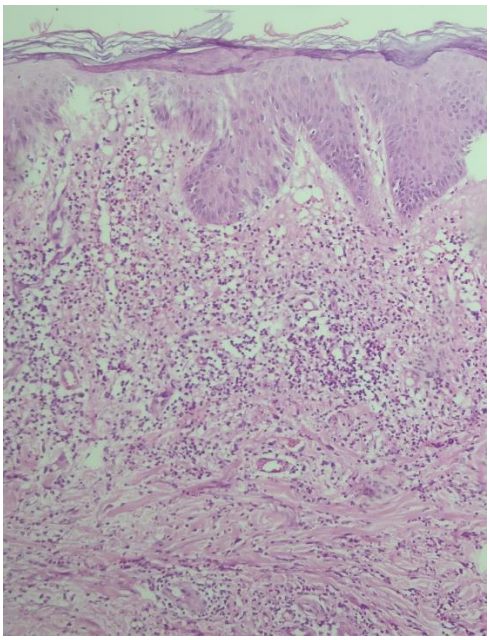

FIG. 3. **Histopathology of a skin biopsy showing dermal edema and a dense neutrophilic infiltrate in the dermis (H&E, ×40).**

Abdominal ultrasonography, tumor markers, peripheral blood smear and culture, coprology, and culture of secretions from hand lesions were requested, all of which were normal.

The patient received treatment with prednisone (0.5 mg), dapsone (100 mg/day), and fluoroquinolone, with improvement in the clinical manifestations 4 weeks after treatment. (FIG. 4). At 12 months, the patient did not experience disease recurrence. Although the causal agent could not be determined, community-acquired pneumonia was assumed to be the probable etiology of acral neutrophilic condition.

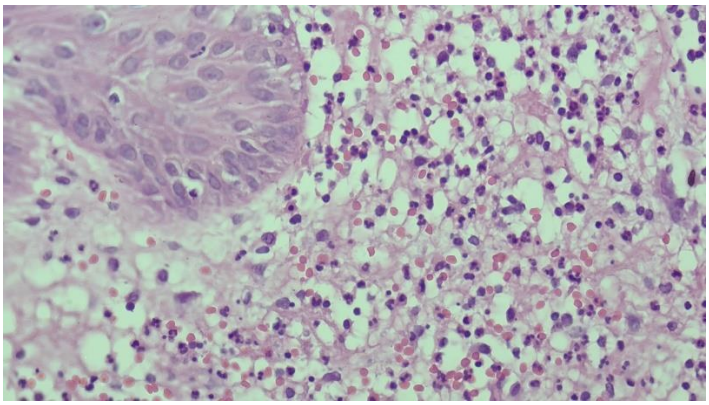

FIG. 4. **Section from the skin showing dense neutrophilic infiltrate, alone with extravasation of red blood cells (H&E, ×100).**

3. Discussion

In this case report, we describe the dermatological findings in a patient with acral neutrophilic dermatosis. Regarding morphology, the main characteristics observed were multiple well-defined erythematous-violaceous plaques and pustules of variable size, concentrated mainly on the backs of both hands. Histologically, acanthosis, spongiosis, and neutrophilic infiltration were observed in the superficial and deep dermis. No findings were found in other topographic sites, such as oral, genital, or ocular involvement, nor were there any histological findings of eccrine gland involvement, consistent with the literature on the subject and with the pathophysiology of acral neutrophilic dermatosis. that does not affect these sites or histological structures, which would suggest a differential diagnosis of other neutrophilic dermatoses such as Behçet's disease or eccrine neutrophilic hidradenitis [1,4,9]. Similarly, no data suggestive of vasculitis were observed, which although it was initially considered as a primary trigger in the physiopathogenesis of certain neutrophilic dermatoses, it is currently considered that any histologically observed vasculitis could actually be a secondary phenomenon (observed in 30% to 60% of cases), and not required for the final diagnosis [10-14].

When comparing what was described with other authors, we found that in the literature, a distribution predominates where the palmar involvement presents with erythematous plaques and blisters, in contrast to the probable presentation of pustules and ulceration in the dorsal area of the hand; however, cases have also been described where this division is not presented in such a clearly delimited manner, as it is in our findings. Some articles also mention a predilection for the radial aspect of the hand, specifically the area between the thumb and index finger, an observation that can be inspected in our case reported by the greater pathological activity present in these regions [11,15,16]. Considering the morphological and topographic variability of this entity, as also described by Brajon et al. and Nofal et al., it was observed that DNA fits within the concept of the continuum that exists between different neutrophilic dermatoses, even within the same entity. Likewise, its designation as acral

neutrophilic dermatosis is more precise and appropriate because it includes acral distribution lesions that are not restricted to the back of the hands, an erroneous classification that could delay the diagnostic process [12,17-19].

Our patient presented with community-acquired pneumonia as a potential trigger. The presence of DNA has been described in association with several other conditions such as chronic glomerulonephritis, diabetes mellitus, sarcoidosis, hepatitis C, Crohn's, urinary tract infections, and streptococcal tonsillitis [13]. This relationship could be explained through the possible pathogenic process where a The inflammatory trigger is coupled with the patient's genetic predisposition and abnormal neutrophil function, manifested as abnormal inflammasome activation, abnormal neutrophilic transformation that predisposes to dermal infiltrate, and overexpression of cytokines and inflammatory markers that possibly trigger an autoinflammation process. Of utmost importance is the relationship between DNA and neoplastic disorders in 27% of the cases according to some reports, which may represent a preneoplastic phenomenon. The most common neoplasms are hematological disorders, such as myelodysplastic syndrome, acute leukemia or lymphoma [10,13].

Sometimes there is no clear infectious trigger, but the DNA process itself is accompanied, in 70% of cases according to some estimates, by systemic involvement, including fever, leukocytosis, and elevated inflammatory markers, which could be confused with a primary infectious process, although it will be distinguished by a lack of response to treatment with antibiotics [11]. In this case, the patient presented an improvement after the use of topical, systemic corticosteroids, and dapsone, which is in accordance with what is described in the literature [12,16]. Other medications reported are the use of colchicine, minocycline, and pentoxifylline, with variable responses [10,16].

4. Conclusion

The intention of this case report, as well as the review of current literature on the subject, is to highlight and highlight a disease that is rarely observed and reported among dermatologists and dermatopathologists, particularly in Mexico. Acute Neutrophilic Dermatoses should be considered in the presence of lesions (plaques, pustules, and ulcerations) that occur on the back of the hands. Timely biopsy must be performed, which is the goal of the diagnostic process. In most cases, the initial clinical diagnosis is an infectious process, which usually leads to the inappropriate use of antibiotics, with little or no clinical response. Patients respond with improvements when using medications, such as corticosteroids. Recognizing this disease should motivate physicians to conduct a thorough investigation to rule out any associated malignancy or systemic disorders.

5. Acknowledgment

The patient authorized the publication of the images, and no further patient information, such as personal data and/or compromising information, was provided.

6. Conflict of Interest

None.

7. Funding Statement

This research received no specific grant from any funding agency in the public, commercial, or not-for-profit sectors.

8. Ethical Compliance

All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki Declaration and its later amendments or comparable ethical standards.

9. Data Access Statement

No new data were generated or analysed during this study.

10. Author Contributions

NMJF and VCLF contributed to the design and implementation of the research, VLSE and MTXI to the writing of the manuscript. RBMR conceived the original and supervised the project.

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