Vasculitis

Introduction

- Classification
- Common Etiologies
- Approach to the patient
  - History
  - Physical Exam
  - Laboratory Work-Up

Introduction

- Vasculitis is inflammation and necrosis of the blood vessel wall producing a wide range of clinical manifestations

Classification

- American College of Rheumatology (ACR)
  - clinical, historical, and histologic
- Chapel Hill Consensus Conference (CHCC)
  - histopathologic
- Classification based on size of vessel involved
  - small: arterioles, capillaries, venules
  - medium: main visceral arteries (renal, hepatic, coronary, mesenteric)
  - large: aorta and its largest branches

Table 26.2 Chapel Hill consensus classification.

CHAPEL HILL CONSENSUS CLASSIFICATION
Large-vessel vasculitis
• Giant cell arteritis • Takayasu's arteritis
Medium-vessel vasculitis
• Classic polyarteritis nodosa • Kawasaki disease
Small-vessel vasculitis
• Wegener's granulomatosis • Churg–Strauss syndrome • Microscopic polyangiitis (polyarteritis) • Henoch–Schönlein purpura • Essential cryoglobulinemia • Cutaneous leukocytoclastic vasculitis

Etiologies

- Idiopathic (45-55%)
- Infection (15-20%)
- Inflammatory disease (15-20%)
- Drugs (10-15%)
- Malignancy (<5%)

Etiologies

- Infections
  - Bacteria, viruses, parasites, and fungi
  - Hep B and PAN
  - Hep C and mixed cryoglobulinemia

Etiologies

- Inflammatory Disease
  - SLE
  - Rheumatoid arthritis
  - Sjogren's
  - Behcet's
  - IBD

Etiologies

- Drugs
  - PCN
  - sulfas
  - quinolones
  - insulin
  - tamoxifen
  - OCP's
  - **propylthiouracil**
  - phenothiazines
  - **hydantoins**
  - **allopurinol**
  - **thiazides**
  - retinoids
  - influenza vaccine
  - interferon
  - leukotriene inhibitors

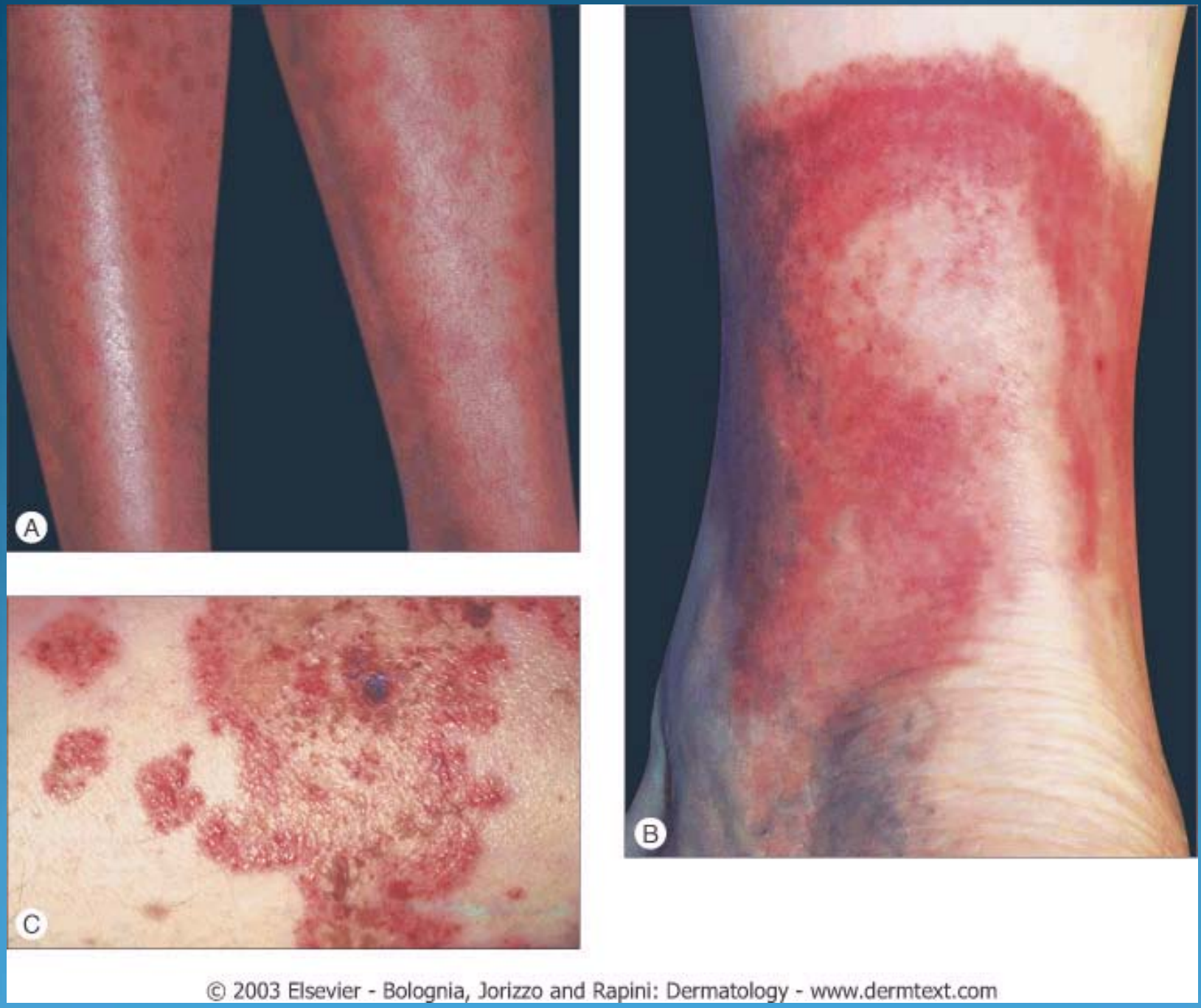
Etiologies

- Malignancy
  - paraproteinemias
  - lymphoproliferative malignancies
  - hairy cell leukemia assoc. with PAN

Small-Vessel Vasculitis

Cutaneous small-vessel vasculitis

- Vasculitis confined only to the skin
- key history: exposure to new meds or infectious agents
- characterized by a single”crop” of lesions that resolve over weeks to months
- lesions consist of palpable purpura, papules, vesicles, and/or urticaria



Cutaneous small-vessel vasculitis

- Lesions occur in dependent areas, areas of trauma, or under tight clothing
- Treatment
  - remove or treat offending agent
  - keep area warm, elevate
  - topical steroids if pruritic
  - NSAIDs, ASA, anti-histamines
  - colchicine, dapsone, systemic steroids

Cryoglobulinemic vasculitis

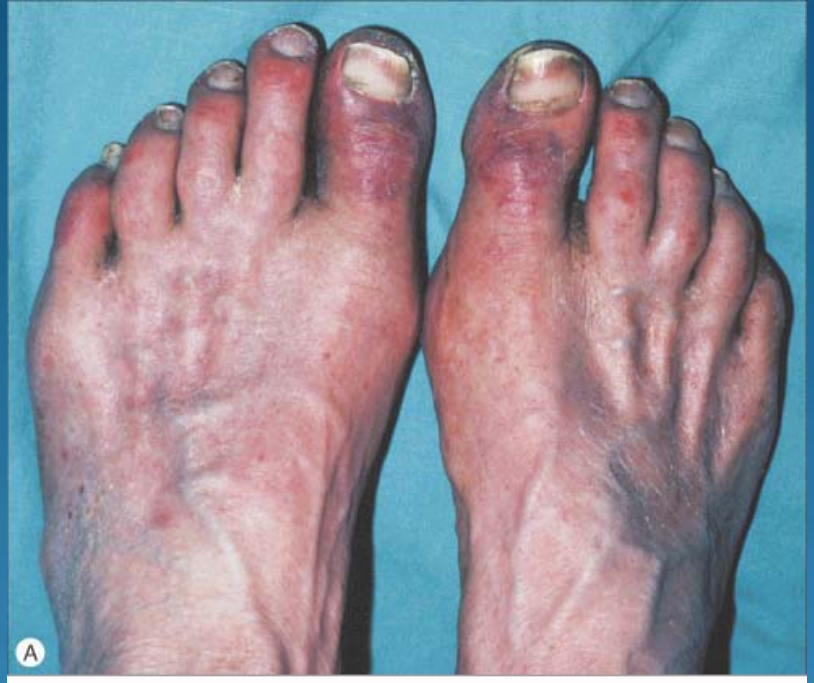
- Type I
  - monoclonal IgM or IgG
  - always associated with hematologic malignancy
- Type II (mixed)
  - monoclonal IgM directed against IgG
- Type III (mixed)
  - polyclonal IgM directed against IgG

