Pennsylvania Allergy, Asthma and Immunology Society
June 25, 2017

Dermatologic Conditions Missed by the Allergist

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Disclosure

Research and Educational Grants (to Winthrop University Hospital)
- Baxter
- Genentech
- Merck

Consultant
- Church and Dwight, Co., Inc
- Regeneron

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Objectives:

1. Discuss common dermatologic conditions presenting in the allergists' practice
2. Discuss some differentiating features of these dermatologic diseases

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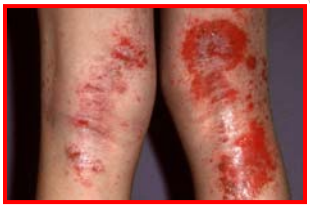
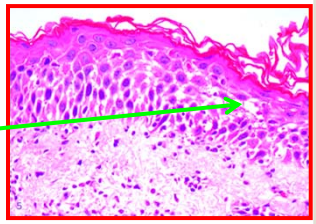
Definition of Eczema

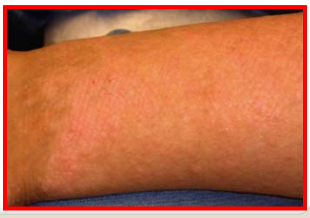
Inflammatory skin reaction
- **Itching**
- **Erythema**
- **Scaling**
- **Clustered papulo-vesicles**



Histology
- **Always present at some stage of eczema:**
  - **spongiosis with acanthosis**
  - **superficial perivascular, lympho-histiocytic infiltrate**



Secondary changes from scratching
- **Excoriations/erosions**
- **Hemorrhage**
- **Lichenification**
- **Secondary infection**



Inflammatory Skin Disorders

- **Dermatitis and Eczema**
  - Atopic D, Contact D, Seborrheic D, Pruritus, Nummular Eczema, Erythroderma, Lichen Simplex Chronicus/Prurigo Nodularis, Dyshidrosis, Pityriasis Alba
- **Papulosquamous disorders**
  - **Psoriasis**
  - **Parapsoriasis**
    - **Acute: Pityriasis lichenoides et varioliformis acuta**
    - **Chronic: Pityriasis Lichenoides Chronica**
    - **Lymphomatoid Papulosis**
  - ***Pityriasis***
    - ***Pityriasis Rosea***
    - **Pityriasis Rubra Pilaris**
  - ***Lichenoid***
    - **Lichen Planus**
    - **Lichen Nitidus**
- **Drug Eruption:** SJS, TEN, E Nodosum
- **Other Erythemas:** E. Annulare, E Centrifugum, E Marginatum, E Toxicum, Necrolytic Migratory Erythema

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Cutaneous T-Cell Lymphoma (Mycosis Fungoides)

Stages:

- **Patch (atrophic or nonatrophic)**
  - **Often goes on for many years**
  - **Patches with thin, wrinkled quality, often with reticulated pigmentation**
  - **Pruritus varies**
    - **minimal or absent**
    - **common in premycotic phase**
    - **may precede MF by years**
  - **Often on lower trunk & buttocks**
- **Plaque**
- **Tumor**

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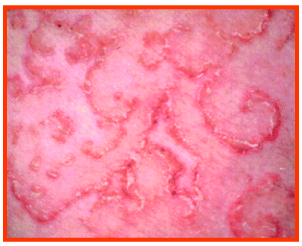
Netherton syndrome



- Rare AR genodermatosis
  - Erythroderma
  - Trichorrhexis invaginata (bamboo hair)
  - Ichthyosis linearis circumflexa (ILC)
  - Atopic diathesis
  - Failure to thrive

- May have immunologic abnormalities
  - Transient neutrophil function defects
  - Impaired cellular & immune responses
  - Raised complement levels (C3 & C4)

circinate, polycyclic plaques bordered by a characteristic double-edged scale

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Irritant Contact Dermatitis

Primary diagnostic criteria

- **Macular erythema, hyperkeratosis, fissuring with less vesiculation**
- **Glazed, parched or scalded**
- **Heal promptly on withdrawal offending agent**
- **Patch test (-)**

