Vasculitis

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Credit: Dr. Bruner and Dr. Kamen

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Objectives

After studying this unit you should be able to:

1. Define vasculitis and describe its classification.
2. Describe differences in pathology between types of vasculitis.
3. Know the clinical features that distinguish different types of vasculitis.
4. Recognize the principles of treatment for the major types of vasculitis.

Definitions

- Vasculitis - “heterogeneous group of disorders characterized pathologically by evidence of blood vessel inflammation [with necrosis] and clinically by a diverse set of symptoms and signs.”

Ruddy: Kelley's Textbook of Rheumatology, 6th ed., 2001

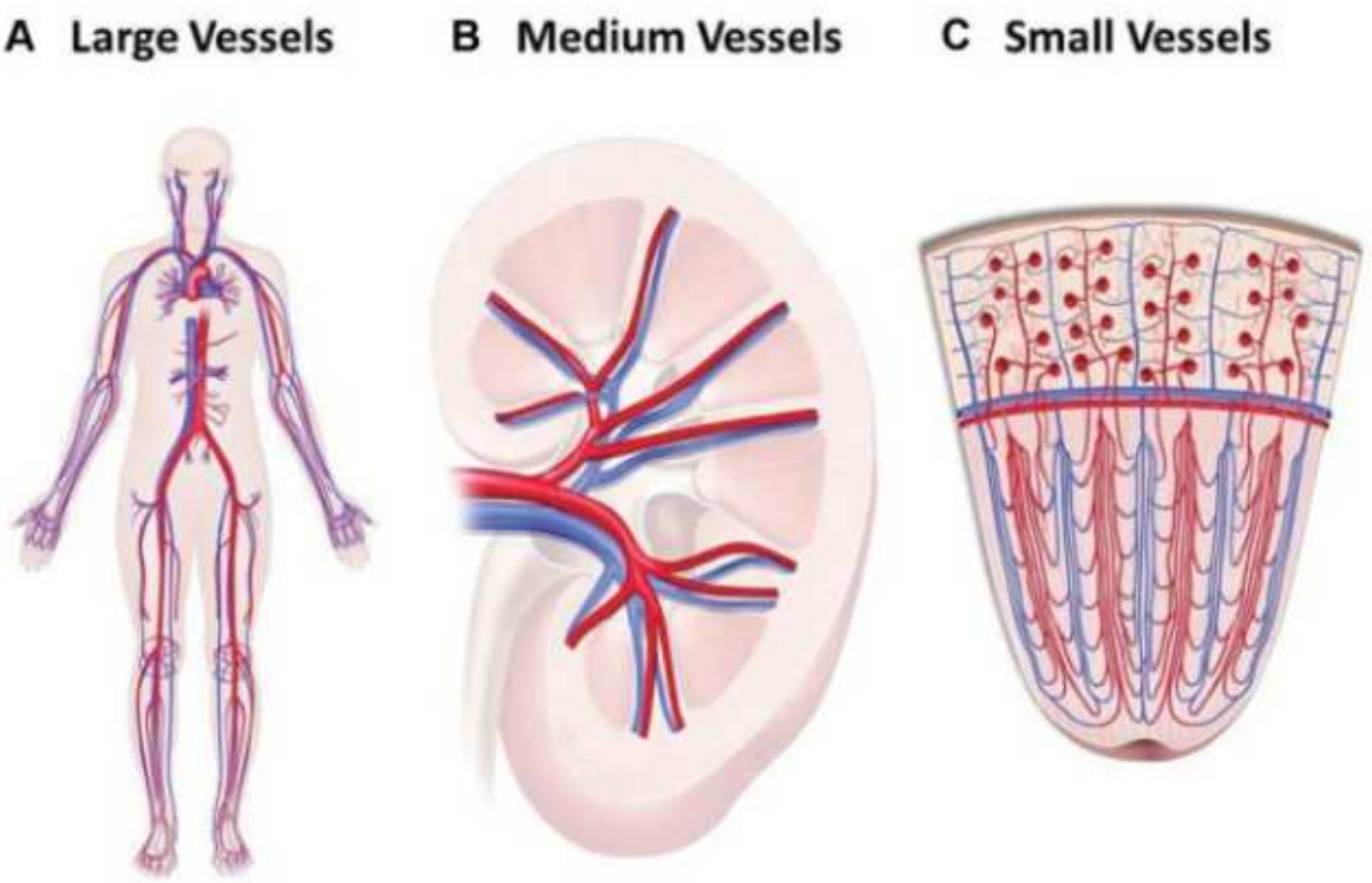
Vasculitis – Pathogenic Mechanisms

- Immune-mediated inflammation
- Direct vascular invasion by infectious organisms
  - Infections can also precipitate an immune mediated vasculitis through cross reactivity or immune complex formation
- Other causes: radiation, trauma, toxins
- Can target any organ/tissue (because they all have vessels…)

- Important to rule out an infectious vasculitis because immunosuppression is given for immune-mediated vasculitis.

