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Sneddon -Wilkinson Disease: A Neutrophilic Autoinflammatory Disorder

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Abstract

Sneddon-Wilkinson disease is a rare, recurrent, benign pustular dermatosis that predominantly affects women between the ages of 40 and 70. It is characterized by hypopyon-like pustules located in proximal areas. The case described here involves a male patient with significant metabolic comorbidities and an atypical lesion distribution, representing a significant deviation from the classic epidemiological profile and constituting a relevant clinical finding.

Keywords: sneddon-wilkinson disease; patient; neutrophilic autoinflammatory disorder

Introduction

This is a 62-year-old male patient from the state of Carabobo - Venezuela, with a history of type 2 diabetes mellitus. He reports the onset of his current illness in 2024, characterized by pustules on the back of his hands, trunk, and lower limbs. He consulted multiple doctors who prescribed antibiotic therapy without improvement, and due to the persistence of the symptoms, he decided to visit the dermatology service in March of this year. On physical examination, the patient presented with skin phototype III and a generalized, bilateral, and symmetrical dermatosis, predominantly affecting the upper extremities. The lesions were characterized by a hypopyon-like pustule with an erythematous base, varying in size and distribution, and mild pruritus. The following presumptive diagnoses were considered: Sneddon -Wilkinson disease. Dermatoscopy revealed a superficial pustule and surrounding erythema with fine scaling at the periphery. Laboratory tests showed neutrophilia and hyperglycemia. A Gram stain was performed , revealing abundant neutrophils and negative bacteria. The patient was evaluated by the Endocrinology service, which also diagnosed grade II obesity. In a histological slide of skin stained with hematoxylin and eosin (H&E) and magnified 10X, subcorneal accumulation of neutrophils is observed in the absence of spongiosis and acantholysis. A superficial perivascular lymphohistiocytic inflammatory infiltrate is evident in the dermis. Based on the clinical and histopathological findings, a definitive diagnosis of Sneddon -Wilkinson disease is established. Systemic treatment with dapsone 100 mg/ day and prednisone 1 mg/kg/ day is initiated, and topical treatment with high-potency steroids is also prescribed. With favorable evolution at 6 weeks.

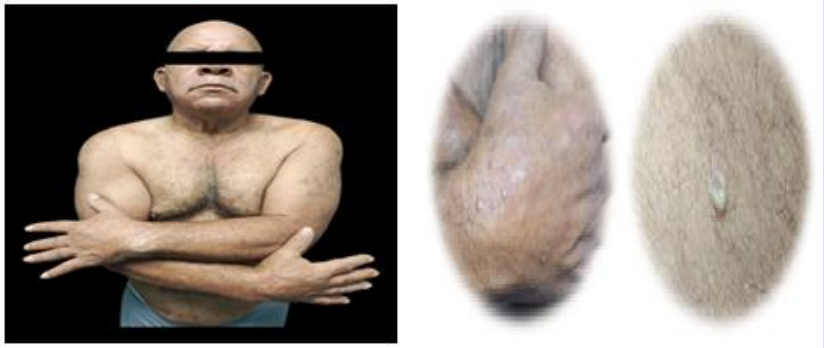

Figure 1: Hypopyon pustules on the back of the hands

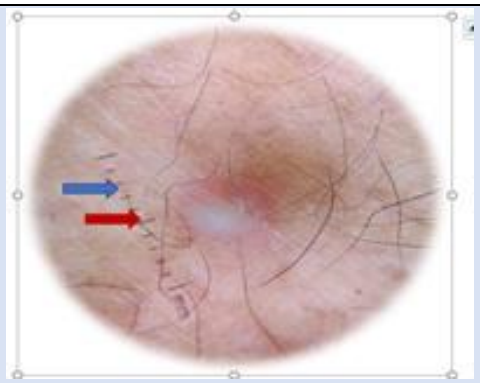

Figure 2: Dermoscopy shows a pustule with an erythematous base

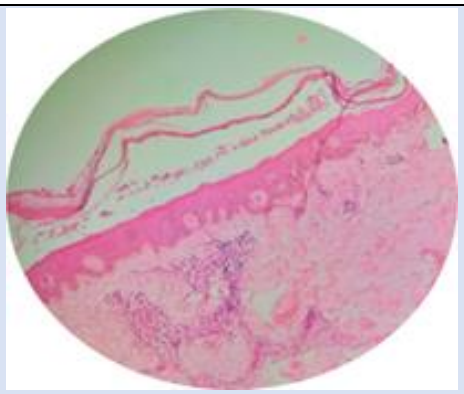

Figure 3: Histological slide of skin stained with H&E and magnified at 10X: Subcorneal accumulation of neutrophils is observed in the absence of spongiosis and acantholysis. In the dermis, a superficial perivascular lymphohistiocytic inflammatory infiltrate is evidente.

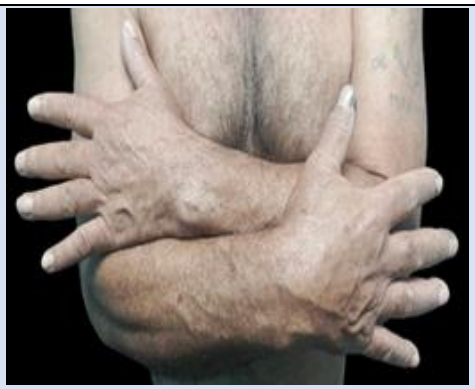

Figure 4: Remission of lesions 6 weeks after starting treatment with dapsone.

Discussion

Sneddon -Wilkinson disease is a rare, recurrent, benign pustular dermatosis that predominantly affects women between the ages of 40 and 70. It is characterized by hypopyon-like pustules located in proximal areas. The case described here involves a male patient with significant metabolic comorbidities and an atypical lesion distribution, representing a significant deviation from the classic epidemiological profile and constituting a relevant clinical finding. From a pathophysiological standpoint, this atypical form is related to its autoinflammatory neutrophilic nature, mediated by IL-8 and tumor necrosis factor-alpha, which is exacerbated in metabolic comorbidities such as diabetes mellitus. The absence of autoantibodies and positive cultures guides treatment toward neutrophil modulation. Dapsone is one such treatment. The therapy of choice , due to its direct action on neutrophils and its rapid clinical response, however, in refractory cases, therapeutic alternatives such as biological agents have been explored, thus expanding the therapeutic spectrum.

Conclusion

The identification of pustules in hypopyon constitutes a distinctive morphological marker that guides the diagnosis towards Sneddon -Wilkinson

disease, allowing its differentiation from other neutrophilic pustuloses. Dapsone remains the conventional treatment of choice due to its effectiveness in inhibiting neutrophilic chemotaxis and its established therapeutic profile.

Summary

Sneddon -Wilkinson disease is a rare, recurrent, benign pustular dermatosis that predominantly affects women between the ages of 40 and 70. It is characterized by hypopyon-like pustules located in proximal areas. The case described here involves a male patient with significant metabolic comorbidities and an atypical lesion distribution, representing a significant deviation from the classic epidemiological profile and constituting a relevant clinical finding.

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