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Collagen Vascular Disease - Considerations for Emergent Management

Joseph H. Hartmann, D.O.

Emergent Management ???

- Chronic conditions with long-term management decisions made by others

but

- Because of their chronicity they present with
  - flares of general disease process
  - end-organ involvement
- Occasionally determine the initial diagnosis

Where are you taking me today?

- Raynaud phenomenon
- Reactive arthritis (Reiter Syndrome)
- Rheumatoid arthritis
- Systemic sclerosis (Scleroderma)
- Systemic lupus erythematosus
- Polymyalgia rheumatica
- Polymyositis / Dermatomyositis
- Vasculitides

Raynaud Phenomenon

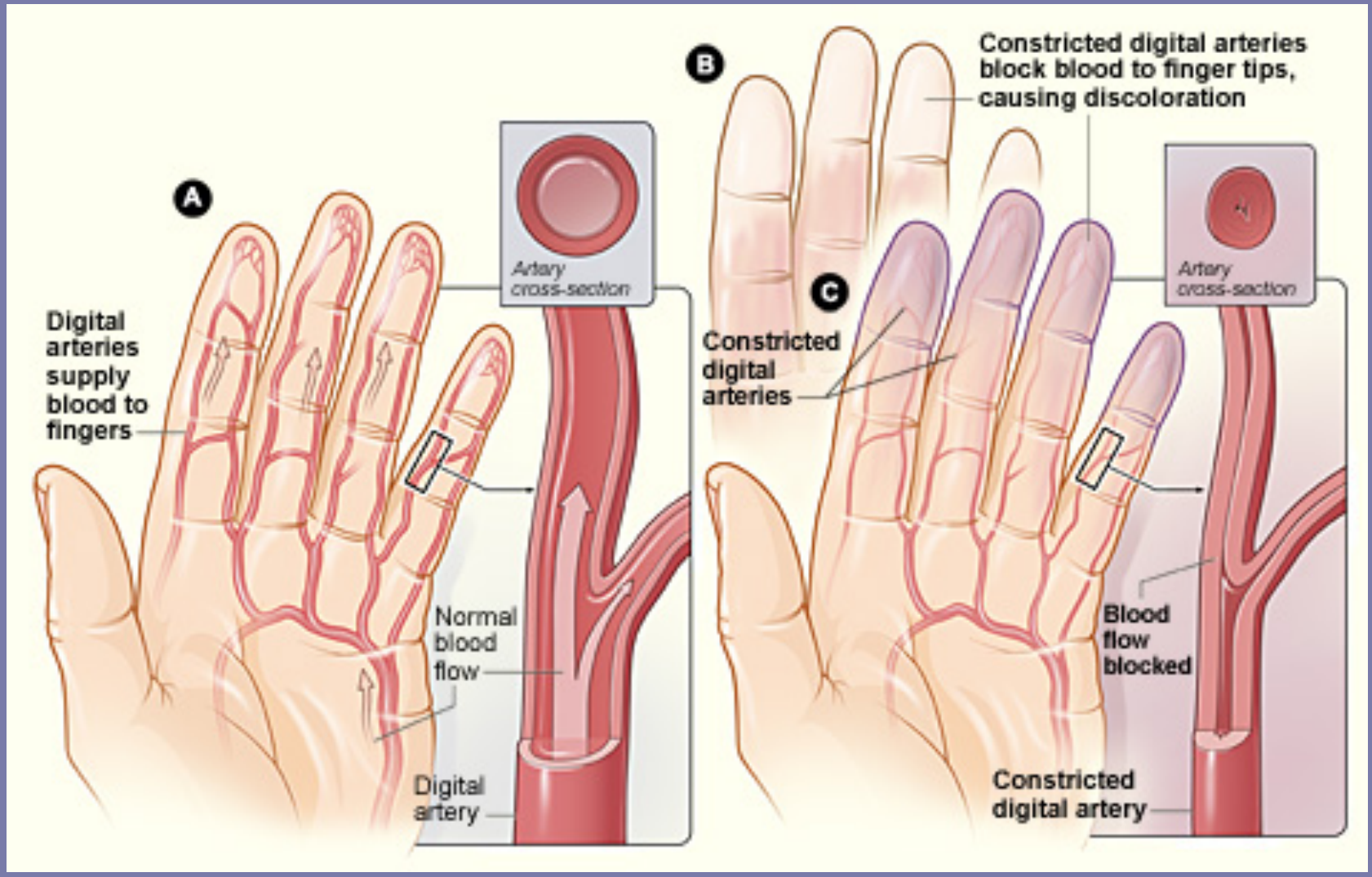
- Definition:
  - Exaggerated vasospasm of digital / precapillary arteries of fingers, toes, ears, nose, knees and nipples
  - Initiated by exposure to cold or emotional stress

Raynaud Phenomenon

- Etiology:
  - Unknown
  - May be first precursor of future connective tissue disorder
- Clinical Presentation:
  - Triphasic
  - Typically begins in one finger then symmetrically spreads to other fingers but usually spares the thumb

Raynaud Phenomenon

- Triphasic progression
  - White – pallor
    - Lack of arterial flow due to vasospasm
  - Blue
    - Cyanosis from blood pooling
  - Red
    - Reactive hyperemia
  - Ischemic phases (white-blue) last 15-20 min



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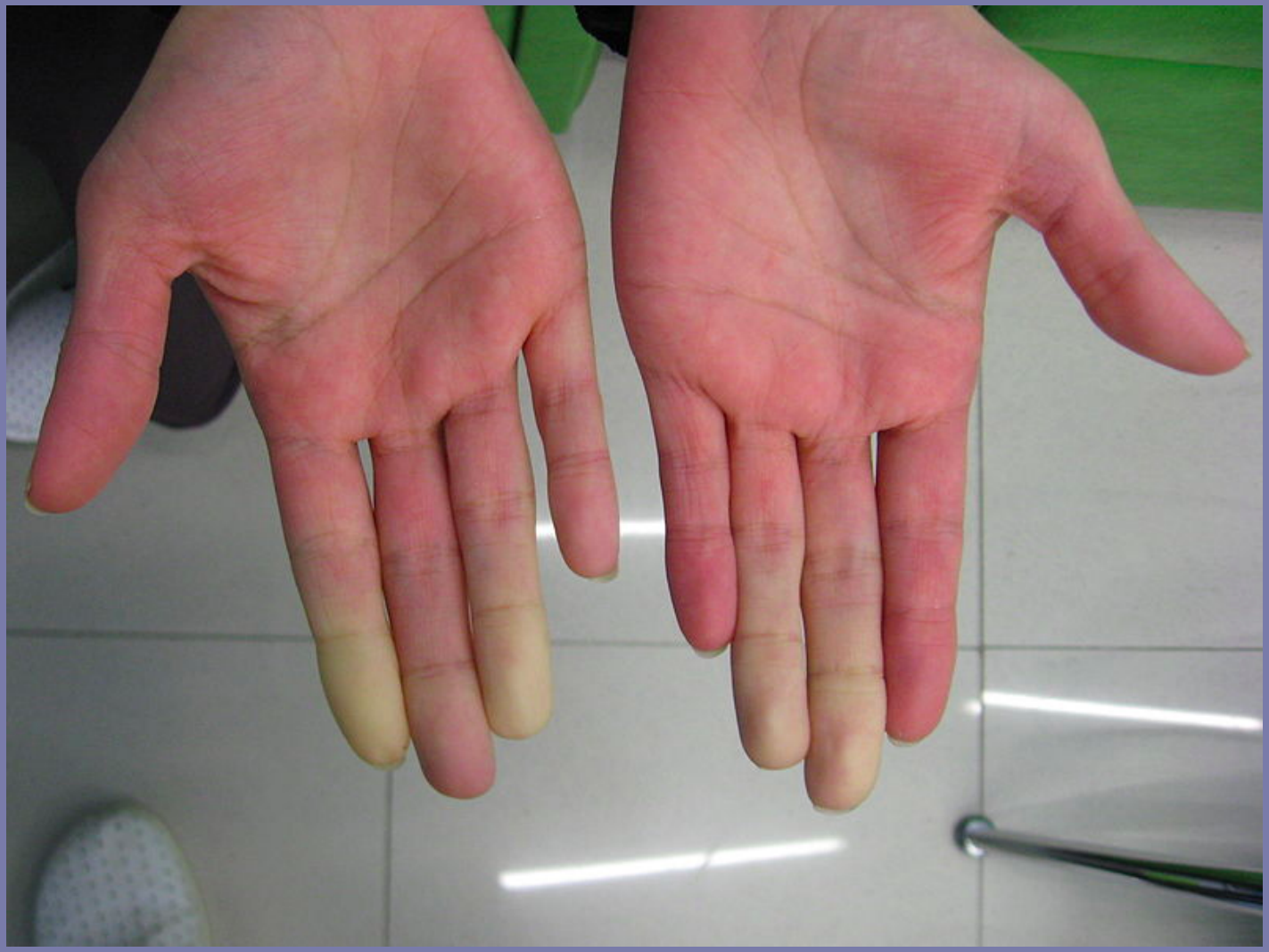

