Dermatologic Manifestations of Internal Disease

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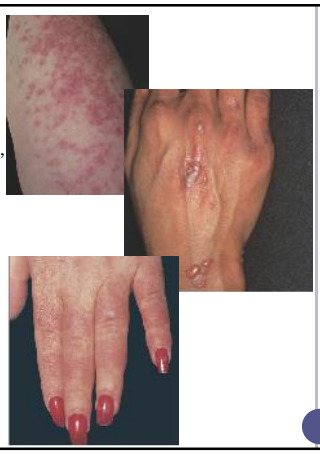
Lupus

- Acute – "butterfly" malar rash, transient, follows sun exposure
- Lesions on hands often spare knuckles
- Evaluate for internal disease (ANA/ENA)
- +/- oral ulcers
- Nephritis concerning complication (usually dsDNA+)



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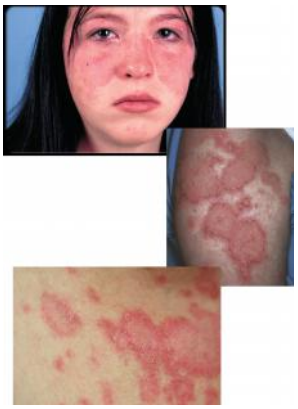
- Bolognia et al. Dermtext.com
- The internet

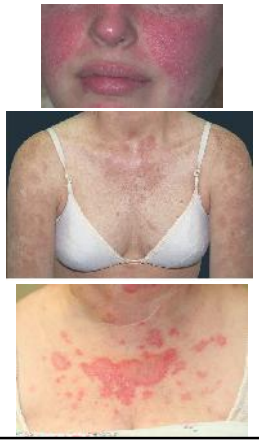
Lupus

- SCLE – photosensitive, photodistributed
- Lesions may appear annular, eczematous, psoriasiform.
- Not scarring
- 15-50% progress to SLE
- Med induced: HCTZ, NSAIDs, diltiazem, and terbenafine
- Usually anti-Ro +



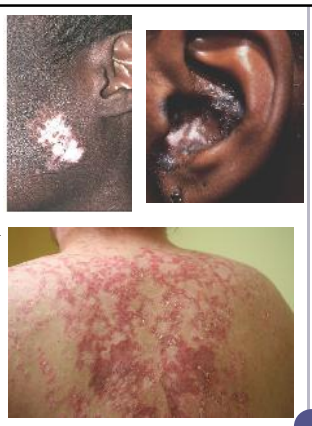
Lupus Erythematosus

- Multisystem, autoimmune disorder
- Female > male 6:1
- SLE prevalence 4 fold higher in AA females
- SLE criteria –rash photosensitivity, oral ulcers, arthritis, serositis, renal disorder, neurologic disorders, hematologic disorder, +ANA.



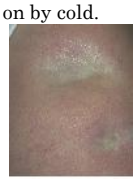
Lupus

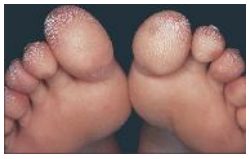
- Discoid Lupus – face, scalp, and ears
- Can occur on mucosa
- Can scar, also follicular plugging and scarring alopecia
- Long standing lesions can dvp SCC
- Only 5-10% dvp SLE



Lupus

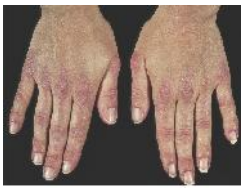
- More rare types –
  - Tumid – no epidermal change. Dermal process. Can appear like urticarial plaques on face and trunk
  - Lupus panniculitis – intense inflammation in the fat with indurated and depressed areas.
  - Lupus pernio – red/dusky/purple papules on fingers, toes, elbows, and lower legs. Brought on by cold.

