Dermatologic Emergencies

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Pemphigus Vulgaris (PV)

- Serious bullous autoimmune disease
- Involves skin and mucous membranes
- Often fatal unless treated with steroids + immunosuppressive drugs
- Age: 40 to 60
- Etiology: autoimmune
- Can start in mouth then skin or generalized acute eruption of both
- Painful mouth erosions (may affect food intake)
PV

- Skin Lesions: Flaccid bullae easily ruptured † erosions, bleeding & crustation
- Nikolysky’s sign: Positive
- Skin Biopsy:
  Intraepidermal blister with acantholysis (separation of keratinocyte from each other)
- Immunofluorescence; (IF)
  (Patient skin) Direct: IgG & C3 positive at intercellular space in epidermis
  (Patient serum) Indirect: autoantibody IgG +Ve against desmoglein III
PV

Course: - Can be fatal unless treated aggressively with immunosuppressive agents

Variants:
- Pemphigus Vegetans: Intertiginous areas with vegetating lesions
- Pemphigus Foliaceous: Erythematous patches and erosions covered with crustations
- Pemphigus erythematosus: erythematous crusted erosive lesions in the butterfly area of face, forehead & chest, ANA + Ve
PV

- Drug Induced pemphigus
  D- Pencillamine
  Captopril
- Paraneoplastic pemphigus: Associated with malignancy

Management:
- Correction Of Fluid & electrolyte loss
- Treatment of infection by antibiotics
- Systemic steroids (high dose)
PV

- immunosuppressive therapy (given with steroids)
- Azathiopurine
- Methotrexate
- Cyclophosphamide
- Plasmapharesis
Bullous Pemphigoid (BP)

Age: 60 to 80 yrs
Often starts as urticarial lesions ‡ bullae
or directly as bullous eruption
Skin findings:
  - Large tense intact bullae
Involvement:
  - skin
  - Mucous membrane (less than pemphigus)
Skin Biopsy:
  - Subepidermal blisters with eosinophils
BP

Immunofluorescence:
- Direct: Linear deposition of IgG & C3 (along the basement membrane)
- Indirect: autoantibodies against BP Ag1 & BP Ag2

Management:
- Superpotent topical steroids (for mild cases)
  - Systemic steroids
  - + Immunosuppressive agents

Prognosis: Better than PV
Compare PV & BP?

- Morphology
- Mucous membrane involvement
- Skin biopsy & IF
- Prognosis.
Steven’s Johnson Syndrome and Toxic Epidermal Necrolysis (SJS and TEN)

Etiology:
  TEN: mostly due to drug
  SJS: due to drugs or infections (commonly mycoplasma)

Drugs:
  Sulfa group
  Allopurinol
  Antiepileptics (carbamazpaine and phenytoin)
  Penicillin's
  Cephalosporins
  NSAIDs
SJS / TEN

Definition: Both can start as target lesion, then diffuse erythema, skin necrosis and detachment
SJS < 10% epidermal detachment
SJS / TEN overlap 10 – 30 %
TEN > 30% epidermal detachment
Time from first drug exposure to onset of disease is 1 to 3 weeks
Prodrome: Fever & flu–like
Skin Pain, burning & tenderness
Mucous membrane: mouth lesions are painful and tender.
SJS / TEN

-Lesions can start as macular erythema and target lesions then detachment of skin
-In TEN : +Ve Nilolsky’s sign
-Mucous membrane erythema, painful erosions of mouth, conjunctiva, genital and anal skin
-Fever, malaise, acute renal failure

Complications:
-Fluid and electrolyte imbalance
-Infection (due to loss of skin barrier)
-Skin: Scarring and dyspigmentation
-Eye: Adhesions & blindness
-Acute renal failure
SJS / TEN

- Diagnosis confirmation: skin biopsy

- Mortality:
  - In TEN 30%
  - In SJS < 5%

*Causes of Death

- Sepsis
- GIT bleed
- Fluid / Electrolyte imbalance
SJS / TEN

Management:
- Stop the causative agent
- Admit to burn unit or ICU
- IVF and electrolyte replacement like burn patient
- IV Abx for infection
- Skin dressing daily
- Eye care by opthalmologist daily
- High calori intake
- Foly’s catheter/ NG tube
SJS / TEN