- In this lecture we will address the **immune-mediated vasculitides.**

Classification (2012 Revised International Chapel Hill Consensus)



JC Jennette, RJ Falk, et al. Arthritis & Rheumatism 2013.

Sites of Vascular Involvement

Variable vessel diameter vasculitis
- Behcet's disease (BD)
- Cogan's

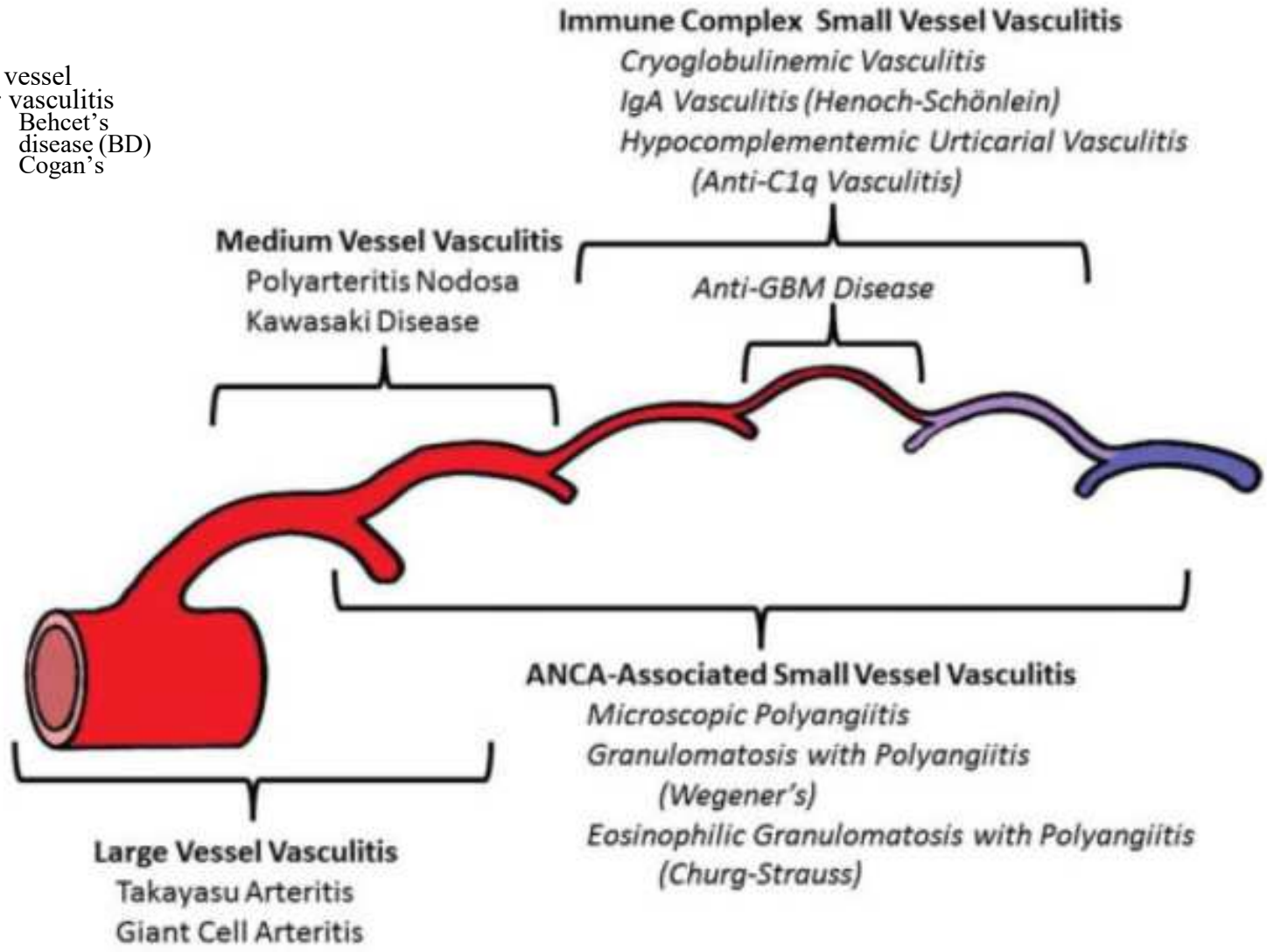


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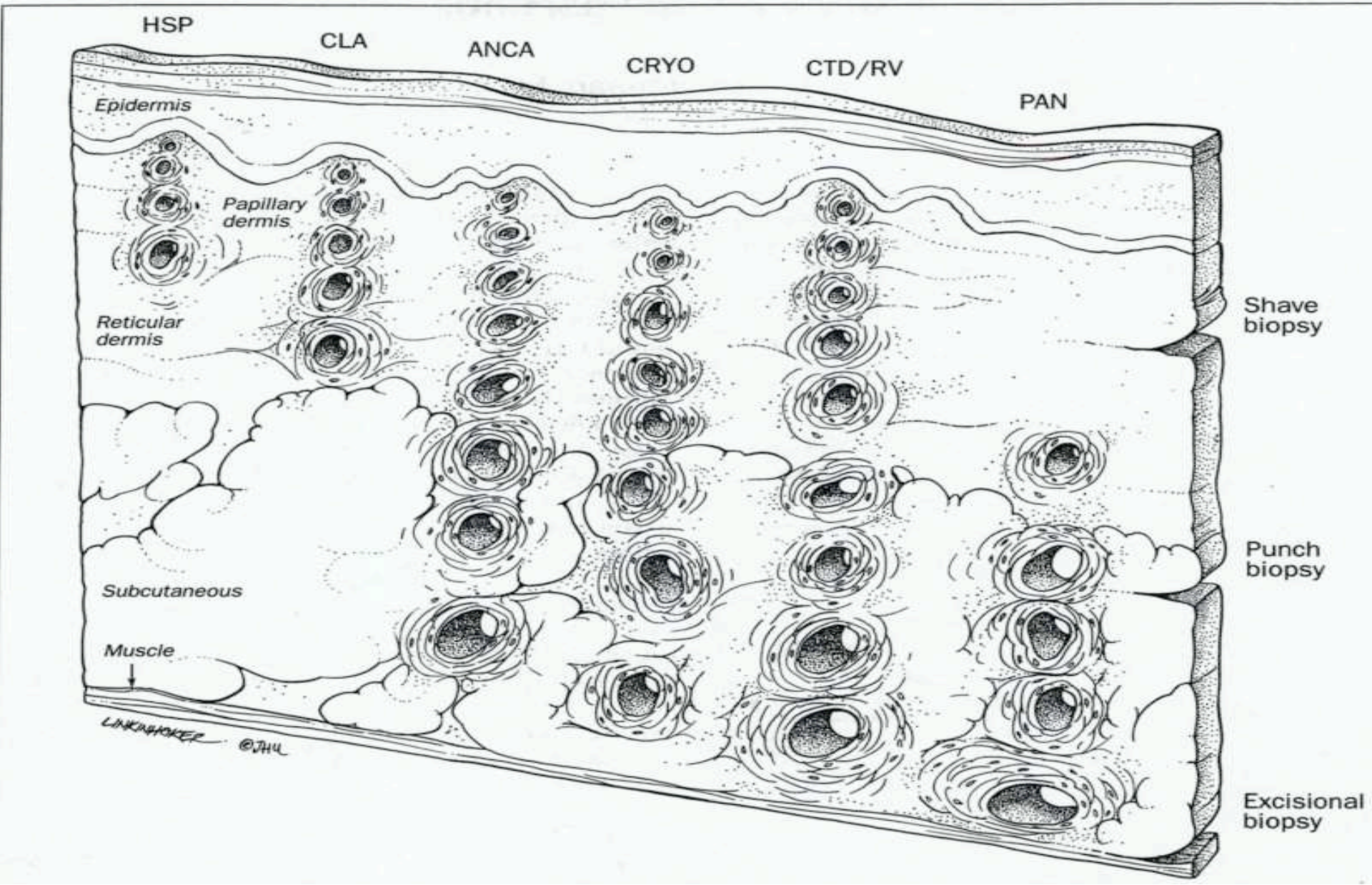
