Dermatologic Emergencies:
What’s That?
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I have no relevant conflicts of interest.

Objectives
• Identify, appropriately work-up, and stabilize urgent/emergent dermatologic conditions
• 3 cases that illustrate:
  – Erythema Multiforme
  – Stevens-Johnson Syndrome
  – Toxic Epidermal Necrolysis
  – Staph Scalded Skin Syndrome
  – Eczema herpeticum
  – Eczema coxsackium
  – Staph superinfection

Clues: When to Worry
• Age (newborn and young infants)
• High fever, toxicity
• Morphology
  – blistersing, mucosal involvement, hemorrhage
• Specific medications
  – anti-convulsants, antibiotics, NSAIDS

How to describe what you are seeing... over the phone
• A picture is worth a thousand words
• Extent: What body surface area is involved?
  – the patient’s palm = 1%

Case 1:
This 5 yo boy with a seizure disorder and language delay has had fever, malaise, lymphadenopathy and a sore throat for 4 days. He has been taking Tylenol for 4 days, and lamotrigine for 6 weeks.
What is the most likely diagnosis?

Case 1: 5 yo with sz d/o and rash.
What is the most likely diagnosis?
1. Erythema multiforme
2. Kawasaki Disease
3. Stevens-Johnson syndrome (SJS)
4. Vasculitis
**Case 1**

SJS vs EM vs TEN

**What is SJS?**

- Severe, life-threatening mucocutaneous disease
- Clinical syndrome - no definitive diagnostic test
- atypical "targetoid" lesions, fragility, denudation ~10%BSA
- ≥2 mucous membranes (mouth, eyes)
- systemic signs: fever, respiratory symptoms

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**The SJS Spectrum**

<table>
<thead>
<tr>
<th>Erythema Multiforme</th>
<th>Stevens Johnson (SJS)</th>
<th>SJS-TEN overlap</th>
<th>Toxic Epidermal Necrolysis (TEN)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Minor and Major</td>
<td>&lt; 10% BSA</td>
<td>10-30% BSA</td>
<td>&gt; 30% BSA</td>
</tr>
</tbody>
</table>

**Infection**

- Low Mortality
- Drug
- High Mortality

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**EM vs SJS vs TEN**

<table>
<thead>
<tr>
<th></th>
<th>EM</th>
<th>EM major</th>
<th>SJS</th>
<th>SJS-TEN</th>
<th>TEN</th>
</tr>
</thead>
<tbody>
<tr>
<td>Rash</td>
<td>Typical targets</td>
<td>Typical targets</td>
<td>Dusky red, target-like, Detachment</td>
<td>Dusky red, target-like, Detachment</td>
<td>Pigmentary delamination, Fluffy plaque, large sheets of detachment</td>
</tr>
<tr>
<td>BSA Detached</td>
<td>&lt;15%</td>
<td>10-30%</td>
<td>&gt;30% with spots, &gt;10% without spots</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Distribution</td>
<td>Extremities, face</td>
<td>Extremities, face</td>
<td>Trunk, back, (+confluence)</td>
<td>Trunk, back, (+confluence)</td>
<td>Face, trunk, ext, (+confluence)</td>
</tr>
<tr>
<td>Mucosal Involvement</td>
<td>None, mild</td>
<td>Severe</td>
<td>Severe</td>
<td>Severe</td>
<td>Yes</td>
</tr>
<tr>
<td>Systemic Symptoms</td>
<td>Absent</td>
<td>Usually</td>
<td>Usually</td>
<td>Absent</td>
<td>Absent</td>
</tr>
<tr>
<td>Progression to TEN</td>
<td>No</td>
<td>No</td>
<td>Possible</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Etiology</td>
<td>HSV, other infections</td>
<td>HSV, Mycoplasma, new drug</td>
<td>Drug</td>
<td>Mycoplasma</td>
<td>HSV</td>
</tr>
</tbody>
</table>

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Bolognia, Dermatology, 2nd Edition.
SJS: Causes

- **DRUGS**
  - Many drugs implicated
  - Anticonvulsants > antibiotics > NSAIDs
  - Typically 7-21 days after start
  - Drugs with longer half-lives more likely to cause a fatal reaction
- **Mycoplasma**
  - up to 25% of pediatric patients with SJS
  - more mucosal, less skin, +cough
- **HSV**
- **Unknown**

Stevens-Johnson Syndrome (SJS) (Mycoplasma Associated)

Why isn’t this EM?

