Emergency Dermatology

Lindy P. Fox, MD
Assistant Professor
Director, Hospital Consultation Service
Department of Dermatology
University of California, San Francisco

Emergency Dermatology
• The "don't miss" fever and rash list
  – Infectious
    • Meningococcemia
    • Rocky Mountain Spotted Fever
    • Toxin-mediated erythemas (Staph Scalded Skin Syndrome and Toxic Shock Syndrome)
  – Drug reactions
    • Drug hypersensitivity syndrome
    • Stevens-Johnson Syndrome
    • Toxic epidermal necrolysis
  – Inflammatory
    • Vasculitis
    • Erythroderma
    • Pustular psoriasis
    • Pemphigus vulgaris
    • Pyoderma gangrenosum
    • Kawasaki disease

Clues are in the primary lesion and distribution
• Purpura
  – Vasculitis
  – Rocky Mountain Spotted Fever
  – Meningococcemia
• Bullae
  – Pemphigus vulgaris
  – Bullous pemphigoid
  – SJS/TEN
• Erythroderma
  – Drug hypersensitivity
  – Pustular psoriasis
  – Toxin mediated erythema
  – TEN
  – Kawasaki disease
• Ulcer
  – Pyoderma gangrenosum
• Acral
  – Rocky Mountain Spotted Fever
• Dependent
• Widespread
  – Drug hypersensitivity
  – Pustular psoriasis
  – TEN
  – Toxin Mediated Erythemas
• Mucosal
  – Pemphigus vulgaris
  – SJS/TEN
• Periorificial
  – Staphylococcal scalded skin syndrome

Case 1
• 42 y.o. HIV+ male brought to the ED
  • febrile
  • rash, rapidly progressive
  • skin is painful
  • gritty sensation in eyes
  • oral pain, difficulty swallowing
  • Severely hypotensive ➔ IV fluids, norepinephrine
  • Sepsis? ➔ antibiotics are started
  • At home has been taking Septra for UTI

Case 1, Question 1
The most likely diagnosis is:
A. Drug Eruption
B. Staphylococcal Scalded Skin Syndrome
C. Autoimmune Blistering Disorder
D. Toxic Shock Syndrome
E. Severe viral exanthem
Skin biopsy

- subepidermal blister
- epidermal necrosis
- sparse dermal inflammatory infiltrate
- Diagnosis: severe bullous drug eruption

Case 1, Question 2

What is the most important consult besides dermatology to get in a patient with SJS/TEN?
A. Renal
B. Ophthalmology
C. Allergy/immunology
D. Wound care
E. GI/liver

Emergency Dermatology: Bullae

1. SJS/TEN
2. Pemphigus vulgaris
3. Bullous pemphigoid

Cutaneous Drug Reactions - Immunologic mechanisms

- IgE dependent (TI)
  - Urticaria, angioedema, anaphylaxis
- Cytotoxic drug-induced reactions (TII)
  - Pemphigus, petechiae 2° drug-induced thrombocytopenia
- Immune complex-dependent (TIII)
  - Vasculitis, serum sickness, certain urticarias
- Delayed-type, cell-mediated (TIV)
  - Exanthematous, fixed, and lichenoid drug eruptions
  - Stevens-Johnson syndrome and toxic epidermal necrolysis

Urticarial Drug Eruption

- Immunologic
  - Mediated by IgE
  - Risk of anaphylaxis
  - Example: Penicillin
  - NEVER GIVE PCN to someone who gets "hives" from PCN
- Non-immunologic
  - Non specific mast cell degranulators
  - Example: opiates, contrast dye
  - OK to rechallenge (but premedicate)
Urticarial Drug Eruption

- Treatment
  - Antihistamines for simple urticaria
  - Anaphylaxis
    - H1 blocker (diphenhydramine)
    - H2 blocker (ranitidine, famotidine)
    - Epinephrine (IM or IV)
    - Methylprednisolone or dexamethasone
    - Cardiovascular support
  - MedAlert bracelet