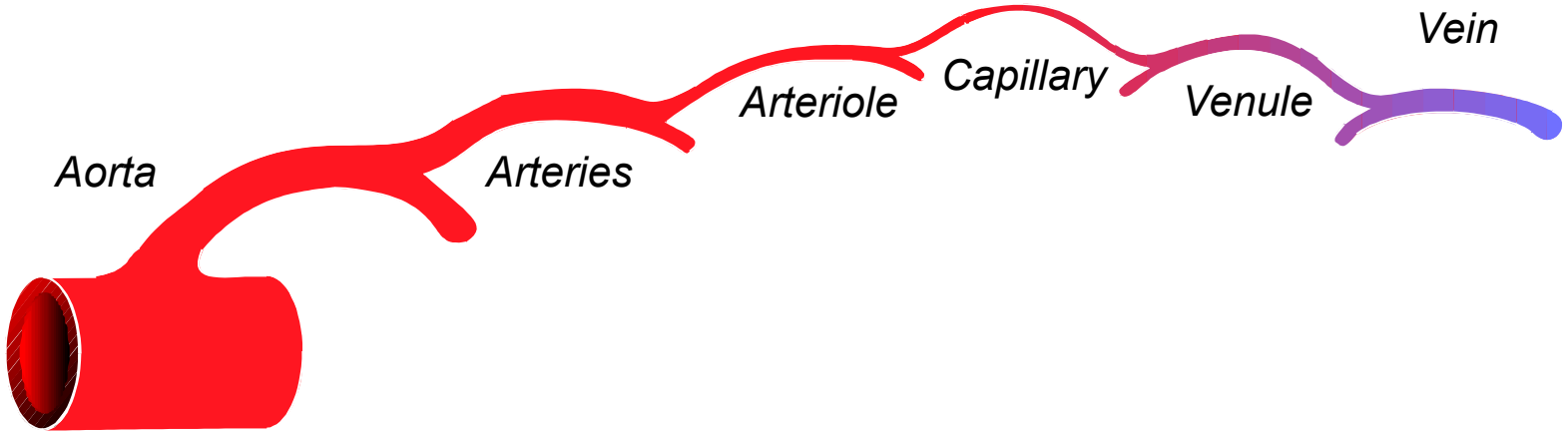
Figure 1. Schematic view of skin layers illustrating the size of blood vessels involved in common forms of cutaneous vasculitis





