Cutaneous Clues to Rheumatic Diseases in Children

Barbara E. Ostrov, MD
Professor of Medicine and Pediatrics
Rheumatology and Pediatric Rheumatology
Penn State Hershey Children's Hospital

Objectives

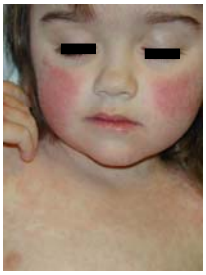
- Recognize subtle cutaneous signs of rheumatic disease
- Recognize different features of psoriasis and psoriatic arthritis
- Recognize cutaneous complications of rheumatic diseases and treatments.

Potential Conflict of Interest Statement

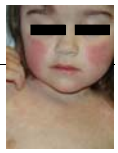
I have no conflict of interest to report.

Case 1- facial rash

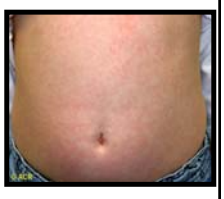
- 4 yo with a facial rash x 5 days
- low grade fever
- LABS:
  - WBC 3800, Hb 9.5
  - LFT nl; ESR 40
- DX ??



Case 1- facial rash

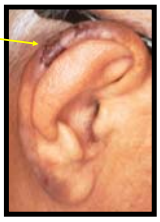


- 4 yo with a facial rash x 5 d
- DX: **Parvovirus B19 infection**
  - = 5th Disease
  - epidemic viral illness
    - facial rash, lacy trunk rash
  - children - low grade fever, arthralgias (20%); diffuse rash
  - adults - fever, small joint polyarthritis (80%); little rash
  - anemia (RBC arrest) >> leukopenia
    - esp. red cell defects
      - ex: Sickle cell disease



Viral arthritis

- Many different viruses
  - Parvovirus, rubella, hepatitis B,C
    - cryoglobulinemia
  - EBV, CMV, adenovirus, coxsackie
- Arthralgia > arthritis
  - "Toxic synovitis"
- Associated with fever, exanthem
- Migratory x 1- 6 wks
  - Rarely persistent
    - except immunocompromised hosts
  - Non-erosive

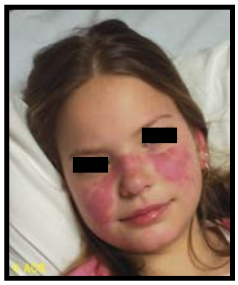


Clues for the PCP

- Arthritis usually transient
  - + associated viral syndrome
  - Quick response to NSAIDs
  - Recurrences uncommon
- Rash
  - Typical viral exanthem to vasculitic
- Labs:
  - near normal or
  - inflammatory markers high

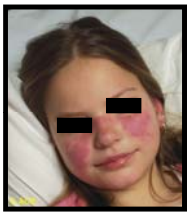
Case 2- facial rash

- 13 yo w/ facial rash x 2 wks
- low grade fever
- joints ache, stiff in a.m. x 4 wks
- fatigue x 6 wks
- LABS:
  - WBC 3800, Hb 9.5
  - LFT nl; ESR 40
- Dx ??



Case 2- facial rash

- 13 yo w/ facial rash + systemic symptoms
- DX: **Systemic lupus erythematosus**
  - 20% onset before 20 yo
  - 70% with photosensitivity
  - 80% with rashes
  - Organ system involvement:
    - 50% develop nephritis
    - 90% have arthritis
    - 80% with cytopenia



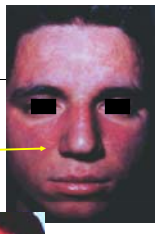
SLE Criteria

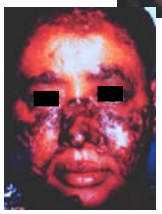
- Malar rash
- Discoid rash
- Photosensitivity
- Oral/nasal ulcers
- + ANA
- + specific serology
  - Anti-DS-DNA
  - APS
  - Anti-Smith ab
- Renal involvement
- CNS involvement
  - Seizures or psychosis
- Non-erosive arthritis
- Serositis
  - Pericarditis
  - Pleuritis
- Hematologic involvement
  - cytopenias

