CASE REPORT

Sneddon-Wilkinson Syndrome: Case Report

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SUMMARY

Sneddon-Wilkinson Syndrome (SWS) is a little-known chronic neutrophilic dermatosis that predominantly affects adult women and is characterized by recurrent subcorneal vesicles and pustules. Although its etiology is unknown, it has been associated with immune system diseases and is believed to have an autoimmune component. Diagnosis is based on clinical presentation and histopathological findings. Managing the disease can be challenging due to its recurrent nature, focusing on symptom control and prevention of exacerbations. The cornerstone of treatment is oral dapsone[1]

INTRODUCTION

Sneddon-Wilkinson Syndrome (SWS), also known as subcorneal pustulosis, is a chronic and recurrent neutrophilic dermatosis of unknown origin, believed to have an autoimmune component.[3] Initially, this disease was considered a rare variant of dermatitis herpetiformis or pustular psoriasis; however, its clinical and histological features are sufficiently consistent to justify its separation from other entities.[2] Most cases have been reported in adult women, typically over 40 years of age. There is no available estimate of global incidence or prevalence, nor any known ethnic or geographic predilection.[1]

SWS is characterized by the appearance of sterile, recurrent subcorneal vesicles and pustules with subsequent superficial desquamation. These lesions usually begin in skin folds and later spread to the trunk and proximal extremities, often presenting in annular or polycyclic patterns.[3,4] Treatments include dapsone, oral corticosteroids, retinoids, colchicine, methotrexate, phototherapy, and more recently, anti-TNF alpha.[3]

CLINICAL CASE

A 21-year-old female patient, with no significant medical history, presented with a one-year history of generalized vesicles and pustules, sparing the palms, soles, and mucous membranes, accompanied by mild pruritus that left residual hyperpigmentation. She had used both topical and systemic corticosteroids without improvement.

Physical examination revealed generalized dermatosis, characterized by symmetrically distributed, hyperpigmented plaques with a polymorphic configuration, well-defined borders, some scaling, and sparse pustules (Figure 1). Dermoscopy showed superficial de-epithelialization secondary to pustules on an erythematous base and residual pigmentation (Figure 2).

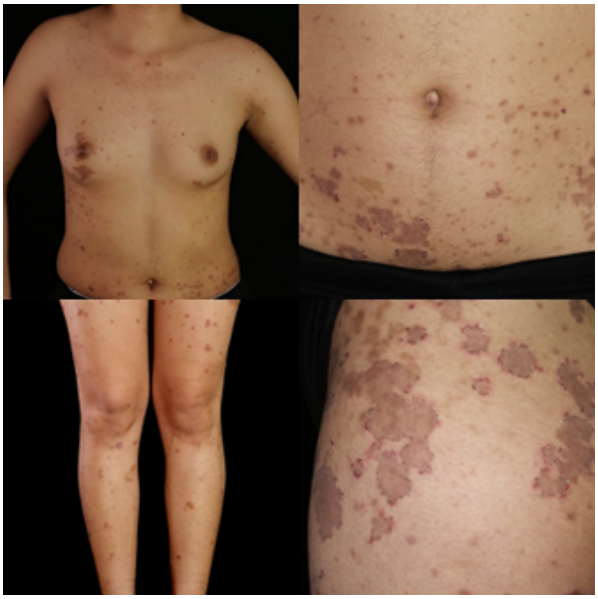

Figure 1. A-B: Distribution of lesions on the chest and abdomen. C-D: Distribution of lesions on the lower limbs. Hyperpigmented plaques with an arcuate, symmetrical pattern, well-defined, and with scaling edges.

A biopsy was performed, with histological findings of a subcorneal pustule containing serous fluid, an inflammatory infiltrate predominantly of neutrophils, and mild spongiosis (Figure 3).

DISCUSSION

In the described case, we observe a young woman with no personal medical history, presenting with dermatosis characterized by recurrent vesicles and plaques spread over the entire body, including folds and flexures, leaving residual hyperpigmentation. Initially, dermatitis herpetiformis was considered; however, the absence of intense pruritus, the lack of blisters, and non-compatible histology ruled this out. Key findings supporting the diagnosis of SWS include: characteristic skin eruption, absence of drug exposure, no history of psoriasis, lack of nail or joint changes,[1] and no general health impairment. These features, combined with histology, were fundamental in reaching the diagnosis. Several diseases have been associated with this disorder, such as multiple myeloma, IgA monoclonal gammopathy, pyoderma gangrenosum, rheumatoid arthritis, hypothyroidism, hyperthyroidism, systemic lupus erythematosus (SLE), multiple sclerosis, and cryoglobulinemia.[6] These were ruled out through patient history, physical examination, and laboratory tests showing normal parameters.