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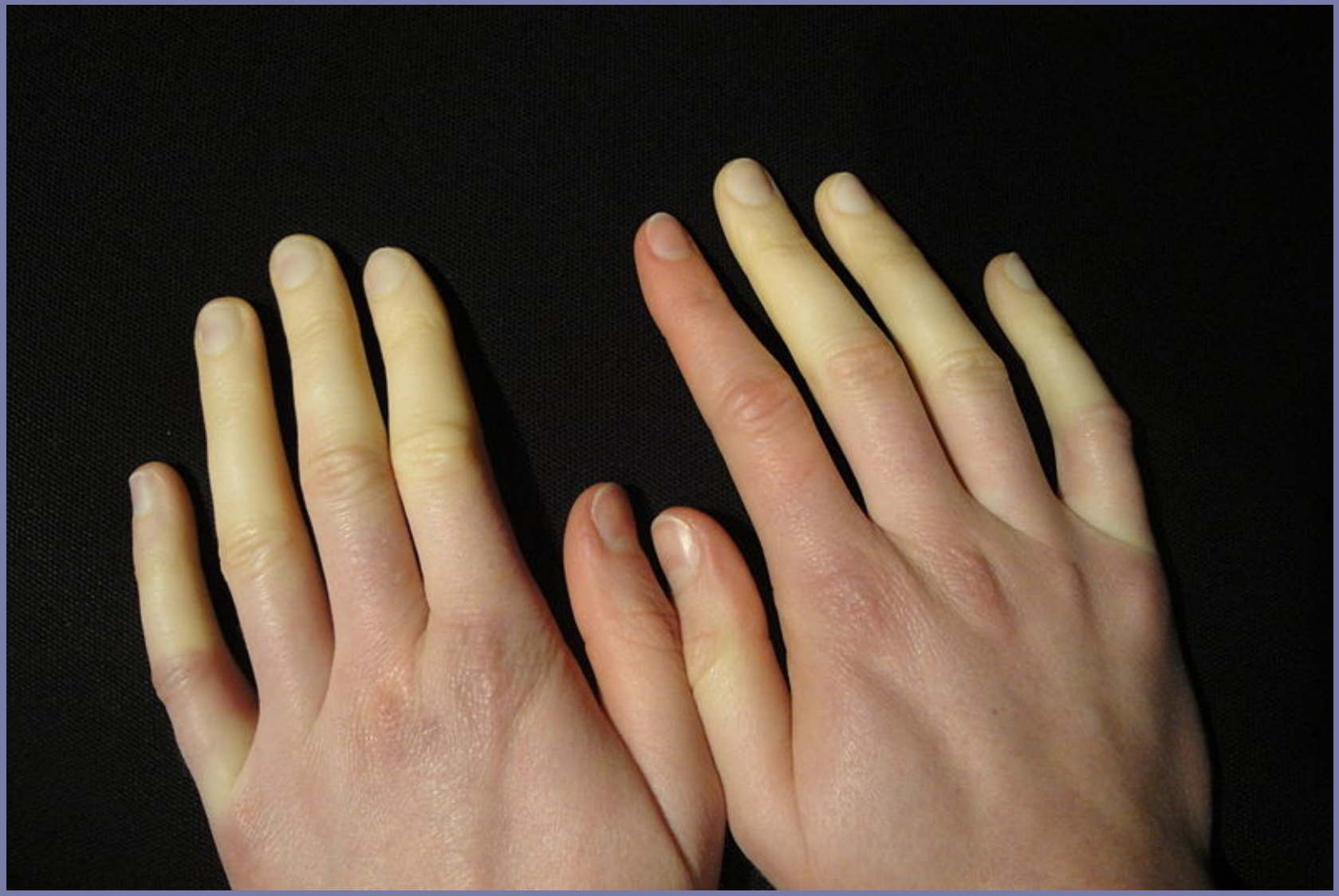

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Raynaud Phenomenon

- Relative temperature shifts may be provocative
- General body chill can trigger
- Fear / anxiety can trigger

Raynaud Phenomenon

- Specific criteria
  - Symmetric episodic attacks
  - No evidence of peripheral vascular disease
  - No tissue gangrene, digital pitting, or tissue injury
  - Negative nailfold capillary examination
  - Negative ANA, normal ESR

Raynaud Phenomenon

- Nailfold capillary microscopy –
  - Place drop of immersion oil on periungual area then examine with ophthalmoscope set at diopter 40 or a dissecting microscope.
  - Should see regularly spaced capillary loops
  - Abnormal findings –
    - Enlarged or distorted capillary loops
    - Relative paucity of loops

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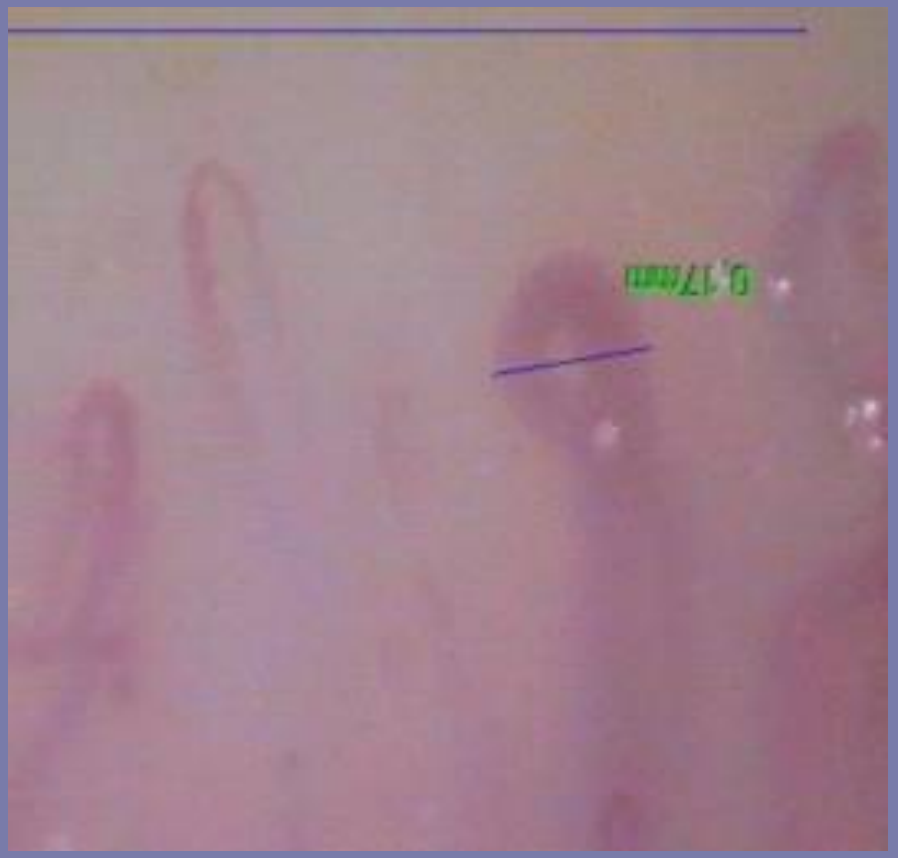

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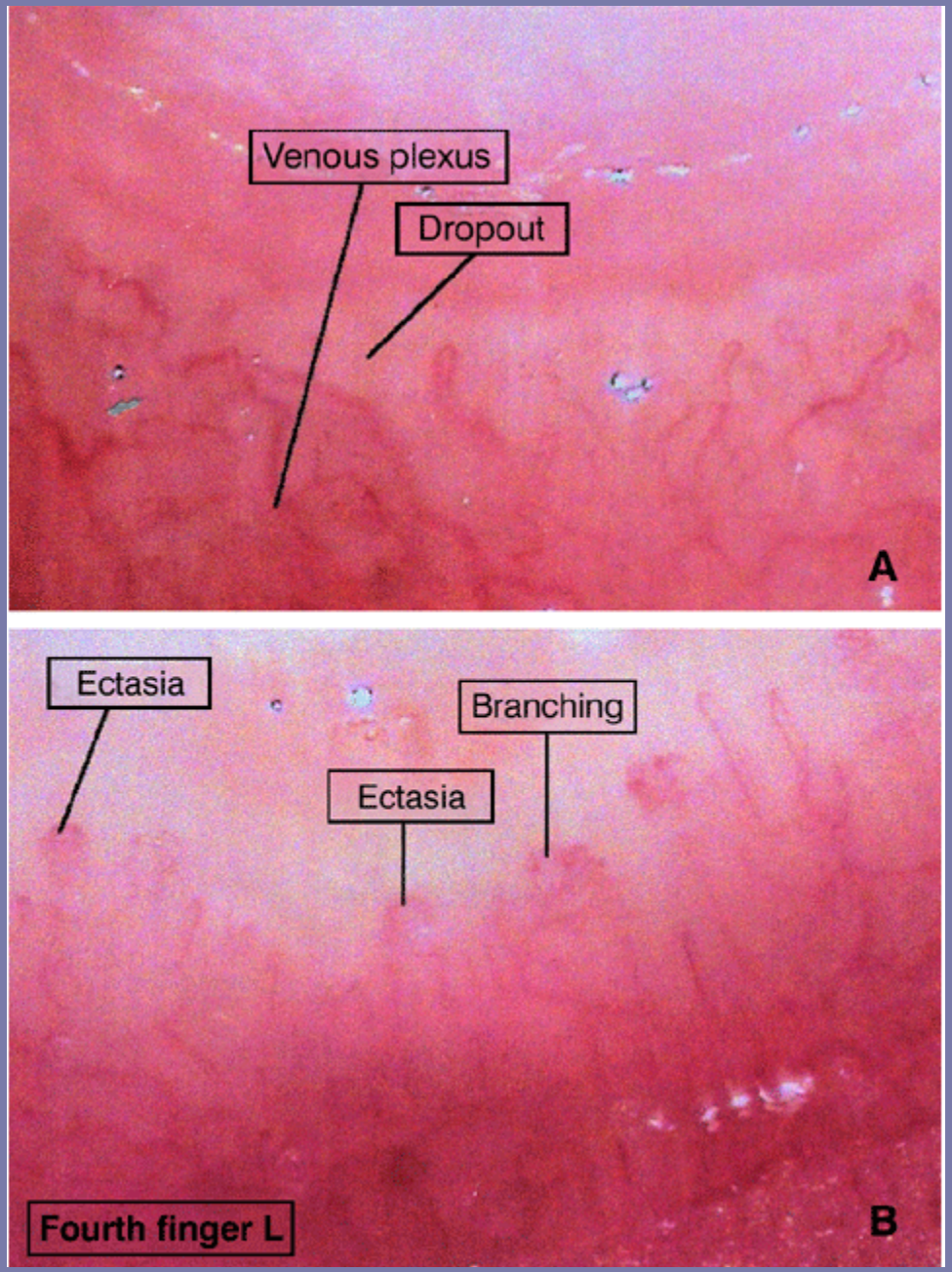

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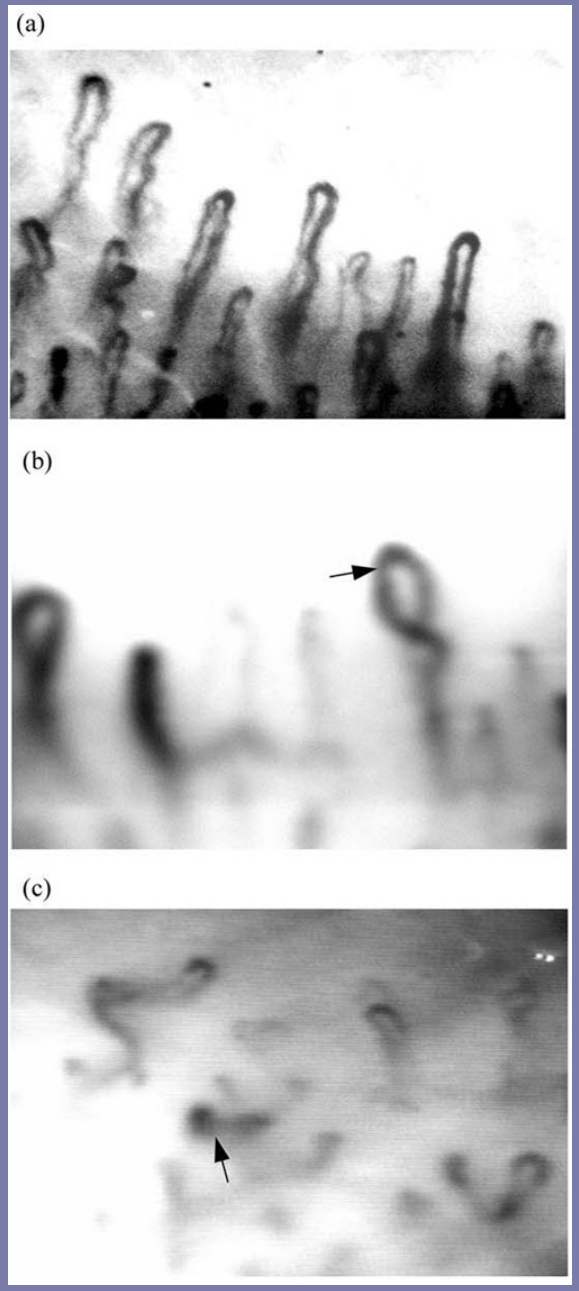