Erythema Multiforme

- Target lesions with 3 zones
  - Dusky center
  - pale edematous ring
  - peripheral erythematous margin
- Discrete lesions
- Usually no/mild systemic signs

Variety of targets in EM

Erythema Multiforme

Bottega, Dermatology, 2nd ed.
EM vs SJS

Typical targets
EM

Atypical targets
SJS

It looks like EM now, but…

• Be more worried if you see:
  – Atypical targets
  – Trunk > Acral lesions
  – Confluent skin lesions
  – Bullous skin lesions
  – Continuing rapid progression

Why isn’t this TEN?

TOXIC EPIDERMAL NECROLYSIS

TEN with spots
>30% BSA detached

TEN without spots
>10% BSA detached

SJS Initial Management & Work-Up

• ABCs
• Stop the causative drug (and all non-essential drugs)
• Admit to ICU or burn unit if >10-20% BSA
• Call dermatology/ophthalmology/urology
• Labs: CBC, Lytes, BUN, Cr, LFTs
• Check for Mycoplasma, HSV (IgM)
SJS Supportive Care

- Meticulous daily wound care
  - Wash with saline, gently remove crust around orifices
  - Provide suction for secretions
  - Cover denuded areas (& corners of mouth) with vaseline gauze
  - Pressure bed
  - Avoid friction, trauma
  - Reverse isolation
- Surveillance cultures (?)
- Hydration (careful not to overload)
- Nutrition (NG)

Practical Treatment

<table>
<thead>
<tr>
<th>Practical Treatment</th>
<th>EM</th>
<th>SJ S</th>
<th>TEN</th>
</tr>
</thead>
<tbody>
<tr>
<td>• Treat infection</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>• Steroids?</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>• Stop drug</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>• Treat Infection</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>• Early steroids</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>• IVIG</td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

What is going to happen to this child?

Outcome of SJS/TEN spectrum

<table>
<thead>
<tr>
<th>Outcome of SJS/TEN spectrum</th>
<th>TABLE 3: Outcome of 56 Children with SJS, SJS/TEN Overlap Syndrome, and TEN</th>
</tr>
</thead>
<tbody>
<tr>
<td>Total Cohort</td>
<td>(n = 56)</td>
</tr>
<tr>
<td>Long-term sequelae</td>
<td>56 (100)</td>
</tr>
<tr>
<td>Skin sequelae, leg</td>
<td>56 (100)</td>
</tr>
<tr>
<td>Hypopigmentation, sunburn</td>
<td>25 (45)</td>
</tr>
<tr>
<td>Eye sequelae (iris, cornea, conjunctiva)</td>
<td>15 (27)</td>
</tr>
<tr>
<td>Defects, iritis, complications</td>
<td>15 (27)</td>
</tr>
<tr>
<td>Phimosis</td>
<td>2 (4)</td>
</tr>
<tr>
<td>Severe angina</td>
<td>1 (2)</td>
</tr>
<tr>
<td>Bronchiolitis shunters</td>
<td>1 (2)</td>
</tr>
<tr>
<td>Stridor</td>
<td>1 (2)</td>
</tr>
<tr>
<td>Venous thrombosis</td>
<td>1 (2)</td>
</tr>
<tr>
<td>Recurrence of SJS</td>
<td>10 (18)</td>
</tr>
<tr>
<td>Recurrence of TEN</td>
<td>0 (0)</td>
</tr>
<tr>
<td>Mortality</td>
<td>1 (2)</td>
</tr>
</tbody>
</table>


Case 2:

An 8 year old otherwise healthy boy presents with a 2 day history of an acute-onset, progressive blistering eruption associated with skin pain, malaise, and low grade fever. He is mildly tachycardic, but other VS are stable.