- > 4/11 are 88% specific

SLE - cutaneous findings

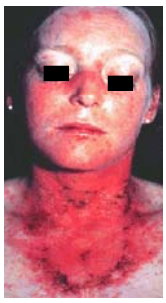
- **Malar rash**
  - over bridge of nose
  - skips nasolabial folds
- similar to rash of:
  - dermatomyositis
  - Fifth disease
  - rosacea (adult acne)
- **Discoid lupus**
  - Scarring rash
  - malar, scalp, ears
  - photosensitive
  - Rx: injections of steroids





SLE - cutaneous findings

- **Photosensitivity**
  - new onset
  - lifelong sun sensitivity does not count
- other causes:
  - other autoimmune illnesses (dermatomyositis)
  - Medications
    - antibiotics, hormones, steroids
  - prior severe sunburn

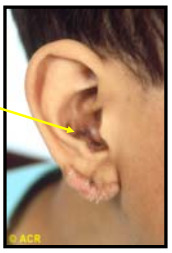


SLE - Treatment

- Avoid sun
  - 70% are photosensitive
  - Suns screen to block UV light
    - > 35-40 SPF preferred
    - Daily sunscreen, sunblock
    - products w/ zinc oxide/titanium dioxide
      - physical blocking agents
      - provide protection from UVa & UVb light
- Topical medications
  - Corticosteroids
  - Tacrolimus, pimecrolimus
- Anti-malarials – usually hydroxychloroquine
- Others: dapsone, colchicine

Clues for the PCP

- Malar rash may present as subtle flush only
- discoid LE lesions:
  - Hide in external ear
  - May mimic psoriasis or ringworm
  - May be sole manifestation in some; only 20% evolve to SLE
- Discoid and malar rashes spare nasolabial fold
- more severe rash requires dermatologist input

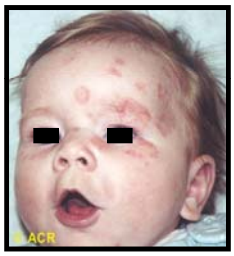


Distinguishing Parvovirus from SLE

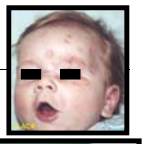
- Age/epidemiology of patient/family
  - 5th dis: younger child; + exposure hx
- Rash characteristics
  - Parvo: slapped cheek, lacy body rash, few other sx
    - vs SLE: older pt; facial rash + systemic sx
- Hematologic parameters
  - Parvo: effects on RBC >> WBC, platelets; no retics
    - vs SLE: WBC >> RBC, platelets; w/ hemolysis - ↑ retics
- Arthritis manifestations
  - Parvo: migratory arthralgias younger; hand arthritis older pt
    - vs SLE: variable inflammatory, persistent polyarthritis
- Hepatitis
  - Parvo: common; transient, occasionally severe
    - vs SLE: occasionally seen
- Time course
  - Parvo: transient; resolves w/in 4-6 weeks in > 90%
    - vs SLE: indolent onset, worsening over 4-12 weeks

Case 3 - infant with facial rash

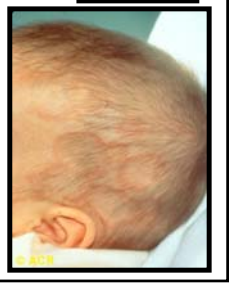
- 3 week old baby has facial rash x 2 wks
- seems well otherwise
- no pregnancy or neonatal complications
- LABS:
  - WBC 3800, Hb 9.5
  - LFT nl; platelets 110,000
- DX ??



Case 3 - infant with rash

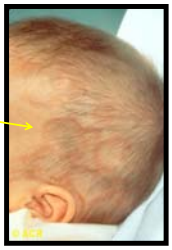


- 3 wk old w/ facial, scalp rash
- DX: **neonatal lupus**
  - 5-10% of mothers w/ SS-A, SS-B ab
  - 50% no hx SLE or Sjogren's
    - 1/2 will go to definite dx in 10 yrs time
  - 10-20% recurrence risk with subsequent pregnancies



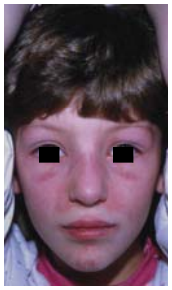
Case 3 - infant with rash

- Neonatal LE
  - rash on face/trunk
    - discoid or SCLE
      - + SS-A, SS-B with SCLE
      - annular or blistering lesions
  - Congenital HB
    - serious - 3rd° → pacemaker
  - hepatitis, serositis, cytopenia
  - Rash ↑ w/ phototherapy
- Known hi risk mother:
  - check SS-A, SS-B
  - High risk OB
  - fetal echo @ 16 wks gestation
  - ? role for high dose steroids



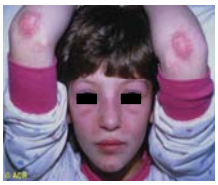
Case 4- facial rash

- 11 yo w/ facial rash x 3 wks
- fever, fatigue x 2 wks
- trouble doing nl activities
  - soccer, karate class
  - going to school
- LABS:
  - CBC nl, ESR 10
  - ALT 90, AST 90
- DX ??