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Raynaud Phenomenon

- Treatment
  - Avoidance strategies
    - Cold, nicotine, sympathomimetics (decongestants, diet pills, herbs containing ephedra)
  - Drug therapy
    - Ca channel blockers, direct vasodilators (NTG, hydralazine, minoxidil), sympatholytics (methyldopa, reserpine, prazosin), prostaglandins, anticoagulation / antithrombotic tx (aspirin, dipyrid-amole, heparin, LMWH)
    - sympathectomy

Raynaud Phenomenon

- Severe ischemia
  - Warm patient (body and digits)
  - Analgesics
  - Antiplatelet tx – aspirin
  - Vasodilator tx
    - Nifedipine extended release 30-60 mg daily
    - Amlodipine 5-10 mg daily
  - Topical NTG
  - Heparin / LMWH 24-72 hrs
  - Temporary chemical sympathectomy
    - Digital or regional block – lidocaine/bupivicaine
  - IV prostaglandin administration

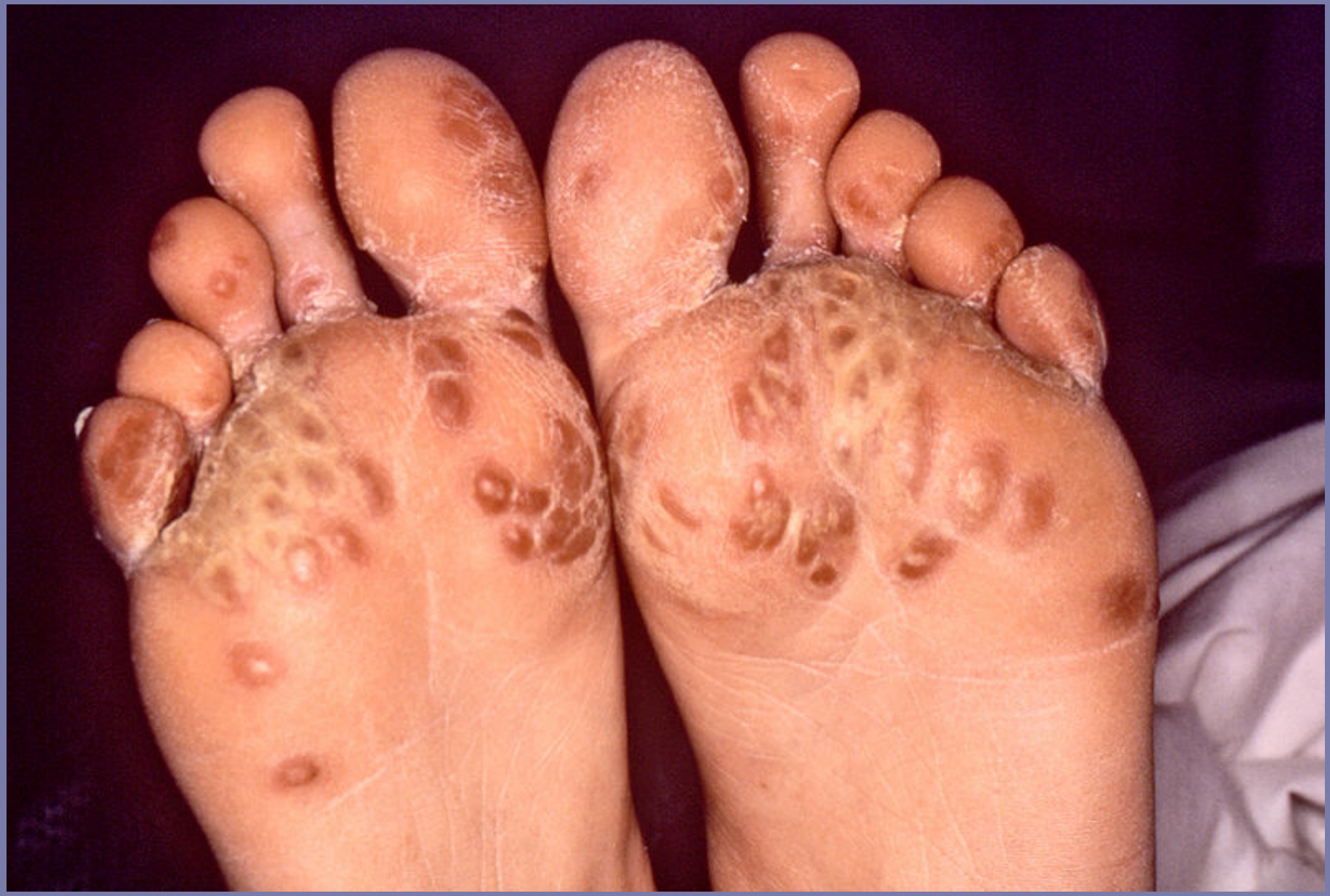
Reactive Arthritis (Reiter Syndrome)

- Definition:
  - Arthritis following a preceding infection without intra-articular presence of the pathogen i.e. not a septic joint

- Etiology:
  - Seronegative (rheumatoid factor negative)
  - Spondyloarthropathy (very likely HLA-B27 pos)
  - Follows a GU or GI infection

Reactive Arthritis

- Clinical Presentation:
  - Male 15 – 35 y/o
  - Asymmetric, oligoarthritis (2-4 joints)
  - Usually involves lower extremeties and sacroiliac joints
  - Skin lesions resembling pustular psoriasis on palms and soles – *keratoderma blennorrhagicum*
  - lesions on glans penis – *balanitis circinata*



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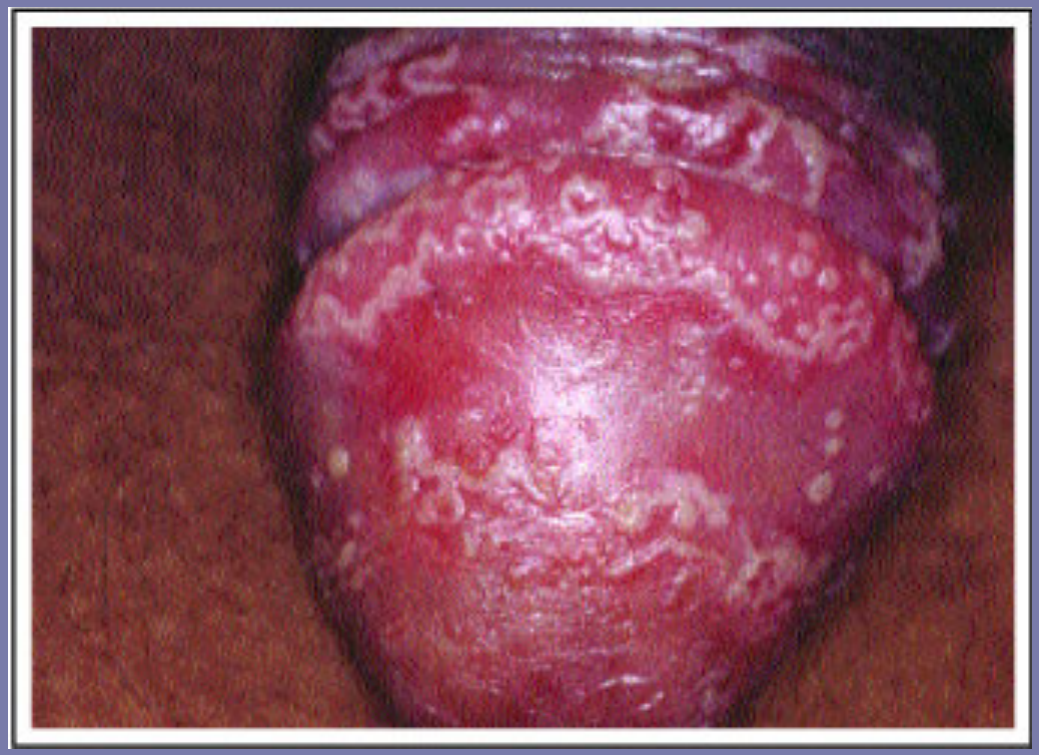

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Reactive Arthritis

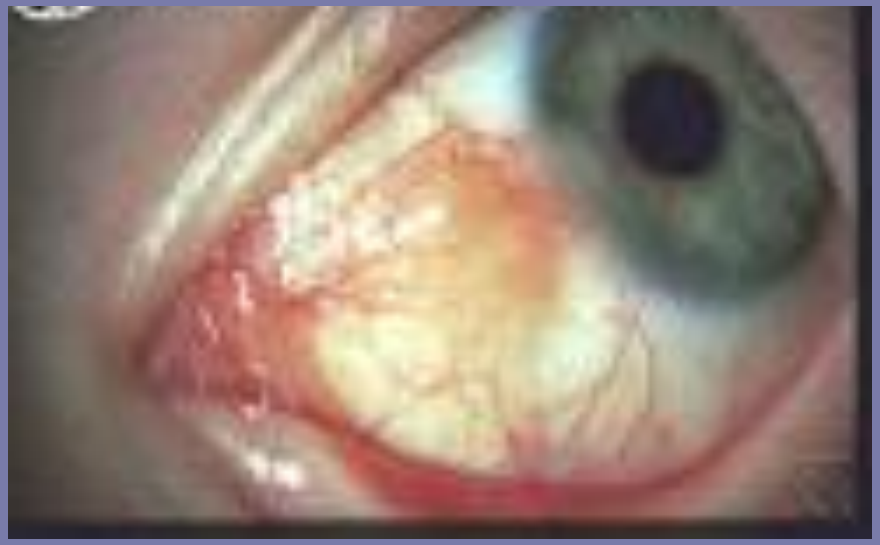
- Classic triad of Reiter syndrome
  - Nongonococcal urethritis (Chlamydia)
  - Conjunctivitis / anterior uveitis
  - Arthritis
- Following GI infection with Shigella, Salmonella, Campylobacter, Yersinia, Clostridium difficile (?)
- Infection precedes arthritis by 2 - 6 wk

Reactive Arthritis

- Treatment:
  - NSAIDs
    - Naproxen 500 mg TID
    - Indomethacin 50 mg TID
  - Intra-articular glucocorticoids
  - Expect resolution 3 – 12 months

Rheumatoid Arthritis

- Definition:
  - Chronic, symmetric polyarticular synovial joint disease
- Nonarticular & Systemic Manifestations
  - HEENT
    - Episcleritis
      - Painless injection of episcleral vessels (self-limiting)
    - Scleritis
      - Dark red / purple discoloration with marked ocular tenderness
      - Potential for visual impairment and scleral rupture