Drug Eruptions: Degrees of Severity

Simple
- Morbilliform drug eruption
- Minimal systemic symptoms

Complex
- Drug hypersensitivity reaction
  - Stevens-Johnson syndrome (SJS)
- Toxic epidermal necrolysis (TEN)
- Systemic involvement
  - Potentially life threatening

Timing is everything: Drug charts

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PMH: h/o “UTI” self-treated with septra, h/o “drug rashes”

Common Causes of Cutaneous Drug Eruptions

- Antibiotics
- NSAIDs
- Sulfa
- Allopurinol
- Anticonvulsants

Morbilliform (Simple) Drug Eruption

- common
- erythematous macules, papules
- pruritus
- no systemic symptoms
- begins in 2nd week
- risk factors: EBV, HIV infection
- treatment:
  - D/C med if severe
  - symptomatic treatment:
    - diphenhydramine, topical steroids
Hypersensitivity Reactions

- Skin eruption associated with systemic symptoms and alteration of internal organ function
- Begins 2-6 weeks after medication started
  - time to abnormally metabolize the medication
- Classic culprits
  - Aromatic anticonvulsants THESE CROSS-REACT
    - phenobarbital, carbamazepine, phenytoin
  - Allopurinol
  - Dapsone
  - NSAIDs

Clinical features (General)

- Rash (morbilliform initially)
- Fever (precedes eruption by day or more)
- Pharyngitis
- Hepatitis
- Hematologic abnormalities
  - eosinophilia
  - atypical lymphocytosis
- Lymphadenopathy
- Facial edema

Cutaneous Features

- Clinical picture is often polymorphic
  - Rash begins as a morbilliform eruption
  - Edematous (vesicles, tense bullae)
  - Pustular
  - Erythroderma
- Face involved
  - Typically spared in morbilliform eruptions

Treatment

- Stop the medication
- Avoid cross reacting medications!!!!
  - Aromatic anticonvulsants cross react (70%)
    - Phenobarbital
    - Phenytoin
    - Carbamazepine
    - Valproic acid and Keppra generally safe
- Systemic steroids (Prednisone 1.5-2mg/kg) tapering dose over 1-3 months
- Allopurinol hypersensitivity may require other immunosuppressive therapy
  - E.g. Cellcept
  - NOT azathioprine (also metabolized by xanthine oxidase)
- Completely recover, IF the hepatitis resolves

Bullous Drug Reactions

- Stevens-Johnson Syndrome (SJS) and Toxic Epidermal Necrolysis (TEN) fall into this category
- Medications
  - Sulfonamides
  - Anticonvulsants
  - Allopurinol
  - NSAIDs

Stevens-Johnson (SJS) versus Toxic Epidermal Necrolysis (TEN)

<table>
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<tr>
<td>SJS/TEN overlap</td>
<td>10-30%</td>
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<tr>
<td>TEN “with spots”</td>
<td>&gt; 30%</td>
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<tr>
<td>TEN “without spots”</td>
<td>Sheets of epidermal loss &gt; 10%</td>
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Stevens-Johnson (SJS) versus Toxic Epidermal Necrolysis (TEN)

<table>
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<td>Mucosal membranes ≥ 2</td>
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<td><strong>Causes:</strong></td>
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<td>Drugs</td>
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**Stevens-Johnson Syndrome (SJS)**

- **Prodrome**
  - Fever, respiratory symptoms, headache, vomiting, diarrhea
- **Clinical morphology:**
  - Round macules and papules, red on the periphery and purple in the center (like a target)
  - Two or more mucous membranes (eyes, mouth, genitalia) involved
  - Can progress to resemble toxic epidermal necrolysis (TEN)