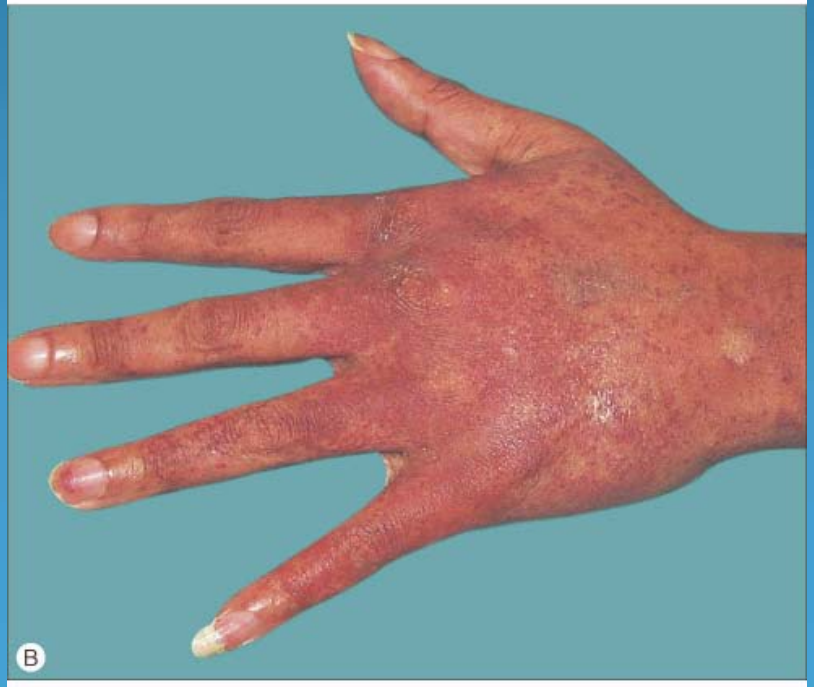
Cryoglobulinemic vasculitis

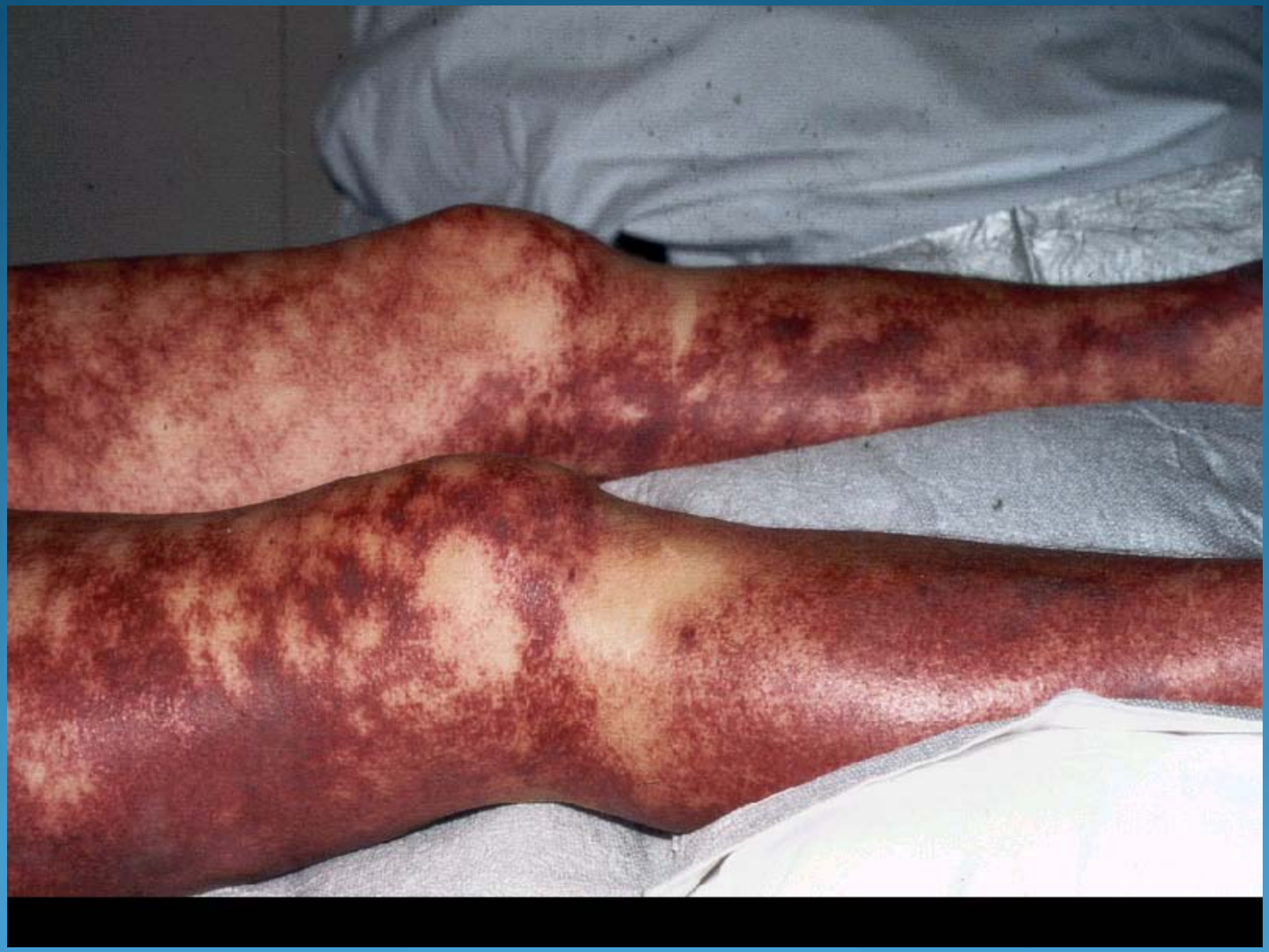
- Systemic inflammation of vessel walls due to deposition of IgM-IgG complexes
- Causes
  - Infections (Hep C, HIV)
  - Autoimmune (SLE, Sjogren's, scleroderma, RA)
  - Lymphoproliferative D/O (non-Hodgkins lymphoma, CLL)

Cryoglobulinemic Vasculitis

- palpable purpura (usually LE), arthralgias, weakness, Raynaud's
- less commonly ecchymoses, papules, and dermal nodules
- trunk and face almost always spared
- sometimes peripheral neuropathy and renal involvement







Cryoglobulinemic vasculitis

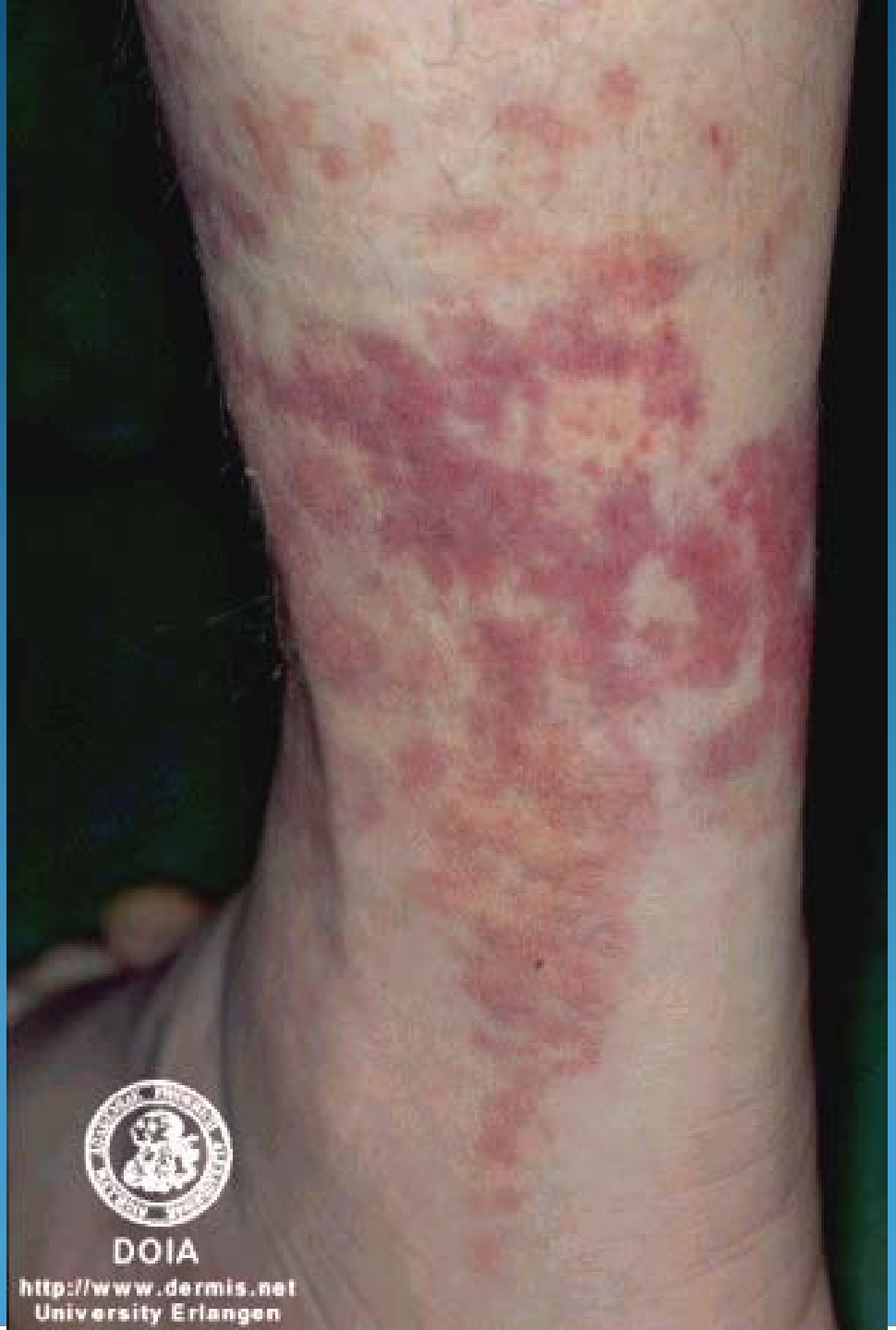
- Labs
  - Hep C panel
  - low C4
  - RF + in 70%
  - elevated LFT's
  - monoclonal spike (15% of Type II pts.)