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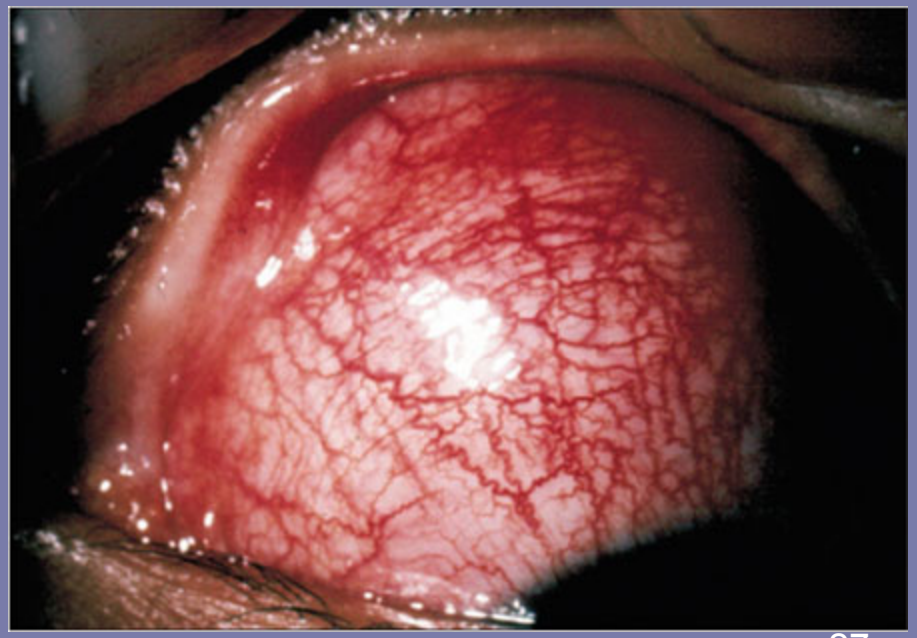

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Rheumatoid Arthritis

- Cricoarytenoid joint
  - Dysphonia, hoarseness, stridor
  - If fix in closed position could require emergent tracheostomy
- Ligamentous destruction of transverse ligament of C-2 with potential for cord compression

Rheumatoid Arthritis

- Pulmonary
  - Pleural effusion *
  - Interstitial fibrosis
  - Pulmonary nodules
- Cardiac
  - Pericarditis *
  - Pericardial effusion 10%
  - Myocarditis
  - CAD – sudden death, MI *

Rheumatoid Arthritis

- Renal
  - Focal glomerulonephropathy
  - Drug toxicity from treatment *
- Vasculitis
  - Distal infarcts, ulcerations, gangrene
- CNS
  - Generally spared

Systemic Sclerosis - Scleroderma

- Definition:
  - Disease process characterized by progressive fibrosis, vascular abnormalities and inflammatory processes that can be manifested quite locally or diffusely systematically with organ system involvement
- Etiology:
  - Poorly understood

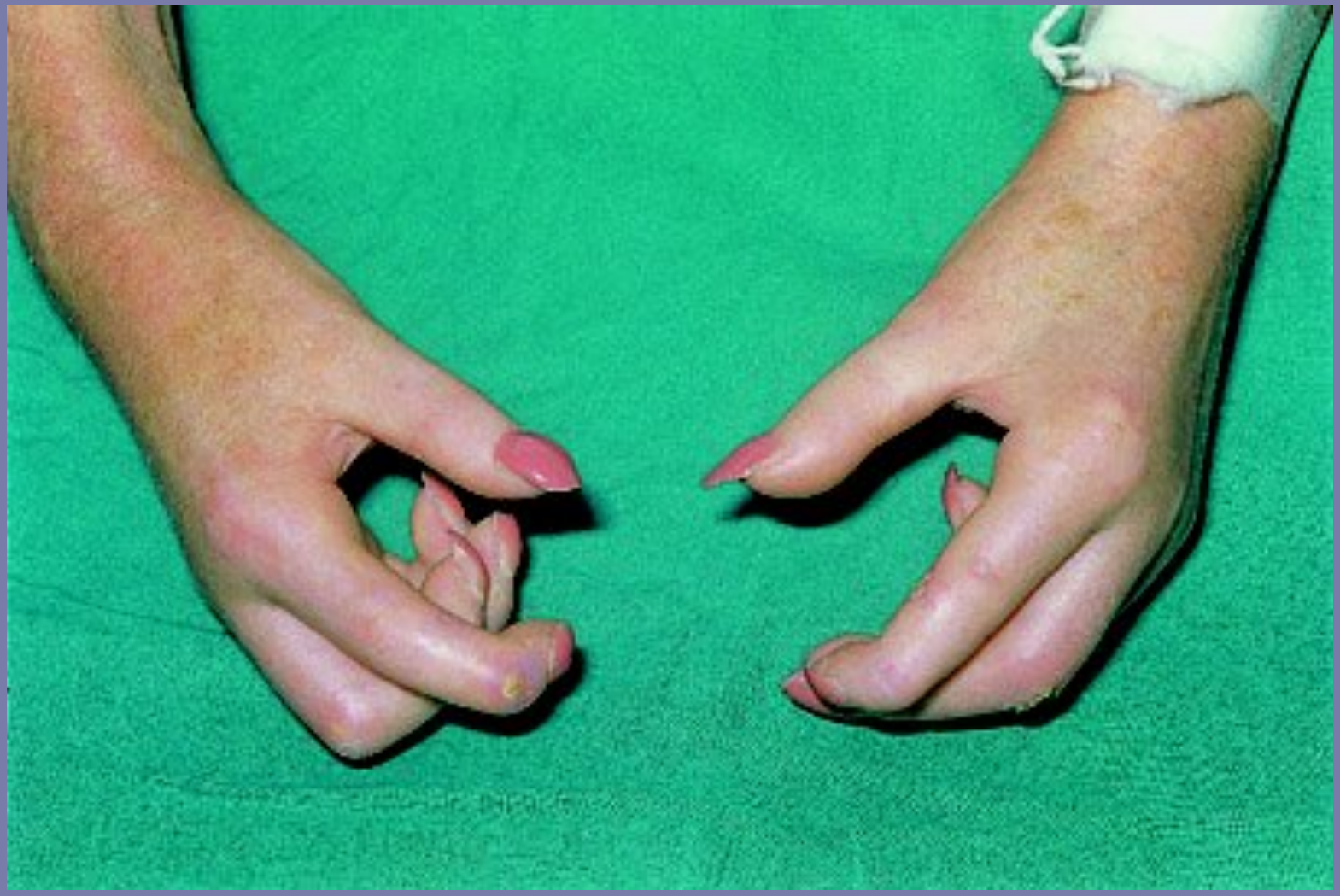
Systemic Sclerosis

- Clinical Presentation:
  - Often initial signs are a thickening, hardening of the skin, usually fingers, hands and face and Raynaud phenomenon
  - Female > Male
  - African-Americans tend to have worse prognosis due to greater likelihood of having a more severe diffuse form

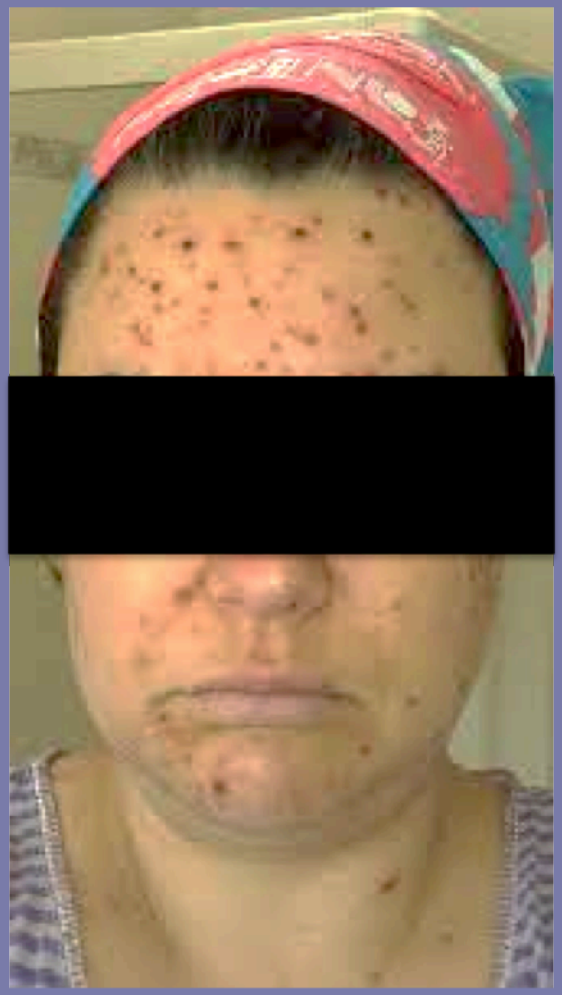
Systemic Sclerosis

- Skin
  - Sclerodactyly
  - Telangiectasias
  - Digital ulcers
  - Calcinosis
  - Raynaud phenomenon

In scleroderma, the abnormal build-up of fibrous tissue in the skin can cause the skin to tighten so severely that the fingers curl and lose their mobility.



