Dermatology for the Internist
Alabama and Mississippi ACP Scientific Meeting 2017

Jeremy D. Jackson, M.D., F.A.A.D
Assistant Professor of Dermatology
Assistant Professor of Medicine
University of Mississippi Medical Center
Disclosures

- Speakers Bureau: Celgene, SunPharma, Actelion, Abbvie
Dermatology in Primary Care

- Skin disorders affect 20-30% of the population in the United States
- 1 in 3 Americans affected with a skin disease at any given time
- National shortage of dermatologists results in referral times on average > 1 month
- In UK, 75% of skin conditions managed exclusively by primary care


Inpatient Dermatology

- Approximately 1/3 of hospitalized patients demonstrate significant skin findings
- 10% of hospitalized patients have skin findings relevant to their admission or indicative of systemic disease
- In one study $900 million dollars in Medicare reimbursement due to dermatology-related diagnosis
Dermatology consultation changed the final diagnosis in 71% of consultation requests.
Case 1

42 yo WM with 10+ year history of rash on arms and trunk. Worse when sweating in the summer.
A skin biopsy is performed. Which of the following dreaded pathology reports is associated with this skin condition?

A. Interface dermatitis with lymphocytes
B. Spongiotic dermatitis
C. Naked granulomas
D. Psoriasiform dermatitis
E. Subepidermal blister with eosinophils

Response Counter
A skin biopsy is performed. Which of the following dreaded pathology reports is associated with this skin condition?

A. Interface dermatitis with lymphocytes  
   LUPUS and other CONNECTIVE TISSUE DZ
B. Spongiotic dermatitis  
   Atopic Dermatitis, Allergic Contact Dermatitis
C. Naked granulomas  
   Sarcoidosis
D. Psoriasiform dermatitis  
   Psoriasis
E. Subepidermal blister with eosinophils  
   Bullous Pemphigoid
Final Pathologic Diagnosis
1. RIGHT ARM, PUNCH BIOPSY: MILD SPONGIOTIC DERMATITIS (SEE COMMENT)
2. BACK, PUNCH BIOPSY: MILD SPONGIOTIC DERMATITIS (SEE COMMENT)
Eczema = Dermatitis

Pathology—Spongiotic Dermatitis

Ddx: Atopic dermatitis, Allergic contact dermatitis, Drug eruption, early Mycosis fungoides

* Erythema and swelling of skin
* Oozing and/or vesiculation
* Crusting and scaling
* Thickening and evidence of repeated excoriation
* Hyperpigmentation, scratch papule formation and/or lichenification
Nummular Eczema
Clinical Characteristics

- **Infantile:** face, scalp, extensors
- **Adult:** more often flexural

- Acute skin lesions - intensely pruritic erythem. papules and thin plaques with secondary excoriations; can see vesicles & serous crusts
- Subacute - erythematous papules/plaques with scaling and excoriations
- Chronic - thickened, hyperkeratotic plaques with lichenification & prurigo nodules
Conventional Therapies for Atopic Dermatitis

- Emollients
- Irritant / allergen identification and avoidance
- Topical corticosteroids
- Sedating antihistamines
- Systemic therapies
  - Phototherapy (PUVA, UVA, UVB)
  - Immunosuppressive agents (Cyclosporine, MTX, Azathioprine, Mycophenolate mofetil)

Which vehicle used in the formulation of topical steroids is the most potent?

A. Ointment
B. Cream
C. Lotion
D. Solution

Response Counter
<table>
<thead>
<tr>
<th>Class</th>
<th>Description</th>
</tr>
</thead>
</table>
| Class 1 (super potent) | Clofertas propionate gel, ointment, cream, lotion, foam, spray and shampoo 0.05%  
Betamethasone dipropionate gel and ointment 0.05%  
Diflorasone diacetate ointment 0.05%  
Flucinonide cream 0.1%  
Flurandrenolide tape 4 mg/cm²  
Halobetasol propionate ointment and cream 0.05%  

Class 2 (high potency) | Aminocorticoid ointment 0.1%  
Betamethasone dipropionate cream, lotion, gel and ointment 0.05%  
Clobetasol propionate solution ("scalp application") 0.05%  
Desoximetasone ointment and cream 0.25% and gel 0.05%  
Diflorasone diacetate ointment and cream 0.05%  
Flucinonide gel, ointment, cream and solution 0.05%  
Halopenicinolone ointment, cream and solution 0.1%  
Mometasone furoate ointment 0.1%  
Triamcinolone acetonide ointment 0.5%  