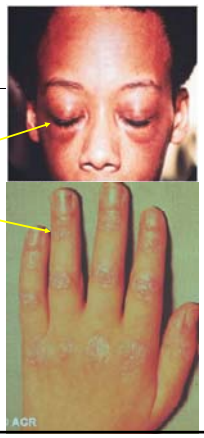
Case 4- facial rash

- 11 yo w/ facial rash, trouble with ADLs
- DX: **Juvenile Dermatomyositis**
  - 20% onset in childhood
  - no malignancy association
    - 15% adult DM
  - muscle and skin features vary
    - muscle involvement alone = polymyositis



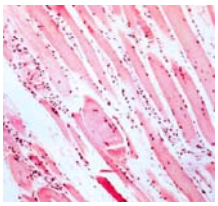
JDM – Labs, Course

- Clinical features:
  - Symmetrical proximal weakness
  - Rash in DM:
    - Heliotrope, facial edema
    - Gottron's papules - violaceous
      - Hands, elbows, knees, extensor
  - Arthritis/arthralgia
  - Dysphagia, dysphonia
    - = medical urgency
- Labs:
  - CK, aldolase, LDH, ALT/AST > 99%
  - ANA in 50%; ESR nl in most



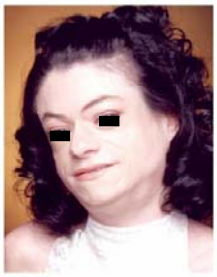
JDM - Criteria

- Inflammatory muscle disease:
  - weakness, ↑ muscle enzymes,
  - abnl EMG, MRI &/or muscle biopsy
- Classification
  - I Primary idiopathic PM
  - II Primary idiopathic DM
  - III PM (7%) or DM (15%) with malignancy
  - IV Childhood DM >>> PM
  - V PM or DM w/ another CTD
  - VI Miscellaneous types
    - inclusion body



JDM - Complications

- No malignancy association
  - 7-15% adult myositis
- > 60% do well w/ long remissions
- calcinosis prominent
  - 30-50% if not treated aggressively in 4 mos of dx
- lipodystrophy - late feature
  - loss of SubQ fat
  - insulin resistance
  - Hyperlipidemia, HTN

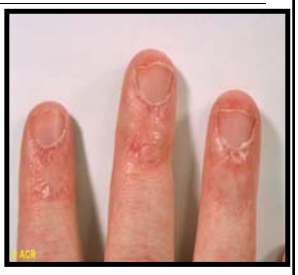


JDM - Treatment

- High dose steroids
  - 2-3 mg/kg/day to start
  - Early Rx w/in 4 mos of onset prevents calcinosis
- anti-malarials
  - for skin
- Steroid sparing early in course
  - MTX
  - Cyclosporine
  - IVIG
  - ?Rituximab

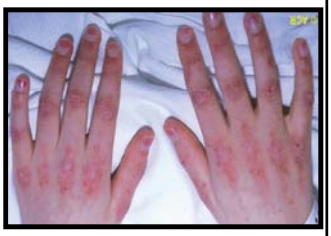
Clues for the PCP

- Diagnosis:
  - Proximal weakness >> muscle pain
  - Heliotrope, periorbital edema may be very subtle
  - Gottron's papules may mimic eczema or psoriasis
  - Cuticle overgrowth and cuticle redness/swelling are VERY suggestive of JDM



Case 5 - hand rash

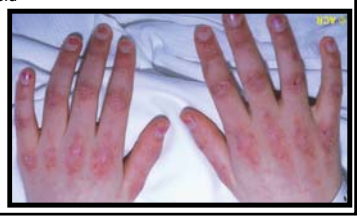
- 8 yo with red hand rash x 2 months
- Minimal response to OTC steroids
- feels fatigued, having trouble with some usual activities
- DX ??