Telangiectasia



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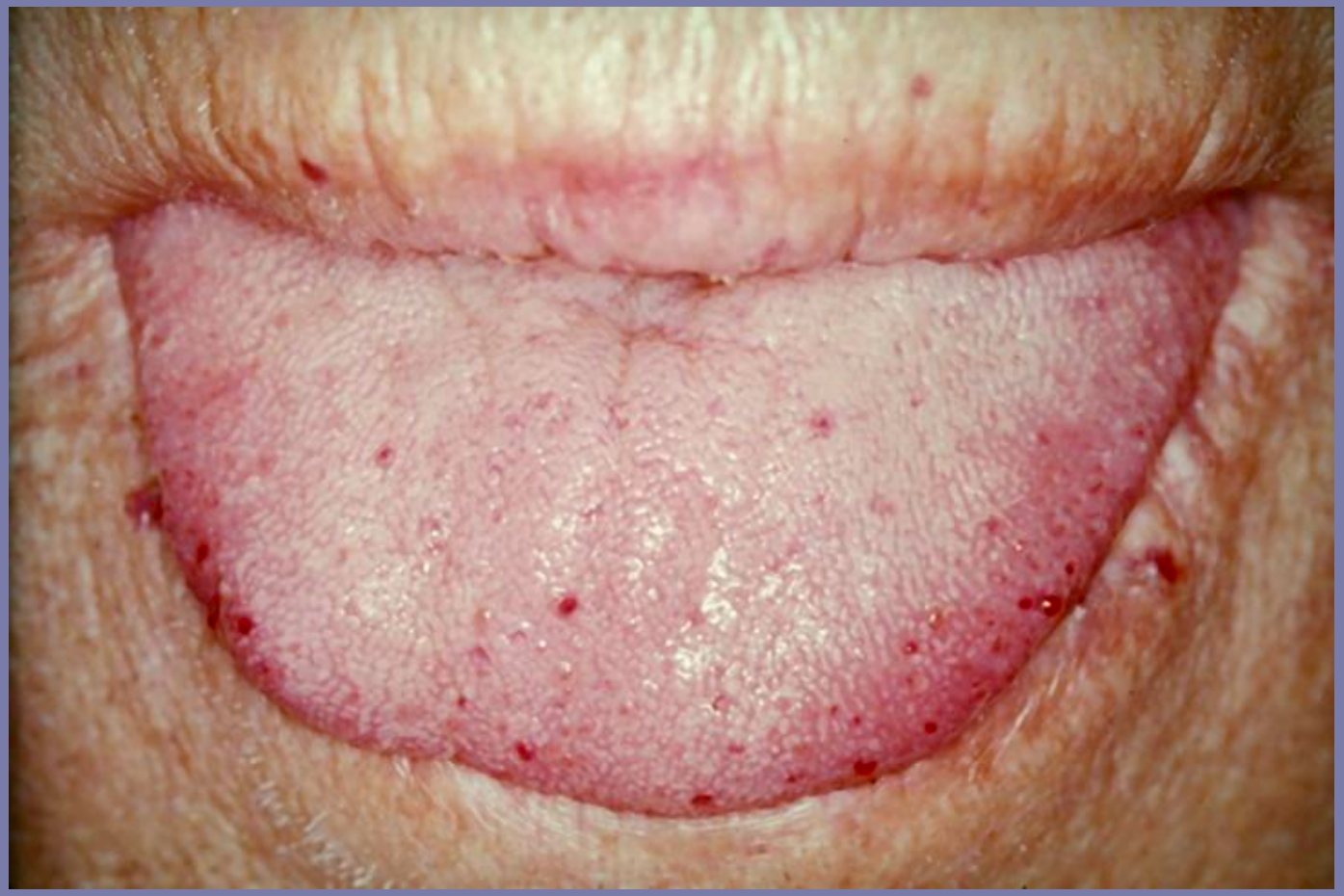
Dilation of small vessels and capillaries cause flat red marks to appear on the skin



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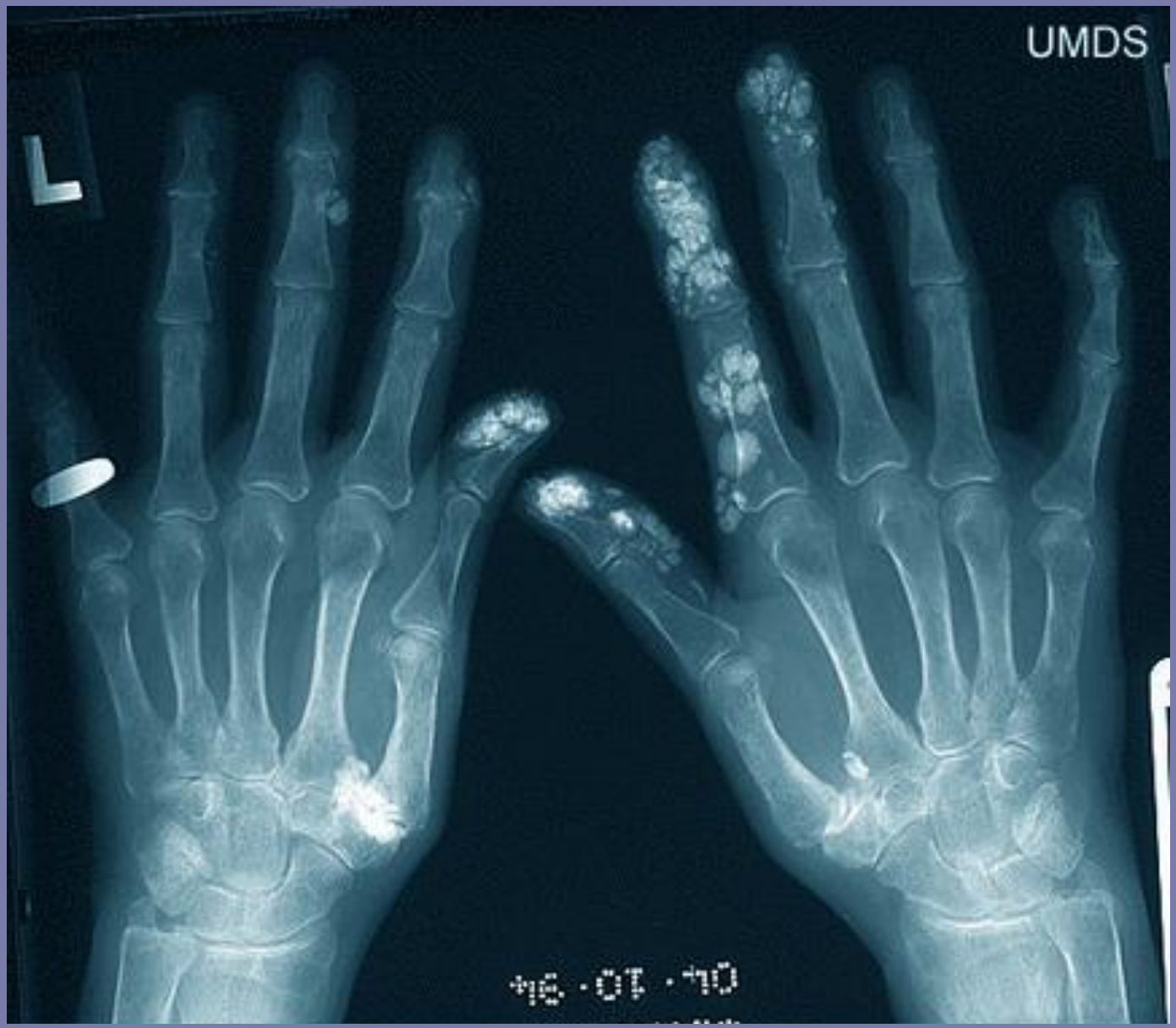

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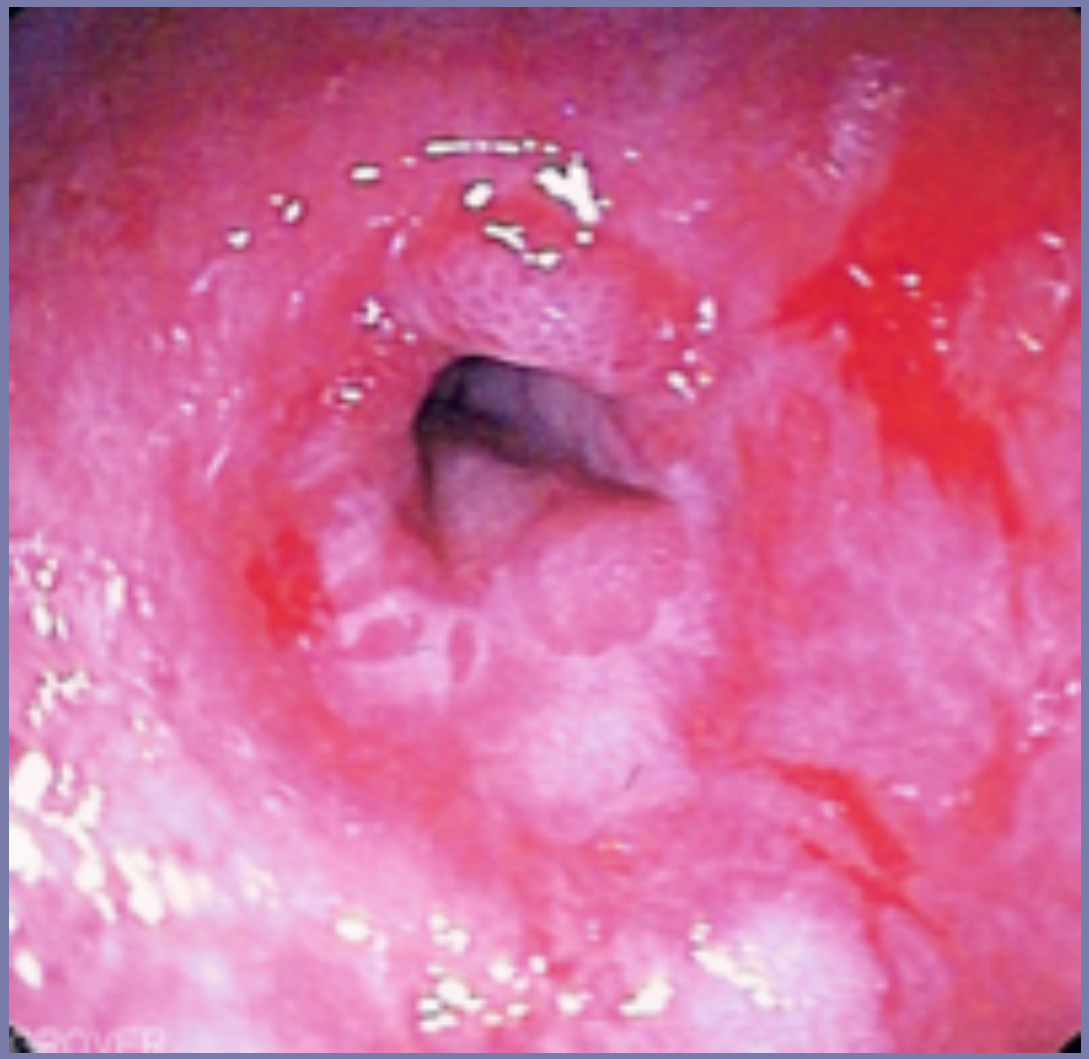
Systemic Sclerosis