Cryoglobulinemic vasculitis

- Treatment
  - treat underlying disorder
    - HepC, malignancy, autoimmune dz.
  - Systemic steroids
    - 0.1-0.3 mg/kg for purpura, arthralgia, weakness
    - 0.5-1.5 mg/kg for renal and CNS dz

Urticarial Vasculits

- wheals lasting longer than 24 hours with evidence of LCV on biopsy
- purpura and post-inflammatory hyperpigmentation may be present
- Causes
  - SLE, Sjogren's, serum sickness
  - infxn, drugs, heme malignancies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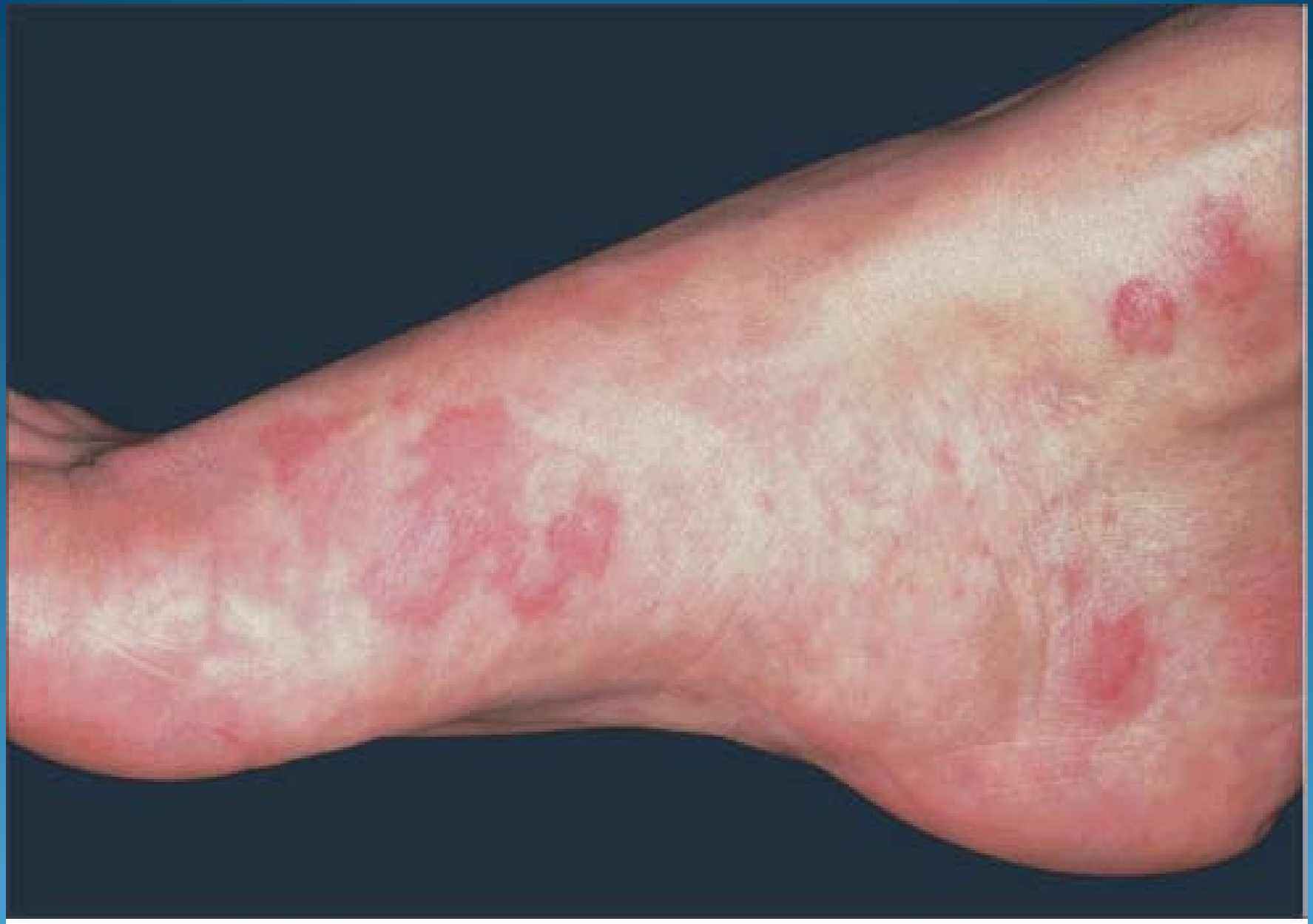
Urticarial vasculitis

- Hypocomplementemic
  - arthritis, GI sx, asthma and obstructive airway, ocular sx, fever, malaise, LAD
  - much more likely to have assoc. systemic dz
  - anti-C1q Ab, +/- low C1 levels, +ANA
- Normocomplementemic
  - limited to the skin
  - idiopathic
  - self-resolving

