Dermatologic Emergencies

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No Disclosures
Objectives

• Identify life threatening and emergent dermatologic conditions

• Discuss clinical clues to help distinguish different diseases

• Review up to date management
Outline

• Introduction

• Emergent conditions:
  • SJS/TEN
  • EM
  • MIRM

• Conclusion
Introduction

- In the United States, about 5-8% of all Emergency Department visits are due to dermatologic complaints

- It is imperative to recognize life-threatening and almost life-threatening conditions that require immediate attention to improve overall prognosis
Steven Johnsons Syndrome/Toxic Epidermal Necrolysis

• Life threatening mucocutaneous eruption

• Both SJS and TEN are regarded as variants on a continuous spectrum of adverse drug reactions

• Mortality rate for SJS is 1% to 5%
• Mortality rate for TEN is 25% to 35%
Surface area of Epidermal Detachment

- Dermatology: French, Lars E; Prins, Christa. Published December 31, 2011. Pages 319-333. ©
SJS/TEN

• Pathophysiology: Immune dysregulation resulting in apoptotic keratinocytes

• Etiology: > 100 causative agents
  • Most common include:
    • Allopurinol
    • NSAIDs
    • Sulfonamides
    • Anticonvulsants

• May sometimes be precipitated by viral illness
Clinical Features

• Timing: Occurs 1 to 3 weeks after starting causative drug
  [aromatic anticonvulsants may take up to 2 months]

• Initially fever and URI symptoms, painful skin → dusky, atypical
targetoid skin lesions → painful mucosal erosions → progresses to
epidermal detachment → bullae and sloughing
• Full thickness epidermal necrosis with subepidermal bulla formation
Diagnosis

• Clinicopathological correlation

• +Nikolsky Sign: epidermal detachment with pressure on area adjacent to blister

• +Asboe-Hansen sign: pressure on bulla causes spreading to uninvolved skin

• Skin biopsy: full thickness epidermal necrosis
Prognosis

- Depends on EARLY Diagnosis

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**SCORTEN**

<table>
<thead>
<tr>
<th>Prognostic factors</th>
<th>Points</th>
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<tbody>
<tr>
<td>Age &gt; 40 years</td>
<td>1</td>
</tr>
<tr>
<td>Heart rate &gt; 120 bpm</td>
<td>1</td>
</tr>
<tr>
<td>Cancer or hematologic malignancy</td>
<td>1</td>
</tr>
<tr>
<td>BSA involved on day 1 above 10%</td>
<td>1</td>
</tr>
<tr>
<td>Serum urea level (&gt;10 mmol/l)</td>
<td>1</td>
</tr>
<tr>
<td>Serum bicarbonate level (&lt;20 mmol/l)</td>
<td>1</td>
</tr>
<tr>
<td>Serum glucose level (&gt;14 mmol/l)</td>
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</tr>
</tbody>
</table>

**Mortality rate (%)**

<table>
<thead>
<tr>
<th>SCORTEN</th>
<th>Mortality rate (%)</th>
</tr>
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<tbody>
<tr>
<td>0–1</td>
<td>3.2</td>
</tr>
<tr>
<td>2</td>
<td>12.1</td>
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<td>3</td>
<td>35.8</td>
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<tr>
<td>4</td>
<td>58.3</td>
</tr>
<tr>
<td>≥5</td>
<td>90</td>
</tr>
</tbody>
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Management

- STOP offending agent
- Transfer to Burn Unit
- Supportive therapy, wound care, nutrition, fluids
- Consult Ophthalmology, Ob/Gyn/Urology
- Adjuvant therapy options remain controversial
- No established guidelines
- Systemic steroids: recent literature showing INCREASE in mortality with use of systemic steroids as sole therapy
- Promising data with IVIG and cyclosporine
- Etanercept
Etanercept therapy for toxic epidermal necrolysis.

Paradisi A¹, Abeni D², Bergamo F³, Ricci F⁴, Didona D⁵, Didona B³.

- Case series of 10 patients with biopsy proven TEN treated with single dose of Etanercept
- All responded to treatment, reaching complete re-epithelialization, with median time to healing of 8.5 days
Erythema Multiforme

• Acute, self-limited eruption
• Seen in 0.01 and 1%, primarily in young adults and children
• EM Major (mucosal) and EM Minor (no mucosal involvement)
• Etiology
  • Most common cause: HSV
  • Other infections i.e., Mycoplasma; rarely drugs
• Does NOT progress to TEN vs SJS
• NOW CONSIDERED SEPARATE ENTITY FROM SJS/TEN
Clinical Features

- **EM Major**
  - Targetoid lesions, atypical targets, +/- bullae
  - Extremities, face
  - Severe mucosal involvement, +systemic symptoms

- **EM Minor**
  - Targetoid lesions, extremities – elbows, knees, wrists hands; face
  - Minimal mucosal involvement, no systemic symptoms
• Vacuolar interface dermatitis with a perivascular lymphocytic infiltrate and necrotic keratinocytes
Management

• Check Mycoplasma serology
• Treat precipitating factor if identified
• If recurrent HSV-associated EM, consider prophylaxis with Acyclovir or Valacyclovir x 6 months
• EM Major: prednisone, dapsone, mycophenolate mofetil
• EM Minor: symptomatic rx, oral antihistamines
• Apremilast
• Recurrent EM defined as at least two episodes, 6 episodes in one year over a course of 6-10 years
• Apremilast has been tried in 3 patients (30-60mg daily)
• Has also been used to treat Behcet’s disease
Mycoplasma Induced Rash and Mucositis (MIRM)

• Previously thought to be a variant of EM/SJS

• Etiology: *Mycoplasma pneumoniae*
  • 25% patients experience extrapulmonary complications
Authors performed literature search, reviewed 202 cases, to characterize morphology and disease course with *M. pneumoniae* and associated mucocutaneous disease.
Clinical Features

- Young patients
- M>F
- Prodrome fever, cough preceding rash x 1 week
- Prominent mucositis: oral mucosa > urogenital, conjunctival
- Cutaneous involvement less common, usually acral distribution
- Polymorphic lesions: Vesiculobullous, targetoid, macules, papules
- Mortality 3%
MIRM

Oscar Tamez-Rivera et al. Pediatrics in Review 2017;38:234
How to Differentiate from SJS/TEN

- Negative Nikolsky sign
- Cutaneous lesions usually acrally distributed
- <10% BSA affected
- Absence of drug exposure
- Evidence of atypical pneumonia i.e. symptoms, CXR, *M. Pneumoniae* serology
- Milder clinical course
Management

- Serology for M. Pneumoniae, CXR
- Skin biopsy: lesser degree of epidermal necrosis vs SJS/TEN
- Supportive care, magic mouthwash
- Ophthalmology, Gyn/Urology consults
- Treatment is controversial; no evidence based guidelines
- Anecdotally, treatment with steroids, antibiotics or both; IVIG in some cases
- Antibiotic therapy helps prevent pulmonary/neurologic complications; unclear if helps with mucocutaneous eruption
Conclusion

• Early identification of dermatologic emergencies is critical

• Steroids are NOT always the answer

• Time is key in order to improve prognosis

• If clinical suspicion is high, treat empirically
  • do not wait for diagnostic testing
Thank You!
References


