SKIN EMERGENCIES???

• Subclassifications:
  – Autoimmune
    (Anaphylaxis, Vasculitis, Pemphigus)
  – Erythroderma
    (AGEP, DRESS, SJS, TEN)
  – Infectious
    (Fournier’s, Immunocompromised –crypto, mucor, zoster, Kawasaki’s, Loxosceles, Necrotizing Fascitis, Rocky Mountain Spotted Fever, Staph-MRSA, TSS)
Approach to the patient

• Presentation- acute, ill appearing
• History
  - all medications taken including OTC
  - time course
  - systemic symptoms
• Skin lesions
• Diagnostic testing
When to worry…

• **Cutaneous:**
  - erythroderma
  - facial involvement
  - mucous membrane involvement
  - skin tenderness
  - purpura

• **Systemic symptoms**
  - fever/B symptoms
  - lymphadenopathy
AUTOIMMUNE CAUSES

- Anaphylaxis
- Vasculitis
- Pemphigus
AUTOIMMUNE CAUSES-
Immunologically Mediated

- Type I (IgE dependent)
  - anaphylaxis, urticaria, angioedema
- Type II (Cytotoxic)
  - pemphigus, thrombocytopenia
- Type III (Immune complex)
  - serum sickness, vasculitis
- Type IV (Delayed-type)
  - lichenoid, fixed, photoallergy
Anaphylaxis

- Type I hypersensitivity
- Skin (urticarial and/or angioedema) plus hypotension and tachycardia
- Causes: PCN, latex
- Treatment: epinephrine, corticosteroids
Angioedema

- Edema of deep dermal, subcutaneous, or submucosal tissues
- Pale or pink subcutaneous swelling
- **ACE inhibitors**, PCN, NSAIDS, contrast media, monoclonal antibodies
- Tx: stop offending med, No ARBs
Vasculitis

- Immune complex (type 3 reaction)
- 3 types - small vessel, medium vessel, large vessel
- Most common type: leukocytoclastic vasculitis
<table>
<thead>
<tr>
<th>CHAPEL HILL CONSENSUS CLASSIFICATION</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Large-vessel vasculitis</strong></td>
</tr>
<tr>
<td>• Giant cell arteritis</td>
</tr>
<tr>
<td>• Takayasu’s arteritis</td>
</tr>
<tr>
<td><strong>Medium-vessel vasculitis</strong></td>
</tr>
<tr>
<td>• Classic polyarteritis nodosa</td>
</tr>
<tr>
<td>• Kawasaki disease</td>
</tr>
<tr>
<td><strong>Small-vessel vasculitis</strong></td>
</tr>
<tr>
<td>• Wegener’s granulomatosis</td>
</tr>
<tr>
<td>• Churg–Strauss syndrome</td>
</tr>
<tr>
<td>• Microscopic polyangiitis (polyarteritis)</td>
</tr>
<tr>
<td>• Henoch–Schönlein purpura</td>
</tr>
<tr>
<td>• Essential cryoglobulinemia</td>
</tr>
<tr>
<td>• Cutaneous leukocytoclastic vasculitis</td>
</tr>
</tbody>
</table>
Table 26.3 Physical examination findings leading to suspicion of vasculitis.

<table>
<thead>
<tr>
<th>Examination</th>
<th>Findings</th>
</tr>
</thead>
<tbody>
<tr>
<td>Vital signs</td>
<td>Fever, weight loss, hypertension</td>
</tr>
<tr>
<td>Cutaneous</td>
<td>Palpable purpura, urticaria, nodules, necrotizing livedo reticularis, superficial thrombophlebitis, ulcerations</td>
</tr>
<tr>
<td>Neurologic</td>
<td>Palpable cutaneous nerves</td>
</tr>
<tr>
<td>Head and neck</td>
<td>Sinusitis, chondritis, otitis, iritis</td>
</tr>
<tr>
<td>Renal</td>
<td>Tenderness to palpation</td>
</tr>
<tr>
<td>Liver</td>
<td>Hepatosplenomegaly</td>
</tr>
<tr>
<td>Pulmonary</td>
<td>Crackles</td>
</tr>
</tbody>
</table>

• History is key- ask about medications (PCN, NSAIDs, sulfas, cephalosporins)
• Workup for systemic involvement
• Treatment: eliminate cause, treat infection, steroids, colchicine, immunosuppressants
PEMPHIGUS

- Autoimmune blistering disease affecting skin and mucous membranes
- 5 types
Table 31.1 Classification of pemphigus.