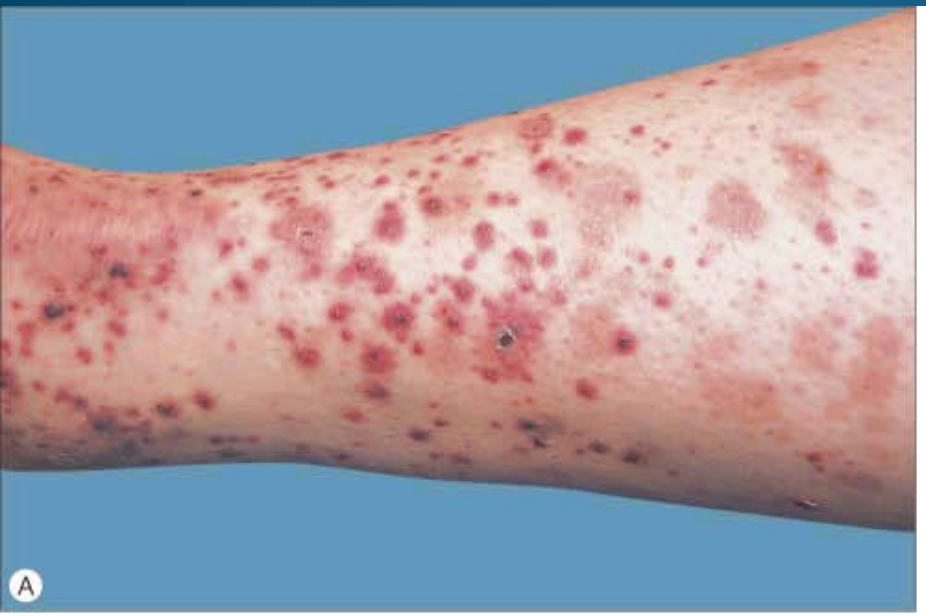


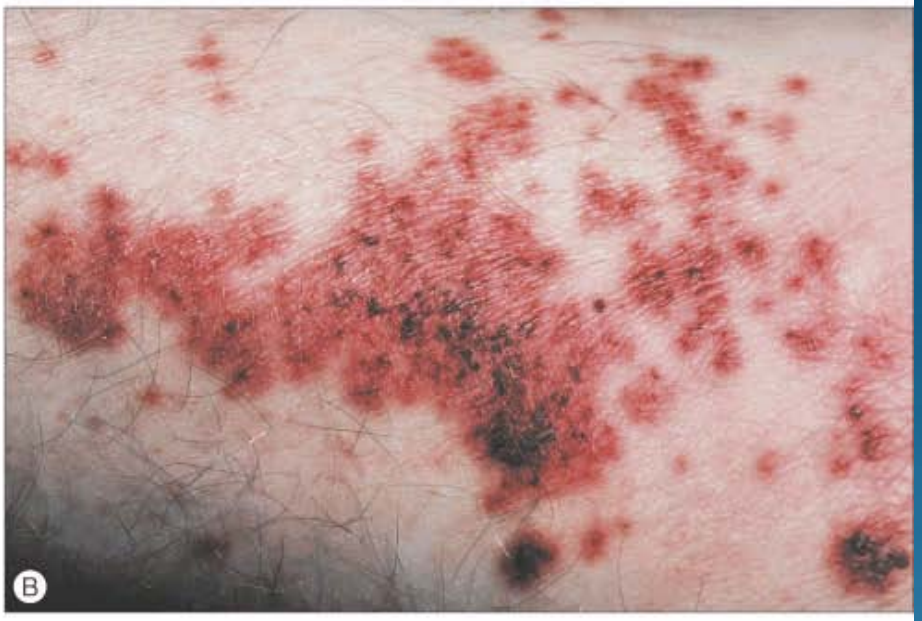
Urticarial vasculitis

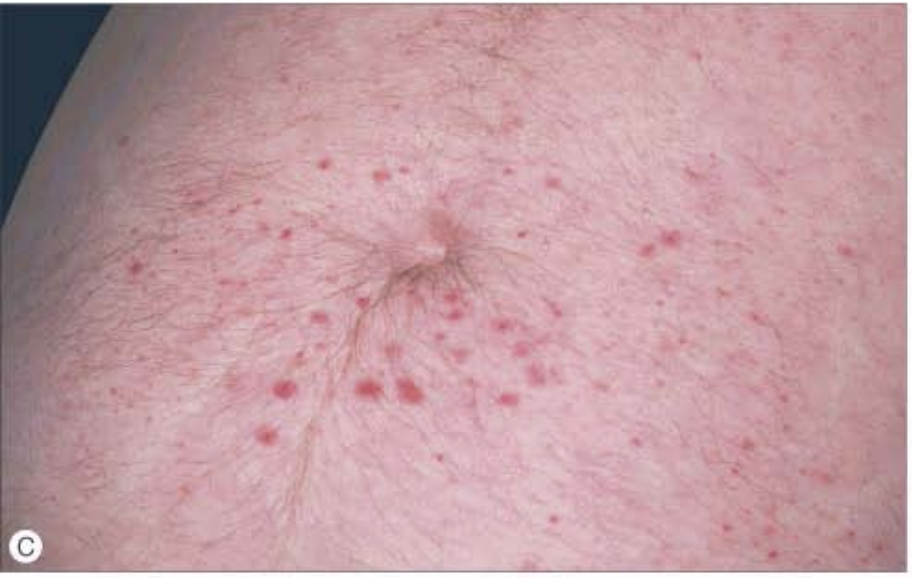
- Treatment
  - anti-histamines
  - NSAIDs (indomethacin)
  - prednisone
  - dapsone, colchicine, plaquenil

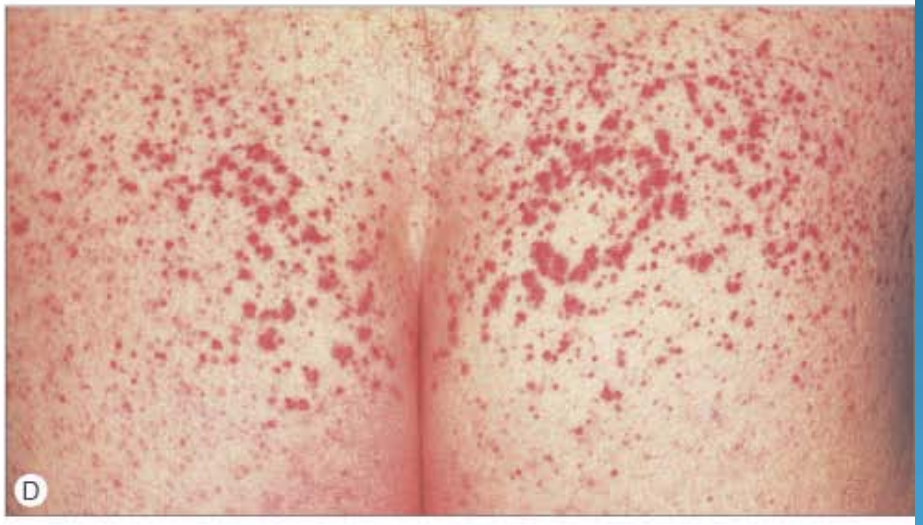
Henoch-Schonlein purpura

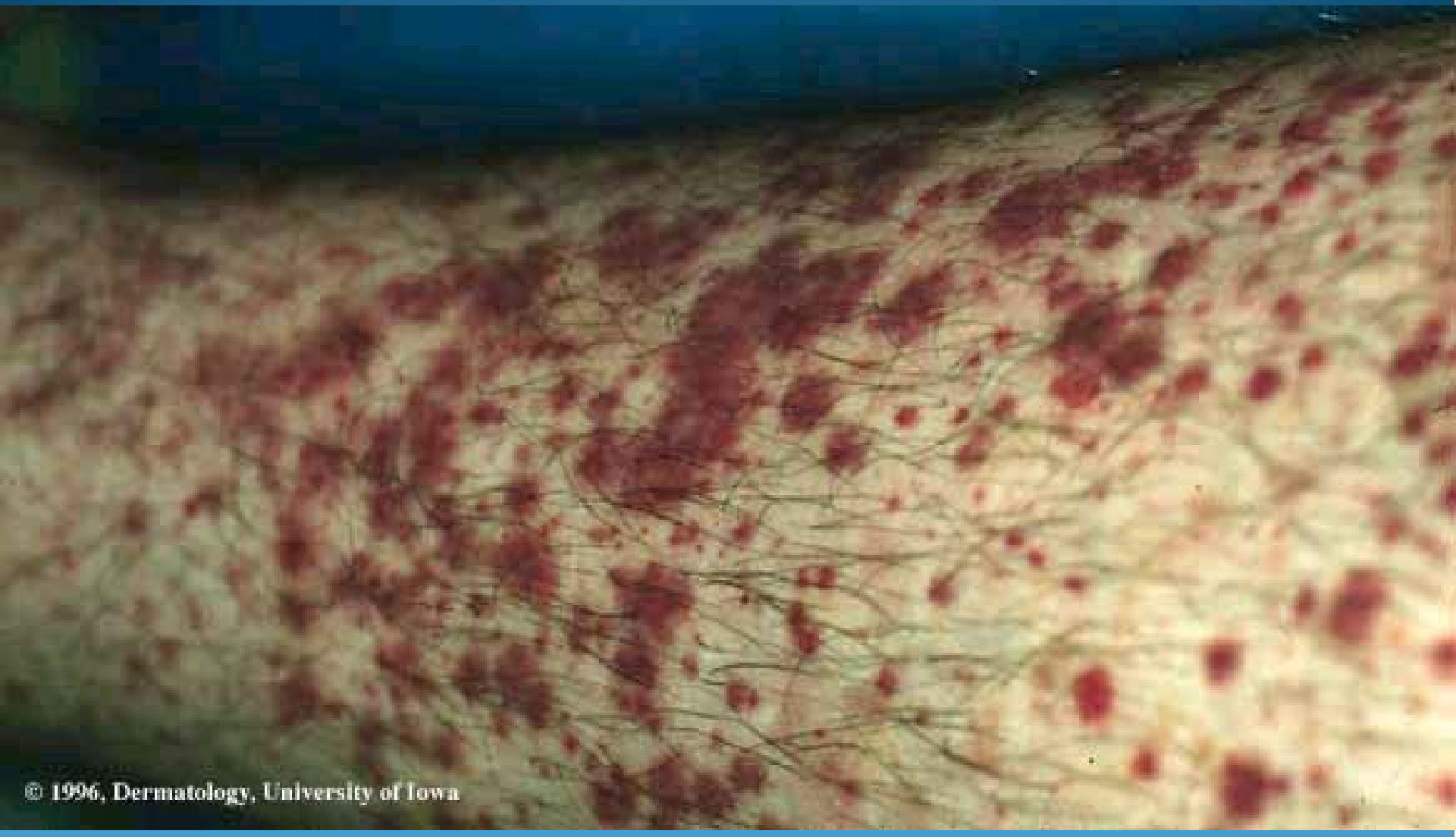
- most common systemic vasculitis in kids
- seen mostly in boys ages 4-8
- usually occurs 1-2 weeks after URI
- palpable purpura on LE and buttocks, arthralgias, abdominal pain
- renal involvement common, 5% ESRD
  - purpura above waist, fever, elevated ESR
- full recovery in weeks to months