Case 5 - hand rash

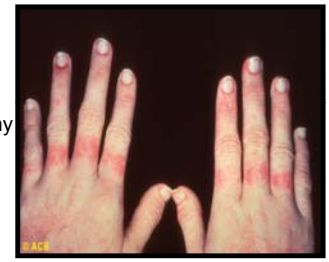
- 8 yo with red hand rash; having trouble w/ ADLs
- DX: **JDM**
  - with Gottron's papules and abnormal nailfold capillaries

Rash <u>on</u> knuckles



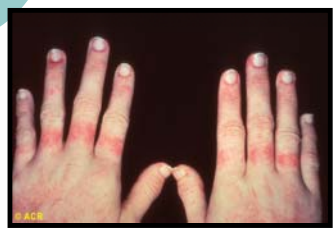
Case 6 - hand rash

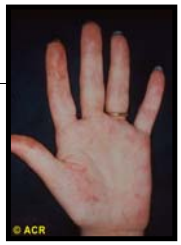
- 15 yo w/ red rash on hands x 2 mos
- minimal response to OTC steroids
- feels fatigued, achy
- no fever, swelling, or edema
- DX ??



Case 6 - hand rash

- 15 yo w/ red rash on hands w/ systemic sx
- DX: **SLE**



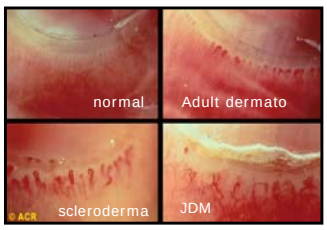


Rash <u>between</u> knuckles

vasculitic rash on palms

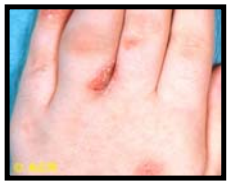
Clues for the PCP

- **<u>SLE hands</u>**
  - red cuticles and rash <u>BETWEEN</u> knuckles
- **<u>JDM hands</u>**
  - cuticles
    - Red, overgrown
  - Rash <u>ON</u> knuckles
  - Gottron's scaly pink papules
  - classic capillary Δ
- **<u>EITHER</u>**
  - telangectasias,
  - vasculitic lesions



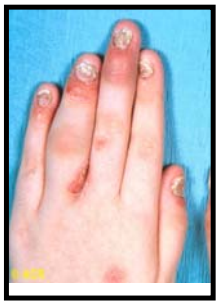
Case 7 - hand rash

- 12 yo w/ red, scaly hand, toe rash x 9 mos
- some pain and stiffness in ankles, knees with a swollen knuckle x 1 month
- feels well otherwise
- LABS:
  - CBC nl, LFT nl; ESR 40
- DX ??



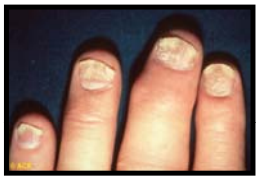
Case 7 - hand rash

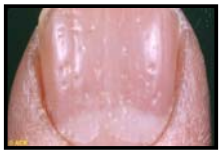
- 12 yo w/ red, scaly hand rash, joint pain
- DX: **Psoriatic arthritis**
  - Psoriasis common
    - 2% of population
  - psoriatic arthritis in 10%
  - pts w/ psoriatic nails ➡ arthritis in 30%
  - 20% w/ sacroiliitis



Diagnosis – Psoriatic arthritis

- 5 clinical subgroups
  - Groups may overlap
    - DIP arthritis w/ dactylitis
    - asymmetric oligoarthritis
    - RA-like distribution
      - RF negative
    - spondylitis
    - arthritis mutilans
      - Destructive, deforming
- extra-articular features:
  - Nails
    - Pits, "oil spots," onycholysis
  - uveitis
  - carditis rare
    - In spondylitis pts

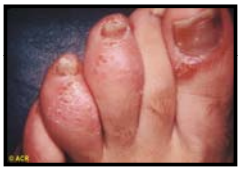




Psoriatic arthritis

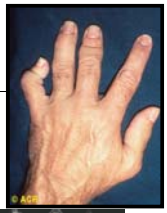
- Laboratory Findings
  - tests nl
  - ESR may be ↑
  - RF absent; ANA + in 30%
  - If RF + consider that RA and psoriasis can co-exist
  - HLA-B27 + in 50% of patients with spondylitis
    - 7-10% in others
    - = population norm