<table>
<thead>
<tr>
<th>CLASSIFICATION OF PEMPHIGUS</th>
</tr>
</thead>
<tbody>
<tr>
<td>• Pemphigus vulgaris</td>
</tr>
<tr>
<td>• Pemphigus vegetans</td>
</tr>
<tr>
<td>• Pemphigus foliaceus</td>
</tr>
<tr>
<td>• Pemphigus erythematous: localized</td>
</tr>
<tr>
<td>• Fogo selvagem: endemic</td>
</tr>
<tr>
<td>• Drug-induced pemphigus</td>
</tr>
<tr>
<td>• Paraneoplastic pemphigus</td>
</tr>
<tr>
<td>• IgA pemphigus</td>
</tr>
</tbody>
</table>

PEMPHIGUS

- Work-up - drug cause, malignancy
- Treatment: initially prednisone then add long term immunosuppressant
AUTOIMMUNE: REVIEW

- Anaphylaxis
- Vasculitis
- Pemphigus
ERYTHRODERMA CAUSES

- AGEP
- DRESS
- SJS
- TEN
AGEP

• Onset-days
• Multiple small nonfollicular sterile pustules on trunk, UE, intertriginous areas
• High fever, edema of face and hands, neutrophilia
• Beta-lactams, macrolides, CCB, antimalarials
• Tx: stop drug, supportive
Drug rash with eosinophilia and systemic symptoms (DRESS)

- “Hypersensitivity syndrome”
- Onset: 2-6 weeks after
- Inability to detoxify toxic 
  arene oxide metabolites
- Anticonvulsants
  (phenytoin, carbamazepine, phenobarbital), lamotrigine, sulfonamides, allopurinol, dapsone
DRESS

• Fever, rash, **facial edema**
• Peripheral blood eosinophilia
• Hepatitis- may be fulminant
• Other: myocarditis, interstitial pneumonitis, interstitial nephritis, thyroiditis, CNS infiltration of eosinophils.
• Tx: Systemic Corticosteroids- takes weeks to months of therapy
# Table 23.5 Characteristics of major drug-induced eruptions.
*Also referred to as hypersensitivity syndrome. **Nonpigmenting.

<table>
<thead>
<tr>
<th>Clinical presentation</th>
<th>Percentage that are drug-induced (%)</th>
<th>Time interval</th>
<th>Mortality (%)</th>
<th>Responsible drugs</th>
</tr>
</thead>
<tbody>
<tr>
<td>Exanthematous eruption</td>
<td>Child: 10–20 Adult: 50–70</td>
<td>4–14 days</td>
<td>0</td>
<td>Aminopenicillins Sulfonamides Cephalosporins Anticonvulsants Allopurinol</td>
</tr>
<tr>
<td>Urticaria Anaphylaxis</td>
<td>&lt;10</td>
<td>Minutes Hours</td>
<td>0</td>
<td>Penicillins Cephalosporins NSAIDs Monoclonal antibodies Contrast media</td>
</tr>
<tr>
<td>Fixed drug eruption</td>
<td>100</td>
<td>&lt;48 hours</td>
<td>0</td>
<td>TMP-SMX NSAIDs Tetracyclines Pseudoephedrine**</td>
</tr>
<tr>
<td>Acute generalized exanthematous pustulosis (ACEP)</td>
<td>70–90</td>
<td>&lt;4 days</td>
<td>1–2</td>
<td>-Lactam antibiotics Macrolides Calcium channel blockers</td>
</tr>
<tr>
<td>Drug reaction with eosinophilia and systemic symptoms (DRESS)*</td>
<td>70–90</td>
<td>15–40 days</td>
<td>5–10</td>
<td>Anticonvulsants Sulfonamides Allopurinol Minocycline</td>
</tr>
<tr>
<td>Stevens-Johnson syndrome (SJS)</td>
<td>70–90</td>
<td>7–21 days</td>
<td>5</td>
<td>Sulfonamides Anticonvulsants NSAIDs Allopurinol</td>
</tr>
<tr>
<td>Toxic epidermal necrolysis</td>
<td></td>
<td></td>
<td>30</td>
<td></td>
</tr>
</tbody>
</table>

EM/SJS/TEN

- Erythema Multiforme Minor (EM)
- Erythema Multiforme Major (Stevens-Johnson syndrome)
- Toxic Epidermal Necrolysis
Erythema Multiforme

- 1860- von Hebra
- 1950- Bernard Thomas
  minor vs major
- Causes: **HSV**, orf, histoplasmosis, ?EBV
- HSV infection m/c precedes EM
- Increased outbreaks in immunosuppressed
Erythema Multiforme

Clinical Features:
• Prodrome HSV
• “Target” lesion with concentric rings with dusky center (bulla) and outer red zone
• Pruritis, burning
• Dorsum hands, forearms, palms, neck, face, trunk
• Koebner
Erythema Multiforme

- Dx: clinically
- Path: r/o vasculitis, LE
- Tx: symptomatic prophylaxis
- Acyclovir not helpful for EM after lesions appear
Stevens-Johnson Syndrome

- EM Major
- 1922 Stevens and Johnson
- Continuum with Toxic Epidermal Necrolysis
- Peaks 2\textsuperscript{nd} decade, spring and summer
Stevens-Johnson Syndrome

Causes:
* Drugs- **NSAIDs**, sulfonamides, anticonvulsants, PCN, TCN, doxycycline (epoxide hydrolase deficient)
* Infections- Mycoplasma, histococcidio, viral
Stevens-Johnson Syndrome