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HSP

- Histo
  - LCV with perivascular IgA deposits
- Treatment
  - mostly supportive
  - systemic steroids may decrease risk of renal dz
  - steroids+Imuran may improve active renal dz
  - dapsone can improve cutaneous eruption

Medium-Vessel Vasculitis

Polyarteritis Nodosa

- Multi-system disease seen in ages 40-60, M>F
- fever, weight loss, arthralgias, malaise
- abdominal pain, HTN, orchitis, CHF, renal failure
- **classic PAN spares the lungs**
- 5-7% of cases due to Hep B

PAN

- Cutaneous findings
  - palpable purpura (20-50%)
  - livedo reticularis
  - large “punched out” ulcers
  - subcutaneous nodules
- Histo
  - necrotizing, obliterative arteritis of small and medium arteries

PAN

- Treatment
  - systemic steroids (1-2mg/kg/day)
  - cyclosporine
  - MTX
  - if HepB +, IFN

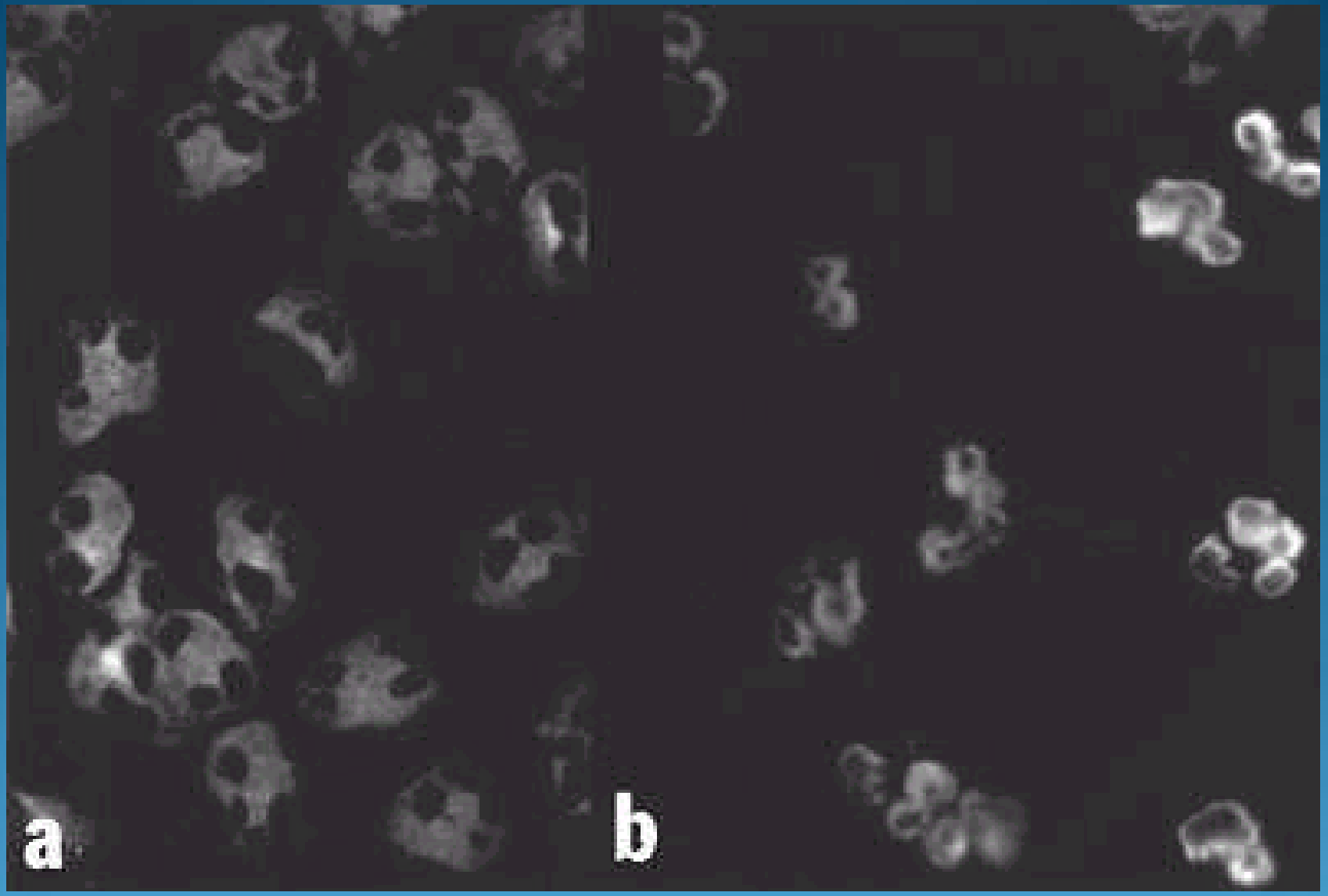
Cutaneous PAN

- Limited to skin (10% of PAN cases)
- fever, myalgias, arthralgias, peripheral neuropathies
- painful dermal/subQ nodules, ulcers, livedo reticularis, atrophie blanche
- most common form of PAN in children
- assoc. with strep, parvo, HIV, Hep B, IBD

Cutaneous PAN

- Treatment
  - NSAIDs, ASA
  - prednisone
  - PCN (in children)
  - IVIG
  - MTX (7.5-15 mg/week)

Small and Medium Vessel Vasculitis (ANCA associated vasculitides)