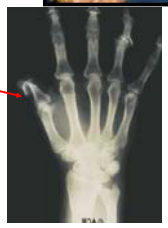
dactylitis



Psoriatic arthritis

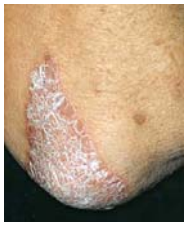
- Outcome
  - Many do well
  - Degree of skin involvement ≠ arthritis or severity
  - Arthritis mutilans uncommon
    - destructive, erosive
  - "pencil in cup deformity"
- Treatment
  - NSAIDs
  - MTX, SSZ
  - Anti-TNF agents
    - Skin and nails improve too





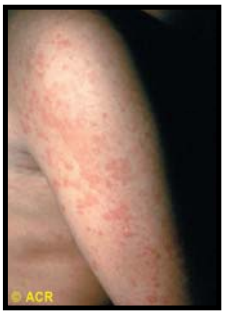
Clues for the PCP

- Mono or oligo arthritis may → psoriatic arthritis
- May take 10 yrs for skin lesions to appear *after* arthritis in kids
  - Adults usually psoriasis 1st
    - umbilicus, ears, gluteal fold
- Family hx psoriasis suggests dx psoriatic arthritis even before rash
  - Consider: fungal vs psoriasis
  - Look for pits



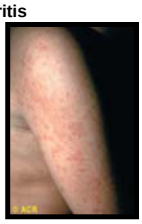
Case 8 – body/extremity rash

- 9 yo with fluctuating rash on trunk, arms
- has had fevers for 3 weeks up to 103˚
  - rash worse when fever is highest
- no joint swelling
- Won't walk in the am
- LABS:
  - WBC 18,000, Hb 9.5; ESR 95
  - Platelets 600,000, LFT nl
- DX ??



Case 8 – body/extremity rash

- 9 yo with fluctuating rash w/ fevers, joint sx
- DX: **Systemic Juvenile idiopathic arthritis**
  - 20% of JIA pts
  - Epidemiology:
    - > 300,000 children in US
    - More common than childhood diabetes
    - incidence 5-15 per 100,000
    - prevalence 60-200 per 100,000
  - Criteria:
    - <16 y, arthritis >6 wks
    - other possible causes ruled out

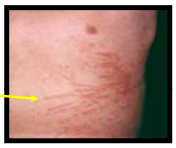


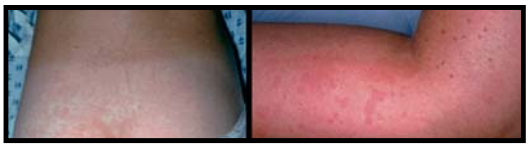
Classification- Juvenile Arthritis

- Juvenile Idiopathic Arthritis
  - Oligoarticular
  - Polyarticular
    - RF+, RF-
  - Systemic
  - Juvenile psoriatic arthritis
  - Enthesopathy associated arthritis
    - Juvenile ankylosing spondylitis
  - Undifferentiated

JIA - Dermatologic Issues

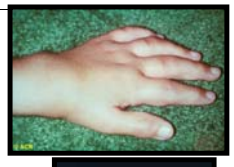
- systemic JIA
  - "salmon colored evanescent rash"
  - Worse with fevers
  - Koebner's phenomena
    - Rash after pressure/scratch
  - May: be pruritic, or coalesce





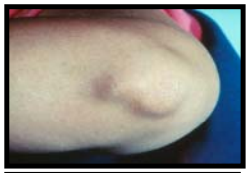
Systemic JIA – Clinical Features

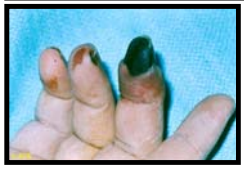
- ≥ 1 joint, fever, rash (95%)
  - serositis (25-50%)
  - anemia, leukocytosis, thrombocytosis
  - hepatitis, coagulopathy
  - ANA + < 5%; RF -
- 50% remit by 1 year
- Late risks:
  - joints often = poly
    - 25% poor articular outcome
  - worse prognosis:
    - age at onset < 4 y
    - active dis 6 mos after onset



