

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

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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.

Síndrome de Stevens-Johnson secundario a prednisona en un paciente con lupus eritematoso sistémico

El síndrome de Stevens-Johnson es una enfermedad mucocutánea sistémica, asociada frecuentemente a medicamentos. Se presenta el caso de un paciente de 35 años de edad con diagnóstico de lupus eritematoso sistémico en tratamiento con prednisona, que desarrolló un síndrome de Stevens-Johnson que se trató con pulsos de metilprednisolona.

Palabras clave: Síndrome de Stevens-Johnson. Lupus eritematoso sistémico. Prednisona.

Introduction

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

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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.^{2,3} One of the medications most frequently used in SJS are oral steroids, though it has been found that they can also unleash the disease.⁴ Intravenous (i.v.) immunoglobulin, methylprednisolone, and cyclophosphamide have also proven successful in treating the disease.^{1,5}

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 where 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); antiDNA 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 Tzank cytodagnosis 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 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.^{6,7} Nonetheless, even though it favorably modifies the course of the disease, in some cases it can unleash it.⁴ 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,⁸ 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,^{9,10} 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.¹¹ The Tzank cytodagnosis is a very important diagnostic resource in an ampoules disease,¹² 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).¹³

Conclusions

Prednisone can unleash a hypersensitivity phenomenon that can endanger the life of a patient. Some autoimmune diseases predispose to the development of these

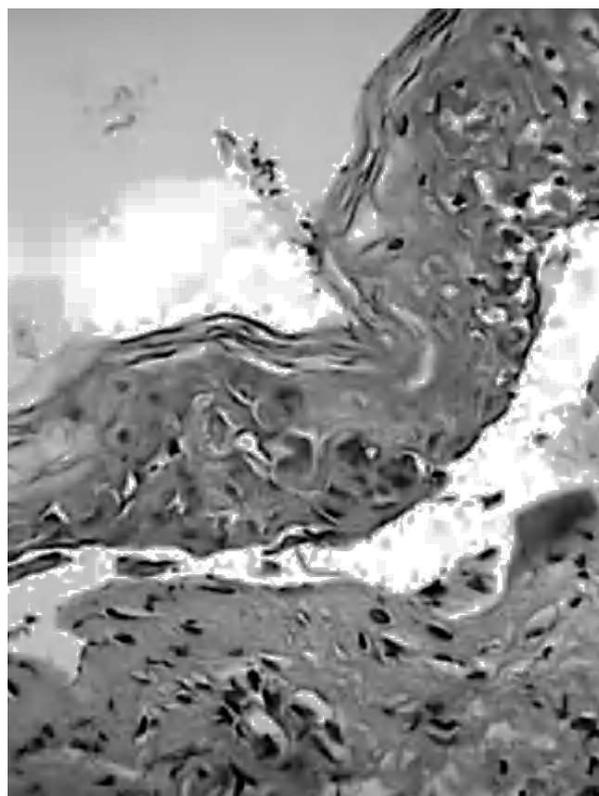


Figure 1. Histologic picture with changes in the interphase and abundant necrotic keratinocytes.



Figure 2. Patient aspect. A) Three days after initiating prednisone. B) After 21 days of treatment.

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.

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