Minor objective criteria

- **Sharply circumscribed**
- **Evidence of gravitational influence (dripping effect)**
- **Less tendency to spread**

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Papulosquamous Disorder

Papules +/- Plaques and scales
(scaly papules and plaques)

- **Psoriasis** (red, scaly lesions)
- **Parapsoriasis** (resembles psoriasis)
  - Large Plaque Parapsoriasis
  - Small Plaque Parapsoriasis
  - Pityriasis Lichenoides
    - Pityriasis lichenoides et varioliformis acuta
    - Pityriasis lichenoides chronica
  - Lymphomatoid papulosis
- ***Pityriasis*** (flaking or scaling)
  - *Pityriasis* Rosea
  - *Pityriasis* rubra pilaris
- ***Lichenoid*** *(resembles lichen: organisms consisting of a symbiotic association of a fungus*
  - Lichen Planus
  - Lichen Nitidus

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Psoriasis

- **Plaque psoriasis**
- **Guttate psoriasis**
- **Pustular psoriasis**
- **Nail psoriasis**
- **Erythrodermic psoriasis**

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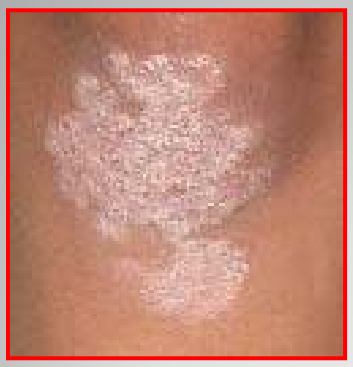
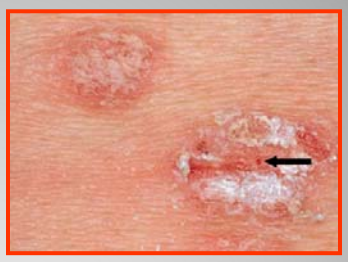
Psoriasis



Plaques typically have dry, thin, silvery-white or micaceous scale



Auspitz sign
Removing scale reveals a smooth, red, glossy membrane with tiny punctate bleeding

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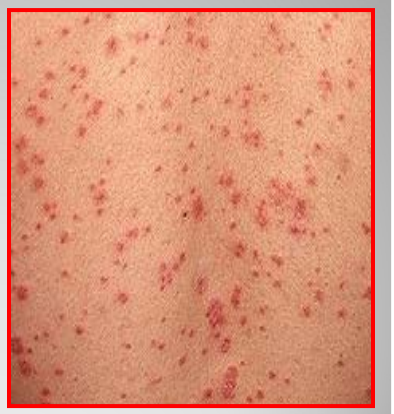
Guttate psoriasis

- Abrupt acute eruption of small (< 1 cm) psoriatic lesions
- Typically child or young adult with no history of psoriasis
- Primarily the trunk
- Strong association with recent strep infection with serologic evidence(26-58 %)



Telfer NR; Chalmers RJ; Whale K; Colman G The role of streptococcal infection in the initiation of guttate psoriasis. Arch Dermatol 1992 Jan;128(1):39-42

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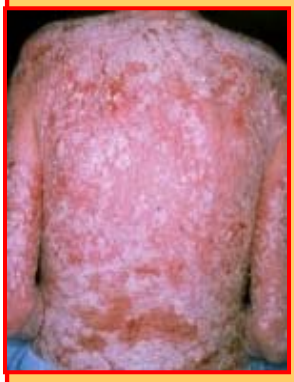
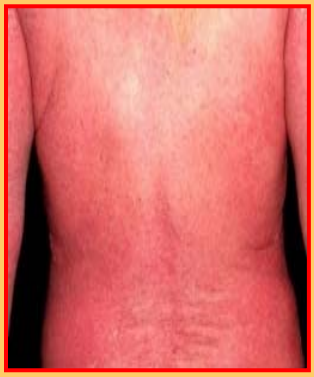
Pustular Psoriasis