JIA – Other Dermatologic Issues

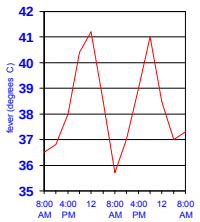
- polyarticular JIA
  - subcutaneous nodules
    - < 10% of JIA pts
    - locations: olecranon, pressure areas, over knuckles
  - rheumatoid vasculitis
    - rare complication of RF + subgroup
    - digital infarcts, purpura, gangrene





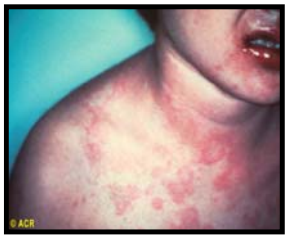
Clues for the PCP

- For systemic JIA
  - Fever pattern w/ high evening spikes
    - subnormal other times
  - r/o virus, malignancy
  - Monitor labs closely for coagulopathy, increased LFTs (MAS)
  - Avoid corticosteroids until dx certain or marrow assessed
  - Early referral to pediatric rheumatologist



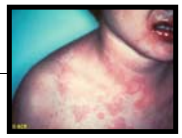
Case 9 – body/extremity rash

- 2 yo has a rash with fevers x 10 days
- hands and feet appear puffy
- diaper area reddened
- lips dry, cracked
- LABS:
  - WBC 18,000, Hb 9.5; ESR 95
  - Platelets 600,000, LFT nl
- DX ??



Case 9 - body rash

- 2 yo has a rash with fevers with mucosal changes
- **DX: Kawaski Disease**
  - 2nd most common childhood vasculitis
  - Usually < 7 yo, M > F
  - Atypical, more severe when pt < 6 mos old, M
  - More risk Asian descent



Diagnosis - KD

- Fever *PLUS* 4 out of 5 criteria.

Spiking fever for at least 5 days

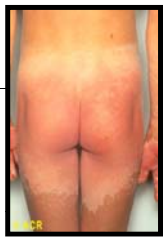
1. Bilateral conjunctival congestion

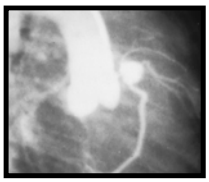
2. Oropharyngeal involvement:
Diffuse erythema, strawberry tongue, redness, lip change

3. Polymorphous erythematous rash

4. Cervical lymphadenopathy

5. One or more of following signs:
Indurative edema hands, feet
Erythema of palms, soles
Desquamation ~ 2 wks after onset
Transverse grooves nails in 2 to 3 mos



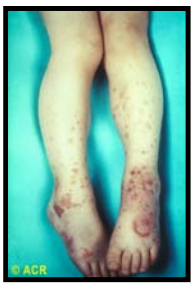


Clues for the PCP - KD

- Diagnostic clues:
  - high index suspicion
    - hi ESR; platelets climbing
    - very irritable, even w/o fever
    - sterile pyuria, rash on perineum
  - Echocardiogram early
    - Also check gall bladder for enlargement (hydrops)
- Rx
  - IVIG 2 g/kg
    - consider repeating if still fever, irritability after 24-36 hrs
  - ASA - hi dose short term; anti-platelet - 81 mg - long term
  - For IVIG failures: repeat IVIG, IV pulse steroids, ? CTX

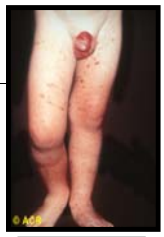
Case 10 - body/extremity rash

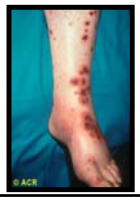
- 13 yo boy w/ knee, ankle pain x 5 days
- now has small reddish purple spots on lower extremities.
- LABS:
  - CBC nl, coags, nl
  - ESR 15
  - LFT nl
- DX ??



Vasculitis – HSP

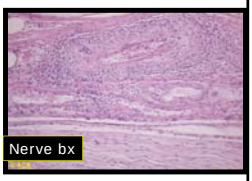
- 13 yo boy w/ joint pain, LE purpura
- DX: **Henoch Schonlein Purpura**
  - Small vessel vasculitis
    - Path: LCV
      - IgA deposits
      - dermal, mesangial, GI lesions
  - Purpura from waist down
  - Arthritis/periarthritis
  - GI vasculitis, bleeding, volvulus
  - Glomerulonephritis – IgA nephropathy
  - Testicular swelling/vasculitis





