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

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

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 affected the face, scalp, trunk, oral mucosa and conjunctiva, and that presented erythema, flacid ampoules, and superficial ulcers. The Tzanck cytodiagnosis did not report any findings. A diagnosis of SJS was considered. Skin biopsy showed changes in the dermoepidermic interphase and abundant necrotic keratinocytes, 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 hydroxychloroquine 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 Martínez 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 Tzanck 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 pemphigus 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