

CLINICAL VIGNETTE

A 37-Year-Old Woman with a Life-Threatening Rash: Stevens Johnson Syndrome

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Introduction

Stevens Johnson syndrome (SJS) is a severe hypersensitive reaction that can be precipitated by infection, vaccination, systemic diseases, or physical agents, including foods and drugs.¹ Many drugs can cause SJS including antibacterials (sulfonamides), anticonvulsants (phenytoin, phenobarbital, carbamazepine), non-steroidal anti-inflammatory drugs (oxicam derivatives) and oxide inhibitors (allopurinol).²

Case Report

A 37-year-old female with asthma, and BiPolar-2 Disorder, presented to the ED with dysphagia and a progressive rash. Three weeks prior, her psychiatrist started Lamotrigine and increased her dose of valproate. She had been taking a lower dose of valproate for years without issues.

After two weeks she noted a rash on her foot, along with high fever and swollen glands. She initially felt like she had the flu and discontinued all new medications. After failure to improve she was evaluated at urgent care and given prednisone for 3 days. She returned to urgent care in day 3 and was found to have oral ulcers and returned to the emergency department.

She reported the rash began on the bottom of her left foot before initially progressing to her chest and arms, followed by involvement of her palms, back, face and mouth. The rash became increasingly confluent and she also noted difficulty swallowing and talking due to throat pain. The pain was described as "burning". She had severe dysphagia for solids for 2 days, while liquids were still tolerable. She also noted blurry vision that improved with blinking or saline drops. There was no shortness of breath, dysuria or vaginal involvement.

Her PMH was significant for Basal Cell Carcinoma, Bipolar Syndrome, and Asthma. She did not have any previously known allergies to medications.

On exam, she had erythematous macules, some coalescing into patches on her face, lower chest and back and arms. There was diffuse erythema of her lower neck and upper chest with early bullae formation. Her legs and dorsal feet had scattered erythematous macules and face and palms had blanchable erythema. Intraoral examination revealed ulcerations of the vermilion surface of the lips. She also had significant facial edema, nasal erosions and conjunctival erythema. Tender neck lymphadenopathy was also noted.

Due to the temporal relationship to starting Lamictal and valproic acid, they were added to her drug allergy list. We discussed never taking these medications again.

She was referred to dermatology who agreed with the diagnosis of SJS versus toxic epidermal necrolysis, and started high dose systemic steroids, topical steroids, and IV Fluid hydration.

Discussion

Stevens Johnson Syndrome (SJS) and Toxic Epidermal Necrolysis (TEN) were originally thought to be two separate disease processes but are now thought to be on the same spectrum.² This severe skin reaction is most often triggered by medications. SJS and TEN differ only in their extent of skin detachment and involvement. SJS/TEN is rare with annual prevalence of 2/1,000,000. It is considered a medical emergency with potentially life-threatening sequelae include infection, shock, organ failure and death.

It often begins with fever and flu-like symptoms and within a few days progresses to involve skin. Usually, the blisters are raw and painful and begin on the face and chest, before spreading to the mucous membranes, including mouth and airways. Occasionally patients have difficulty swallowing and breathing. SJS/TEN also can affect the eyes causing irritation and redness. Table 1 lists the features that differentiate SJS from TEN.

"Several drugs are considered "high" risk of inducing TEN/SJS. These include: Allopurinol, Trimethoprim-sulfamethoxazole and other sulfonamide-antibiotics, aminopenicillins, cephalosporins, quinolones, carbamazepine, phenytoin, phenobarbital and oxicam-type NSAID's."³

Diagnosis relies heavily on clinical signs and occasional skin biopsy. Due to the high morbidity, early diagnosis with prompt recognition and withdrawal of all potential causative drugs is needed for a favorable outcome. Management relies heavily on supportive care. Cessation of the offending agent, IV Fluids, and wound care remain the cornerstone of therapy. Additional therapeutic options are controversial.

In a single center 15-year study at LAC+USC burn unit, 40 patients were identified with biopsy proven SJS/TENS with 10 % mortality rate. Their treatment algorithm added Nutritional

supplementation to the mainstay of early discontinuation of the offending agent, IV Fluids, and wound care.¹

Antiseptic wound care and liberal used antibiotic eye drops are recommended as infection is thought to be the largest risk. Corticosteroids were the mainstay of therapy for SJS but this has become more controversial. The popular belief was that steroids suppress the reaction intensity. However, some studies show that steroids increased the risk of infection. A recent European study showed short term steroids have beneficial role in reducing mortality.¹ Other controversial systemic treatments include IVIG, Cyclosporine, Plasmapheresis and TNF inhibitors. Additional studies are needed to validate and better define usefulness.¹

Conclusion

Our patient was treated with IV Fluids, wound care, eye drops and IV steroids. She remained stable with no involvement of other organs. Kidney and renal function remained normal and she had no respiratory or vaginal symptoms. Her main symptom beside the skin rash were oral sores which improved with treatment. After dramatic improvement, and she was discharged home with close dermatology follow-up. High clinical suspicion for SJS and close management is needed to prevent sequela.

Clinical entity	SJS	SJS-TEN overlap	TEN
Primary lesions	Dusky red lesions	Dusky red lesions	Poorly delineated erythematous plaques
	Flat atypical targets	Flat atypical targets	Epidermal detachment
			Dusky red lesions
			Flat atypical targets
Distribution	Isolated lesions	Isolated lesions	Isolated lesions (rare)
	Confluence (+) on face and trunk	Confluence (++) on face and trunk	Confluence (+++) on face, trunk, and elsewhere
Mucosal involvement	Yes	Yes	Yes
Systemic symptoms	Usually	Always	Always
Detachment (%body surface area)	< 10	10-30	> 30

Table 1.

REFERENCES

1. **McCullough M, Burg M, Lin E, Peng D, Garner W.** Steven Johnson Syndrome and Toxic Epidermal Necrolysis in a burn unit: A 15-year experience. *Burns*. 2017 Feb;43(1):200-205. doi: 10.1016/j.burns.2016.07.026. Epub 2016 Aug 21. PMID: 27554629.
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