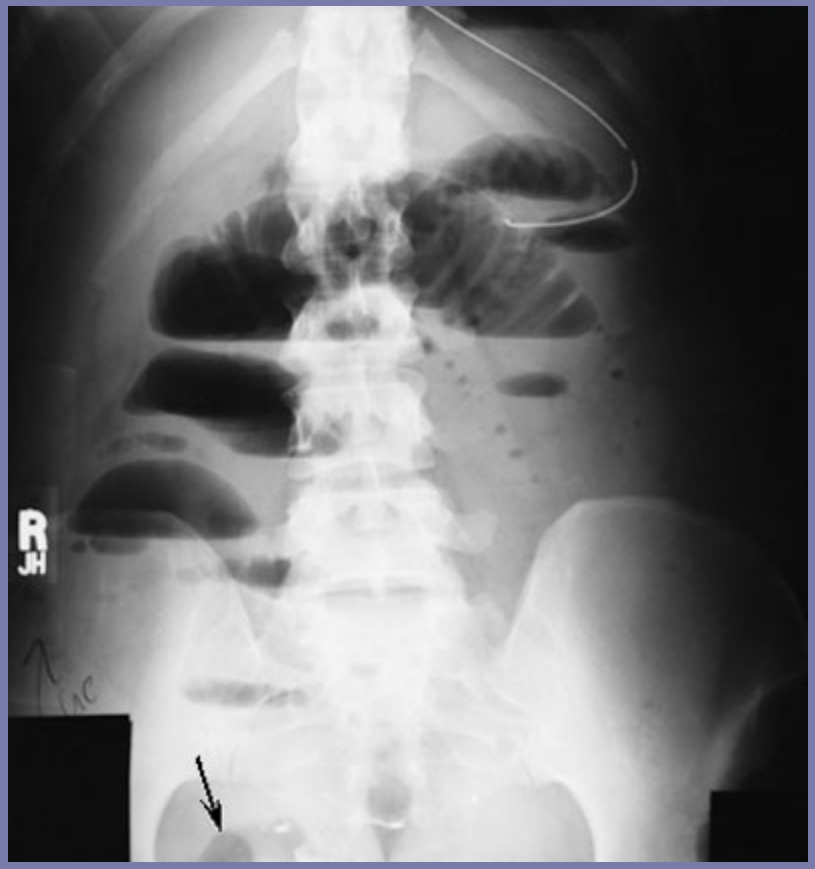
- Pulmonary
  - Most common cause of death
  - Interstitial lung disease
    - Alveolitis leading to eventual fibrosis
  - Pulmonary vascular disease
    - Pulmonary hypertension
- Cardiac
  - Pericarditis w/wo effusion
  - Myocardial fibrosis
    - Resultant ventricular dysfunction with diminished cardiac output
  - Dysrhythmias
    - Fibrosis of conduction system resulting in sudden death

Systemic Sclerosis

- Renal
  - Renal crisis
    - Acute onset of renal failure
    - Proteinuria and microscopic hematuria
    - Abrupt onset of hypertensive emergency
- Gastrointestinal
  - Hypomobility
    - Esophageal dysmotility / GERD
    - Pseudo-obstruction, constipation
    - Teleangectasias with bleeding
    - Pneumatosis intestinalis



Lipothymia, Wikimedia Commons



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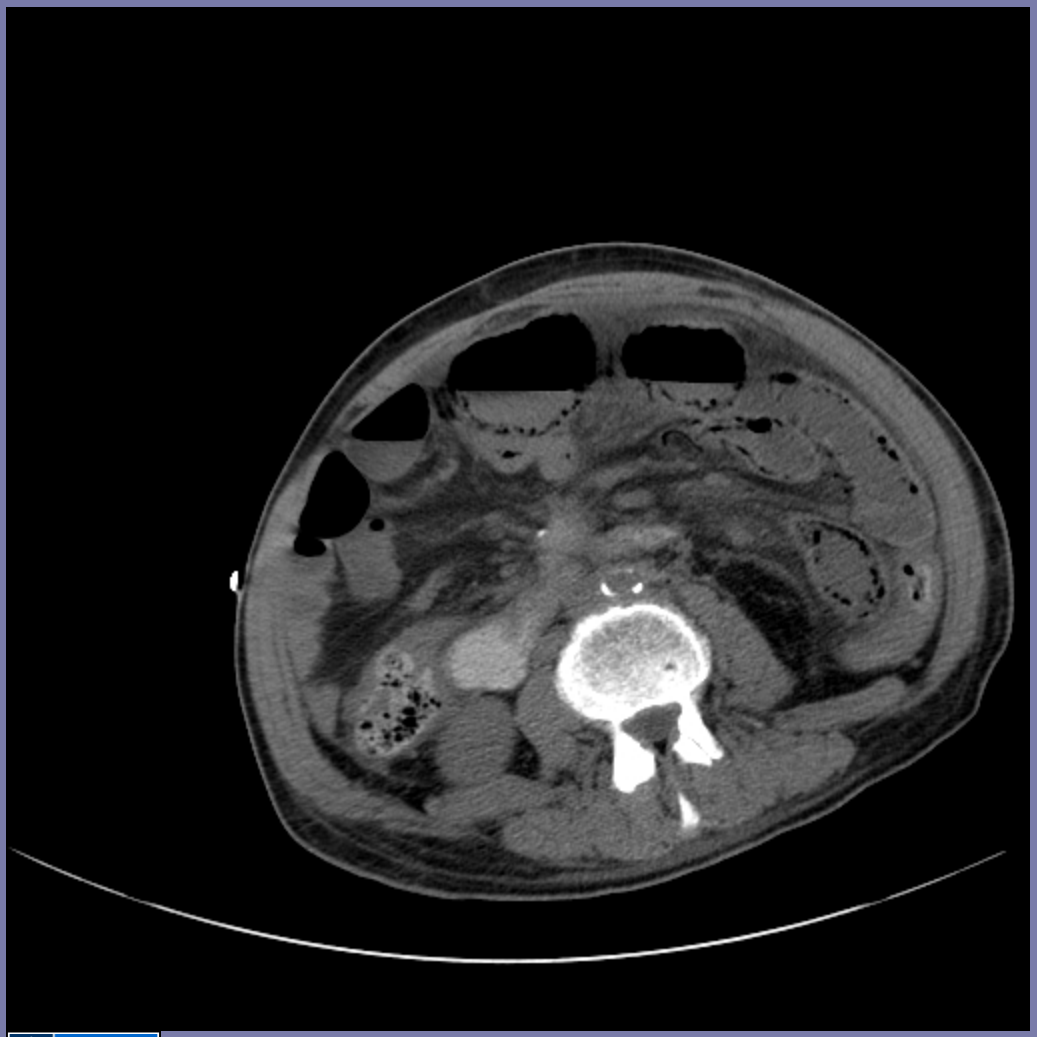

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Systemic Sclerosis

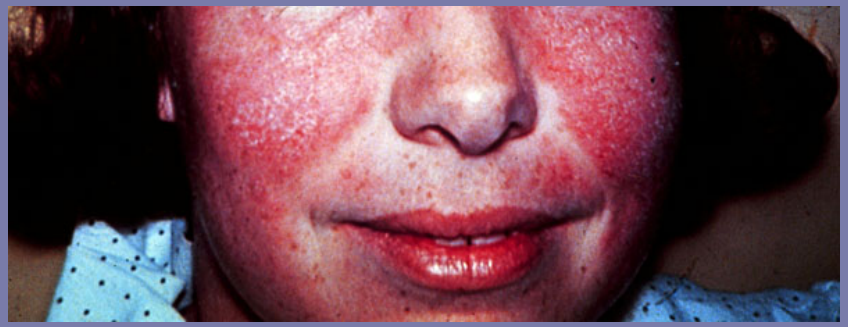
- Treatment
  - Generally immunosuppressive therapy
  - Based on specific organ system involved
  - Emergently
    - Pulmonary decompensation
    - Cardiac – effusion, failure, dysrhythmia
    - Renal crisis
      - ACE inhibitor is first line antihypertensive agent – Captopril
      - Captopril + Ca channel blocker
      - Angiotensin receptor blocker for those who can not tolerate ACE inhibitor

Systemic Lupus Erythematosus

- Definition:
  - Chronic autoimmune disease characterized by presence of autoantibodies with multi-organ system involvement
- Etiology:
  - Genetic predisposition and nebulous factors combine to alter immune cell function resulting in production of autoantigens and thereby auto-antibodies with systemic consequences.

SLE

- Clinical Presentation:
  - Female > Male 10 : 1
  - Typical presentation 21 – 45 yrs of age
  - African American > Caucasians
  - Constitutional
    - Wt loss, fever, myalgias, arthralgias
    - Fatigue often the most debilitating
  - Skin
    - Butterfly malar rash – may be fleeting
    - Oral and nasal ulcerations



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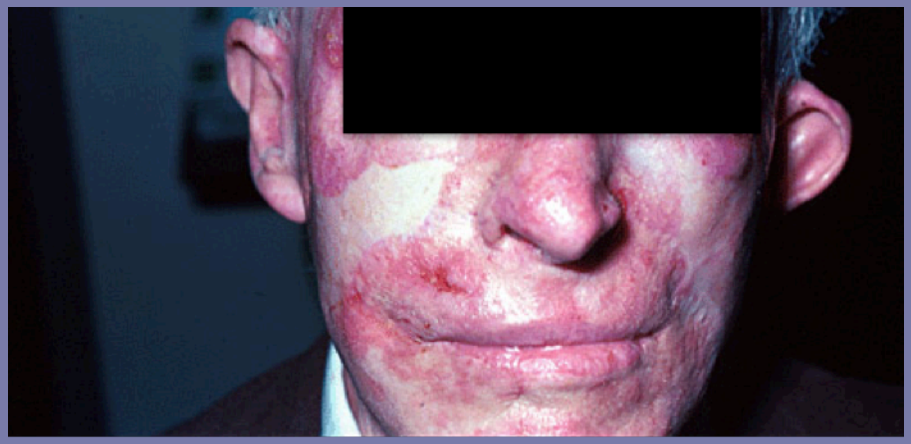

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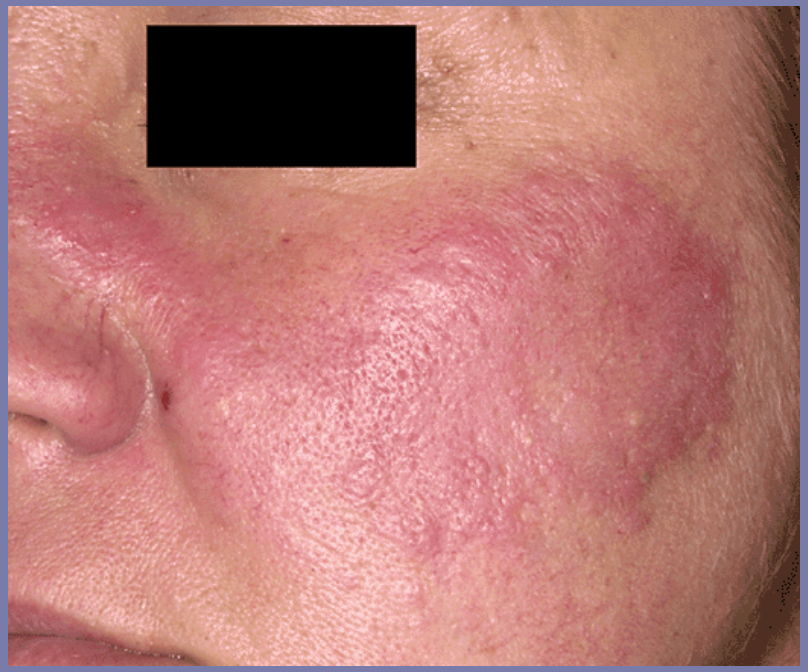

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SLE

- Pulmonary
  - Pleurisy, effusion, interstitial lung disease, pulmonary hypertension
  - “lupus lung” – alveolar hemorrhage
- Cardiac
  - Pericarditis *, effusion
  - Increased risk for CAD
- Renal
  - Lupus nephritis
    - Elevated creatinine, proteinuria, hypertension