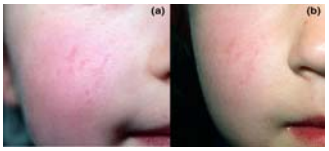
Vasculitis: Clues for the PCP

- Systemic illness with:
  - Rash, Fever
    - Small vessel = purpura
    - Medium vessel = organ problems
      - GI, brain, heart, nerve
      - renal artery = HTN
    - Large vessel = claudication
  - Labs - non-specific inflammation
    - Except ANCA
      - cANCA = Wegener's in 90%
  - Biopsy = dx
  - Angiogram
    - medium or large vessel vasculitis
- Treatment
  - varies w/ dx, organ involvement
  - Steroids, CTX, MTX



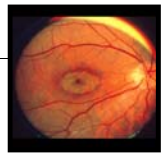
Dermatologic Complications - Medications

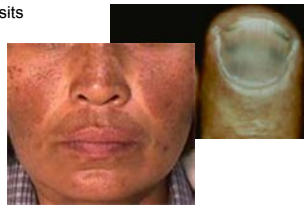
- **NSAIDs -** commonly used drugs
  - allergic rash
    - serum sickness w/ fever, arthritis
    - urticaria, maculopapular rash
  - pseudoporphyria
    - naproxen, nabumetone; others less common
    - Fair complexion; blistering in sun exposed areas
    - small scars fade to freckling



Complications - Medication

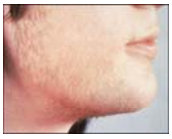
- **Anti-malarial agents**
  - hydroxychloroquine, chloroquine
  - for SLE, dermatomyositis, JIA
  - Rare, worrisome toxicity:
    - retinal pigment deposits
      - Chloroquine > risk
    - myopathy
  - hyperpigmentation:
    - face, mucosa, nails
      - Adjust dose
      - Minimize sun

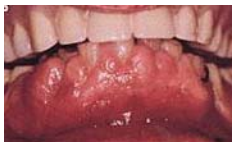




Complications - Medication

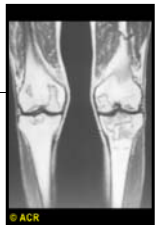
- **Cyclosporine**
  - used for systemic JIA, occ. Poly JIA, JDM
    - controls systemic JIA
    - Synergistic w/ MTX
  - most worrisome SE:
    - renal dysfunction
    - hyperkalemia
    - infections
    - hirsutism
    - gingival hypertrophy

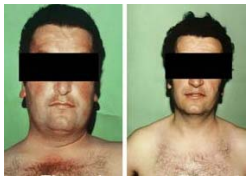




Complications - Medication

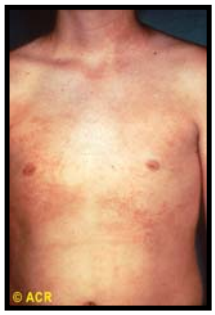
- **Corticosteroids**
  - for JIA, SLE, JDM, others
  - Toxicity: dose, route related
    - low dose < 0.25 mg/kg/d or < 10 mg/d
    - hi dose > 1 mg/kg/d or > 40 mg/d = ↑ risk SE
      - weight ↑, acne, striae, cataracts, HTN, DM
      - Hypopigmentation at site of steroid injections
      - Late: AVN, osteoporosis
    - topical ophthalmic steroids
      - use 2-3 x a day →glaucoma, cataracts





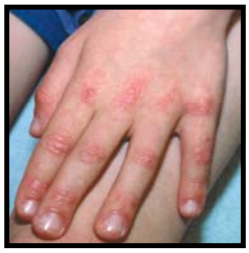
Unknown Case

- 11 yo boy with rash on and off x 6 weeks
- Not itchy or tender
- Fevers to 102˚ nightly w/ arthralgias for past 4 wks
- About 5 # weight loss
- No CP, SOB, abdominal pain, joint swelling
- LABS ordered ?
- DX ??



Unknown Case

- 8 yo girl w/ hand rash x 3 mos
- Minimal response to topical OTC steroids
- No other rash noted
  - did burn more easily during the summer (2 months ago)
- Slight ↓ activity level (not as fast when running)
- LABS ordered?
- DX ??



Summary

- Skin manifestations often clue to rheumatologic dx
- Subtle differences in distribution of rash, associated sx can give definitive dx
- Cutaneous complications of the illnesses and medication complications important part of disease monitoring
- PCP can work together w/ pediatric rheumatologist and dermatologist
- Optimize pt care and outcome

Any questions?