Class 3 (high potency) | Aminocorticoid cream and lotion 0.1%  
Betamethasone dipropionate cream and lotion 0.05%  
Betamethasone valerate ointment 0.1%  
Diflorasone diacetate cream 0.05%  
Flucinonide propionate ointment 0.005%  
Triamcinolone acetonide ointment 0.1% and cream 0.5%  

Class 4 (medium potency) | Betamethasone valerate foam 0.12%  
Desoximetasone cream 0.05%  
Flucinonide acetonide ointment 0.025%  
Flurandrenolide ointment 0.05%  
Hydrocortisone valerate ointment 0.2%  
Mometasone furoate cream and lotion 0.1%  
Triamcinolone acetonide ointment (Kenalog®) and cream 0.1% or spray 0.2%  

Class 5 (medium potency) | Betamethasone dipropionate lotion 0.05%  
Betamethasone valerate cream and lotion 0.1%  
Clocortolone pivalate cream 0.1%  
Flucinonide acetonide cream 0.025% or oil and shampoo 0.01%  
Fluticasone propionate cream and lotion 0.05%  
Flurandrenolide cream and lotion 0.05%  
Hydrocortisone butyrate ointment, cream and lotion 0.1%  
Hydrocortisone probutate cream 0.1%  
Hydrocortisone valerate cream 0.2%  
Prednicarbate ointment and cream 0.1%  
Triamcinolone acetonide ointment 0.025% and lotion 0.1%  

Class 6 (low potency) | Aclopiemetasone dipropionate ointment and cream 0.05%  
Triamcinolone acetonide cream 0.1% (Aristocort®)  
Betamethasone valerate lotion 0.1%  

Class 7 (low potency) | Topicals with hydrocortisone, dexamethasone and prednisolone  
Desonide gel, ointment, cream, lotion and foam 0.05%  
Flucinonide acetonide cream and solution 0.01%  
Triamcinolone acetonide cream and lotion 0.025%
Steroid-Free Topical Therapies

- Tacrolimus ointment 0.03%, 0.1%
  - Topical formulation of oral transplant drug
  - Approved December 2000

- Pimecrolimus cream 1%
  - Specifically developed to treat inflammatory skin diseases
  - Approved December 2001
What’s new?

- **Crisaborole 2% ointment—PDE-4 inhibitor** approved in December 2016 for mild-to-moderate atopic dermatitis
  - ISGA score success (clear/almost clear with ≥2-grade improvement; AD-301: 32.8% vs 25.4%, P = .038; AD-302: 31.4% vs 18.0%, P < .001)
  - Clear/almost clear (51.7% vs 40.6%, P = .005; 48.5% vs 29.7%, P < .001).
  - Crisaborole-treated patients achieved success in ISGA score and improvement in pruritus earlier than those treated with vehicle (both P ≤ .001).
What’s New?

- Dupilumab—First biologic approved for atopic dermatitis (March 2017)
- IL-4/IL-13 inhibitor
- 18 and older
Case 2

- 45 yo obese WF with hx of multiple sclerosis presents with complaints of rash on abdomen and elbows. It does not itch. Pt reports joint pains, particularly of the DIPs of the hands. She also has morning stiffness lasting about an hour. Your exam shows well demarcated plaques on the bilateral elbows and abdomen. BSA >15%
Which of the following is the best choice in management for this patient?

A. Topical Calcitriol and triamcinolone 0.1% cream
B. Oral prednisone taper
C. Ustekinumab
D. Cyclosporine
E. Infliximab

Response Counter

C. Ustekinumab
Psoriasis

- Sharply demarcated plaques with silvery scale
- Affects 2% of population; Two peaks 20-30 and 50-60
- Auspitz sign—Bleeding upon removal of scale
- Elbows, Knees, Scalp, and Non sun exposed areas
- Nail pits more frequently associated with psoriatic arthritis
- Sausage digits, pencil-in-cup deformities, inflammation of DIP joints, enthesitis associated with arthritis
Inverse Psoriasis
Psoriatic Arthritis/Nail Dystrophy
Nail Psoriasis
Psoriatic Erythroderma
Comorbidities and Psoriasis

- People with psoriasis more likely to have a cardiovascular event and stroke
- 46% more likely to Type II Diabetes
- Up to 30% have psoriatic arthritis
- Also associated with the metabolic syndrome
Psoriasis Pearl

Systemic steroids are rarely used as patients with risk of severe rebound (pustular) psoriasis after discontinuation.
Psoriasis Pearl

Always ask about joint pain/morning stiffness.
Psoriasis pathogenesis
Treatment

- Topicals (Vit D analogs, topical steroids, tar compounds)
- Phototherapy—nbUVB
- Systemic therapy
  - Moderate-to-severe psoriasis
  - Psoriatic arthritis
  - Psoriasis in difficult locations (scalp, palms/soles, inverse)
Systemic Treatments

1. Methotrexate
2. Acitretin
3. Cyclosporine
4. Biologics
   - Etanercept, Infliximab, Adalimumab, Ustekinumab
What’s new?