**SCORTEN**

- **Criteria**
  1. Age > 40 yrs
  2. Presence of malignancy
  3. BUN > 27 mg/dL
  4. Glucose >252 mg/dL
  5. Pulse > 120 bpm
  6. Bicarbonate <20mEq/l
  7. BSA > 10%
- **Mortality rates**
  - 0-1 3.2%
  - 2 12.2%
  - 3 35.3%
  - 4 58.3%
  - ≥5 90%

**Toxic Epidermal Necrolysis (TEN)**

- Life threatening blistering reaction
- Early on, patients complain of skin pain
- Skin becomes red, then develops bullae that slough to reveal denuded dermis
  - Nikolsky sign present
- Medical emergency- call dermatology immediately

**SJS/TEN: Emergency Management**

- Stop all unnecessary medications
  - The major predictor of survival and severity of disease
- **Treatment**
  - Topical
    - Aquaphor and Vaseline gauze
  - Systemic
    - Consider antivirals
    - Check for Mycoplasma- 25% of SJS in pediatric patients
    - Controversial
      - SJS: high dose corticosteroids
      - TEN: IVIG 0.5-1g/kg/d x 4d
      - Refer to burn unit early
      - Reduces risk of infection and reduces mortality to 5%
  - Call Ophthalmology

**Pathogenesis of TEN**

- Normal skin
  - Express Fas (CD95)
- TEN
  - Induction of Fas L → Fas: Fas L binding induces widespread apoptosis of keratinocytes

IVIG (intravenous immunoglobulin) as a treatment for TEN

Human IVIG has antibodies against Fas L

IVIG blocks Fas mediated apoptosis in vitro & Arrests development of TEN in vivo


IVIG for TEN

Dose and Response

- Recommended dose: 0.5-1.0g/kg/d over 3-5 days
- Arrest in disease progression in 24-48 hours
- Complete re-epithelialization within 4-10 days
- Decreases mortality?*
  - Decreases to 6-12% in some studies
  - Other studies report increased mortality
- 7 of 8 studies (non-controlled clinical studies with ≥ 10 pts)
  - Overall mortality benefit of IVIG in doses > 2g/kg
- Risk factors for failing to respond to IVIG**
  - Delayed use of IVIG (≥ day 10), lower dose (2g/kg total), underlying chronic diseases, higher BSA involved (>65%), older age
- Also batch-to-batch variation in anti-Fas activity

*Semin Cutan Med Surg 2006. 25:91-3
^ Allergology Int 2006. 55: 9-16
**Arch Derm 2003. 131:26-32

Bullous Drug Reactions: Supportive Care

- Protect exposed skin
- Prevent and treat secondary infection (sepsis)
- Monitor fluid and electrolyte status
- Nutritional support
  - Hyperglycemia assoc with increased morbidity/mortality
- Warm environment
- Refer to burn unit early
  - Reduces risk of infection and reduces mortality to 5%
- Respiratory care
- Ophthalmology consult
- Death (up to 25% of patients with more than 30% skin loss, age dependent)

Signs of a Serious Cutaneous Drug Eruption

- Cutaneous
  - Facial involvement
  - Confluent erythema
  - Skin pain
  - Epidermal detachment
  - Nikolsky sign
  - Mucous membrane involvement
- Systemic
  - High fever
  - Lymphadenopathy
  - Arthralgias/arthritis
  - Shortness of breath, wheezing, hypotension
- Laboratory
  - Eosinophilia
  - Lymphocytosis with atypical lymphocytes
  - Elevated liver function tests
  - Renal failure

Bullous Pemphigoid

- Most common autoimmune bullous disease
- Favors elderly (65-75)
- Unilocular, tense, bullae, some on erythematous base
- Bullae usually large (>1 cm)
- Favors inner arms, thighs, and flanks
- 1/3 of patients have oral erosions
- Diagnosis: Biopsy for histology and direct immunofluorescence

Drug Induced Bullous Pemphigoid

- Drug “unmasks” patients predisposition to develop BP?
- Drugs
  - Penicillamine
  - Furosemide
  - Captopril, enalapril
  - Penicillin
  - Sulfasalazine
  - Nalidixic acid
  - Beta blockers
Pemphigus Vulgaris