Large Vessel Vasculitis

Chronic granulomatous arteritis

- granulomatous arteritis in a patient >50 yo → Giant Cell Arteritis
- granulomatous arteritis in a patient <50 yo → Takayasu Arteritis

Giant Cell Arteritis (GCA)

- AKA temporal arteritis or cranial arteritis
- Only in patients > 50 years old
  - mean 70 years old
  - Women : Men 2:1

GCA Vessel Involvement

- Highest incidence of severe involvement
  - **superficial temporal arteries (classic presentation)**
  - vertebral arteries
  - ophthalmic and posterior ciliary arteries
- Somewhat less common
  - internal and external carotids
  - central retinal arteries
  - subclavian and brachial arteries
  - abdominal arteries

GCA Clinical Presentation

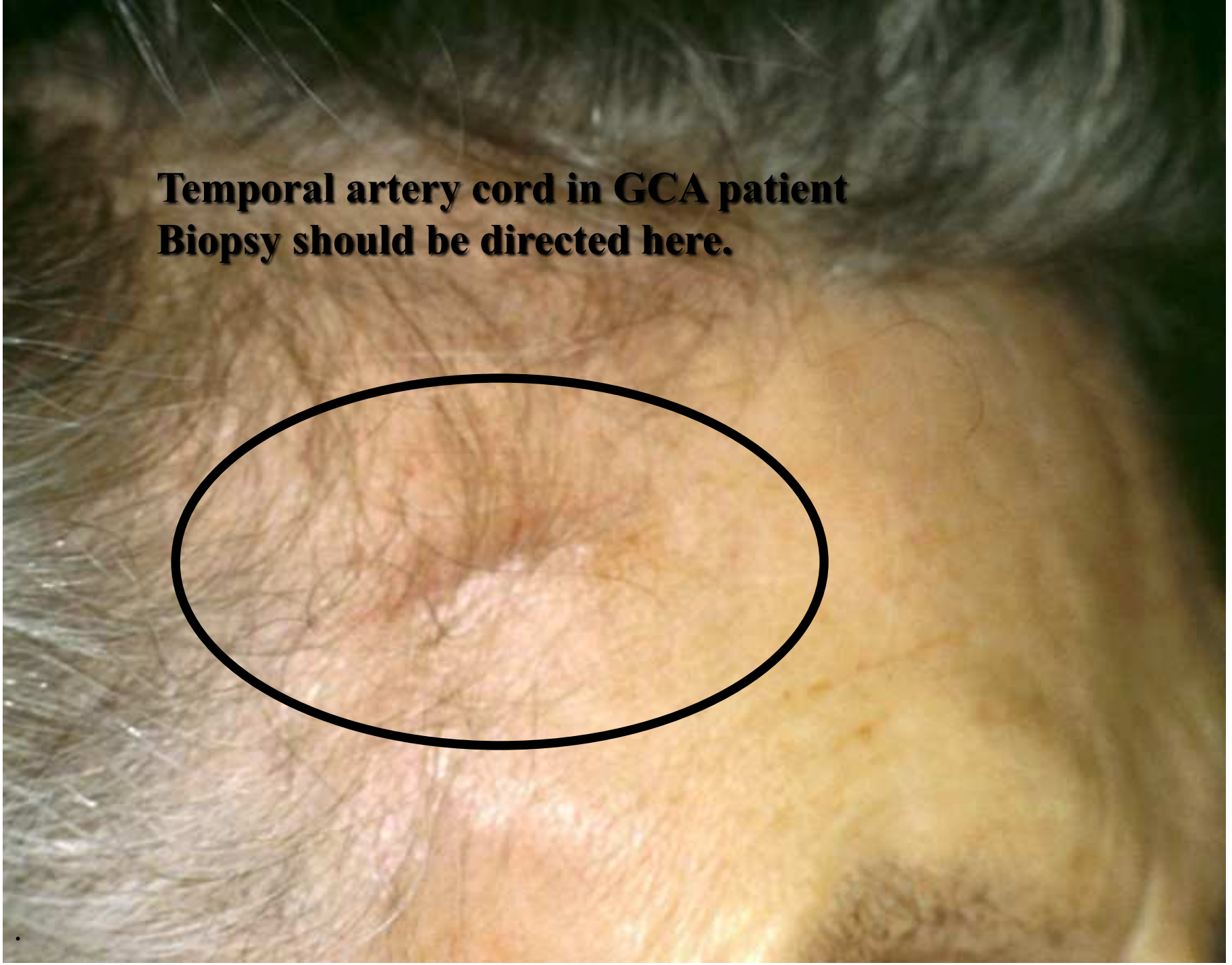
- Insidious onset
  - Fatigue
  - Anorexia / weight loss
  - Fever (10% of FUO cases are GCA)
  - Polymyalgia Rheumatica (PMR) overlap in 40%
    - (16% of PMR patients have GCA)
  - Elevated erythrocyte sedimentation rate (ESR) and/or C-reactive protein (CRP)
  - Anemia of chronic disease (mild – moderate)