- IL-17 inhibitors (secukinumab, ixekizumab, brodalumab)
- Apremilast (PDE-4 inhibitor)—oral medication
- Biosimilars for TNF-alpha inhibitors (etanercept, adalimumab, infliximab) are FDA approved but caught up in court systems over patents
- In phase III trials—JAK inhibitors, IL-23 inhibitors
Case 3

24 yo college student presents with 2 week history of rash on the arm. She denies any joint pain and takes no medicines...And she is getting married in 4 weeks!!
Which of the following is the most likely diagnosis?

A. Granuloma annulare
B. Tinea corporis
C. Subacute cutaneous lupus erythematosus
D. Erythema multiforme
E. I don’t know but just put Lotrisone on it (betamethasone/clotrimazole)
What percent of patients with SCLE meet criteria for SLE?

A. 5%
B. 25%
C. 40%
D. 60%
E. 80%

Response Counter
What positive lab test is associated with patients with SCLE?

A. Anti-histone
B. Anti-centromere
C. dsDNA
D. Jo-1
E. SS-A

Response Counter

- Anti-histone: 20%
- Anti-centromere: 20%
- dsDNA: 20%
- Jo-1: 20%
- SS-A: 20%
What positive lab test is associated with patients with SCLE?

A. Anti-histone
B. Anti-centromere
C. dsDNA
D. Jo-1
E. SS-A -(75-90% SSA/Ro; 30-40% SSB/La)
Subacute Cutaneous Lupus

Often associated with *Anti-Ro*

Sun-exposed sites
- Sides of face, V of neck, forearms
- Mid-face usually spared

Annular patches/plaques with raised, red borders, central clearing, slight scale

Roughly 75% have arthralgia or arthritis
Tinea Corporis
Granuloma annulare
Variants of Cutaneous Lupus

**Acute cutaneous lupus**
- Epidermis and upper dermis
- Associated with *systemic disease*

**Subacute cutaneous lupus**
- Epidermis and upper dermis
- Not significant systemic involvement
- Associated with *anti-Ro* autoabs and *photosensitivity*

**Discoid lupus**
- Epidermis, dermis, and *adnexal structures*
- *Scarring*
- No significant systemic involvement

**Lupus erythematosus tumidus**
- Dermis only
- No adnexal involvement

**Lupus panniculitis**
- Subcutaneous tissue
- *Depressed*, disfiguring scars

### Different Forms of Cutaneous Lupus and Their Association with Systemic Lupus Erythematosus (SLE)

<table>
<thead>
<tr>
<th>Type of cutaneous lupus</th>
<th>Association with SLE</th>
</tr>
</thead>
<tbody>
<tr>
<td>Acute cutaneous lupus erythematosus (ACLE)</td>
<td>++++</td>
</tr>
<tr>
<td>Subacute cutaneous lupus erythematosus (SCLE)</td>
<td>++</td>
</tr>
<tr>
<td>Chronic cutaneous lupus erythematosus (CCLE)</td>
<td></td>
</tr>
<tr>
<td>- Discoid lupus erythematosus (DLE)</td>
<td>++</td>
</tr>
<tr>
<td>- Localized (head and neck)</td>
<td>++</td>
</tr>
<tr>
<td>- Widespread/disseminated</td>
<td>+</td>
</tr>
<tr>
<td>- Hypertrophic</td>
<td>+</td>
</tr>
<tr>
<td>- Lupus erythematosus tumidus (LET)</td>
<td>+/−</td>
</tr>
<tr>
<td>- Lupus panniculitis</td>
<td>+</td>
</tr>
<tr>
<td>- Chilblain lupus</td>
<td>++</td>
</tr>
<tr>
<td>Other variants</td>
<td></td>
</tr>
<tr>
<td>- Bullous eruption of SLE</td>
<td>++++</td>
</tr>
<tr>
<td>- Rowell’s syndrome</td>
<td>++ to +++</td>
</tr>
</tbody>
</table>
Discoid Lupus Erythematosus

Face, scalp, ears, occasionally widespread
Sun-exposed and sun-protected sites, even mucosal
Scarring and dyspigmentation

About 5% develop systemic disease

SCC can develop in long-standing lesions
Discoid Lupus
Discoid Lupus
Acute Cutaneous Lupus