Clinical Features:
- Prodrome URI
- 1-14 days: symmetric red macules, vesicles/bulla, epidermal necrosis
- 2 or more mucosal sites (always oral)
- Systemic symptoms
Stevens-Johnson Syndrome

- DDx: TEN, Kawasaki, PNP, GVHD
- Treatment: ICU
  - stop drugs
  - ophtho exam
  - fluid/electrolytes
  - infection
  - GI-strictures
  - GU
- IVIG
Toxic Epidermal Necrolysis

- Lyell’s syndrome
- At risk: women, elderly, slow acetylates, immunocompromised
- Mortality 25-50%
- Almost always drug related
Toxic Epidermal Necrolysis

- Associated Meds:
  Antibiotics (sulfa), NSAIDs, anticonvulsants

- Risk highest during 1st week (typically within 1-3 weeks).
Toxic Epidermal Necrolysis

S/S:

- High fever, skin pain, anxiety, asthenia
- Erythematous, dusky macules, coalescing, progressing to full thickness necrosis with bulla formation and detachment
- (+) Nikolsky
Toxic Epidermal Necrolysis

Diagnosis:
- SJS: <10% BSA
- TEN: >30% BSA
- Overlap exists
- Scorten

Pathology
- Bx: r/o SSSS, AGEP
Table 23.7 SCORTEN. It represents a prognostic scoring system for patients with TEN. The score is based upon the number of prognostic factors in a particular patient. BSA, body surface area.

<table>
<thead>
<tr>
<th>Prognostic factors</th>
<th>Points</th>
</tr>
</thead>
<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>
<td>1</td>
</tr>
</tbody>
</table>

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

Toxic Epidermal Necrolysis

Treatment:

• Withdrawal medications
• Prevention of complications
• **IVIG** - antibodies against Fas, **blocks binding of FasL to Fas**
ERYTHRODERMA: REVIEW

- AGEP
- DRESS
- SJS
- TEN
INFECTIOUS CAUSES

- Fourniers gangrene
- Immunocompromised (crypto, mucor, zoster)
- Kawasaki’s
- Loxosceles
- Menningococcemia
- Necrotizing fascitis
- Rocky Mountain Spotted Fever
- Staph- MRSA, TSS
FOURNIER’S GANGRENE

• Necrotizing fascitis of genitalia
• Middle-aged men
• Cause: predisposing trauma, mixed bowel organisms
• Treatment: supportive, antibiotics, surgery
IMMUNOCOMPROMISED

• 2 Groups:
  HIV/AIDS (Tcell < 50)

• IMMUNOSUPPRESSED
  (ANC-<1000, <500, <100)

• Discuss:
  Cryptococcus
  Mucormycosis
  Herpes Zoster
CRYPTOCOCCUS

- Cryptococcus neoformans
- Soil, pigeon droppings
- CNS (meningitis), pulmonary, skin (mimics molluscum)
- Cutaneous disease preceds CNS infection
MUCORMYCOSIS

- Mucorales - Mucor, Rhizopus, Absidia and Cunninghamella
- Diagnosis: biopsy
- Treatment: surgery, IV antifungals
HERPES ZOSTER

• Varicella-zoster virus, herpes type 3
• Varicella pneumonia, encephalitis, hepatitis, purpura fulminans
• Tx- IV acyclovir
• Prophylaxis
KAWASAKI DISEASE

• “Mucocutaneous lymph node syndrome”
• Cause: unknown, ? Bacteria/viral
• Fever, conjunctivitis, rash, “strawberry tongue”, lymphadenopathy
KAWASAKI DISEASE

- Risk factors:
  age <5, boys, Asian
- Concern: coronary aneurysms, myocarditis, dysrhythmias
- Diagnosis: exclusion
- Treatment: ASA, gamma globulin
LOXOSCELES

- Loxosceles reclusa
- “Brown recluse”
- Erythema, edema, necrosis
- Systemic symptoms-loxoscelism include HA, fever, n/v/d, rash, hypotension, shock, DIC
NECROTIZING FASCITIS

- Bacterial infection (usually mixed)
- Sudden onset of symptoms - painful skin/edematous skin, fever, n/v/d
- LRINEC score
- Tx: surgery, antibiotics, support
ROCKY MOUNTAIN SPOTTED FEVER

- Rickettsia rickettsii
- Transmitted by tick
- Incubation: 2-14 days
- Fever, HA, myalgia/arthralgia, rash
- Mortality: 70% without tx
- Tx: Doxycycline
STAPH- MRSA/TSS

• Staphylococcus aureus
• MRSA: skin-boils/abcesses, bones, joints, blood, heart valves, lungs
• TSS: fever, n/v/d, HA, pharyngitis, myalgia, hypotension, exanthenm
INFECTIOUS CAUSES: REVIEW

- Fournier's gangrene
- Immunocompromised (crypto, mucor, zoster)
- Kawasaki’s
- Loxosceles
- Meningococcemia
- Necrotizing fascitis
- Rocky Mountain Spotted Fever
- Staph- MRSA, TSS
REVIEW

• 3 categories: autoimmune, erythroderma, infectious
• How to approach the patient
• Work-up and treatment
THE END