LIFE-THREATENING DERMATOSES

Patricia Treadwell, M.D.
Professor of Pediatrics
IU School of Medicine
UK Health Care
Faculty Disclosure

- Novartis - PI for research study
- Eli Lilly & Co - spouse has stocks

- I do intend to discuss an unapproved/investigative use of FDA approved products in my presentation.
Practice Gap

- Cutaneous findings are sometimes the first clue to a life-threatening disorder. Practitioners are not always cognizant of the diseases associated with the cutaneous findings and proper diagnosis may be delayed.
Objective

- This presentation will highlight the cutaneous findings in staphylococcal scalded skin syndrome, toxic shock syndrome, meningococcemia, RMSF, Steven’s Johnson Syndrome and Kawasaki disease. Recognition of the findings will allow for prompt diagnosis.
Expected Outcome

- The attendees should be able to recognize skin changes in these disorders and appropriately recommend further work-up and treatment following this session. Patient outcomes will be improved through the acquisition of this knowledge.
STAPHYLOCOCCAL SCALDED SKIN SYNDROME

- Exfoliation toxin
- Colonization with *S. aureus* usually phage type II
- Primarily children under 5 years of age
- Renal disease contributes to poor clearance of the toxin
SSSS
CLINICAL FINDINGS

- Generalized erythema with flexural accentuation
- Skin tenderness
- Flaccid bullae in the intertriginous areas
- Exfoliation
- Positive Nikolsky’s sign
- Later desquamation
SSSS
THERAPY

- Maintain fluid status
- Prevent secondary infection
- Systemic anti-staphylococcal antibiotic
REFERENCES

REFERENCES


TOXIC SHOCK SYNDROME

- Toxic shock syndrome toxin TSST-1
- Staphylococcal enterotoxins
- Streptococcal toxin
- Other toxins
TOXIC SHOCK SYNDROME

CASE DEFINITION

- Fever
- Erythema
- Desquamation, 1-2 weeks after the onset of the illness, particularly of the palms and soles
- Hypotension (systolic BP <90 for adults and <5th percentile for age for children <16 years of age, or orthostatic syncope)
Involvement of 3 or more of the following:

- Gastrointestinal (vomiting or diarrhea)
- Muscular (severe myalgia or high CK)
- Mucous membrane hyperemia
- Renal (sterile pyuria, high BUN or CR)
- Hepatic (high bili, SGOT< or SGPT)
- Hematologic (low platelets)
- CNS (disorientation)
TOXIC SHOCK SYNDROME

- Cutaneous findings
  - erythema
  - conjunctival injection
  - necrolysis
  - multiple pustules
  - desquamation
TOXIC SHOCK SYNDROME - TREATMENT

- Supportive therapy-including maintaining fluid status and use of vasoactive agents as necessary
- Adequate drainage of suppurative sites
- Anti-staphylococcal antibiotics
REFERENCES


REFERENCES

MENINGOCCEMIA

- Patients present with fever, myalgias and malaise
- Sometimes may see meningismus
- Skin lesions - macules, petechiae, and purpuric lesions with jagged edges
- Profound hypotension and shock can occur with overwhelming infections
MENINGOCCEMIA

- DIC may develop
- Complications of DIC include thromboses or gangrene
MENINGOCCEMIA - TREATMENT

- Isolation
- Supportive therapy including fluids and vasoactive agents as necessary
- Systemic penicillin
- Cefotaxime and ceftriaxone are alternatives
- If patient has anaphylactoid-type penicillin reaction, may use chloramphenicol
MENINGOCCEMIA

- Evaluate need for treatment of household members and close contacts
REFERENCES

ROCKY MOUNTAIN SPOTTED FEVER

- Caused by *Rickettsia rickettsii*
- Typically history of tick exposure
- Incubation 2-14 days
ROCKY MOUNTAIN SPOTTED FEVER

- Fever
- Severe headache
- Confusion
- Nausea and vomiting
- Photophobia
ROCKY MOUNTAIN SPOTTED FEVER - exanthem