- Butterfly/malar rash, poikiloderma, papules, occasional scale
- Follow sun exposure
- Transient (hrs-weeks)
- Non-scarring
- May have oral ulcerations
- Spares knuckles (Unlike dermatomyositis which involves the knuckles)
ACR Diagnostic Criteria

Need 4/11 Criteria

S- Serositis
O- Oral ulcers
A- Arthritis
P- Photosensitivity

B- Blood disorders (cytopenias)
R- Renal disorders
A- Anti-nuclear antibody
I- Immunologic (dsDNA, Sm, antiphospholipid)
N- Neurologic disorders (Sz/psychosis)

M- Malar rash
D- Discoid rash

4 of the 11 Criteria Involve Mucocutaneous Findings
Treatment

- Topical or intralesional CS: mainstay of therapy
- Systemic: Anti-malarials are gold standard
  - Hydroxychloroquine 200mg qd or bid
  - Chloroquine 250 mg qd
  - Resistant cases: oral retinoids, thalidomide, gold, clofazimine, sulfasalazine, azathioprine, systemic CS, dapsone, MTX
- Adjuvant therapy
  - High SPF Sunscreen (UVA and UVB)
  - Protective clothing
  - Stop smoking
Case 4

- 56 yo AAF with history of discoid lupus since 1999 presents with 4 month history of pruritic rash.

- Pt given trimethoprim/sulfamethoxazole initially without improvement. Started on **prednisone 20mg** for past 3 months along with triamcinolone cream.

- She has been on hydroxychloroquine 400mg daily for over 15 years

- She has joint pains but denies any recent worsening
What is the most likely diagnosis?

A. Lupus with co-existent psoriasis
B. Mixed connective tissue disease
C. Crusted scabies
D. Pityriasis rubra pilaris
E. Lupus with tinea capitis

Response Counter
• Crusted (Norwegian) scabies first described in 1848 by Boeck and Danielssen among lepers

• Causative agent is the mite, *Sarcoptes scabiei var hominis*

• Female lays 2-3 eggs daily and the eggs hatch in 3-4 days

• Average mite in normal scabies is around 10 but thousands can be present in crusted scabies
Disorders Predisposing to Crusted Scabies

- AIDS, T-cell leukemia, Lymphoma, organ transplantation
- Leprosy
- Parkinsons
- Kwashiorkor
- Down syndrome, Mental retardation
- SLE, dermatomyositis
- Diabetes mellitus
Permethrin and Ivermectin for Scabies

Bart J. Currie, F.R.A.C.P., and James S. McCarthy, F.R.A.C.P.

### Table 1. Therapies for Scabies.

<table>
<thead>
<tr>
<th>Purpose of Therapy</th>
<th>Recommended Therapy</th>
<th>Alternative Therapy</th>
<th>Comments</th>
</tr>
</thead>
<tbody>
<tr>
<td>Treatment for classic scabies</td>
<td>Two applications — one on day 1 and one between day 8 and day 15 — of topical permethrin 5%, applied in the evening and left on overnight</td>
<td>Two doses of oral ivermectin (200 μg/kg/dose), taken with food — one on day 1 and one between day 8 and day 15</td>
<td>Keratolytic creams should be used for skin crusts; maintain vigilance for the development of sepsis; apply appropriate measures to control the spread of scabies infection</td>
</tr>
<tr>
<td>Treatment for crusted scabies</td>
<td>Both topical permethrin 5% every 2 to 3 days for 1 to 2 weeks and oral ivermectin (200 μg/kg/dose), taken with food, administered as three doses (days 1, 2, and 8), five doses (days 1, 2, 8, 9, and 15) or seven doses (days 1, 2, 8, 9, 15, 22, and 29), depending on severity of infection*</td>
<td>Topical benzyl benzoate 25% (with or without tea-tree oil 5%) instead of permethrin</td>
<td></td>
</tr>
<tr>
<td>Prevention of infection in close contacts of patients with scabies</td>
<td>A single application of topical permethrin 5% applied in the evening and left on overnight</td>
<td>Oral ivermectin (200 μg/kg/dose), taken with food, administered as a single dose*</td>
<td></td>
</tr>
<tr>
<td>Management of institutional outbreak of scabies</td>
<td>Treat persons with clinic cases as recommended above for classic and crusted scabies and treat all potentially exposed residents, staff, and visitors as recommended above for contacts</td>
<td>For refractory outbreaks, consider treatment of all residents with oral ivermectin*</td>
<td>Look for “core transmitter” index cases with crusted scabies; give attention to planning and logistics of therapy; apply appropriate measures to control the spread of scabies infection</td>
</tr>
<tr>
<td>Prevention in communities where scabies is endemic or management of community outbreak</td>
<td>Adopt multifaceted approach that includes education and community involvement; treat clinical cases as recommended above for persons with classic and crusted scabies and all family and household members as recommended above for contacts; consider treating all other community members as recommended above for contacts</td>
<td>Treat persons with classic and crusted scabies, as well as contacts in the community, as recommended above</td>
<td>Look for “core transmitter” index cases with crusted scabies; give attention to planning and logistics of therapy; be aware that maintaining control of scabies requires addressing underlying issues of overcrowding and access to health hardware (e.g., functioning taps with clean water, sinks, and toilets in the house), health care, and education</td>
</tr>
</tbody>
</table>