- Elderly
- Widespread, larger friable blisters, erosions
- 50% present with oral erosions and 100% develop oral lesions at some time
- Flaccid blisters anywhere on the skin
- Blisters do not heal, but leave very painful erosions up to 10 cm in diameter
- Gradually worsening, progressive course in most patients
  - Until prednisone became available, considered a fatal disease
- Treated with systemic immunosuppressants

Images courtesy of Siegrid Yu, MD

Case 2

- 37 yo man with hepatitis C infection presents with fever, joint pain, and rash
- A skin biopsy confirms leukocytoclastic vasculitis

Case 2, Question 1

In this patient, the test most likely to be abnormal is:
A. Antinuclear antibody
B. Rheumatoid factor
C. Cryoglobulins
D. Urinalysis
E. Stool guaiac

Case 2, Question 1

In this patient, the test most likely to be abnormal is:
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Emergency Dermatology: Purpura

1. Vasculitis
2. Rocky Mountain Spotted Fever
3. Meningococcemia
### Leukocytoclastic Vasculitis

**Etiology**
- Conditions associated with LCV
  - Idiopathic (45-55%)
  - Infection (15-20%)
  - Inflammatory diseases (15-20%)
  - Medications (10-15%)
  - Malignancy (<5%)
  - Other
    - Hypergammaglobulinemic purpura of Waldenström
    - HIV
    - Cocaine use (p-ANCA +)

**Differential Diagnosis**
- Infection
  - Post strep GN
  - Hepatitis B
  - SBE
- Hypersensitivity
  - Henoch-Schönlein purpura
  - Serum sickness
  - Medication
- Rheumatic disease
  - SLE
  - RA
  - Sjögren’s syndrome
- Mixed cryoglobulinemia (HCV)
- Malignancy associated
  - CLL
  - Multiple myeloma
  - Lymphoma
  - Hodgkin’s disease
- ANCA associated vasculitis
  - Wegeners granulomatosis
  - Microscopic polyangiitis

**Clinical Presentation**
- Degree of purpura increases from cephalad to caudad
- Favors dependent areas (lower legs)
- May itch, sting, or burn
- Associated symptoms: fever, malaise, arthralgias/arthritis
- May affect blood vessels in many organs
  - kidneys, joints, and gut most frequently

**Evaluation**
- H+P, including medications and ROS
- Skin biopsy for H+E, DIF
- Lab tests (initial):
  - Blood culture
  - CBC with differential
  - Urinalysis with micro
  - Creatinine
  - Stool guaiac
  - ASO, throat culture
  - Hepatitis B, C serologies
  - ANA, Complement, ANCA
  - Cryoglobulins
  - SPEP/IFE

**Treatment**
- Treat underlying cause
- First line
  - NSAIDS
  - Colchicine 0.6 mg BID
  - Dapsone 50-100 mg BID
  - Prednisone (60-80mg/day) for short course
- Second line
  - Mycophenolate mofetil, methotrexate, azathioprine, cyclophosphamide
Meningococcemia

- **Organism:** *N. meningitidis*
- Skin lesions typically associated with acute sepsis
- Acutely ill
- Widespread eruption
  - petechiae
  - palpable purpura
  - stellate, gunmetal gray
- Can progress to DIC/purpura fulminans

Higher risk:
- Military recruits
- Close contact with an index case
- Travel to an endemic area
- Asplenia
- College students living in dormitory

Diagnosis
- Culture blood, skin, CSF
- Skin lesions demonstrate organism in 70% cases
- Latex agglutination tests
  - Group A,B,C,Y, and W-135 antigens in CSF and urine

