SEPTEMBER 2015 VOLUME 2 ISSUE 7

EM CASE OF THE WEEK

BROWARD HEALTH MEDICAL CENTER DEPARTMENT OF EMERGENCY MEDICINE

Involvement of the oral mucosa in a case of Stevens-Johnson Syndrome.

SJS is a mucocutaneous reaction to an inciting factor that causes fever, necrosis and detachment of the epidermal layer.

EM CASE OF THE WEEK

EM Case of the Week is a weekly "pop quiz" for ED staff. The goal is to educate all ED personnel by sharing common pearls and pitfalls involving the care of ED patients. We intend on providing better patient care through better education for our nurses and staff.

Stevens-Johnson Syndrome

A 49 year old male with a history of depression comes into the ED with a 10 day history of painful desquamating rash involving palms, arms, legs, back, tongue and lips. Vital signs: Afebrile HR 76, RR 18, BP 144/94, O2 sat 98%. He denies URI symptoms, aggravating or alleviating factors. The rash has improved, leaving only mild crusting of the lips. No ocular or urethral involvement noted. Patient noted that he was started on lamotrigine (Lamictal®) and mirtazapine (Remeron®) 1 month ago by his PCP, which he stopped on day 1 of the rash. He also takes Venlafaxine (Effexor®) for his depression as well as Advil and Tylenol as needed for aches and pains. Which of the following is the most likely precipitating factor for his presentation?

A. Venlafaxine (Effexor®)

B. Lamotrigine (Lamictal®)

C. Ibuprofen (Advil®)

D. Acetaminophen (Tylenol®)

E. Mirtazapine (Remeron®)

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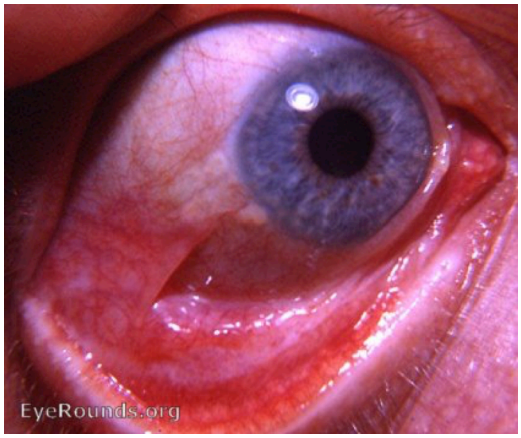


Take Home Points

- Medications are the leading trigger of SJS/TEN in both adults and children, watch out for patients who have recently been started on a new anticonvulsant, allopurinol, or sulfonamide.
- Typically, viral URI symptoms or flu symptoms may precede the rash.
- In severe cases, you may see massive loss of fluids and electrolyte imbalance, hypovolemic shock with renal failure, bacteremia, insulin resistance, hypercatabolic state, and multiple organ dysfunction syndrome.
- It is important to stop the offending agent early on, treatment is mostly supportive.

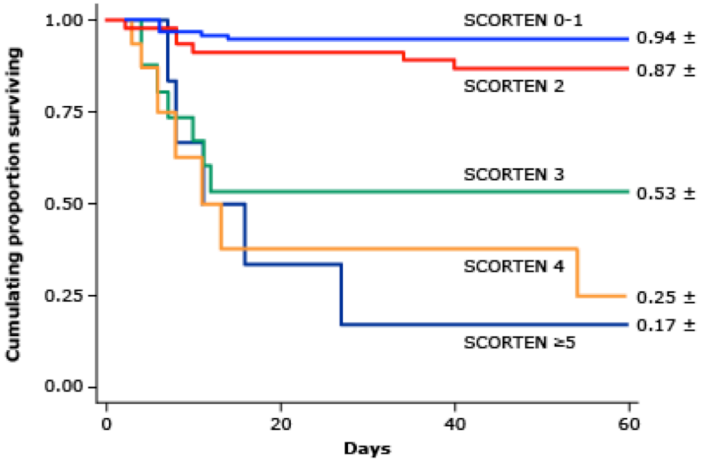
Stevens-Johnson Syndrome

The correct answer is B. Lamotrigine (Lamictal®) is one of the more common causes of Stevens-Johnson Syndrome.