* Ivermectin is not approved for this indication by the Food and Drug Administration; there are insufficient data on the safety of ivermectin in pregnancy and in children younger than 5 years of age.
Case 5

A 40yo AAF presents with a 2 month history of plaques on her face. She denies joint pain or SOB.
What is the name when cutaneous disease involves the nose and eyes?

A. Chilblain’s lupus
B. Lupus pernio
C. Lupus erythematosus
D. Lupus profundus
E. Lupus nosiosus

Response Counter

Chillblain’s lupus: 20%
Lupus pernio: 20%
Lupus erythematosus: 20%
Lupus profundus: 20%
Lupus nosiosus: 20%
Sarcoidosis
Sarcoidosis

- Systemic granulomatous dz that commonly involves the lungs
- F > M; AA; bimodal age distribution, peaks between 25-35 years and again between 45-65 years
- Pathogenesis:
  - An unknown antigen elicits upregulation of CD4+ Th1 cells which leads to formation of epitheliod granulomas
  - Genetic susceptibility alleles: HLA-1, HLA-B8, HLA-DR3, HLA-DRB1, and HLA-DQB1
- Cutaneous manifestations are seen in 33% of patients
Erythema Nodosum
<table>
<thead>
<tr>
<th>Affected organ</th>
<th>Frequency of occurrence (%)</th>
<th>Common findings</th>
</tr>
</thead>
<tbody>
<tr>
<td>Lungs and thoracic lymph nodes</td>
<td>&gt;90</td>
<td>Dyspnea, cough, chest pain, pulmonary hypertension, mixed pulmonary function test abnormalities (obstruction, restriction, diffusion deficits)</td>
</tr>
<tr>
<td>Skin</td>
<td>20–30</td>
<td>Nodules, plaques, lupus pernio, erythema nodosum (a non-granulomatous panniculitis)</td>
</tr>
<tr>
<td>Eyes</td>
<td>20–25</td>
<td>Uveitis, conjunctivitis, lacrimal gland enlargement, sicca syndrome, optic neuropathy</td>
</tr>
<tr>
<td>Liver and/or spleen</td>
<td>10–20</td>
<td>Hepatosplenomegaly, jaundice, elevated liver function tests, cirrhosis, hyperplasmia</td>
</tr>
<tr>
<td>Cardiovascular system</td>
<td>10–20</td>
<td>Ectopy, heart block, arrhythmias, cardiomyopathy, sudden death</td>
</tr>
<tr>
<td>Central nervous system</td>
<td>10–25</td>
<td>Cranial neuropathy, mass lesions, aseptic meningitis and/or encephalitis, myelitis, spinal cord and peripheral neuropathy, small fiber neuropathy, pain, hypothalamic-pituitary involvement</td>
</tr>
<tr>
<td>Sinuses and upper respiratory tract</td>
<td>5–10</td>
<td>Chronic sinusitis, laryngeal involvement, parotid gland involvement</td>
</tr>
<tr>
<td>Bones, joints, muscle</td>
<td>5–15</td>
<td>Chronic arthritis, dactylylitis, lytic bone lesions, myopathy</td>
</tr>
<tr>
<td>Hematologic system</td>
<td>&gt;50</td>
<td>Peripheral lymphopenia, hypergammaglobulinemia</td>
</tr>
<tr>
<td>Renal system (including calcium abnormalities)</td>
<td>5–10</td>
<td>Hypercalcemia and/or hypercalciuria, nephrocalcinosis, nephrolithiasis</td>
</tr>
<tr>
<td>Endocrine system</td>
<td>5–10</td>
<td>Hypothalamic-pituitary involvement, pancreatic mass, Herfordt syndrome (nephropathic diabetes)</td>
</tr>
<tr>
<td>Gastrointestinal and reproductive tract</td>
<td>&lt;1</td>
<td>Gastric nodules, ovarian or testicular masses</td>
</tr>
</tbody>
</table>
Initial Evaluation