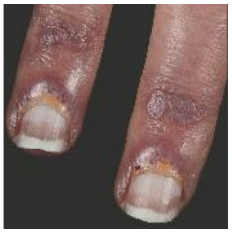




Dermatomyositis

- ¼ of adult pts have assoc malignancy
- Females 2x> males
- can overlap with other collagen vascular diseases
- Poikiloderma, violaceous color (elbows, knees with fine scale)
- Photodistribution
- Periungal telangiectasia and dilated capillary loops. Ragged cuticles



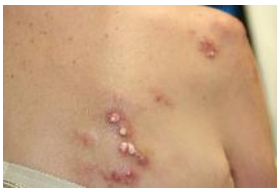


Lupus

- Diagnosis – Clinical, skin biopsy and DIF
- Eval for SLE – diffuse non-scarring alopecia, periungal telangiectasia, Raynauds, livedo retic, vasculitis, lymphadenopathy, ANA (and ENA), UA, CBC with diff, platelets, CMP, ESR.
- Rheumatology referral
- TX: topical/intralesional steroids, SUN PROTECTION,hydroxychloroquine, dapsone, methotrexate. With SLE - prednisone azathiaprine, cyclosporine, …

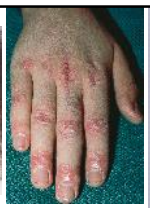
Dermatomyositis

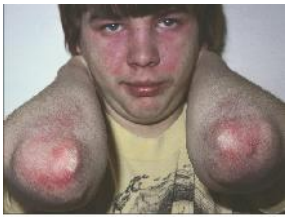
- Heliotrope sign - eyelids violaceous and puffy
- Gottrons sign – violaceous poikiloderma over knuckles, elbows, and knees
- Gottrons papules – over knuckles
- Erosions and bullae are a bad sign
- Calcinosis cutis



Dermatomyositis

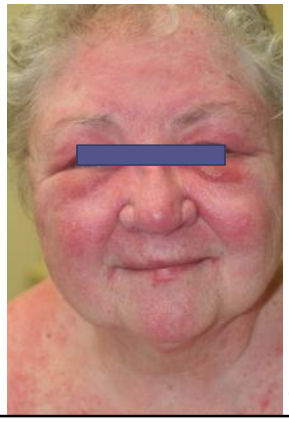
- Connective tissue-vascular disease
- Autoimmune
- Symmetric proximal, extensor, inflammatory myopathy and characteristic skin eruption.
- Polymyositis – muscle only, spares the skin
- Amyopathic – skin only, no muscle





Dermatomyositis

- Malaise and fatigue
- Muscle soreness/pain and decreased strength
- Can be assoc with interstitial lung dz and cardiac dz
- Rarely drug-induced
- DX: skin biopsy, ANA, ENA, muscle enzymes, MRI, LFT's, LDH, 24 hour creatinine, MI2
- Malignancy eval

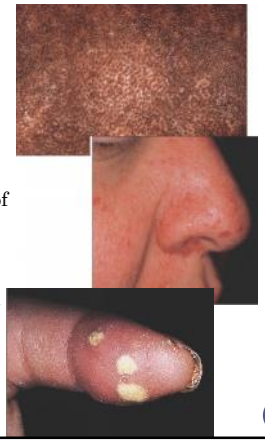


Dermatomyositis

- Characteristics of subtypes:
  - Juvenile – Calcinosis cutis is common, usually no malignancy
  - Adult – Common malignancies ovarian, breast, colon, lung, and pancreas
  - Amyopathic – no myopathy, skin only. Can have late onset myopathy after 15mo-6 years. 14% have malignancy, 13% have ILD.
- Treatment: sunscreen, topical steroids, hydroxycholorquine, methotrexate, azathioprine, IVIG

Scleroderma

- Skin findings:
  - Diffuse hyperpigmentation, can have leukoderma except perifollicular ("salt and pepper")
  - Matted telangiectasias of face, lips and palms
  - Sclerodactyly
  - Calcinosis cutis – distal
  - Raynaud's and ulcers on fingers common
  - Dry skin
  - Pruritus



Systemic Sclerosis (Scleroderma)

