Stevens-Johnson Syndrome Due to Prednisone in a Patient With Systemic Lupus Erythematosus

Andrés Tirado-Sánchez
Dermatology Department, Hospital General de México, OD, Cuauhtémoc, México.

Stevens-Johnson syndrome is a systemic, mucocutaneous disease, frequently related to drugs. We present the case of a 35-year-old patient with a diagnosis of systemic lupus erythematosus treated with prednisone who developed a Stevens-Johnson syndrome and was treated with methylprednisolone.

Key words: Stevens-Johnson syndrome. Systemic lupus erythematosus. Prednisone.

Introduction
Stevens-Johnson syndrome (SJS) is a mucocutaneous vesicular and ampoule forming generalized disease of unknown etiology in 50% of cases.¹

It is characterized by a hypersensitivity reaction due to viruses, bacterial infection, drugs, vaccines, hormonal changes of menses, Reiter’s syndrome, and sarcoidosis, among other causes.² Three of the medications most frequently used in SJS are oral steroids, though it has been found that they can also unleash the disease.³ Intravenous (i.v.) immunoglobulin, methylprednisolone, and cyclophosphamide have also proven successful in treating the disease.⁴,⁵ The case of a SJS secondary to the use of prednisone in a patient with systemic lupus erythematosus (SLE), that remitted after treatment with pulse methylprednisolone and a suspension of the suspected causal drug is presented.

Case Presentation
A 35-year-old male patient sought medical attention due to malaise, joint pain, and a 2 week old fever. When interrogated the patient referred joint pain in the fingers and hand joints, reported having photosensitivity, even when exposed briefly to direct sunlight and a higher than 39°C fever, without a specific time of onset in various occasions. No previous consumption of medication or addictive substances was recorded, nor were infections or drug hypersensitivity; the patient had not received prior medical treatment and had no previous illness. The findings on physical exploration were facial erythema, asymptomatic superficial oral ulcers that presented in episodes with intervals in between of 1 to 2 weeks duration, as well as symmetrical arthritis in distal interphalangeal joints. The decision to admit the patient to hospitalization was made. In the initial laboratory workup an erythrocyte sedimentation rate of 56 mm/h (normal up to 20 mm/h) was found; antinuclear antibodies >1:640 (normal <1:40); anti-DNA antibodies >200 UI/mL (normal <35 UI/mL). To study the origin of the fever urine cultures, pharyngeal, and serial blood cultures were done when the patient had a fever spike, but no pathogen growth was observed; serology for herpesvirus 1 and 2 was negative, as was the ELISA test for the human immunodeficiency virus (HIV). A diagnosis of SLE was made. The patient was
initiated on oral prednisone 1 mg/kg daily. At 7 days of 
treatment he presented fever and a skin eruption that 
affecting the face, scalp, trunk, oral mucosa and 
conjunctiva, and that presented erythema, flacid 
ampoules, and superficial ulcers. The Tzank cytodiagnosis 
did not report any findings. A diagnosis of SJS was 
considered. Skin biopsy showed changes in the 
dermoepidermic interphase and abundant necrotic 
k eratinocytes, compatible with SJS (Figure 1). Direct 
immunofluorescence was negative. Prednisone was 
suspended, and pulse methylprednisolone (1 g i.v. daily 
for 3 days) and hydroelectrolytic reposition, with 
improvement of the mucocutaneous lesions at 3 weeks 
(Figure 2). Epycutaneous tests were carried out (TRUE 
test) with microdose prednisone with a response at 72 
hours of ++ (strong positive reaction), being interpreted 
as hyperreactive to the drug, with a definite diagnosis 
of SJS secondary to prednisone. The patient was 
discharged and treatment with hydroxicloroquine 400 
mg/d was started.

Discussion

The use of steroids as initial treatment of SJS is 
controversial, even though its usefulness has been 
demonstrated for the stabilization of hypersensitivity in 
SJS, and can increase survival when used early in the 
course of the disease. Nonetheless, even though it 
favorably modifies the course of the disease, in some cases 
it can unleash it. There are cases, as the one presented, 
where the use of oral steroids is contraindicated, and the 
use of pulse methylprednisolone is recommended, as 
established by Martinez and Atherton, who even 
consider that the treatment with i.v. pulse steroid 
modifies the prognosis of SJS. Some studies mention 
that corticosteroids does not reduce mortality in SJS or 
even the days of hospital stay, but they are few and 
inconclusive. It has been found that immunologic disease 
such as SLE, can predispose to drug hypersensitivity, 
because they can act as SJS presentation cofactors. The 
Tzank cytodiagnosis is a very important diagnostic 
resource in an ampoulous disease, due to the fact that it 
can distinguish an illness related with pharmacologic 
hypersensitivity like SJS, from other illnesses that 
present intraepidermic ampoules such as the pemphigo 
group, particularly Senear Usher syndrome (the 
association of pemphigus erythematosus and SLE), or 
with viral diseases (herpesvirus).

Conclusions

Prednisone can unleash a hypersensitivity phenomenon 
that can endanger the life of a patient. Some autoimmune 
diseases predispose to the development of these
phenomena with a greater frequency, such as the case of SLE. Even though methylprednisolone can be a useful treatment in these cases, more studies to prove it are needed.

References