Treatment
- Penicillin
- Chloramphenicol
- Ceftriaxone

Rocky Mountain Spotted Fever

- **Organism:** *Rickettsia rickettsii*
- Tick: *Dermacentor or Ixodes*
- Summer, early fall
- Tick bite typically painless
- Incubation period: 6-8 days
- Initial symptoms:
  - Flu-like syndrome: fever, chills, HA, myalgia, malaise
  - GI symptoms: nausea, vomiting, diarrhea, abdominal pain
- Cutaneous lesions begin 2-4 days after fever

Diagnosis
- Laboratory tests non-specific
  - Normal CBC or leukocytosis, leukopenia, anemia (5-25%)
  - Thrombocytopenia (30-50%)
  - Hyponatremia common
  - Elevated LFTs, bil, CK, LDH
  - Skin biopsy- organisms present in vessels
- Serology
- Mortality (untreated) 20-25%

Treatment
- Doxycycline
- Chloramphenicol

RMSF: cutaneous eruption

- Erythematous macules- wrists and ankles
- Lesions develop central petechiae
- Spreads centripetally
- Involves trunk, extremities, palms, soles; spares face
Case 3

- 55 yr old male
- COPD, HTN, non-small cell lung cancer and mild psoriasis
- Presents with low grade fever, shaking chills, and diffuse erythema (erythroderma)
- Meds:
  - ACE inhibitor x 3 months
  - 1 week of pulsed prednisone with rapid taper for COPD flare

Case 3, Question 1
The most likely diagnosis is:
A. Drug eruption due to ACE inhibitor
B. Paraneoplastic syndrome due to non-small cell lung cancer
C. Sézary syndrome (cutaneous T-cell lymphoma)
D. Flare of psoriasis due to prednisone taper
E. Staphylococcal Scalded Skin Syndrome

Case 5, Question 1
The most likely diagnosis is:
A. Drug eruption due to ACE inhibitor
B. Paraneoplastic syndrome due to non-small cell lung cancer
C. Sézary syndrome (cutaneous T-cell lymphoma)
D. Flare of psoriasis due to prednisone taper
E. Staphylococcal Scalded Skin Syndrome

Emergency Dermatology: Erythroderma

1. Pustular psoriasis
2. Toxin mediated erythemas
3. Kawasaki disease
4. Drug eruptions (hypersensitivity, TEN)

Pustular Psoriasis

- Often occurs when known psoriatrics are given systemic steroids
- When the steroids are tapered, the psoriasis flares, often with pustules
- Can be life threatening
  - High cardiac output state
  - Electrolyte imbalance
  - Respiratory distress
  - Temperature dysregulation
Psoriasis Aggravators

- Medications
  - Systemic steroids
  - Beta blockers
  - Lithium
  - Hydroxychloroquine
- Strep infections
  - Guttate psoriasis in children
- Trauma
- Sunburn
- Severe life stress
- HIV
  - Up to 6% of AIDS patients develop psoriasis
- Alcohol for some
- Smoking for some

Treatment for Psoriasis

- Topical therapy
  - Steroid ointment (start mid-potency)
  - Calcipotriene (Dovonex)
  - Tar
- Phototherapy- refer to dermatologist
  - Broadband UVB or Narrowband UVB
  - PUVA: psoralens + UVA
- Systemic therapy- refer to dermatologist
  - Acitretin (oral retinoid)
  - Methotrexate, cyclosporine
  - Biologics
    - etanercept, infliximab, adalimumab, alefacept, efalizumab
**Systemic steroids are NOT on this list!**

Toxin Mediated Erythemas

- Staphylococcal Scalded Skin Syndrome
- Streptococcal Toxic Shock Syndrome
- Staphylococcal Toxic Shock Syndrome

Staphylococcal Scalded Skin Syndrome

- Caused by Staphylococcal exfoliative exotoxins A and B of Phage group II strains 55, 71
- Most common in children < 6 years of age
- Rare in adults unless immunosuppressed (HIV) or renal failure (can’t clear toxin, which is renally excreted)
- Mortality
  - Children 3-4%, adults >50%