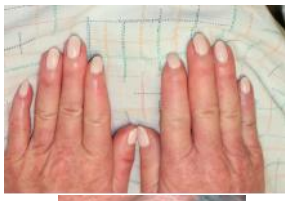
- Collagen vascular dz primarily affecting females (30-50yo)
- Symmetric induration of the skin with involvement of distal areas
- Systemic involvement – esophagus, lungs, heart, and kidneys
- May be fatal (lung involvement)

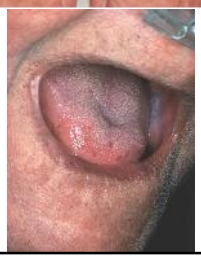
Scleroderma

- Labs:
  - ANA +, and Scl-70 (diffuse) or anti-centromere (limited)
- TX: CCB's for Raynauds (nifedipine). Avoid cold and tobacco.
- ACE inhibitors (for renal involvement?)
- ?Prednisone, ?methotrexate, imatinib in trials

Systemic Sclerosis

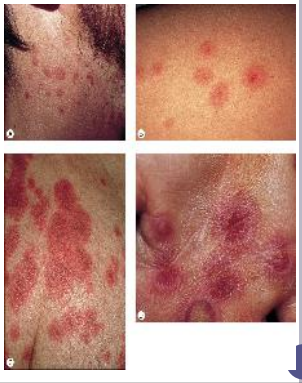
- Limited vs Diffuse
- CREST: Calcinosis cutis, Raynaud's phenomenon, esphogeal dysmotility, sclerodactyly, and telangiectasia (matted).





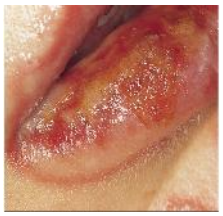
Erythema Multiforme

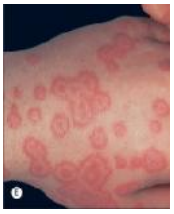
- Strongly assoc. with a preceding infection Recurrent cases likely HSV.
- Can also be drug induced, radiation induced, and idiopathic
- Appears targetoid with dusky center. Can be bullous. Common on the palms and soles, +/- oral involvement, acral distribution.



Erythema multiforme

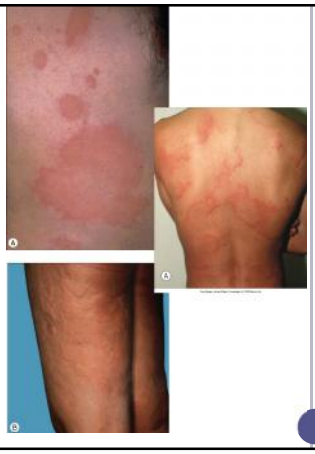
- Minor – self limited, recurrent, usually in young adults (spring and fall) and lasts 1-4 weeks.
- Major – more extensive w/ mucosal involvement. Previously thought to be a precursor to SJS.
- Treatment is supportive (prednisone may provoke recurrences).





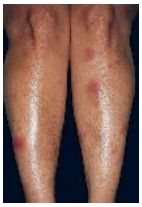
Urticaria

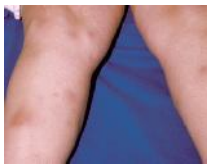
- Vascular rxn characterized by the appearance of wheals, generally surrounded by a red halo or flare
- Severe itching, stinging, or prickling sensation.
- May be annular or polycyclic.
- Individual wheals rarely last >12 hours.



Erythema Nodosum

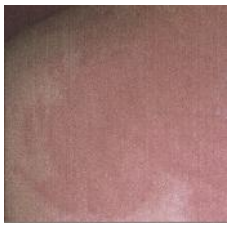
- Bilateral, symmetric panniculitis with deep, tender, bruise-like nodules
- Pre-tibial
- Can have malaise, leg edema, arthralgias, fever, HA, conjuctivitis, GI complaints

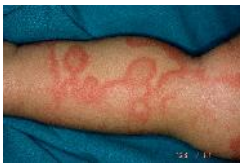




Urticaria

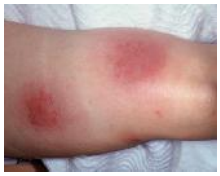
- Acute – resolves in days to weeks, with complete resolution of symptoms within 6 weeks.
- Chronic - >6 weeks.
- Females 2x more affected than males. More common in adults