Localized

Generalized



Erythrodermic



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Parapsoriasis

A Complex Issue

- **Resembles Psoriasis (red, scaly)**
- **Unrelated to pathogenesis, histopathology or treatment**

- **Large Plaque Parapsoriasis**
- **Small Plaque Parapsoriasis**
- **Pityriasis Lichenoides**
  - **Pityriasis lichenoides et varioliformis acuta**
  - **Pityriasis lichenoides chronica**
- **Lymphomatoid papulosis**

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Parapsoriasis

T-cell–predominant skin infiltrates

- Large plaque parapsoriasis
  - indolent & progresses over years, sometimes decades
  - Treatment may prevent progression to CTCL (~ 10%)
- Small plaque parapsoriasis
  - benign; rarely progresses
  - lasts several months to years
  - can spontaneously resolve

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Mucha-Habermann disease

Pityriasis lichenoides et varioliformis acuta

- Abrupt onset of multiple papules on trunk, buttocks, proximal extremities
- Rapidly progress to vesicles & hemorrhagic crusts
- Minor constitutional symptoms fever, malaise & myalgias

Pityriasis lichenoides chronica

- May develop over days
- Same distribution

Lichenoid skin eruptions

- Subcategory of papulosquamous skin disease
- Scale often subtle; papules tend to remain small & discrete
- Occasionally, confluent plaques may form

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Lichen Planus

A disease characterized by "P-words":

- Plentiful
- Pruritic
- Polished
- Purple
- Polygonal
- Planar
- Papules

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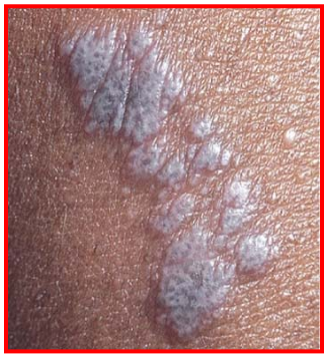
Lichen planus

- Arranged in groups of lines or circles
- Flexor surfaces of upper extremities
- Wickham stria: fine, white lines on papules
- Pruritus common but varies in severity
- > 50% resolve within 6 months
  85% subside within 18 months
- Other areas of involvement:
  - Mouth: white or gray streaks forming linear or reticular pattern
  - Genital
  - Nail plate thinning, grooving, ridging, pterygium
  - Cicatricial alopecia



Pyoderma Gangrenosum

- **50% have systemic illnesses**
- **Arthralgias & malaise often present**
- **Commonly associated diseases**
  - inflammatory bowel disease (ulcerative colitis or Crohn's)
  - seronegative or seropositive polyarthritis
  - hematologic disorders (leukemia, preleukemia, monoclonal gammopathies (primarily immunoglobulin A)
  - less common: psoriatic arthritis, osteoarthritis, spondyloarthropathy; hepatitis; SLE

Dermatitis Herpetiformis

- Young to middle age
- Intensely pruritic
- Symmetrically grouped papules & vesicles
- Elbows, knees, buttocks, scapula, scalp

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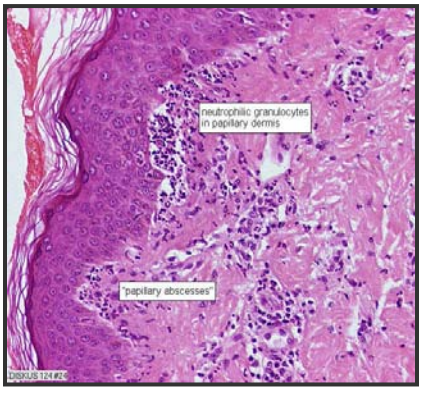
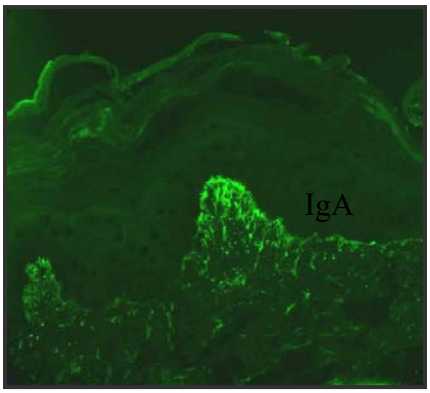
Dermatitis Herpetiformes