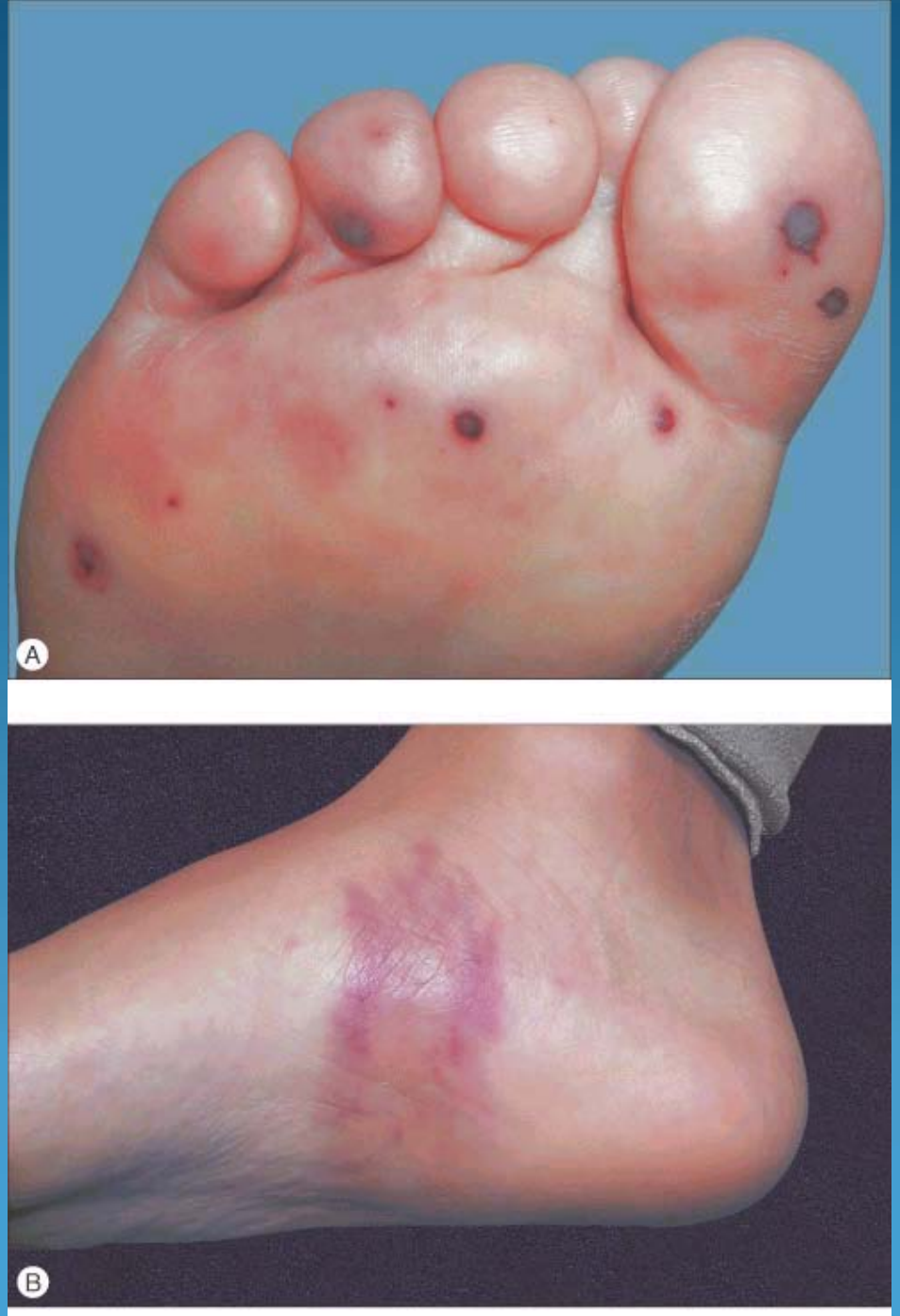


C-ANCA

P-ANCA

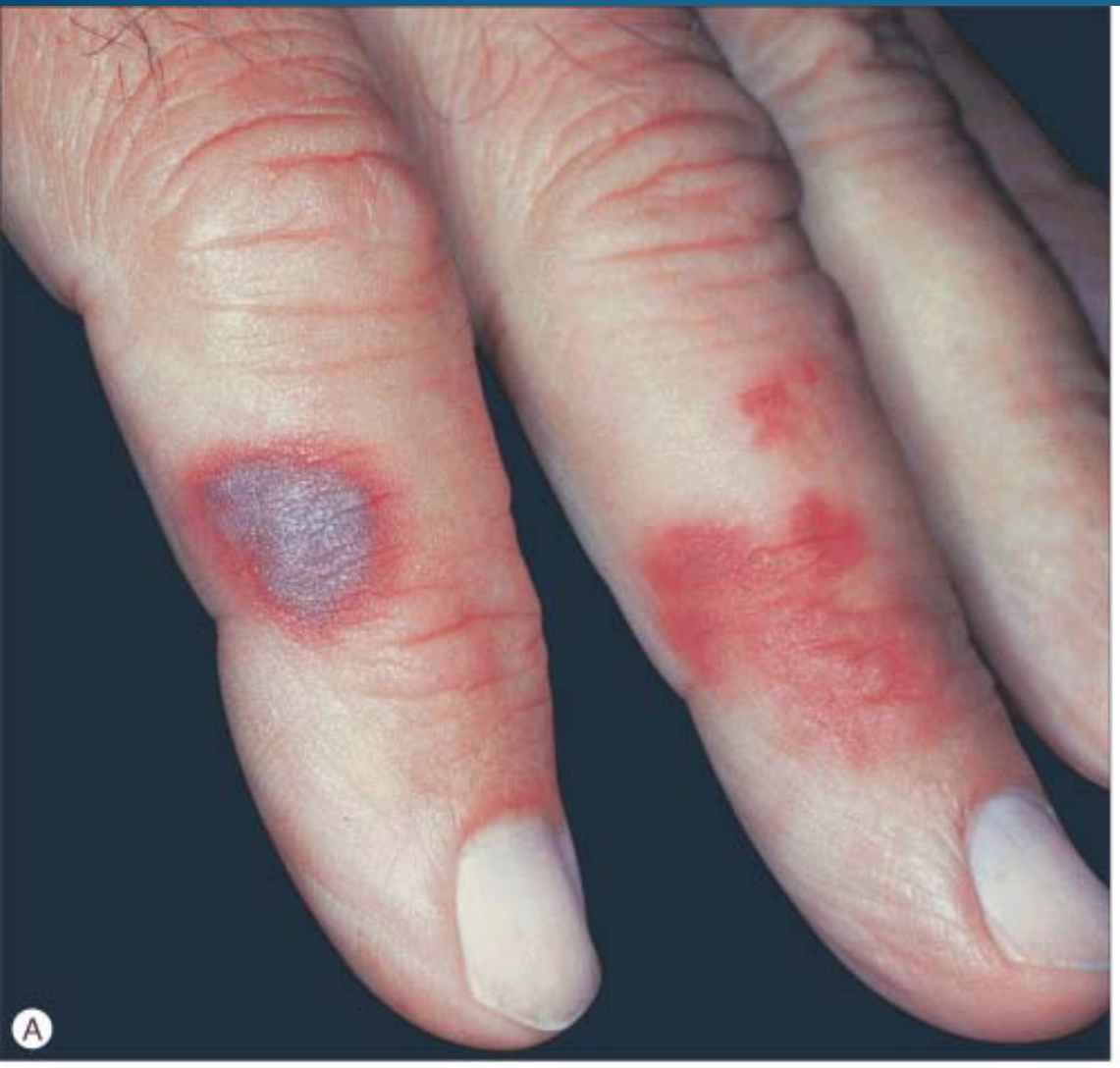
Microscopic Polyarteritis

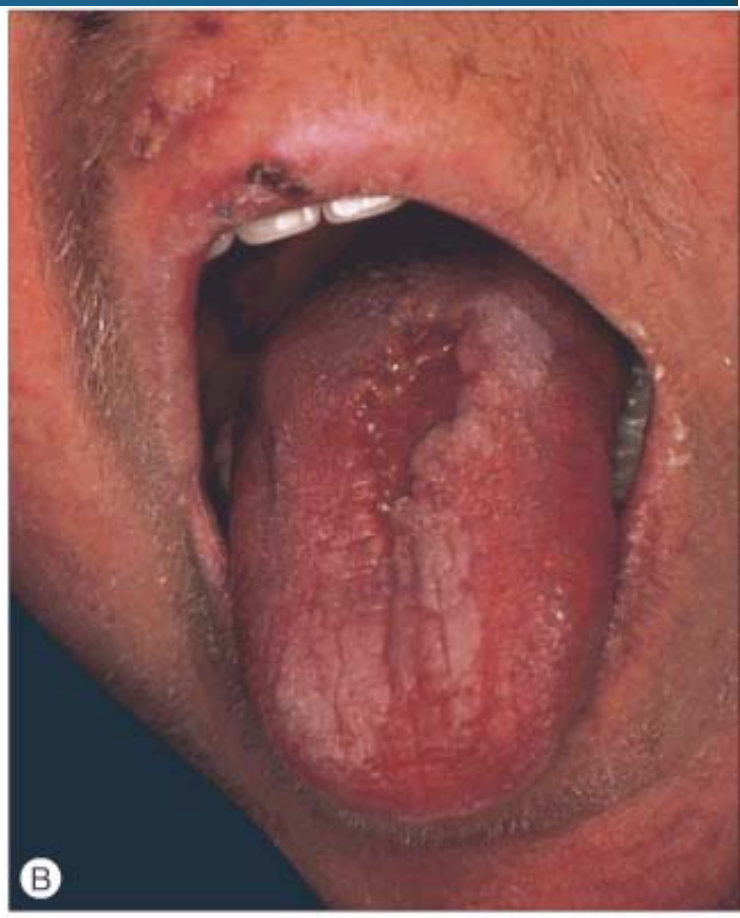
- Fever, weight loss, myalgias, arthralgias
- necrotizing GN (80-90%)
- pulmonary infiltrates, hemorrhage (25-50%)
- palpable purpura (46%)
- + ANCA (mostly p-ANCA, myeloperoxidase)
- Treat with po steroids and cyclophosphamide

