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A rapidly progressive rash with mucosal involvement

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Key words: skin necrosis, exfoliation, skin pain.

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A 40-year-old man, recently diagnosed with systemic lupus erythematosus, presented to the emergency department with a one-week history of high fever, headache, cough, and keratoconjunctivitis. Two weeks earlier, he had started treatment with hydroxychloroquine 200 mg twice daily (400 mg/day orally). After the onset of fever, he self-administered paracetamol 1 g three times daily. Within 48 hours, a rapid worsening of pre-existing cutaneous lesions occurred. Macular lesions rapidly coalesced into large flaccid blisters involving the face, chest, palms, and soles. The lesions exfoliated within three days. The patient reported severe pain affecting the skin and the mucous membranes of the mouth, genitals, and eyes. A positive Nikolsky sign was observed. On admission, he was febrile (38.5°C), with blood pressure 100/70 mmHg, heart rate 100 bpm, and respiratory rate 25 breaths per minute. Cutaneous detachment with areas of necrosis involved approximately 8% of the body surface area, predominantly involving the extremities, particularly the hands. Within a few days, all the lesions became necrotic. Laboratory tests showed leukocytosis and elevated C-reactive protein, while procalcitonin was negative. Pain severity was rated 9/10 on a visual analogue scale, requiring intravenous tramadol 100 mg every 8 hours for analgesia.

Question

Based on the clinical history and presentation, what is the most likely diagnosis?

- A. Toxic shock syndrome (TSS)
- B. Toxic epidermal necrolysis (TEN)
- C. Erythroderma in systemic lupus erythematosus
- D. Stevens-Johnson syndrome

Answer

The clinical presentation was consistent with Stevens–Johnson Syndrome (SJS), confirmed by skin biopsy showing full-thickness epidermal necrosis with keratinocyte apoptosis.¹

The differential diagnosis mainly includes Toxic Epidermal Necrolysis (TEN), erythroderma, and Toxic Shock Syndrome (TSS). TEN was excluded because epidermal detachment involved less than 10% of the body surface area, whereas TEN typically involves more than 30%.^{2–4} Erythroderma is characterized by diffuse erythema and scaling involving $\geq 90\%$ of the body surface area, usually associated with pruritus rather than severe pain and typically without significant mucosal involvement.^{5–7} TSS typically presents with fever and a diffuse erythroderma-like rash associated with systemic manifestations and late desquamation occurring after 1–2 weeks.⁸ These features were not observed in our patient.

Key clinical features supporting the diagnosis of SJS include acute onset, recent exposure to potential triggering drugs, involvement of multiple mucosal sites, predominant skin pain rather than pruritus, and limited epidermal detachment.

The patient was admitted to a sub-intensive care unit. Hydroxychloroquine and paracetamol were immediately discontinued. Among the drugs taken by the patient, paracetamol has been reported as a possible trigger of SJS, although hydroxychloroquine has also been rarely associated with severe cutaneous adverse reactions.

Intravenous methylprednisolone (1 mg/kg/day) was initiated, together with supportive management including fluid therapy, wound care, and ophthalmologic monitoring. Early systemic corticosteroid therapy has been reported to reduce disease progression in selected cases of SJS.

Clinical improvement was observed after five days, with progressive re-epithelialization of the skin lesions. The patient was discharged after 14 days in stable condition. No secondary infections or systemic complications occurred during hospitalization.

Early recognition and prompt discontinuation of the suspected triggering drug are essential for improving outcomes in patients with Stevens–Johnson syndrome.

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Received: 10 February 2026; Accepted: 21 April 2026; Early view: 2 July 2026

Contributions: all the authors equally contributed to the work and approved the final version.

Conflicts of interest: the authors declare no conflict of interest.

Availability of data and materials: the datasets used during the current case report are available from the corresponding author on reasonable request.

Ethics approval and consent to participate: as this was a descriptive case report and data was collected without patient identifiers, ethics approval was not required under our hospital's Institutional Review Board guidelines.

Informed consent: the patient provided consent for the access to medical records at the time of admission.

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