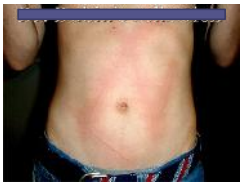
Erythema nodosum

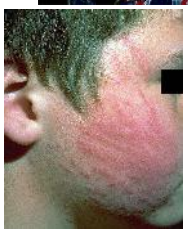
- Causes: strep infection, TB, intestinal infection, fungal infection, sarcoidosis, IBD, drugs
- Tx: identify trigger, rest, elevation, anti-inflammatories



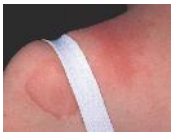
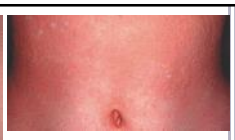
Urticaria

- Triggers: drugs are most common
- Acute >50% idiopathic followed by URI, drugs, and foods
- Chronic 60% "ordinary" (autoimmune, pseudoallergic, infection related, and idiopathic) then physical and vasculitic causes



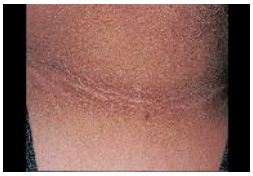


Urticaria

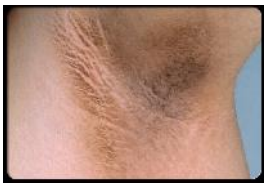



- Physicial urticarias: dermatographism, cold, heat, cholinergic, aquagenic, solar, vibratory, and exercise induced.
- +/- allergic rhinitis, asthma, nasal polyps and food induced anaphylaxis
- Food: acute.
- Can do food diary
- Infections: URI, strep, Hep B/C, mono
- Emotional stress, menthol, neoplasms, pollen, and alcohol

Acanthosis Nigricans



- Type II – familial, present at birth
- Type III – neck, axillae, groin. Occurs in obesity, acromegaly and gigantism, Cushing's, DM, hypothyroidism, Addison's, hyperandrogenic states.
- Drugs: OCP's, testosterone, and glucocorticoids.



Urticaria

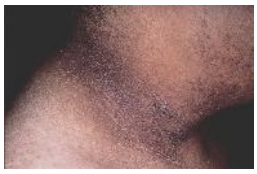
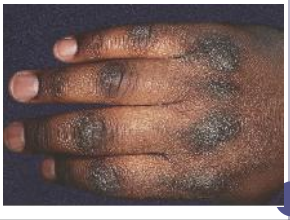
- D/DX: urticarial vasculitis, BP, EM, GA, Sarcoidosis, CTCL which would all last >24 hours
- TX: antihistamines. Consider Singulair. Cold baths, sarna, phototherapy, nifedipine, dapsone, gold, methotrexate.

Acantosis Nigracans

- Tripe palms – thickened velvety palms with pronounced dermatoglyphics. 95% occur in pt's with cancer.
- Lung CA most common if only palms affected. Tripe palms + AN = gastric cancer.

Acanthosis Nigricans

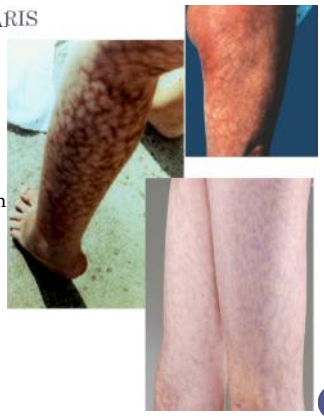
- Grey, brown, or black hyperpigmentation and velvety textured plaques.
- Symmetric distribution.
- Type I – Rare. Assoc with malignancy. Increased extent of involvement.
  - Increased suspicion >40yo, not obese with rapid onset

Acanthosis Nigracans

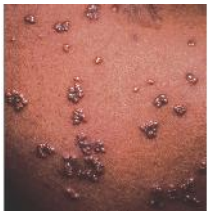
- Treatment – remove malignancy, treat DM, weight loss, Oral retinoids, metformin, tretinoin, calcipotriol, urea, sal acid, CO2 laser and pulsed dye laser may be of benefit.