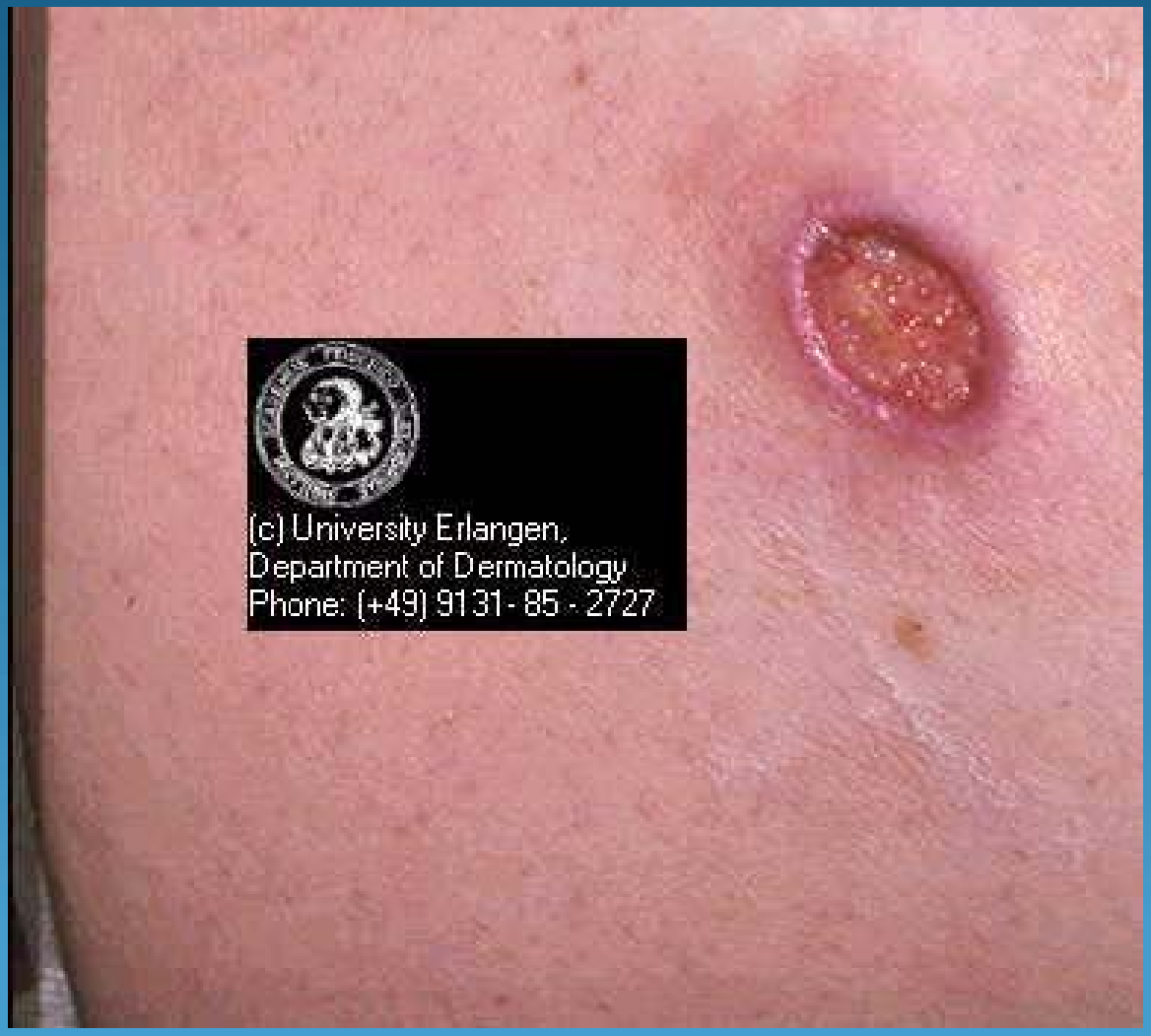


Wegener's granulomatosis

- Triad of necrotizing granulomatous inflammation of airways, systemic necrotizing vasculitis, and pauci-immune GN
- palpable purpura, oral ulcers, "PG-like" lesions, papulonecrotic lesions esp. on elbows can be mistaken for rheumatoid nod.
- 80% c-ANCA+ (proteinase-3)
- tx- steroids+cyclophosphamide, Imuran



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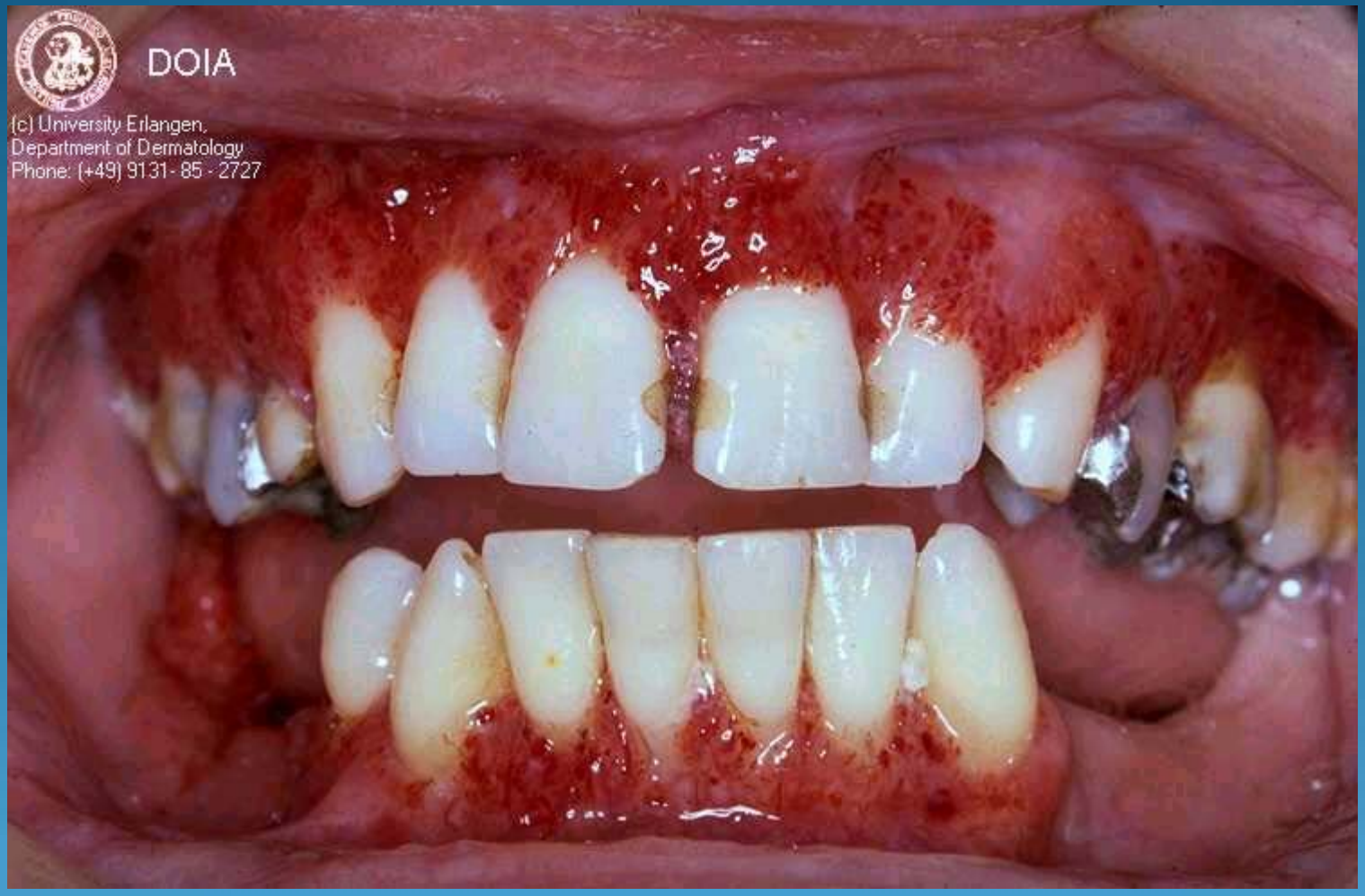
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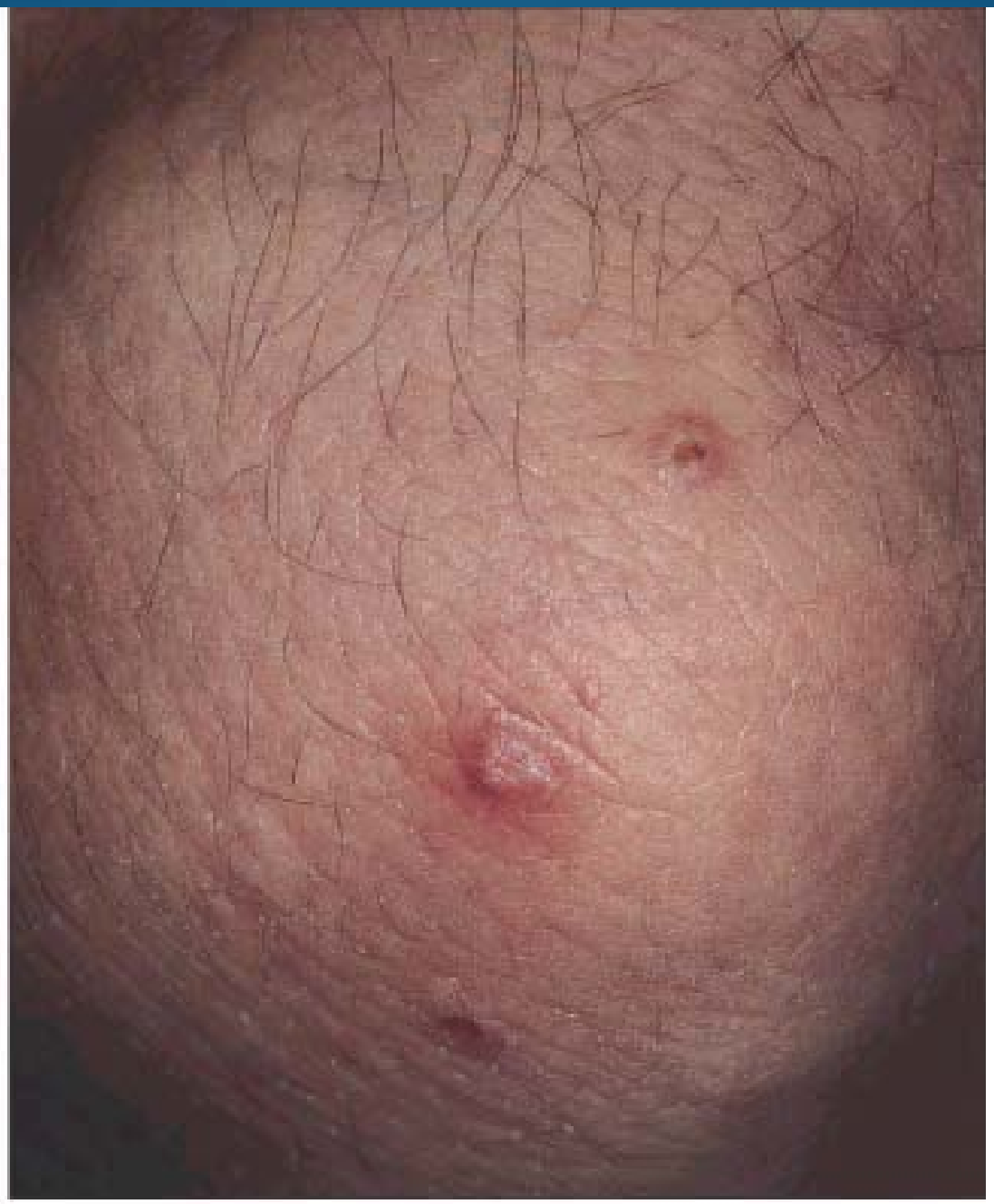
Churg-Strauss syndrome