Stevens-Johnson syndrome and toxic epidermal necrolysis are mucocutaneous adverse reactions, most commonly triggered by medications. They are characterized by fever, necrosis and detachment of the epidermis. The risk of SJS/TEN appears to be limited to the first 8 weeks of treatment. The most common causative agents are allopurinol, anticonvulsants, sulfonamides, lamotrigine, nevirapine and Oxicam NSAIDs. In children, SJS/TEN can be caused by M. Pneumoniae and CMV infections. Rarer causes include vaccines, contrast mediums and herbal medicines. Certain immunocompromised populations may be at increased risk as well.

Discussion:

Stevens-Johnson syndrome is the less severe of the two, in which skin detachment is <10 percent of the body surface. Mucous membranes are affected in over 90 percent of patients, usually at two or more sites (ocular, oral, and genital). Toxic Epidermal Necrolysis involves detachment of >30 percent of the body surface area.
Patients may present with fever and flu-like symptoms 1-3 days prior to the development of lesions. Photophobia, conjunctival burning or painful swallowing may be present.
The Pathogenesis of SJS/TEN is incompletely understood. It is thought to involve a cytotoxic T-cell reaction against the native forms of the causative drugs. This leads to eventual keratinocyte apoptosis, indirectly due to release of cytotoxic proteins like granulysin. Genetic factors may play a role as well.



Prognosis and SCORTEN Score

Prognosis can be evaluated on admission by applying a prognostic scoring system called SCORTEN.

Age	≥40 years	1
Malignancy*	Yes	1
Body surface area detached	≥10 percent	1
Tachycardia	≥120/min	1
Serum urea	>10 mmol/L	1
Serum glucose	>14 mmol/L	1
Serum bicarbonate	<20 mmol/L	1
SCORTEN#		7

* Malignancy: evolving cancer and hematological malignancies.

Treatment

Patients with a limited skin involvement, a SCORTEN score of 0 or 1, and disease that is not rapidly progressing may be treated in nonspecialized wards. Patients with more severe disease and a SCORTEN score ≥2 should be transferred to intensive care units or burn units if available.

These patients may present prior to any hemodynamic instability, therefore it is important for ER physicians to recognize SJS/TEN promptly. Early withdrawal of the causative drug is important and has been shown to reduce mortality.

Treatment is usually supportive and symptom-based. The use of corticosteroids, IVIG or immunosuppresants remains controversial. In the ED it is important to address fluid status and electrolyte imbalances that occur due to water loss from the exposed dermis. Keep temperatures warmer to decrease caloric loss.

Open wounds are typically treated like burns. Some hospitals surgically debride wounds and use whirlpool therapy to remove necrotic epidermis, others use the epidermis to act like a biologic dressing.

A baseline ophthalmologic consultation should be obtained soon after admission. Ocular care includes appropriate cleaning of eye lids and daily lubrication.

Patients with SJS/TEN are at high risk of infection and sepsis remains a prominent cause of death. Because of this, it is important to regularly culture blood, gastric tubes or any catheters placed.

Sterile handling is essential, antiseptic solutions containing octenidine, polyhexanide (eg, Octenisept, Lavasept, Prontosan), or chlorhexidine or silver nitrate preparations may be used for disinfection. Silver Sulfadiazine is not often used in SJS because of the association of sulfa drugs.

UpToDate: Stevens-Johnson syndrome and toxic epidermal necrolysis: Pathogenesis, clinical manifestations, and diagnosis
University of Iowa Healthcare: EyeRounds Online Atlas of Ophthalmology: Stevens-Johnson Syndrome
Medscape: Stevens-Johnson Syndrome Treatment and Managment

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ABOUT THE AUTHOR:
This month's case was written by Alisa Hussain. Alisa is a 4th year medical student from NSU-COM. She did her Emergency Medicine rotation at BHMC in September 2015. Jane plans on pursuing a career in Internal Medicine after graduation.