SLE

- Neurologic
  - Cognitive defects, cephalgia, seizures, peripheral neuropathies (stocking / glove), psychosis, stroke (antiphospholipid antibody syndrome)
- Musculoskeletal
  - Arthritis, atrophy, tendon rupture

SLE

- Treatment:
  - Immunosuppression
    - Glucocorticoids
    - Methotrexate, cyclophosphamide, azathioprine, mycophenolate, rituximab
  - Causes of death
    - Early deaths – first few years
      - Active lupus (cardiac, renal, CNS dz)
      - Infection due to immunosuppression
    - Late deaths
      - Chronic effects of lupus (ESRD, CAD)
      - Infection, malignancy

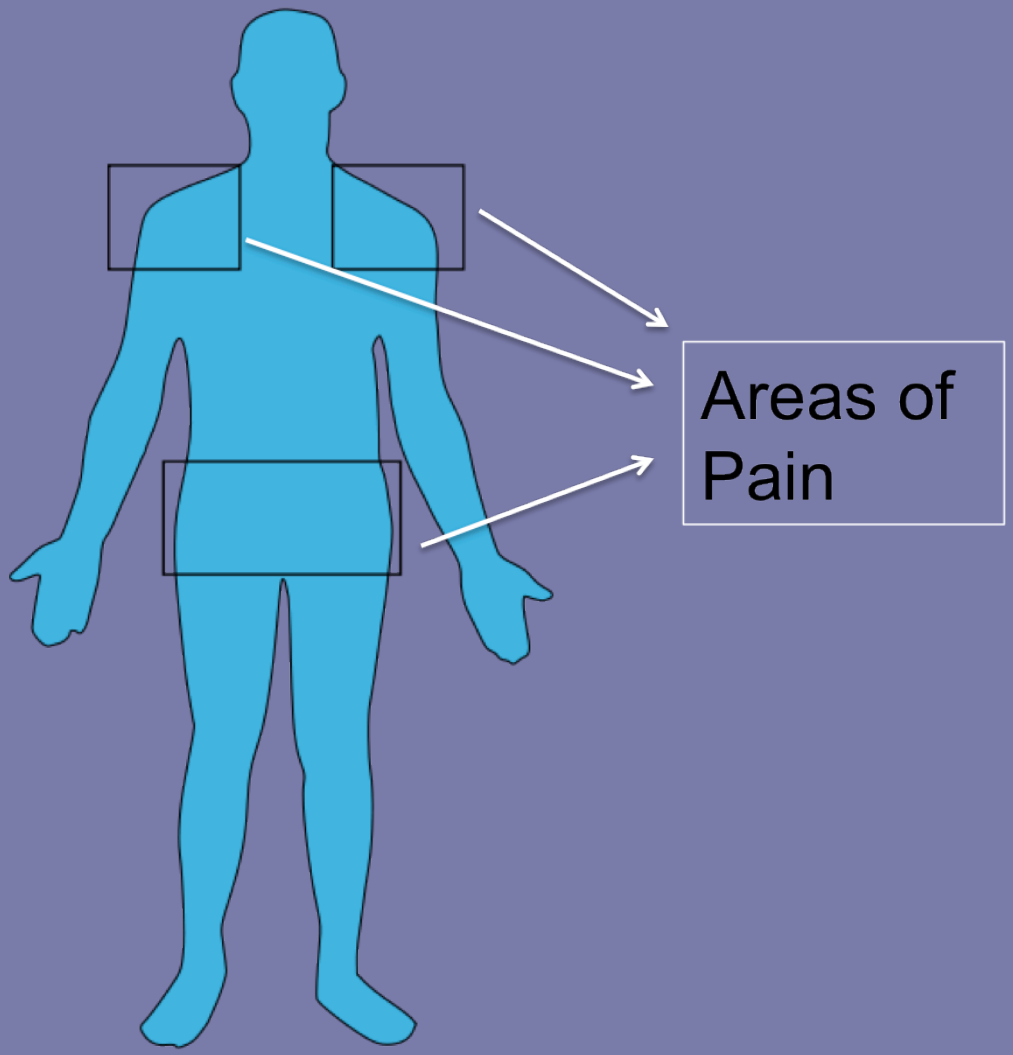
Polymyalgia Rheumatica

- Definition:
  - Rheumatic condition frequently associated with giant cell (temporal) arteritis
- Etiology:
  - Genetic predisposition
  - Relatively common
  - 50% of pts with GCA will develop PMR
  - 15% of pts with PMR will develop GCA

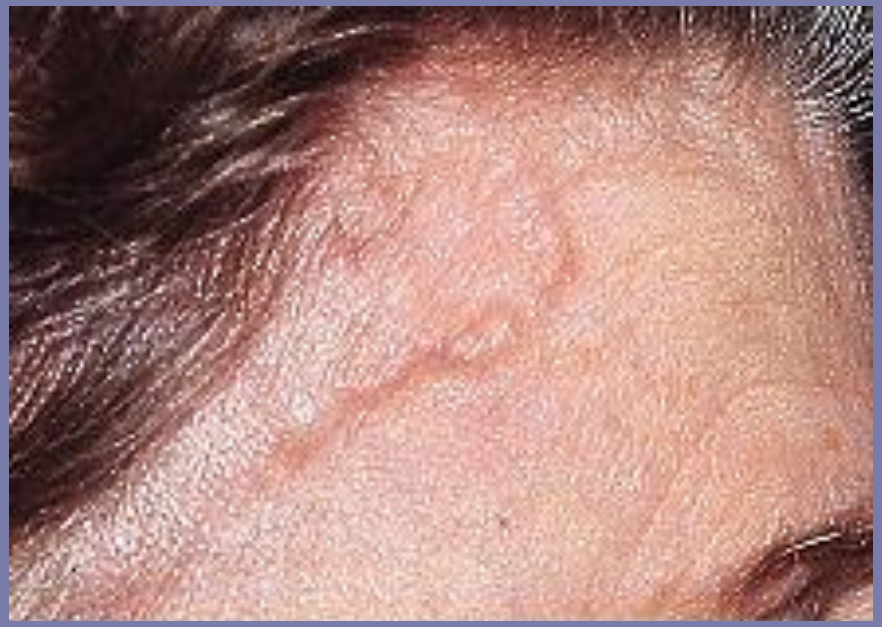
Polymyalgia Rheumatica

- Clinical Presentation:
  - Age 50 or older at onset
  - Bilateral aching and morning stiffness which lasts 30 min or more for 1 month or more involving at least 2 of 3 areas
    - Neck or torso
    - Shoulders or proximal arms
    - Hips or proximal thighs
  - ESR = 40 mm/hr or greater

Polymyalgia rheumatica



Giant Cell Arteritis



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Polymyalgia Rheumatica

- treatment:
  - Prednisone 10 – 20 mg / day
    - (compared to 60 mg / day dose for GCA)
  - Rapid response is characteristic (often after first dose)
  - Relapse is commonly seen requiring increase in prednisone

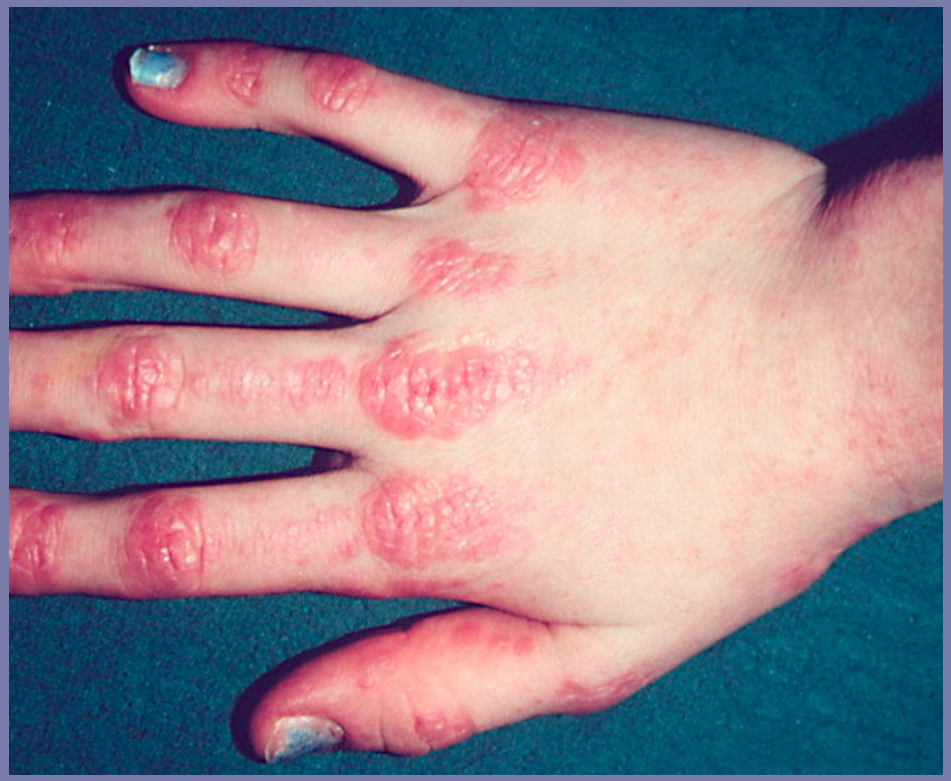
Polymyositis / Dermatomyositis

- Definition:
  - Idiopathic inflammatory myopathies

- Etiology:
  - Genetic component with presumed environmental triggers
  - Peak incidence between 40 – 50 yrs of age
  - Female : Male 2 : 1

Polymyositis / Dermatomyositis

- Clinical Presentation:
  - Muscle weakness
    - Onset is insidious
    - Gradually worsening over months
    - Typically symmetric and proximal
  - Myalgias / muscle tenderness occurs in 25-50% but is mild compared to PMR or fibromyalgia
  - Dermatologic findings in DM
    - Gottron's sign
      - Erythematous, often scaly exanthem occurring symmetrically over MCP and IP joints and / or over extensor surfaces of elbows and knees resembling psoriasis



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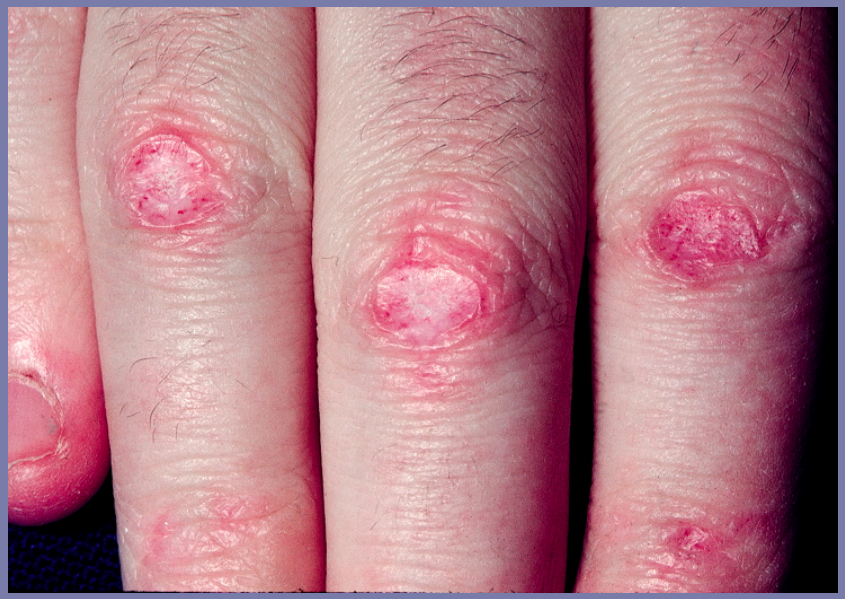