- History (occupational, environmental exposure, symptoms)
- Physical exam
- Chest X-ray, PA and Lateral
- Pulmonary function tests (including DLCO)
- Routine ophthalmologic examination
- Complete blood count
- Comprehensive serum chemistries (Ca, LFTs, creatinine)
- Urinalysis
  - If history of stones, 24h urine calcium
- EKG
  - If history of palpitations, additional testing
- Tuberculin skin test or IFNγ release assay
- Thyroid testing
- Vitamin D 25, Vitamin D 1,25

Adapted from: Statement on sarcoidosis
Am J Respir Crit Care Med 1999; 160: 736-755
Cutaneous Sarcoidosis Treatment Algorithm

- **Topicals**
  - Intralesional

- **Intralesional**
  - Antimalarials
  - Minocycline

- **Antimalarials**
  - Methotrexate
  - Thalidomide

- **Methotrexate**
  - Adalimumab
  - Infliximab

_Wanat KA, Rosenbach M. Am J Clin Dermatol 2014_
Pearl

Be an advocate for your patients.

Don’t give up in our current climate of

“Insurance Based Medicine”
Case 6

- 81 yo AAF with hx of cicatricial pemphigoid admitted with worsening blisters, mental status changes, and hypotension.
- Diagnosed 6 months earlier with cicatricial pemphigoid after initially presenting with oral erosions and blisters.
- Previously treated with dapsone and methotrexate, but did not tolerate due to worsening anemia.
• As outpatient was on prednisone 40mg daily and mycophenolate mofetil 750mg BID

• Admitted 2 weeks prior to this admission with worsening pemphigoid flare, hypotension, and worsening fatigue

• Diagnosed with UTI/urosepsis and improved on IV Rocephin

• Discharged at that time on prednisone 40mg, mycophenolate mofetil, and ciprofloxacin 500mg BID
• Her daughter reports that she had continued to have poor PO intake since her discharge but that 24 hours prior to admission, she became significantly more lethargic.

• Her skin also had significant worsening of erosions on her flank, hands, and feet.
Vitals

- BP 75/56
- P: 119
- T: 33.9C (93F)
- O2 Sats 82% on room air, 94% on 4L O2
Which of the following is the most likely diagnosis?

A. Flare of Cicatricial Pemphigoid
B. DIC
C. Herpes Zoster
D. Vasculitis from cryoglobulins
Hospital Course

- Patient admitted to ICU
- Started on IV vancomycin, aztreonam, levofloxacin
- Aggressive IVF resuscitation
- Transfused 1 unit PRBCs
- Stress dose hydrocortisone
- Consulted derm the following day
Hospital Course

- Fibrinogen 84, Coags elevated—DIC
- Echo, MRI brain obtained
• Echocardiogram
  - 50% EF
  - Valves normal
  - Large thrombus in Left atrium
Multiple embolic infarcts
• CSF studies-
  Initial slight elevation in protein, 4-5 WBCs (lymphs)
  VZV PCR- Positive +
• HSV and VZV PCR from Wound (DFA unavailable at our institution)
  HSV- Negative
  VZV- Positive
Hospital Course

- Pt initially responded well to IVFs with improved mental status
- Renal function continued to worsen—hemodialysis eventually initiated
- Cryoprecipitate given initially to correct fibrinogen and coags
- Once thrombi found, heparin infusion was initiated
- Acyclovir started- renally dosed for presumed Herpes Zoster infection—Not started until day #4
Hospital Course

- Pt became more unresponsive
- Intubated on day 5 of hospitalization
- Repeat CT Head showed worsening ischemic

- Pt went into Vfib on day 7 and expired after
  ACLS measures
• Presentation of 3 distinct cutaneous morphologies made this case difficult
• Possible availability bias from recent admission
• Delay in initiation of proper treatment might have affected outcome
• Cutaneous findings of VZV infection are often the best indicator for diagnosis and dermatologist can play a significant role in diagnosis and management.
Herpes Zoster

- Zoster-associated encephalitis and vasculopathy is most common in immunosuppressed patients

- VZV is known to produce stroke syndromes and has been associated with increased risk of stroke in patients with prior episodes of zoster. Pathogenesis if thought to be secondary to infection of the cerebral arteries.

- VZV is a known cause of purpura fulminans in children but very few documentations in the literature about DIC in adults with VZV
A review of 38 cases of disseminated VZV infection in renal transplant recipients revealed 47% (18/38) developed DIC; however a majority of cases were primary varicella infection.