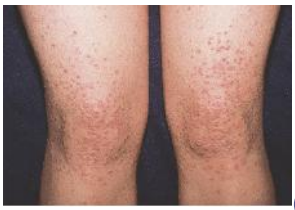
Livedo Reticularis



- Netlike mottled or reticulated pink or reddish blue discoloration.
- Exposure to cold can accentuate.
- +/- coldness, numbness, paresthesia.

Eruptive Xanthoma



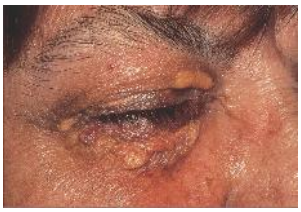


- Small yellowish orange to red-brown papules that appear in crops over entire body
- Markedly elevated triglycerides.
- Also seen with DM, obesity, pancreatits, chronic renal failure, hypothyroidism, estrogens, corticosteriods, or systemic retinoids.

Livedo Reticularis

- Most not assoc with systemic disease, but may be a manifestation of lupus, dermatomyositis, scleroderma, rheumatic fever, rheumatoid arthritis, Hep C, parvovirus B19, syphilis, meningococcemia, pneumococcal sepsis, TB, pancreatitis, breast CA, and on and on……
- Cholesterol emboli can cause. May be uni or bilateral. Usu with cyanosis, purpura, nodules, ulceration or gangrene.

"Xanthelasma" palpebrarum

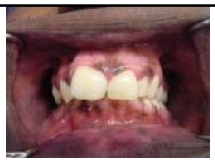


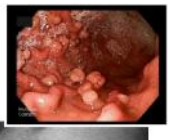
- Most common. Soft yellowish orange oblong plaques on eyelids
- Assoc with other types of xanthoma, but are typically present without any other disease
- >1/2 pt's with normal lipid profile

Xanthomatosis

- Cutaneous manifestation of lipidosis in which plasma lipoproteins and free fatty acids are changed quantitatively.
- Names based on clinical morphology
- Can be associated with genetic diseases
- Morphologies fairly specific for the associated elevated lipid

Peutz-Jegher Syndrome





- AD. Mucocutaneous pigmented macules and hamartomatous polyps of the GI tract.
- Dark brown to black round to oval macules on central face, lips and oral mucosa. Can be seen on hands, feet, tongue, and periumbilical.

Peutz-Jeghers Syndrome

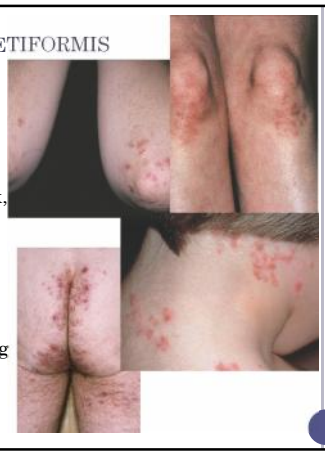
- Polyps in GI tract usually in jejunum and ileum. +/- abdominal pain, obstruction, hemorrhage, and anemia.
- Increased risk for malignancy
- Tx: check stool for blood, colonoscopy, upper endoscopy

Thanks for your Attention!

- Questions???

Dermatitis Herpetiformis

- Symmetric distribution favoring elbows, extensor forearms, upper back, buttocks and knees.
- Urticarial papules, vesicles and blisters.
- INTENSE pruritis, burning or stinging. May only see crusting



Dermatitis Herpetiformis

- Cutaneous manifestation of celiac disease
- Only about 20% have intestinal symptoms
- Males > females 2:1
- Uncommon in AA and Asian populations
- Can be associated with Hashimoto's thyroiditis
- Dx: biopsy and DIF (perilesional), anti-endomysial antibioties, tissue transglutaminase
- TX: gluten free diet!!! Dapsone, topical steriods for itching. GI eval?? Probably