Staphylococcal Scalded Skin Syndrome

- Prodrome
  - Fever, malaise, irritability, severe skin tenderness
- Erythema begins in head and neck area, then rapidly progresses to the rest of the body
- Flaccid bullae develop, giving the skin a wrinkled appearance
- 1-2 d later, bullae are sloughed, leaving moist skin, sometimes a yellow crust is present
- Exfoliation begins in the flexural areas
- Perioral crusting and fissuring is common
- Re-epithelialization without scarring occurs in 10-14 days

Staphylococcal Scalded Skin Syndrome

- Diagnosis
  - Clinical
  - Culture any suspected site of infection
    - Skin foci- pustule, furuncle, erosions, etc
    - Intact bullae will be culture negative (unlike bullous impetigo)
    - Conjunctiva, nasopharynx, feces
    - Blood cultures
      - Typically negative in children, can be positive in adults.
  - Skin biopsy (to differentiate from TEN)
- Treatment
  - Admit
  - β-lactamase-resistant antibiotic (dicloxacillin, cephalaxin)
  - Addition of clindamycin can help clear the toxin
  - Neonates need isolation to avoid outbreaks in other neonates
Streptococcal Toxic Shock Syndrome
• Criteria
  • Isolation of group A streptococci from normally sterile site OR
  • Isolation of group A streptococci from non-sterile site
  • AND
  • Hypotension (SBP<90mmHg for adults)
  • AND
  • Two or more
    – Renal impairment
    – Coagulopathy (platelets < 100000/mm³ or DIC)
    – Elevated LFTs
    – ARDS
    – Generalized erythematous macular rash +/- desquamation
    – Soft tissue necrosis (necrotizing fasciitis, myositis, gangrene)

Streptococcal Toxic Shock Syndrome
• Due to exotoxin producing strains of Group A, β-hemolytic streptococcus (S. pyogenes)
• Affects healthy people, ages 20-50
• Skin portal of entry 80%
• 50% have necrotizing fasciitis
• Mechanism of disease
  – Streptococcal M proteins and exotoxins act as “superantigens”
  – Bind to MHC class II APCs and T cell receptors
  – Leads to T cell activation, cytokine induction (TNF-α, IL-1, IL-6)

Staphylococcal Toxic Shock Syndrome
• Historically associated with menstruating women and tampon use in 1980s
• Currently most commonly seen
  – after surgical procedure, with focal pyodermas or deep abscesses, postpartum, nasal packing, insulin pump infusion site
• Due to infection or colonization with strain of S. aureus that produces toxic shock syndrome toxin-1 (TSST-1)
• TSST-1
  – Acts as a superantigen
  – Is directly toxic to organs
  – Impairs clearance of endogenous exotoxins derived from gut flora
Staphylococcal Toxic Shock Syndrome

- Sudden onset high fever, myalgias, vomiting, diarrhea, headache, pharyngitis
- Rapid progression to shock
- Diffuse scarlatiniform exanthem
  - Starts on trunk and spreads to extremities
  - Erythema and edema of palms and soles
  - Erythema of mucous membranes
    - Strawberry tongue, conjunctival erythema
  - 1-3 weeks later, desquamation of hands and feet

Staphylococcal Toxic Shock Syndrome

- Diagnosis
  - High index of suspicion
  - Criteria

- Treatment
  - Admit
  - Supportive care (IV fluid, pressors)
  - Remove packing, etc
  - IV antibiotics
  - Clindamycin
  - IVIG

Kawasaki Disease Criteria

- Fever > 39.6°C
- Rash- diffuse macular erythroderma
- Desquamation: 1-2 weeks after the onset of the illness (typically palms and soles)
- Hypotension (SBP<90 mmHg for adults)
- Involvement of 3 or more of the following organ systems
  - GI, muscular, CNS, renal, hepatic, mucous membranes (erythema), hematologic (platelets < 100000/mm³)
- Lack of evidence for another cause
  - Blood, throat, CSF cultures negative
  - Serologies for RMSF, leptospirosis, measles negative