- Intravenous immunoglobulins (very useful)
- ? Systemic steroid (controversial)
- Plasma exchange
- Prevention: In future patient must not take the causative drug or other drugs which can cause cross-reaction.
- SJS:
  - More mucosal involvement but less skin involvement than TEN
  - Better prognosis than TEN. (SJS is usually self-limited disease within 3 wks)
Necrotizing Fasciitis

- Known in the media as "Flesh eating Bacteria"
- Infection of subcutaneous tissue\(\}
  destruction of fascia and fat.
- Rapidly progressive bacterial infection
- Polymicrobial group A strept, other G+Ve & G-Ve
- Pain, erythema edema, fever \(\}
  severe pain with limb swelling \(\}
  high fever, Bluish discoloration & blisters \(\}
  Gangrene & muscle necrosis.
Necrotizing Fasciitis

• Can lead to death or loss of a limb

• Treatment:
  - Surgical debridement
  - I V antibiotics.
Pyoderma Gangrenosum (PG)

- Rapidly progressive causing severe tissue destruction
- Start as small pustule ‡ enlarge to form ulcer with necrotic center involving skin & subcutaneous tissue down to fascia & muscle
- Border is undermined with a purple hue.
- Severe pain & tenderness
- Causes:
  - Inflammatory Bowle disease
  - Rheumatoid Arthritis
  - Leukemia'
  - CTD
  - Idiopathic
PG

- DDX: Infection

Treatment:
- Treat underlying disease if any
- High dose of oral or IV steroids ± immunosuppressive
Acute Urticaria

Urticaria is a common disease
- Recurrent, transient, cutaneous swelling and erythema due to fluid transfer from the vasculature to the dermis
- Typical lesion is the “Wheal”
- Severe itching
- Can be acute or chronic
- Acute < 6 wks - Chronic > 6 wks
Acute Urticaria

Causes:
- Drugs: due to release of histamine
  - e.g. Aspirin, Coedine, Morphine, Indomethacin
- Food and Food additives
- Food additives like tartrazine dye
- Benzene preservative
Acute Urticaria

- Contact Urticaria: e.g. site of contact with food (Lips)
- Physical Urticaria: Caused by pressure, heat, cold and sunlight.
- Infection: Parasitic infection
  - Hepatitis
  - Sinusitis
  - UTI
  - Gum and tooth infection.
Acute Urticaria

• Medical conditions: Thyrotoxicosis, SLE and Lymphoma

• Management:
  - Complete hx & examination are important
  - Invx: Basic: CBC & D, ESR, LFT, U/A
  - Additional According to hx and exam:
    Hepatitis B&C, thyroid function test, thyroid antibodies, ANA
Acute Urticaria

Consider Allergen Testing: Prick test RAST

Treatment:
1. Avoid Specific Allergens if known
2. Oral H1 antihistamine
e.g. Loratidien, cetrizine, fexofenadine
3. Add H2 blocker for resistance cases
4. Short Course of oral prednisone is helpful.
<table>
<thead>
<tr>
<th>Urticaria</th>
<th>Anigoedema</th>
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<tbody>
<tr>
<td>Itchy</td>
<td>Mild or non-itchy (burning or tightness feeling)</td>
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<tr>
<td>Edema in the dermis</td>
<td>Edema in the subcutaneous fat</td>
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<tr>
<td>Well defined</td>
<td>ill defined</td>
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<td>Only skin</td>
<td>Can involve skin and mucous membranes like lip, larynx (upper airway obstruction)</td>
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<td>GI mucousa (abdominal pain)</td>
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<td>Acute anigoedema is a real emergency case</td>
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Anigoedema

Usually Non-Itchy
C/0 burning & painful swelling
Involvement of
GI mucosa ‡ dysphagia, colicky abdominal pain, diarrhea & vomiting
Respiratory mucosa ‡ SOB
Upper airway ‡ Horaseness
Upper airway obstruction (might need tracheostomy)
• Can be acute Vs chronic/ hereditary vs acquired
• Can be due to enzyme deficiency (C1 esterase inhibitor)
• Acute anigoedema can occur with acute urticaria
• Can be caused by
  Drug (like ACEIs, Aspirin, NSAIDs)
  Contrast dye used in radiology
  Insect Stings
Anigoedema

Management
- Patient with history of acute anigoedema has to wear a bracelet that identifies the diagnosis.
- Patient needs to carry an Epi-Pen (auto injector to deliver epinephrine subcutaneously in emergency cases).
- Laryngeal involvement may need tracheostomy or intubation.
- Enzyme replacement.