- Allergic rhinitis, asthma that starts avg. age of 35
- eosinophilia and gastroenteritis
- systemic granulomatous necrotizing vasculitis
- palpable purpura, subQ nodules
- myocardial involvement leading cause of death
- rare renal involvement, unlike WG
- 60-70% p-ANCA+ (myeloperoxidase)
- Tx with systemic steroids

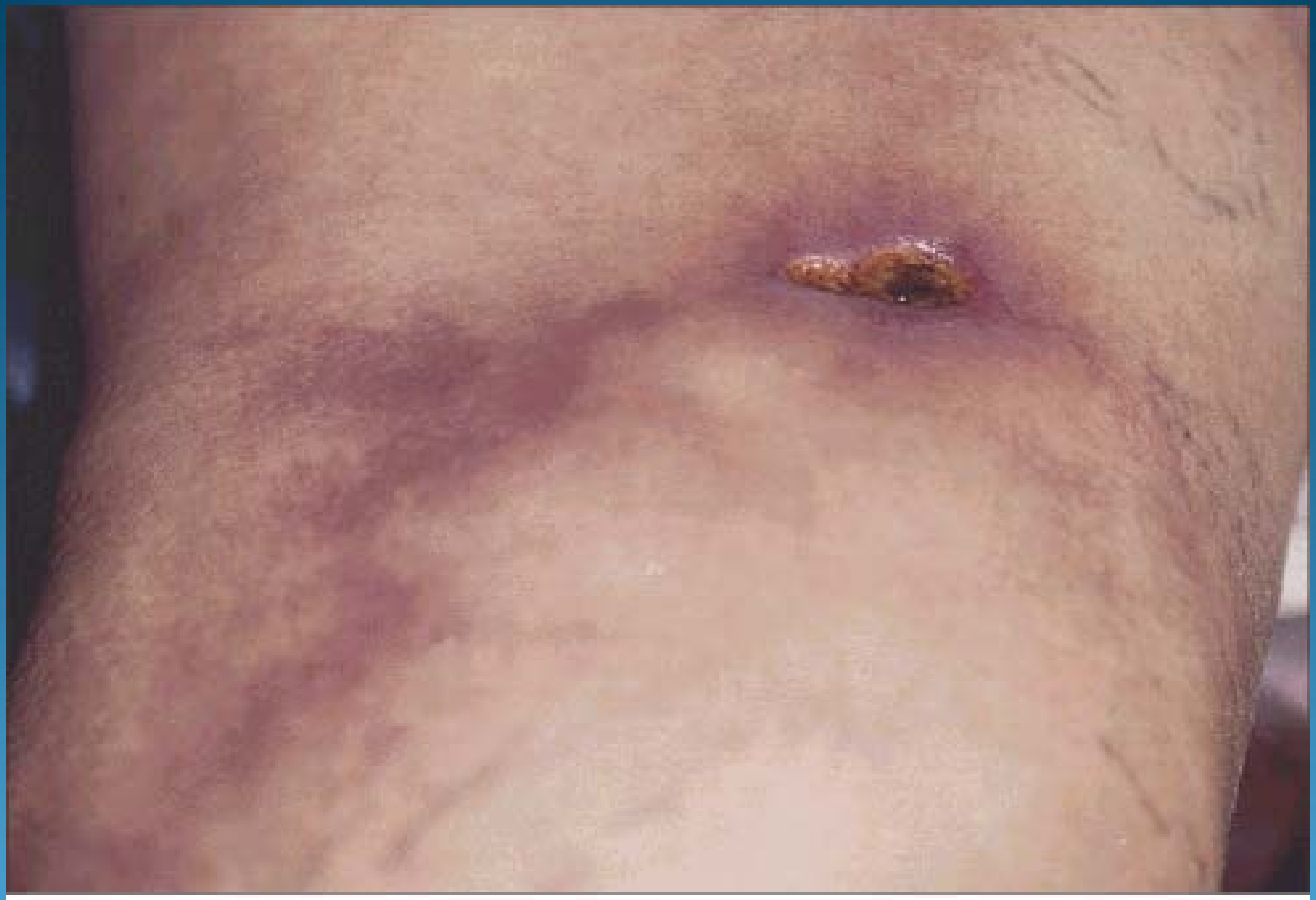


Drug-induced ANCA vasculitis

- Hydralazine
- propylthiouracil
- minocycline
- leukotriene inhibitors
- acral purpuric plaques and nodules esp. on extremities, face, breast and ears
- may develop pulmonary hemorrhage, GN, and digital gangrene

Rheumatoid vasculitis

- 5-15% of patients with RA
- seen mostly in middle-aged smokers with end stage RA and high RF titers
- palpable purpura, digital infarcts, nailfold infarctions (Bywaters lesions)
- systemic vasculitis rare (<1%) involves GI tract, heart, lungs, kidneys



SLE vasculitis

- can involve any size blood vessel
- represents flare of disease
- palpable purpura, uritcaria, livedo reticularis, microinfarcts of digits
- punched-out ulcers suggest systemic vasculitis

Scleroderma associated vasculitis

- Commonly affects skin and CNS
- palpable purpura, ecchymoses, Raynaud's, finger tip ulcaration and scarring

Approach to the Patient with Suspected Vasculitis

History

- chronicity (acute vs. chronic)
- preceding illness
- exposures to drugs, vaccines, chemicals
- systemic involvement?
  - Arthralgias, fever, hemoptysis, SOB, cough, wheezing, eye or ear sx, sinusitis, numbness, abd pain, melena, hematuria
- malignancy?- weight loss, night sweats
- CTD?- photosensitivity, oral lesions, muscle wkns

Physical Exam

- Small vessel
  - palpable purpura, pinpoint papules, hemorrhagic vesicles, petechiae, splinter hemorrhages, urticaria
- Medium vessel
  - subQ nodules, livedo reticularis, ulcers, papulonecrotic lesions, digital infarcts
  - HTN (may indicate renal involvement)

Laboratory Evaluation

- CBC with diff
- BUN/Cr
- LFT's
- U/A, stool guiac
- Hep B/Hep C panels
- cryoglobulins
- complement levels (CH50, C3, C4)
- RF
- +/- ANA
- +/- ANCA
- +/- CXR

The Biopsy

- Biospy newest lesion
- Document LCV
- size of vessels involved
- granulomatous inflamm (CSS or WG)
- lymphocyte rich infiltrate (CTD)
- immunofluorescence?