Kawasaki Disease

- Most severe complication is cardiac
  - Coronary artery aneurysms (10%)
  - EKG changes (PR, QT prolongation; ST, T wave changes)
  - Angina, myocardial infarction

Kawasaki Disease

- Laboratory findings
  - Leukocytosis, anemia, elevated ESR, sterile pyuria
  - Thrombocytosis
    - Highest in second week, same time as highest risk of coronary artery thrombosis

- Echocardiogram: coronary artery aneurysms

Clin Exp Dermatol. 2001; 26
Kawasaki Disease

• Treatment
  – IVIG
    • 2g/kg single infusion
  – Aspirin
    • Must be given within 10d of fever onset
    • 80-100mg/kg/d during acute febrile phase, then decrease to
      3-5mg/kg/d after fever subsides

• Prognosis (untreated)
  – 75% resolution without sequelae
  – 25% abnormal coronary arteries with 1-2% mortality
    in acute phase
  – Leading cause of acquired heart disease in children
  – Risk factor for adult ischemic heart disease and
    sudden death in young adults

Kawasaki Disease-Like Syndrome (KDLS) in HIV

• Reported in 13 patients
  – 11 adults
  – 2 children
• Moderate-to-severe immune dysfunction
  (CD4 10-298 cells/mm²)

Kawasaki Disease-Like Syndrome (KDLS) in HIV- Clinical Features

• Classic KD
  – Fever ≥ 5 days
  – Conjunctivitis
  – Exanthem
  – Cervical LAD
  – Hand/foot edema
  – Oropharyngeal changes

• KDLS of HIV
  – More GI complaints
  – Less prominent cervical LAD
  – Laboratory parameters may be normal
    • ESR or CRP
    • Platelet count
  – Coronary artery aneurysm not reported

Kawasaki Disease-Like Syndrome in HIV- Course and Treatment

• Similar therapies as used in classic KD
  – Aspirin: start at 80mg/kg/d X 2 weeks
  – Pooled IVIG: 2g/kg over 10-12 hours
• Initiate or optimize HAART
• Untreated, course similar to classic KD
• Higher rate of relapse

http://www.pediatrics.ucsd.edu/kawasaki/kdhiv.asp

Case 4

• 37 yo woman with inflammatory bowel disease
• Rapidly progressive, painful ulceration of lower leg appears 3 days after bumping her leg on a chair
Case 4, Question 1

- The most appropriate treatment for this disorder is
  A. Systemic steroids
  B. Intravenous antibiotics
  C. Surgical debridement
  D. Compression dressing
  E. Wet to dry dressings

Pyoderma Gangrenosum

- Rapidly progressive (days) ulcerative process
- Begins as a small pustule which breaks down forming an ulcer
- Undermined violaceous border
- Expands by small peripheral satellite ulcerations which merge with the central larger ulcer
- Occur anywhere on body
- Triggered by trauma (pathergy) (surgical debridement, attempts to graft)

Case 4, Question 2

- All of the following underlying diseases are strongly associated with this condition except:
  A. Rheumatoid arthritis
  B. Inflammatory bowel disease
  C. Acute myelogenous leukemia
  D. IgA monoclonal gammopathy
  E. Tuberculosis infection

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Pyoderma Gangrenosum

- Most cases have no underlying cause
- Associations:
  - Inflammatory bowel disease (1.5%-5% of IBD patients get PG)
  - Rheumatoid arthritis
  - Seronegative arthritis
  - Hematologic abnormalities

Pyoderma Gangrenosum Treatment

- AVOID DEBRIDEMENT
- Refer to dermatology
- Treatment of underlying disease may not help PG
  - Topical therapy:
    - Superpotent steroids
    - Topical tacrolimus (up to .3%)
  - Systemic therapy:
    - Systemic steroids
    - Cyclosporine or Tacrolimus
    - Celcipt
    - Thalidomide
    - TNF-blockers (Remicade)

The end. (whew!)