- Exanthem present in 90% patients
- Erythematous macules and papules initially
- Later, petechial or purpuric lesions
- Lesions occur initially on the palms and soles, then spread centrally
ROCKY MOUNTAIN SPOTTED FEVER

- Supportive therapy may be necessary
- Doxycycline
- Chloramphenicol
ROCKY MOUNTAIN SPOTTED FEVER

References


REFERENCES

HENOCK-SCHONLEIN PURPURA

- A hypersensitivity reaction that occurs typically following an infection.
- Infection most often streptococcal or viral.
HENOCH-SCHONLEIN PURPURA

- Palpable purpura
- Petechial lesions
- Acral distribution
- Vesicular or bullous lesions
- Infants tend to have more involvement of the face and scalp with accompanying edema
HENOCH-SCHONLEIN PURPURA

- Gastrointestinal abnormalities – including abdominal pain, vomiting and bloody stools
- Arthralgias and/or arthritis
- Renal abnormalities
HENOCH-SCHONLEIN PURPURA

- Immune complex formation
- Histology shows leukocytoclastic vasculitis with extravasated RBC’s
- Immunofluorescence shows IGA
HENOCH-SCHONLEIN PURPURA

- Treatment
  - Elevation
  - Anti-inflammatory medication
  - Corticosteroid controversy

- Renal status should be monitored if there is evidence of renal disease in the acute stages
REFERENCES


REFERENCES


STEVEN JOHNSON SYNDROME

- Stevens-Johnson Syndrome and Toxic Epidermal Necrolysis (TEN) considered part of a spectrum
- Hypersensitivity (allergic) reaction to medications or infectious agent
- Toxic injury to keratinocytes and mucosal epithelial cells
STEVENS-JOHNSON SYNDROME - ETIOLOGY

- Sulfa drugs
- Anti-convulsants
- Non steroidal anti-inflammatory agents
- Other drugs
- Mycoplasma
STEVENS-JOHNSON SYNDROME

◆ Clinical findings
  Rapid onset of symptoms
  Mucous membrane erosions
    - Oral
    - Ocular
    - Genital
  Subepidermal bullous formation
  Nikolsky’s sign
  Can see atypical target lesions
STEVENS-JOHNSON SYNDROME – OTHER FINDINGS

- Fever
- Malaise
- Headache
- Respiratory distress
- GI Involvement - dysphagia, malnutrition, abdominal pain, diarrhea
A frozen section of the blister roof will show several layers of cell.
STEVENS-JOHNSON SYNDROME

- May be a genetic predisposition
- Family history pertinent
- Avoid triggers
STEVE N S-JOHNSON
SYNDROME - TREATMENT

- Discontinue medication
- Supportive care
- ?Burn center
- Ophthalmology consult if eye involvement
STEVENS-JOHNSON SYNDROME - TREATMENT

- Corticosteroid controversy
- IVIG
- Cyclosporine
REFERENCES


REFERENCES


KAWASAKI DISEASE

- Searching many for etiologic infectious agent
- Role of superantigens
KAWASAKI DISEASE

- Evidence for superantigens
  - activation of T/B cells
  - induction of immunologic tolerance
KAWASAKI DISEASE
CASE DEFINITION

- Fever persisting 5 days or more
- Erythema and swelling of the palms and soles- later desquamation
- Polymorphous exanthema
- Conjunctival injection
- Erythema of the lips and oral pharynx, strawberry tongue
- lymphadenopathy
KAWASAKI DISEASE

- CUTANEOUS FINDINGS
  - Conjunctival injection
  - Erythema of the lips and oral pharynx
  - Strawberry tongue
  - Fissures of the lips
  - Swelling of the hands and feet
  - Perineal desquamation
  - Desquamation of the hands and feet
KAWSASKI DISEASE

- Cardiac consultation
  - Echocardiogram
- IVIG
REFERENCES


REFERENCES