Which of the following is the most likely diagnosis?
Case 2: 8 yo with blistering. Diagnosis?
1. Kawasaki Disease
2. Staph Scalded Skin Syndrome
3. Toxic Epidermal Necrolysis
4. Toxic Shock Syndrome

Case 2
SSSS vs TEN vs TSS

SSSS
- Begins as a localized, often occult infection
  - Nasopharynx
  - Perioral
  - Conjunctiva
  - Umbilicus
  - Paronychia
  - Wound
  - Urine
  - Middle Ear
- Progresses to generalized erythema and skin fragility

SSSS: Etiology
- Staph produces an exfoliative exotoxin
- Exotoxin cleaves desmoglein 1 → superficial epidermal cleft, acantholysis

Staphylococcal Scalded Skin Syndrome
- Clinical Presentation
  - Neonates: Widespread erythema, superficial erosions
  - Toddlers & Children: erythema, periorificial scale and erosions, skin fragility and pain
  - Adults: rare - protective antitoxin

SSSS
Source: blistering dactylitis & conjunctivitis
- Perioral furrowing, scale
- Skin pain
Why isn’t this TEN?

Why isn’t this TEN?

- Not shiny = SSSS
  Superficial epidermal split

- Shiny = TEN
  Subepidermal split

Why isn’t this toxic shock syndrome?

Toxic Shock Syndrome

- Rarely a primarily cutaneous disease

- Staph produces superantigens that cause:
  - fever
  - rash
  - hypotension
  - organ system involvement

SSSS: Management

- Admit (especially in younger pts)
- Dermatology consult
- Culture potential sources
- Empiric anti-staph antibiotics (cover for MRSA) +/- Clindamycin
  - Clindamycin inhibits toxin production
  - d/c with abx based on cx results
- Careful FEN management
- Pain management

Case 3:

A 13 yo girl with a history of atopic dermatitis presents with 1 day of a new rash around her eyes and mouth, and low grade fever.

What is the best diagnosis?
Case 3: Best Diagnosis?
1. Contact Dermatitis
2. Eczema coxsackium
3. Eczema herpeticum
4. Staph superinfection

Eczema Herpeticum
- Disseminated HSV in pts with chronic skin dz
- Abrupt onset fever, malaise
- Painful
- History of HSV exposure or prior infection
- *Delay in Dx common*

Eczema Herpeticum: Morphologic Clues
- Monomorphous erosions > vesicles
- Lesions favor
  - Areas of active dermatitis
  - Head, neck & trunk

Eczema Herpeticum vs. Contact Dermatitis

Eczema Herpeticum vs. Staph Superinfection
Strep Superinfection

Eczema Herpeticum vs. Eczema Coxsackium

Eczema Herpeticum

Treatment
• Culture, DFA or PCR
• Culture for bacteria
• Ophthalmology consult (for periocular involvement)
• Dermatology consult
• Prompt high dose acyclovir
• Empiric antibiotics if signs of bloodstream infection
• Topical steroids okay
• Avoid systemic steroids


Sequelae
• Scarring
• Ocular complications
• Recurrent infections
• Prolonged hospital stays

Summary
• Case 1: Stevens-Johnson Syndrome
  – Watch for atypical targets, classic mucous membrane involvement, calculate BSA
• Case 2: Staph Scalded Skin Syndrome
  – Look for a superficial epidermal split, non-toxic child, culture potential sources, can do a frozen section
• Case 3: Eczema Herpeticum
  – Look for monomorphous erosions in a patient with AD, consult ophtho if close to eyes, prompt acyclovir

Thank You!
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References


• Metry DW, Jung P, Levy ML. Use of intravenous immunoglobulin in children with stevens-johnson syndrome and toxic epidermal necrolysis: seven cases and review of the literature. Pediatrics 2003 Dec;112(6 Pt 1):1430-6


References (cont)


