

Background & Methodology

Stevens-Johnson Syndrome (SJS) and Toxic Epidermal Necrolysis (TEN) are believed to be two ends of a disease spectrum involving vast keratinocyte death that causes mucocutaneous lesions, often resulting in severe morbidity and mortality.^[1,2,3] Currently, little is known regarding the frequency with which certain diseases mimic SJS/TEN in the pediatric population, and it is believed that SJS/TEN has been misdiagnosed as Staphylococcal Scalded Skin Syndrome (SSSS) and Erythema Multiforme Major (MMM), among others.^[2,3] Misdiagnosis increases a patient's risk for suffering and long-term sequelae. On the one hand, both SJS/TEN and certain mimickers may have similar treatment approaches, but there are important differences, such as the use of antibiotics in SSSS, which might otherwise be neglected if SJS is mistakenly diagnosed.

This study is a descriptive, sub-analysis of the complete data published in 2019 by Weinkle et al. on distinguishing SJS/TEN from mimickers.^[4] In that study, 208 patients were sampled from dermatology consults for suspicion of SJS, SJS-TEN overlap, or TEN. Patients ≤ 18 years old were included in this descriptive analysis of SJS/TEN and mimickers. Simple descriptive statistics were calculated. Analyses and graphics were created in R v3.6.1 (R Core Team, 2019).

Table 1: Patient Characteristics

	Mimicker (n=11)	SJS/TEN (n=2)
Age		
Mean (SD)	8.05 (6.44)	10.0 (11.3)
Median [Min, Max]	7.00 [0.583, 18.0]	10.0 [2.00, 18.0]
LOS		
Mean (SD)	3.45 (2.34)	16.5 (2.12)
Median [Min, Max]	3.00 [0.00, 8.00]	16.5 [15.0, 18.0]
Sex		
Female	6 (54.5%)	2 (100%)
Male	5 (45.5%)	0 (0%)
Transferred in		
No	6 (54.5%)	0 (0%)
Yes	5 (45.5%)	2 (100%)
Admit location		
Floor	6 (54.5%)	0 (0%)
MICU	4 (36.4%)	2 (100%)
Not Admitted	1 (9.1%)	0 (0%)
Fever		
No	4 (36.4%)	0 (0%)
Yes	7 (63.6%)	2 (100%)
High-risk medication		
Antibiotic	6 (54.5%)	0 (0%)
Anticonvulsant	1 (9.1%)	1 (50.0%)
Antiviral	1 (9.1%)	0 (0%)
Multiple	0 (0%)	1 (50.0%)
None	3 (27.3%)	0 (0%)
CBC		
No Labs	2 (18.2%)	0 (0%)
Normal	8 (72.7%)	2 (100%)
Other	1 (9.1%)	0 (0%)
CMP		
Abnormal hepatic labs	0 (0%)	1 (50.0%)
Multiple abnormalities	1 (9.1%)	0 (0%)
No labs	5 (45.5%)	0 (0%)
Within Normal Limits	5 (45.5%)	1 (50.0%)

Results & Discussion

Thirteen patients from the original study met inclusion criteria: 2 SJS/TEN, and 11 mimickers. The median age for the mimickers was 7 years with the youngest patient as 7 months and 18 years as the oldest; the SJS patients were 2 and 18 years old. Just under 46% (5) of mimickers were transferred from an OSH compared to both SJS patients transferring in for care. Both SJS patients were admitted to MICU while 36% (4) of mimickers were admitted to the MICU; roughly 55% (6) of mimickers were female. Median LOS was 3 days (max of 8) for mimickers; the two SJS patients stayed 15 and 18 days. Nearly 64% (7) of mimicker patients were febrile and 55% (6) had antibiotic exposure. **Table 1** provides more detail.

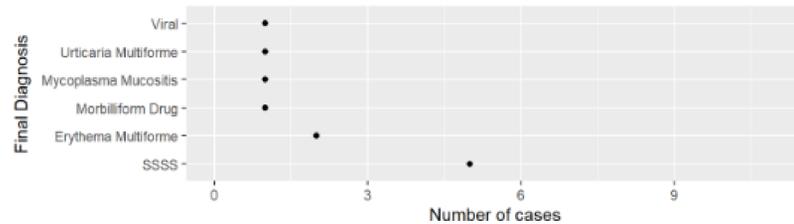
Rash characteristics by final diagnosis are presented in **Table 2**. SSSS was the most common mimicker (5/11). About 70% (9/13) of all patients presented in a 24-120 hour window; both SJS cases appeared in this period. Eighty percent (4) of SSSS cases presented during this time as well. Vesicle/bullae was the most common primary lesion, presenting only in SSSS patients, followed by atypical targets; both SJS patients had atypical targets while the only non-SJS patient with atypical targets was EM. Skin pain was present in both SJS patients and in 80% (4) of SSSS patients as well as one EM patient. Excluding SSSS, mimickers tended not to report skin pain. Mucosal involvement occurred in roughly half (7) of all patients with oral involvement seen in UM, MM, EM. Eighty percent of SSSS patients did not have mucosal involvement and 40% (2) had a positive Nikolsky sign recorded.