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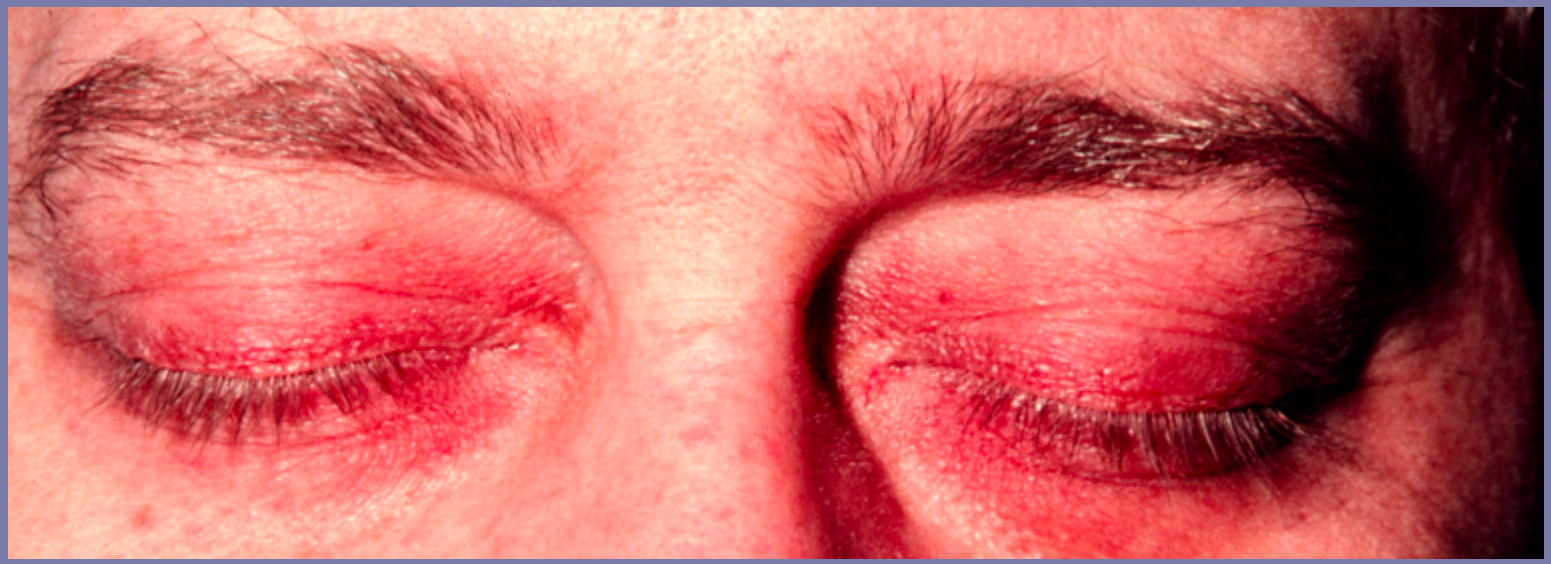
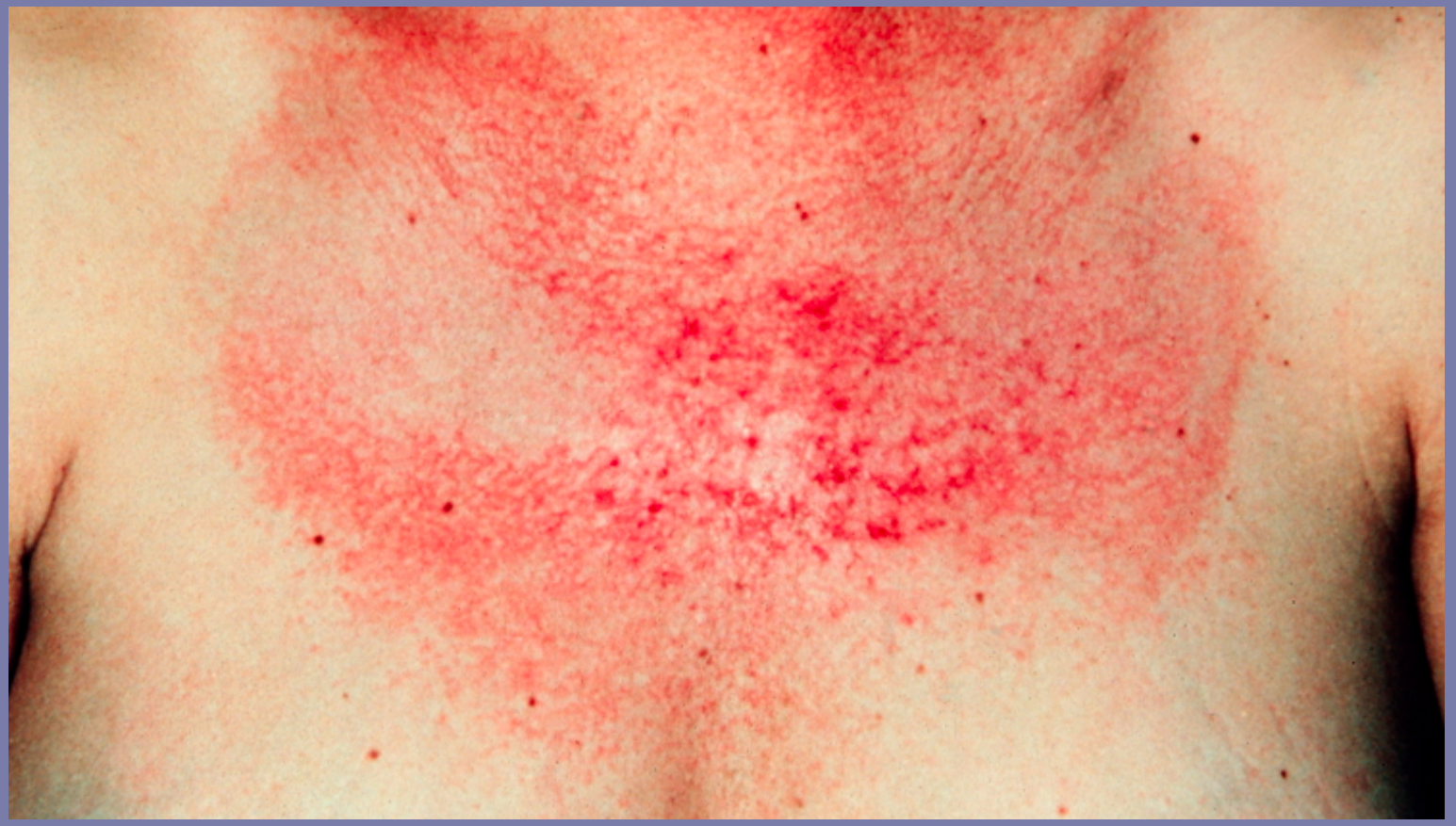
Polymyositis / Dermatomyositis

- Heliotrope rash
  - Violaceous eruption on upper eyelids, often with swelling
- Shawl sign and V sign
  - Diffuse flat erythematous lesion over chest and shoulders (shawl sign) or over anterior neck and chest (V sign)
- Erythroderma
  - Extensive areas of skin redness (malar, forehead)
- Mechanic's hands (DM or PM)
  - Rough, cracking skin at tips and lateral aspects of fingers with irregular dirty appearing lines





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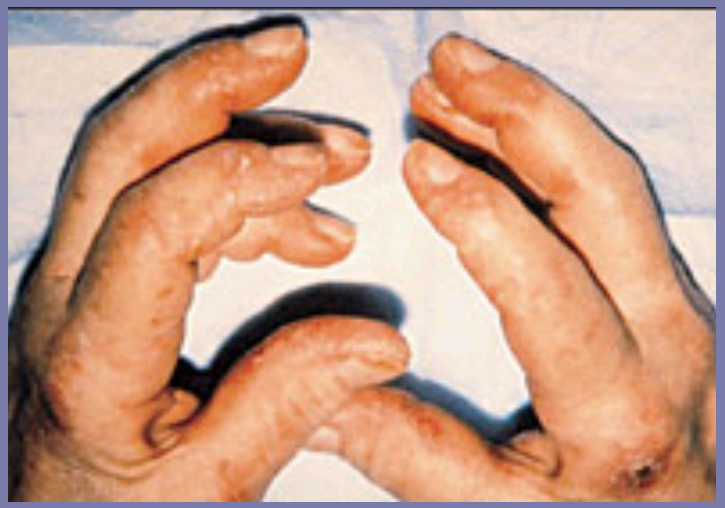

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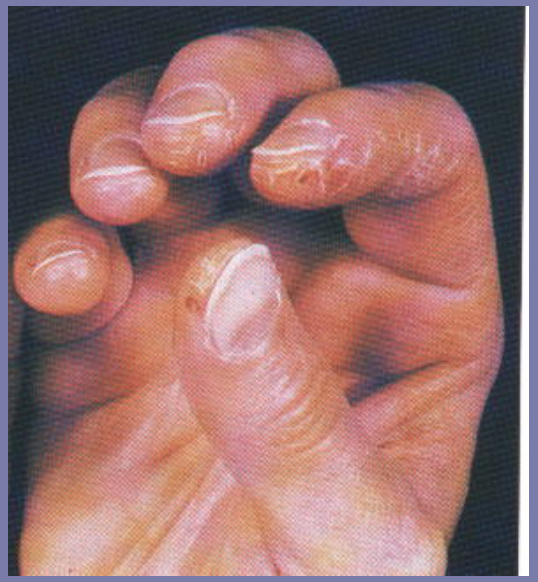

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Polymyositis / Dermatomyositis

- Diagnostic testing
  - Elevated CK, LDH, aldolase, aminotransferases
- Increased incidence of malignancy associated with dermatomyositis
- Treatment:
  - Glucocorticoid regimen
    - initiate with high doses for several months to establish disease control
    - Slow taper to lowest effective dose for 9 – 12 months
  - Glucocorticoid-sparing regimen
    - Azathoprine / methotrexate

Vasculitides

- Large vessel
  - Takayasu arteritis
  - Giant cell (temporal) arteritis
- Medium vessel
  - Polyarteritis nodosa
  - Kawasaki disease
- Small vessel
  - Churg-Strauss arteritis
  - Wegener's granulomatosis
  - Henoch-Schonlein purpura