Neutrophils in tips of dermal papillae
Microabscesses containing neutrophils & eosinophils



DIF: ~90% (+) Granular deposition of IgA at dermal papillae & occ along the DE border

DERMATOPATHOLOGY REPORT

DIAGNOSIS:

RIGHT LOWER LEG - **LEUKOCYTOCLASTIC (ALLERGIC) VASCULITIS**
Note: Please refer to the companion immunofluorescence report (HI09-832).

CLINICAL IMPRESSION:
POSSIBLE VASCULITIS

GROSS DESCRIPTION :
PUNCH, 0.4X0.4X0.6CM

MICROSCOPIC DESCRIPTION:
There is a superficial and mid-dermal perivascular and interstitial mixed inflammatory cell infiltrate that contains lymphocytes, histiocytes, eosinophils and many neutrophils. There are nuclear dust, fibrin within the vessel walls and numerous extravasated erythrocytes. (82A-MT)

DIAGNOSIS:

RIGHT LOWER LEG - POSITIVE DIRECT IMMUNOFLUORESCENCE STUDY FOR VASCULITIS
Note: There are deposits of C3 and IgM about vessels. This could represent leukocytoclastic vasculitis. Please refer to the companion report of light microscopic findings (HD09-176034).

CLINICAL IMPRESSION:
RASH/POSSIBLE VASCULITIS

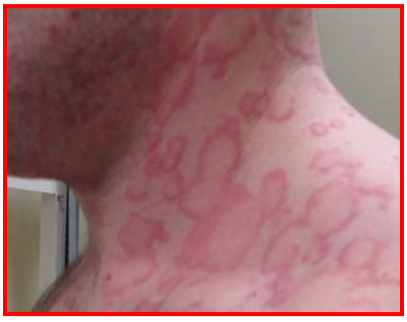
GROSS DESCRIPTION :
PUNCH, 0.3X0.3X0.4CM

MICROSCOPIC DESCRIPTION:
Specimen is treated with a panel of four immunoglobulins (IgG, IgA, IgM, and C3).

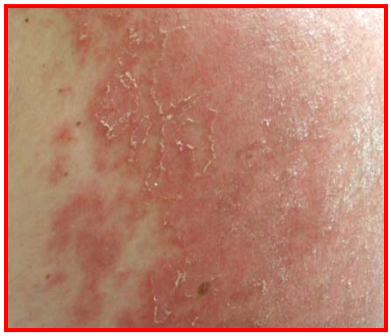
An H&E stained section shows perivascular dermatitis. There are deposits of C3 and IgM about vessels. IgG and IgA are not present in this area. All four antibodies tested are not present in the epidermis, basement membrane zone, or dermis.

Subacute Cutaneous Lupus Erythematosus

Annular Polycyclic



Papulosquamous



- 60 % of SACL have (+) DIF on lesional skin
- Some (usually those with SLE features) have (+) Lupus Band Test

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Chronic Pruritus

Dermatologic
With Primary Rash

- Xerosis
- Atopic Dermatitis
- Contact dermatitis
- Scabies
- Psoriasis
- Lichen Planus

Neurologic

- Notalgia Paraesthetica
- Brachioradial Pruritus
- Multiple sclerosis
- Post Herpetic itch

Systemic

- Thyroid disease
- Primary biliary cirrhosis
- Chronic renal failure
- Hodgkin's disease
- Polycythemia vera
- HIV
- Metabolic states
- Drugs

Psychogenic

- OCD
- substance abuse
- delusional parasitosis

Idiopathic

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Severity of Pruritus 0-10:

6 distracts from activities
8 awakens from sleep
10 is the worst imaginable

SEVERE	MODERATE
Scabies, mite infestation	Psoriasis
Pediculosis, insect bites	Seborrheic dermatitis
Contact & atopic eczema	Pityriasis Rosea
Urticaria	Sunburn
Prickly Heat	Fungal disease
Lichen Planus	Asteatotic skin
Dermatitis Herpetiformis	Urticaria pigmentosa

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Brachioradial Pruritus

- Localized to outer aspect of elbow & adjacent lower & upper arms but can generalize
  - may be on ACE-I
  - may have cervical pain
  - sun exacerbated but worse at end of summer rather than the beginning

- Causes:
  - Sunlight induced chronic episodic pruritus ? "solar pruritus"
    - nerve damage of cutaneous branch of radial nerve or cervical spine irritation

Walcyk PJ *Br J Dermatol* 1986;115:177-80
Bech-Thomjsen N *Acta Derm Venereol (Stockh)* 1995;75:488-9

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Neurologic Causes of Intense Itching

Notalgia Paraesthetica

- Persistent burning pruritus localized in mid-scapular area
  - but often widespread including scalp
- Mild lichenification & pigmentation
- May be a type of localized sensory neuropathy
  (nerve entrapment of posterior rami of spinal nerves at T2-T6)
- Capsaisin cream may be effective

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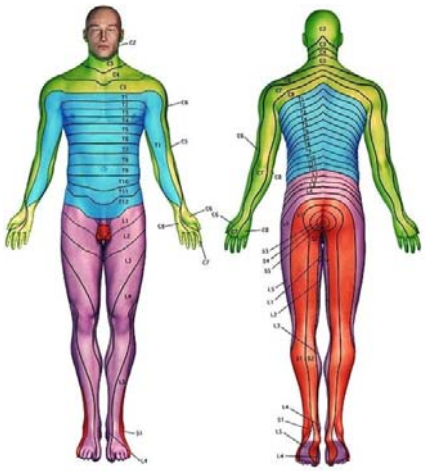
Neurologic Causes of Intense Itching

- Association with other sensory symptoms
- Dermatomal distribution
- Presence of other neurologic sensory signs
- Presence of nerve damage
  - Motor deficits
  - Autonomic dysregulation



Yosipovitch and Samuel. Dermatol Ther 2008

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Systemic Causes of Pruritus without Primary Skin Lesions

- Chronic renal disease
- Cholestasis
- Gluten enteropathy
- Hematologic disease
  - Iron deficiency
  - Polycythemia vera
- Endocrine diseases
  - Hypothyroidism
  - Hyperthyroidism
  - Diabetes mellitus
- Infections:
  - HIV
  - Hep B, Hep C
- Malignancy
  - Leukemia
  - Lymphoma
  - Multiple myeloma
- Pregnancy
- Food/Drug

***The incidence of generalized pruritus associated with significant internal disease is difficult to assess but is estimated to be ~10%**

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Drugs reported to cause pruritus

- Opium Alkaloid
- Calcium channel blockers
- ACE-inhibitors
- Hydrochlorothiazide
- Simvastatin
- Niacinamide
- CNS stimulant/depressant
- Cimetidine
- Aspirin
- Allopurinol
- Chloroquine
- Sulfonamides
- Amiodarone,
- Quinidine
- Estrogens

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Generalized Pruritus That Can Precede Skin Disease

- Bullous Pemphigoid
- Mycosis Fungoides
- Polycythemia vera
- Hodgkin's Disease
- Dermatitis herpetiformis
- Dermatomyositis

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Pruritus workup: Labs

- Screening labs that are reasonable:
  - CBC with Diff, CMP, HIV, Hep B/C, TSH
- Screening labs that may be reasonable:
  - Peripheral smear, Iron studies
- As indicated:
  - B12, Folate, Stool O&P, SPEP/UPEP, drug screen, CXR, colonoscopy, 5-HIAA, MRI of brain
  - Skin biopsy
  - Indirect serum immunoflorescence
  - Sinus X Ray, ANA, specific IgE, UA

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