- VZV pneumonia/pneumonitis and hepatitis also common
- Only 7% of cases of DIC present with thromboembolisms
Case 7

- 52 yo AAM with no prior medical history presented to ER with face pain, right eye pain, and lip swelling.
- Symptoms began 1 week prior. He was given Clindamycin with no improvement after 3 days and pt returned to ER.
- Pt admitted for failed outpt antibiotics and started on IV Vanc. Ophthalmology consulted for preorbital cellulitis.
- Dermatology consulted for evaluation
Case 8

- 74 yo WM s/p BMT for multiple myeloma now relapsed and on chemotherapy presents with new rash and fever.
Case 7 and 8

Which of the following is the most likely diagnosis?

1. Herpes/Zoster
2. Herpes/Zoster
3. Herpes/Zoster
4. Herpes/Zoster
Case 7 and 8

Which of the following is the most likely diagnosis?

1. Herpes/Zoster
2. Herpes/Zoster
3. Herpes/Zoster
4. Herpes/Zoster
Herpes/Zoster Pearls

- It’s Herpes/Zoster until proven different!!!
- Order HSV and VZV PCR from swab—use viral medium but does not have to be on ice
- Go ahead and place patient under appropriate isolation if suspect zoster
Disseminated Zoster Pearls

- > 20 lesions outside of 2 contiguous dermatomes
- Elderly and immunocompromised are at increased risk
- Viscera can be affected
- Call ophthalmology
- Contact and droplet isolation
- IV Acyclovir
Case 9

• 24 yo WM admitted with 5 day history of abd pain, nausea/vomiting. Found to have adrenal mass/hemorrhage on CT scan.

• Had discoloration and pain of right great toe 6-7 weeks prior. “Was infected and bruised”

• Derm consulted for possible onychomycosis!! STAT!!
Which of the following is the most likely diagnosis?

A. Vasculitis
B. Vasculopathy
C. Chilblain lupus
D. Onychomycosis

Response Counter
PURPURA

- Skin biopsy
- PRIMARY VASCULITIS
  - Small-sized vessel
- THROMBOTIC VASCULOPATHY
  - Medium-sized vessel
  - Pauci-Inflammatory
  - Inflammatory
Retiform Purpura

- Due to thrombosis and occlusion of the vasculature
- Early on, lacks the erythema of palpable purpura; purpuric early
- Purpura tracks along tack of dermal vessels leading to retiform appearance
- Over time, ischemia leads to necrosis and eschar formation
- Palpable purpura can become retiform
- Ddx: meds, infection, coagulation defect, embolic
Causes of cutaneous thrombosis

- Hypercoagulable states – numerous!
  - Hyperhomocysteinuria
  - Protein C/S deficiency
- Atrophie blanche/livedoid vasculopathy
- Calciphylaxis
- Degos disease
- Type I cryoglobulinemia
- Sepsis
- Coumadin necrosis
- Antiphospholipid syndrome
Case 9

- Extensive work up of vasculopathy revealed:
  - Positive lupus anticoagulant and anti-cardiolipin antibodies
  - Also with autoimmune hemolytic anemia
  - Did not meet full criteria for SLE
  - Pt did well on corticosteroids and anti-coagulation
Small Vessel Vasculitis

- **Infections (15-20%)**
  - Bacterial - Group A strep, Mycobacteria, Mycoplasma, Chronic meningococcemia
  - Viral - Hep C > B >> A; HIV Parvo B19

- **Inflammatory d/o (15-20%)**
  - Al - SLE, RA
  - IBD
  - Seronegative spondyloarthropathies

- **Drugs (10-15%)**
  - Anti-TNF agents, COX-2 inhibitors, G-CSF, Hydralazine, MCN, NSAIDS, PCNs, antithyroid agents, Quinolones

- **Neoplasms** (usually blood d/o)

- **Idiopathic** ~ 50%
Basic laboratory evaluation for patients with confirmed cutaneous vasculitis

- CBC with diff
- ESR
- CRP
- SPEP/ UPEP
- Cryos
- LFTs
- Stool guiac
- BUN/ Cr
- UA
- Electrolytes
- Hep B/ C
- ASLO/ anti-DNaseB
- HIV
- RF
- Compliment (C3, C4, CH50)
- ANA
- ANCAs
Case 10

- 62 y/o AAF w/ PMH DM, HTN, CHF transferred from OSH with diffuse rash initially thought to be SJS. She also had renal failure and leukocytosis.

- Only new medication was allopurinol started 1 month prior to onset of rash

- She was admitted to the OSH for ~2 weeks prior to transfer and managed on IVFs, vancomycin, and meropenem
Case 10

- T: 97.1 BP 143/76  P 93 SpO2 93%
- Labs at OSH:
  - **CBC 11/11**: wbc 21k, 26% segs, 17% lymphs, 4.9% monos, 50% eos
  - **CBC 11/18**: wbc 29k, 35% segs, 22% lymphs, 10.8% monos, 31% eos
Which of the following labs should be checked next?