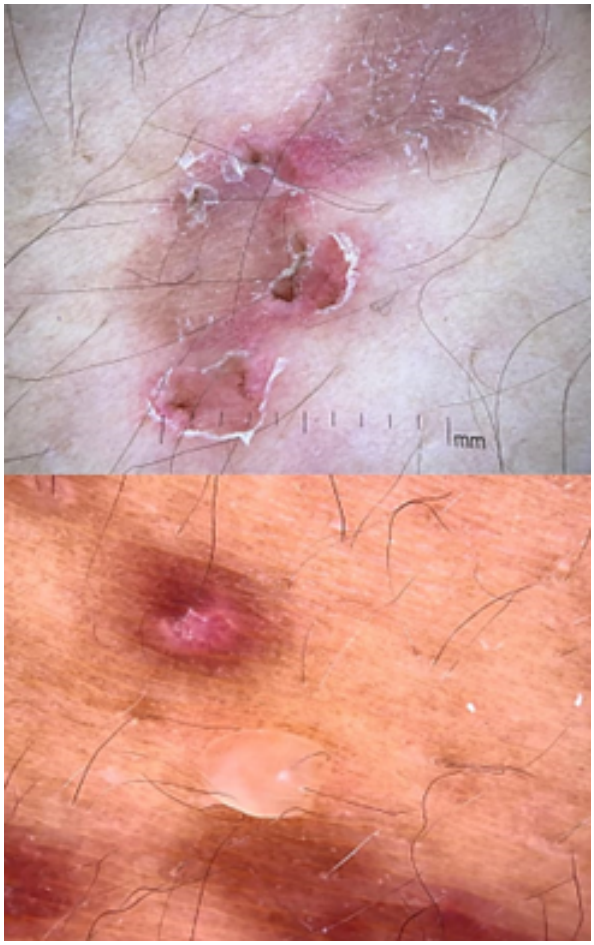

Figure 2. Dermoscopy: A. Superficial scaling on an erythematous base. B. Pustule (Arrow) and residual pigmented lesions.

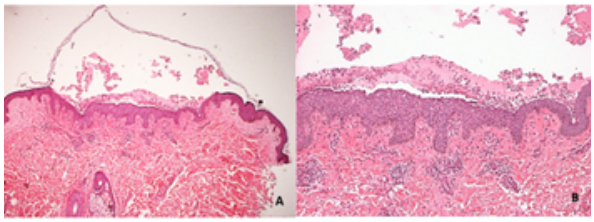

Figure 3. A. Subcorneal pustule with serous content. B. Inflammatory infiltrate, predominantly neutrophils, with mild spongiosis.

In the limited literature available on SWS, some case reports present findings that align with the clinical and histopathological features of SWS: an erythematous-pustular rash on the proximal region of the upper limbs, later spreading to the chest and abdomen, with a recurrent course evolving into hyperchromic macules.

Histopathologically, it is described as a subcorneal spongiform pustule with abundant neutrophils, few eosinophils, and papillary dermal edema[5,8] A case has been reported with acute vesicles and pustules diagnosed as SWS eight days after COVID-19 vaccination.[9] The findings in these case reports correspond with the physical examination of our patient, as well as the histology.

The patient was treated with dapsone at a dose of 50 mg daily, with prior glucose-6-phosphate dehydrogenase testing yielding normal results, showing good response and confirming our diagnosis. A case of SWS during pregnancy has also been reported, treated with dapsone without complications for either the patient or the fetus;[10] we consider this information important, given that adult women are the primary demographic for this condition.

CONCLUSION

Sneddon-Wilkinson Syndrome is a chronic, recurrent neutrophilic dermatosis that can significantly impact patients' quality of life. Although advances have been made in understanding the disease, its exact etiology remains unknown, and management can be challenging.

CONFLICT OF INTEREST

None declared by the authors.

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