These data suggest that lack of mucosal involvement may be useful for differentiating SSSS from SJS/TEN; atypical targets were found in only one mimicker which may also continue to benefit the assessing physician with diagnosis. These findings are congruent with the literature.^[1,3]

Table 2: Rash Characteristics by Final Diagnosis

	Erythema Multiforme (n=2)	Morbiliform Drug (n=1)	Mycoplasma Mucositis (n=1)	SSSS (n=5)	Urticaria Multiforme (n=1)	Viral (n=1)	SJS/TEN (n=2)	Overall (n=13)
Admitted for rash								
No	0 (0%)	1 (100%)	0 (0%)	0 (0%)	0 (0%)	0 (0%)	0 (0%)	1 (7.7%)
Yes	2 (100%)	0 (0%)	1 (100%)	5 (100%)	1 (100%)	1 (100%)	2 (100%)	12 (92.3%)
Rash duration								
<24 hours	1 (50.0%)	1 (100%)	0 (0%)	1 (20.0%)	0 (0%)	0 (0%)	0 (0%)	3 (23.1%)
24 to 48 hours	0 (0%)	0 (0%)	0 (0%)	2 (40.0%)	0 (0%)	1 (100%)	1 (50.0%)	4 (30.8%)
48 to 120 hours	1 (50.0%)	0 (0%)	0 (0%)	2 (40.0%)	1 (100%)	0 (0%)	1 (50.0%)	5 (38.5%)
>120 hours	0 (0%)	0 (0%)	1 (100%)	0 (0%)	0 (0%)	0 (0%)	0 (0%)	1 (7.7%)
Primary lesion								
atypical targets	1 (50.0%)	0 (0%)	0 (0%)	0 (0%)	0 (0%)	0 (0%)	2 (100%)	3 (23.1%)
erythroderma	0 (0%)	0 (0%)	0 (0%)	1 (20.0%)	0 (0%)	0 (0%)	0 (0%)	1 (7.7%)
macule or patch	0 (0%)	1 (100%)	0 (0%)	0 (0%)	0 (0%)	1 (100%)	0 (0%)	2 (15.4%)
papule or plaque	0 (0%)	0 (0%)	1 (100%)	0 (0%)	0 (0%)	0 (0%)	0 (0%)	1 (7.7%)
typical targets	1 (50.0%)	0 (0%)	0 (0%)	0 (0%)	1 (100%)	0 (0%)	0 (0%)	2 (15.4%)
vesicles or bullae	0 (0%)	0 (0%)	0 (0%)	4 (80.0%)	0 (0%)	0 (0%)	0 (0%)	4 (30.8%)
Skin pain								
No	1 (50.0%)	1 (100%)	1 (100%)	1 (20.0%)	1 (100%)	1 (100%)	0 (0%)	6 (46.2%)
Yes	1 (50.0%)	0 (0%)	0 (0%)	4 (80.0%)	0 (0%)	0 (0%)	2 (100%)	7 (53.8%)
Mucosal involvement								
Multiple	1 (50.0%)	0 (0%)	0 (0%)	1 (20.0%)	0 (0%)	0 (0%)	0 (0%)	2 (15.4%)
Oral	1 (50.0%)	0 (0%)	1 (100%)	0 (0%)	1 (100%)	0 (0%)	2 (100%)	5 (38.5%)
None	0 (0%)	1 (100%)	0 (0%)	4 (80.0%)	0 (0%)	1 (100%)	0 (0%)	6 (46.2%)
Nikolsky sign present								
No	2 (100%)	1 (100%)	1 (100%)	3 (60.0%)	1 (100%)	1 (100%)	0 (0%)	9 (69.2%)
Yes	0 (0%)	0 (0%)	0 (0%)	2 (40.0%)	0 (0%)	0 (0%)	2 (100%)	4 (30.8%)

Staphylococcal Scalded Skin Syndrome: the most common mimicker



Mimickers tend not to have mucosal involvement

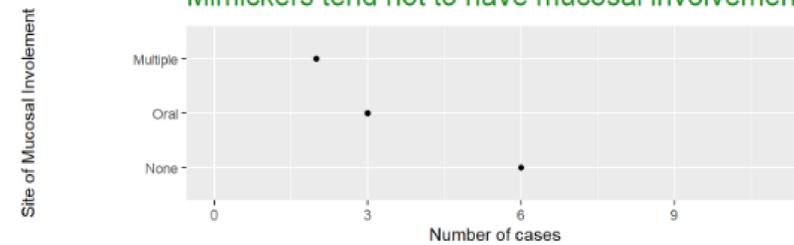


Figure 1: SJS/TEN mimicker photos obtained with consent during dermatologic consultation. A. SSSS; the ears, upper back, and upper extremities with superficial erosions and desquamation. B. SSSS; perioral oral accentuation of erythema with radial scale-crusts. C. Urticaria Multiforme (UM); arcuate and annular edematous plaques. Some with dusky center but without signs of erosion or necrosis, others with central clearing.

Conclusion

- ❖ SSSS was the most common SJS mimicker representing 45% of pediatric, non-SJS cases
 - DRESS and other severe cutaneous drug eruptions were the most common SJS/TEN mimicker in adult patients (25% all cases)^[4]
- ❖ The discernment of SSSS from SJS may be aided by the absence of mucosal involvement in pediatric cases
- ❖ Presence of atypical targets may continue to prove useful for distinguishing a mimicker from SJS in pediatric patients
- ❖ Treatment of SJS/TEN involves primarily supportive care while SSSS needs antimicrobial therapy^[1,3]
 - SJS/TEN may utilize immunosuppressive medications that are unwarranted in a mimicking disease
- ❖ Awareness about timely and accurate diagnosis of SJS mimickers, particularly SSSS, is imperative as it may reduce morbidity and mortality from misguided treatment

References

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