# Don’t Rash the Diagnosis!

**Learning objectives:**

1. Diagnose Stevens-Johnson-Toxic-Epidermal-Necrolysis Spectrum with its multi-mucosal and multi-organ involvement.
2. Recognize the importance of multidisciplinary care for patients with Stevens-Johnson-Toxic-Epidermal-Necrolysis Spectrum.

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# Case Presentation

A 64-year-old man presents with a one-month history of rash. Significant past medical history includes alcohol use disorder, congestive heart failure and chronic renal insufficiency.

He was seen by an outpatient dermatologist. He had a skin biopsy demonstrating spongiotic psoriasiform dermatitis and was prescribed acitretin.

The dermatitis did not resolve so he subsequently presented to his local emergency department twice and received two courses of cephalexin.

His dermatitis then significantly worsened, extending to 90% body surface area. He had painful erythroderma over 90% body surface area with superficial scaling and sloughing, including on the palms and soles. Nikolsky's sign was positive.

# Investigations & Initial Management

- Skin biopsy was repeated.  Working diagnosis was erythrodermic psoriasis vs Stevens-Johnson-Toxic-Epidermal-Necrolysis Spectrum (SJS/TEN).
- The patient received supportive management including intravenous fluids, antibiotics for presumed secondary bacterial infection, and temperature re-warming methods.
- Dermatology was consulted. Based on the painful erosions and mucositis present, there was a high suspicion for SJS/TEN. His treatment expanded to include clobetasol cream, wound care with non adherent dressing, methylprednisolone, cyclosporine, guselkumab and etanercept .
- The Ophthalmology team was also consulted and noted cicatrizing conjunctivitis.
- Skin biopsy showed extensive ulceration and necrosis with complete loss of epidermis and necrotic keratinocytes. SJS/TEN was confirmed and suspected to be secondary to cephalexin. His risk of mortality was estimated to be > 90% based on SCORTEN of 5[1].

## SCORTEN

**Scoring Criteria (1 point each)**

| Risk Factor | Criteria |
|---|---|
| Age | > 40 years |
| Heart Rate | > 120 bpm |
| Cancer/Hematologic Malignancy | Present |
| Body Surface Area Involved | > 10% |
| Serum Urea/BUN | > 28 mg/dL (> 10 mmol/L) |
| Serum Bicarbonate | < 20 mEq/L (< 20 mmol/L) |
| Serum Glucose | > 252 mg/dL (> 14 mmol/L) |

**Mortality Risk by SCORTEN Score**

| SCORTEN Score | Predicted Mortality Rate |
|---|---|
| 0-1 | 3.2% |
| 2 | 12.1% |
| 3 | 35.8% |
| 4 | 58.3% |
| 5 | 90.0% |
| ≥ 6 | > 90% |

Figure 1: SCORTEN (Score for Toxic Epidermal Necrolysis) severity-of-illness scale for SJS/TEN.

# Discussion & Conclusion

Multidisciplinary discussions between Internal Medicine, Dermatology, Ophthalmology, Critical Care and the Burn Unit at a quaternary care center were necessary to guide treatment as the patient developed sepsis from Raoultella bacteremia, worsening hypothermia and multi-organ failure secondary to lack of epidermis and persistent inflammation. Despite treatment, the patient had worsening liver failure, renal failure and heart failure, leading to anasarca and severe pulmonary edema requiring oxygen delivery via Optiflow Nasal High Flow. Intubation was not within the patient's goals of care, so eventually, he decided he wanted to pursue comfort care and passed away surrounded by his family.

KEY POINTS:

- SJS/TEN is a spectrum of potentially life-threatening severe cutaneous adverse reactions. It is a type IV hypersensitivity reaction typically mediated by an immunologic response to a trigger drug[1].
- There should be a low threshold to treat bacterial infections in SJS/TEN because it causes widespread detachment of the skin and mucosa, predisposing patients to superimposed bacterial infections[1].
- As demonstrated in this case, SJS/TEN can lead to multi-organ failure, and having a multidisciplinary approach is essential to providing optimal care.
- In severe cases, Burn Units should be consulted as they can provide unique expertise. Transfer to a Burn Unit can allow for advanced wound care and specialized controlled environments to prevent heat loss, as patients with SJS/TEN lose their skin barrier and cannot regulate their temperature adequately[2].
- Finally, this case highlights the importance of antibiotic stewardship as the patient had received two courses of cephalexin to treat a non-infectious rash initially, leading to this life-limiting reaction.

# Evidence of Conflict of Financial Interest

| | Co-author | Conflict disclosures |
|---|---|---|
| 1 | Shaonie Ton-Leclerc | No conflict of interest to disclose. |
| 2 | Lourdes Ramirez-Hobak | No conflict of interest to disclose. |
| 3 | Marie Leung | No conflict of interest to disclose. |

## References


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