

Systemic lupus mimicking Stevens-Johnson syndrome: About one case

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ABSTRACT

The Stevens–Johnson Syndrome (SJS) is a severe cutaneous adverse reaction with high morbidity and mortality, often drug-induced. In rare cases, other factors such as infections and vaccinations have been implicated. This report describes the case of a young adolescent who presented with cutaneous involvement characterized by target-like lesions and erosions covered with hemorrhagic crusts, predominantly acral in distribution. There was erosive involvement of the oral and nasal mucosa, as well as hemorrhagic cheilitis, progressing over one month before the consultation. No medication was found. The evolution was marked by the appearance of a lupus mask on the face. Immunological assessment revealed positive homogeneous antinuclear antibodies and anti-native DNA antibodies associated with hypocomplementemia and bicytopenia, leading to the diagnosis of systemic lupus. This case highlights the importance of considering SLE in the differential diagnosis of severe dermatopathies, especially in atypical clinical contexts.

Key words: Stevens–johnson Syndrome, Systemic Lupus, Chronic Progression, Photodistribution

INTRODUCTION

Systemic lupus erythematosus (SLE) is a chronic autoimmune disease affecting multiple organs and is characterized by the aberrant production of autoantibodies. Among its numerous clinical manifestations, dermatological involvement is common and varied. However, atypical presentations, such as skin lesions mimicking Stevens–Johnson syndrome (SJS), are rarely reported. SJS is a severe mucocutaneous distress syndrome, often triggered by medications or infections, and is associated with a high mortality rate. The clinical resemblance between SLE and SJS may complicate the diagnosis, leading to therapeutic challenges. This article examines a case of systemic lupus mimicking SJS, highlighting the underlying mechanisms, diagnostic approach, and therapeutic implications.

CASE REPORT

Herein, we report the case of an eighteen-year-old patient with no prior medical history, who presented

with painful acral skin lesions and erosive involvement of the oral and nasal mucosae, evolving for one month. Upon admission, clinical examination revealed a cachectic patient in poor general condition, febrile at 40°C. Cutaneous findings included target lesions on the extremities, erosive pulpitis, diffuse paronychia, a hemorrhagic crusted plaque on the nose, and bilateral auricular impetiginization. Mucosal examination showed hemorrhagic cheilitis and erosions of the oral and nasal mucosae (Figs. 1a – 1c).

Initial diagnoses of major erythema multiforme and Stevens–Johnson syndrome were considered; however, no history of drug intake was identified. The patient reported no systemic symptoms such as arthralgia, photosensitivity, sicca syndrome, or Raynaud’s phenomenon. Three days post-admission, the patient developed an erythematous, butterfly-shaped plaque with hemorrhagic crusts on the face (Fig. 2). A skin biopsy was taken but was nonspecific. Immunological workup revealed positive antinuclear antibodies (ANA) with a homogeneous pattern and positive anti-DNA

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Figure 1: (a-c) Clinical images showing oral and nasal erosions, target lesions, and diffuse erosive pulpitis.



Figure 2: Erythematous butterfly-shaped plaque topped with hemorrhagic crusts.

antibodies. Systemic evaluation showed bicytopenia, altered liver function tests, and hypocomplementemia. The diagnosis of lupus erythematosus mimicking Stevens–Johnson syndrome was retained. The patient was prescribed synthetic antimalarials and oral corticosteroids, leading to significant improvement. After eighteen months of follow-up, the patient remained in good health.

DISCUSSION

Stevens–Johnson syndrome (SJS) is a rare yet serious condition characterized by severe cutaneous and mucosal involvement. While historically associated with drug reactions, it may also occur in response to infections or idiopathically. Systemic lupus

erythematosus (SLE), on the other hand, is a multisystem autoimmune disease with varied clinical manifestations, including dermatological ones. SLE may mimic SJS due to the presence of severe skin lesions and mucosal involvement. This clinical similarity results from generalized inflammation and epidermal apoptosis, which are also characteristic of SJS. However, there are fundamental differences in the pathogenesis [1,2]:

- In SLE, skin lesions are mediated by immune complexes, antibody deposits, and complement activation, leading to exaggerated keratinocyte apoptosis.
- In SJS, lesions are primarily related to a type-IV hypersensitivity reaction, triggered by drugs or infections.

There are diagnostic criteria that help to distinguish the two entities. SLE is clinically characterized by an insidious onset and a prolonged evolution, significantly longer than drug-induced SJS, which progresses more rapidly, often with signs of epidermal detachment within hours to days [1]. While our patient presented with buccal ulcers and hemorrhagic cheilitis, there was no genital or perineal involvement. This absence has also been noted in previous studies in this form of SLE [2,3]. Moreover, the initially affected and most severely impacted areas in our patient were the photodistributed regions, suggesting UV radiation as a triggering or aggravating factor [4].

In the literature, there are cases of SJS where no drug was clearly responsible for the symptoms [5,6]. In such idiopathic cases, ANA screening is recommended

to exclude SLE, as it could represent the initial manifestation, as seen in our patient.

The presence of target lesions and positive ANA could suggest Rowell syndrome. However, the homogeneous ANA pattern, negative anti-SSA and anti-SSB antibodies, and normal rheumatoid factor failed to meet the necessary criteria for this diagnosis [7].

The therapeutic management of these two conditions differs significantly, highlighting the necessity of an accurate diagnosis. In SJS, the immediate discontinuation of the suspected drug is crucial, followed by symptomatic management, including hydration, the prevention of secondary infections, and the treatment of ulcers. In severe cases, immunomodulators such as intravenous immunoglobulins (IVIg) or TNF- α inhibitors may be considered [1]. Conversely, SLE often requires systemic corticosteroids to control acute flares, combined with synthetic antimalarials such as hydroxychloroquine for long-term management. In severe forms, immunosuppressants such as mycophenolate mofetil or azathioprine are indicated [4,8].

Thus, in cases where SLE mimics SJS, a delayed diagnosis may lead to rapid disease progression with multiorgan involvement. A multidisciplinary approach involving dermatologists, internists, and immunologists is often required.

CONCLUSION

Systemic lupus erythematosus may exceptionally mimic Stevens–Johnson syndrome, complicating diagnosis and management. Early recognition and appropriate treatment are crucial to improving prognosis. This case highlights the importance of considering SLE in the differential diagnosis of severe dermatopathies, especially in atypical clinical contexts.

Consent

The examination of the patient was conducted according to the principles of the Declaration of Helsinki.

The authors certify that they have obtained all appropriate patient consent forms, in which the patients gave their consent for images and other clinical information to be included in the journal. The patients understand that their names and initials will not be published and due effort will be made to conceal their identity, but that anonymity cannot be guaranteed.

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