

**Risky Rashes: A Review of Dermatologic Emergencies**  
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**Stevens - Johnson Syndrome (SJS) & Toxic Epidermal Necrolysis (TEN)**

- 1) Overview
  - a) Severe mucocutaneous reaction resulting in necrosis and detachment of the epidermis
  - b) SJS & TEN are variants of same disease continuum
  - c) Typically triggered by medications; onset within 8 weeks of starting medication
    - i) Anticonvulsants, allopurinol, sulfonamides, nevirapine, and –oxicam NSAIDS most frequently implicated
- 2) Presentation
  - a) “Influenza-like” prodrome 1-3 days prior to skin changes
  - b) Exanthematous rash may be initial symptom
  - c) Cutaneous findings:
    - i) Painful, coalescing, erythematous/purpuric macules or diffuse erythema; macules may have targetoid appearance
    - ii) Face and thorax first → widespread
    - iii) Vesicles and bullae form → skin sloughing
    - iv) Nikolsky sign: epidermal sloughing with lateral pressure on unblistered skin
  - d) Mucosal lesions:
    - i) Seen in majority of cases
    - ii) Can precede or follow the skin eruption
    - iii) Oral, ocular, and urogenital are most commonly involved locations
- 3) Diagnosis
  - a) Clinical
  - b) Laboratory abnormalities may include anemia, lymphopenia, elevated LFTs, hypoalbuminemia, electrolyte imbalance, elevated BUN, hyperglycemia
  - c) Skin biopsy may be beneficial for appropriate management and disposition
- 4) Management
  - a) Remove causative drug
  - b) Stabilize with supportive care; maintain normal body temperature; treat infection only if present
  - c) Admit all cases; most will require treatment in an ICU or burn unit
  - d) Consider treatment with cyclosporine early in the course of illness
  - e) Recurrence is possible; educate patient on avoiding causative drug for life.

## **Drug Reaction with Eosinophilia and Systemic Symptoms (DRESS) Syndrome**

- 1) Overview
  - a) Drug-induced hypersensitivity reaction
    - i) Typically occurs 2-8 weeks after drug exposure
    - ii) Allopurinol, anticonvulsants, sulfonamides, vancomycin, minocycline, and dapsone most frequently implicated
  - b) Potentially life-threatening because of internal organ failure and/or epidermal sloughing
  - c) Prolonged course with frequent relapses is common
- 2) Presentation
  - a) Prodrome – fevers, malaise, lymphadenopathy
  - b) Mucous membrane pain without ulceration
  - c) Morbilliform rash → diffuse, infiltrated erythema with follicular accentuation
  - d) DRESS suggested when eruption is  $\geq 50$  percent of the BSA and/or includes 2+ of:
    - i) Facial edema
    - ii) Infiltrated lesions
    - iii) Scaling
    - iv) Purpura
  - e) Exfoliative dermatitis & desquamation of palms/soles possible
  - f) Internal organ involvement in majority of patients
    - i) Liver, kidneys, and lungs are most commonly involved, but other organs may be affected
- 3) Diagnosis
  - a) High clinical suspicion
  - b) Lab evaluation to confirm diagnosis and evaluate visceral involvement
    - i) Eosinophilia on CBC differential & peripheral smear
    - ii) Skin biopsy: perivascular eosinophils and lymphocytes
    - iii) Obtain a CMP (possible LFT increase), UA (possible nephritic syndrome or eosinophils in sediment), EKG & troponin (evaluate for myocarditis)
- 4) Management
  - a) Remove causative drug
  - b) Admission – ICU or burn unit preferred
  - c) Systemic corticosteroids
    - i) Taper over 3-6 months to prevent relapse
  - d) Treat cutaneous lesions with high or super-high potency steroids
  - e) Cases may persist for months with relapses and remissions
  - f) Patients may develop autoimmune endocrine diseases months or years after resolution

## Necrotizing Fasciitis

- 1) Overview
  - a) Deep tissue infection causing progressive destruction of muscle fascia and overlying subcutaneous fat
  - b) Systemic toxicity develops rapidly
  - c) Associated with significant mortality
  - d) Risk factors: diabetes, drug use, obesity, immunosuppression, recent surgery, traumatic wounds
- 2) Pathophysiology
  - a) Type I
    - i) Mixed infection – aerobic and anaerobic
    - ii) Risk factors: diabetes, peripheral vascular disease, immunocompromised, recent procedure or surgery
  - b) Type II
    - i) Group A *Streptococcus* (GAS) or other beta-hemolytic streptococci
    - ii) *A. hydrophila* or *V. vulnificus* infection in a seawater-exposed injury
    - iii) Affects all ages and without specific past medical history
    - iv) Risk factors: skin injury, blunt trauma, recent surgery, childbirth, injection drug use, varicella
- 3) Presentation
  - a) Initially overlying tissue can appear unaffected
  - b) Early skin changes:
    - i) Erythema, warm, shiny
    - ii) Edema, induration
    - iii) Exquisitely tender with pain out of proportion to exam
  - c) Later skin changes:
    - i) Patches of red-purple to blue-gray
    - ii) Skin breakdown, bullae (pink/purple fluid), gangrene
    - iii) Skin becomes anesthetic
  - d) Subcutaneous gas may result in palpable crepitus
  - e) Triad of warning signs: rapid progression, pain out of proportion to exam, crepitus
- 4) Diagnosis
  - a) Have a low threshold for consideration of diagnosis
  - b) Diagnosis only confirmed with surgical exploration
  - c) Do not delay surgical debridement for diagnostics
  - d) Laboratory studies:
    - i) Laboratory studies alone are not predictive
    - ii) CBC with differential, CMP, ESR and/or CRP, CK, Lactic Acid, Blood Cultures x 2 sets, Gram Stain & Culture in OR
  - e) Imaging
    - i) Radiographs or noncontrast CT can accurately identify soft tissue gas

## 5) Treatment

- a) Surgical emergency – immediate surgical consult
- b) IV antibiotics
  - i) Initial antibiotic regimen:
    - (1) A carbapenem or beta-lactam-beta-lactamase inhibitor *plus*
    - (2) *MRSA* coverage - Vancomycin, daptomycin, linezolid, etc. *plus*
    - (3) Consider adding clindamycin for antitoxin effects
- c) Hemodynamic support
- d) Treat sepsis per guidelines
- e) Early surgical exploration and debridement of necrotic tissue
  - i) “Dishwater” fluid drainage & necrotic fascia are classic findings
  - ii) Aggressive debridement until only viable tissue remains
  - iii) Surgical site left open & regularly debrided in OR until necrosis ceases
- f) Antibiotic therapy alone is insufficient for treatment and is associated with high mortality
- g) Tailor antibiotics to gram stain, culture, and sensitivities when available
- h) Confirmed GAS treatment: Penicillin G & Clindamycin
- i) IV immune globulin in GAS complicated by toxic shock syndrome
- j) Continue antibiotics until no further debridement and hemodynamics normalized

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