A. HSV/VZV PCR
B. HIV
C. HCV Ab
D. LFTs

Response Counter
• **CMP:** Na 139, K 4.3, Cl 108, HCO3 18, BUN 81, sCr 3.9, gluc 230,
• Alk phos 166
• Bili 2.4
• **AST 380**
• **ALT 433**
DRESS

- Drug Reaction with Eosinophilia and Systemic Symptoms
  - AKA:
    - Drug Induced Hypersensitivity Syndrome
    - Anticonvulsant Hypersensitivity Syndrome
Which of the following labs should also be checked?

A. TSH
B. HCV Ab
C. Fasting lipids
D. ANA
DRESS

- Commonly implicated drugs: anticonvulsants, sulfonamides, allopurinol, NSAIDs, azithromycin, azathioprine, anti-retrovirals
- Most cases 2-8 weeks after drug
Clinical Features of DRESS

- Fever, leukocytosis, eosinophilia
- **Facial Edema**
- Generalized eruption: erythematous papules, morbilliform, bullae, purpura, erythroderma, pustules
- Lymphadenopathy
- Myositis
- **LFT Abnormalities**
- Lung, kidney, heart, thyroid involvement
Fig 1. Algorithm for the diagnosis, management, and treatment of drug reaction with eosinophilia and systemic symptoms syndrome.

Treatment of DRESS

- Withdraw offending medication
- High dose systemic steroids (1-2mg/kg tapering dose over 1-3 months)
- LFTs should be followed until normal
- Supportive care
Case 11

• 56 yo WM presents with onset of complete body rash. States that he was given a steroid shot for a rash he had on his hip three weeks ago. Rash improved but returned one week ago. Only medication is lisinopril for HTN. No changes to detergent or soap.
Which of the following is the most common cause of erythroderma?

A. Atopic Dermatitis
B. Psoriasis
C. Cutaneous T-cell Lymphoma
D. Drug induced
E. Idiopathic
Erythroderma

- Psoriasis
- Pityriasis rubra pilaris
- Atopic Dermatitis
- Seborrheic Dermatitis
- Drug Eruption
- CTCL
- Infectious
- Autoimmune bullous disorder
<table>
<thead>
<tr>
<th>Primary Skin Diseases</th>
<th>Malignancy</th>
<th>Other</th>
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<tbody>
<tr>
<td>Psoriasis</td>
<td>Cutaneous T-cell lymphoma (CTCL)</td>
<td>Drug hypersensitivity reaction</td>
</tr>
<tr>
<td>Atopic dermatitis</td>
<td>Systemic lymphoma</td>
<td>Dermatomyositis</td>
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<tr>
<td>Pityriasis rubra pilaris</td>
<td>Leukemia</td>
<td>Hepatitis</td>
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<td>Contact dermatitis</td>
<td>Myelodysplastic syndrome</td>
<td>HIV</td>
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<tr>
<td>Chronic actinic dermatitis</td>
<td>Solid organ malignancy</td>
<td>Graft vs Host Disease</td>
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<tr>
<td>Bullous pemphigoid</td>
<td></td>
<td>Omenn syndrome</td>
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<tr>
<td>Pemphigus foliaceus</td>
<td></td>
<td>Dermatophyte infection</td>
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<tr>
<td>Ichthyoses</td>
<td></td>
<td>Crusted scabies</td>
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</tbody>
</table>
Erythroderma Pearls

- Etiology can be difficult to determine. History is most important. Can be idiopathic but most commonly associated with prior dermatoses.

- Biopsy necessary but often not helpful. Flow cytometry might be needed to rule out cutaneous T-cell lymphoma.

- Treatment will vary based on etiology.

- Wet wraps with topical steroids ok for almost any etiology.

- Cyclosporine instead of prednisone for erythrodermic psoriasis.
Summary

- Primary care physicians will manage a large of number of common skin diseases. It is important to be comfortable managing mild atopic dermatitis and psoriasis with topical regimens and recognizing when referral to a specialist for systemic treatment might be indicated.

- It is important to understand the systemic work-up needed for cutaneous presentations of psoriasis, sarcoidosis, and lupus.

- Purpura can result from vasculopathy and/or vasculitis. It is important to understand the different etiologies and work-up needed for patients presenting with purpura.

- It is Herpes until proven different in the hospital setting!

- DRESS—check LFTs, thyroid studies, and EKG/Echo

- Etiology of erythroderma is difficult to determine on biopsy. History, history, history!!