GCA Clinical Presentation

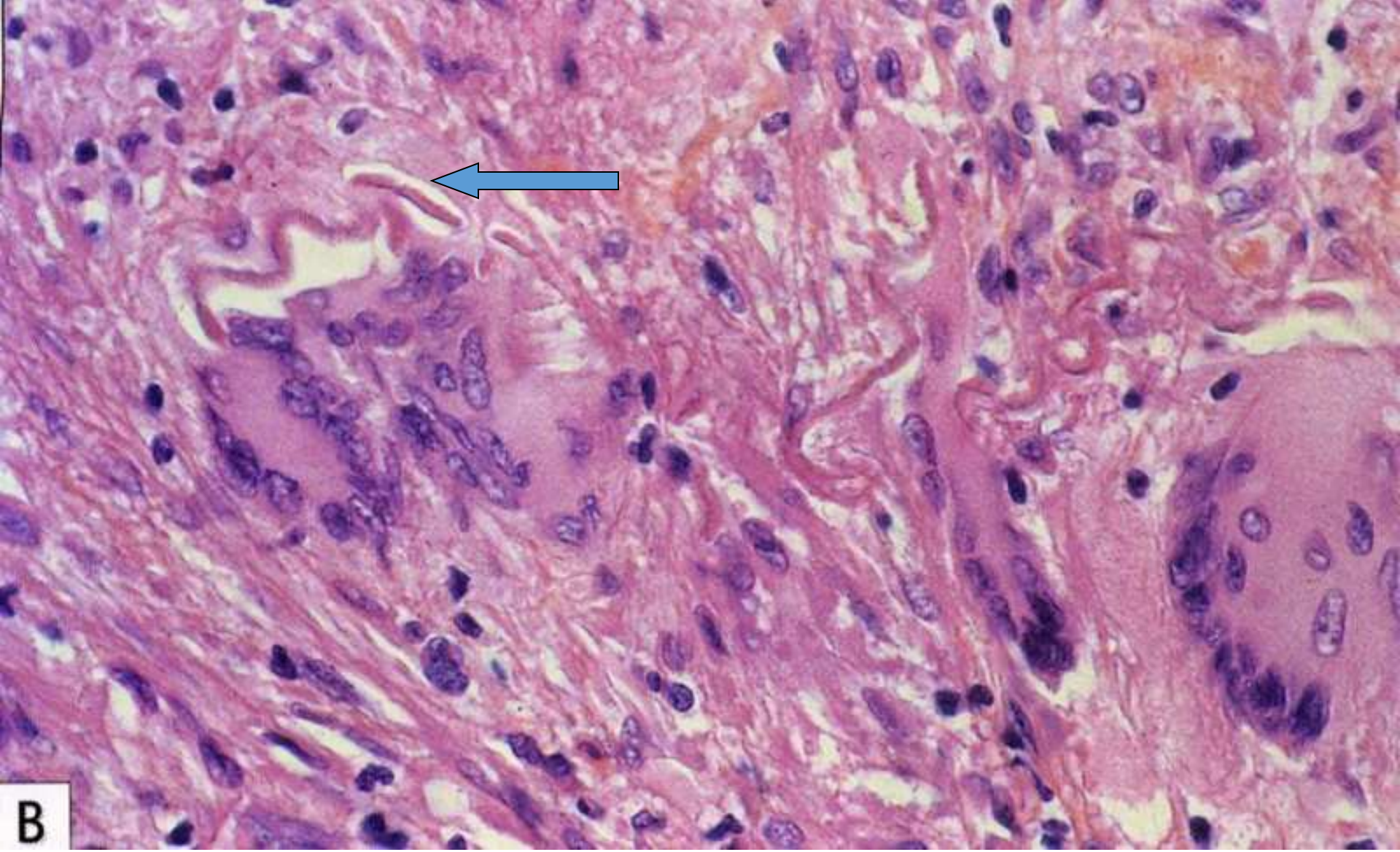
- More abrupt onset
  - Headache, unilateral (66% of GCA patients)
  - Scalp tenderness
  - Visual symptoms
    - diplopia, ptosis, and transient or permanent blindness (amaurosis fugax)
    - ischemic optic neuritis with pallor/edema of optic disk
  - Jaw claudication
  - Tongue claudication

Best Diagnosis is by Biopsy

- Can use vascular ultrasound to help guide biopsy
- Temporal artery biopsy is the gold standard
  - 3 - 5 cm section of vessel
  - several cross sections/cm
  - 5% increase in sensitivity with bilateral biopsy
- Don’t delay treatment before obtaining biopsy
  - If treatment delayed, can result in permanent blindness
- Biopsy specimens may be positive after up to 2 weeks of corticosteroid therapy


Temporal artery cord in GCA patient
Biopsy should be directed here.

H&E stain of section of temporal artery showing giant cells at the degenerated internal elastic membrane in active arteritis (arrow).



Higher magnification with giant cell associated with elastica (arrow)

GCA Treatment

- Initially 40-60 mg prednisone or equivalent
  - Increased, IV doses if visual loss
  - Reduce dose by 10% every 2 weeks
    - follow symptoms
    - follow ESR, CRP
  - May slow down, stop or reverse taper if ESR rises or symptoms return
- Steroid-sparing agents
  - Methotrexate
  - Tocilizumab (anti-IL6 receptor monoclonal antibody)

Course of GCA

- Treated cases usually remit in 1-2 years
- PMR symptoms may return at lower doses of prednisone
- Unilateral blindness followed rapidly by bilateral blindness if not treated
- Recurrence much less likely with normal ESR

Takayasu Arteritis

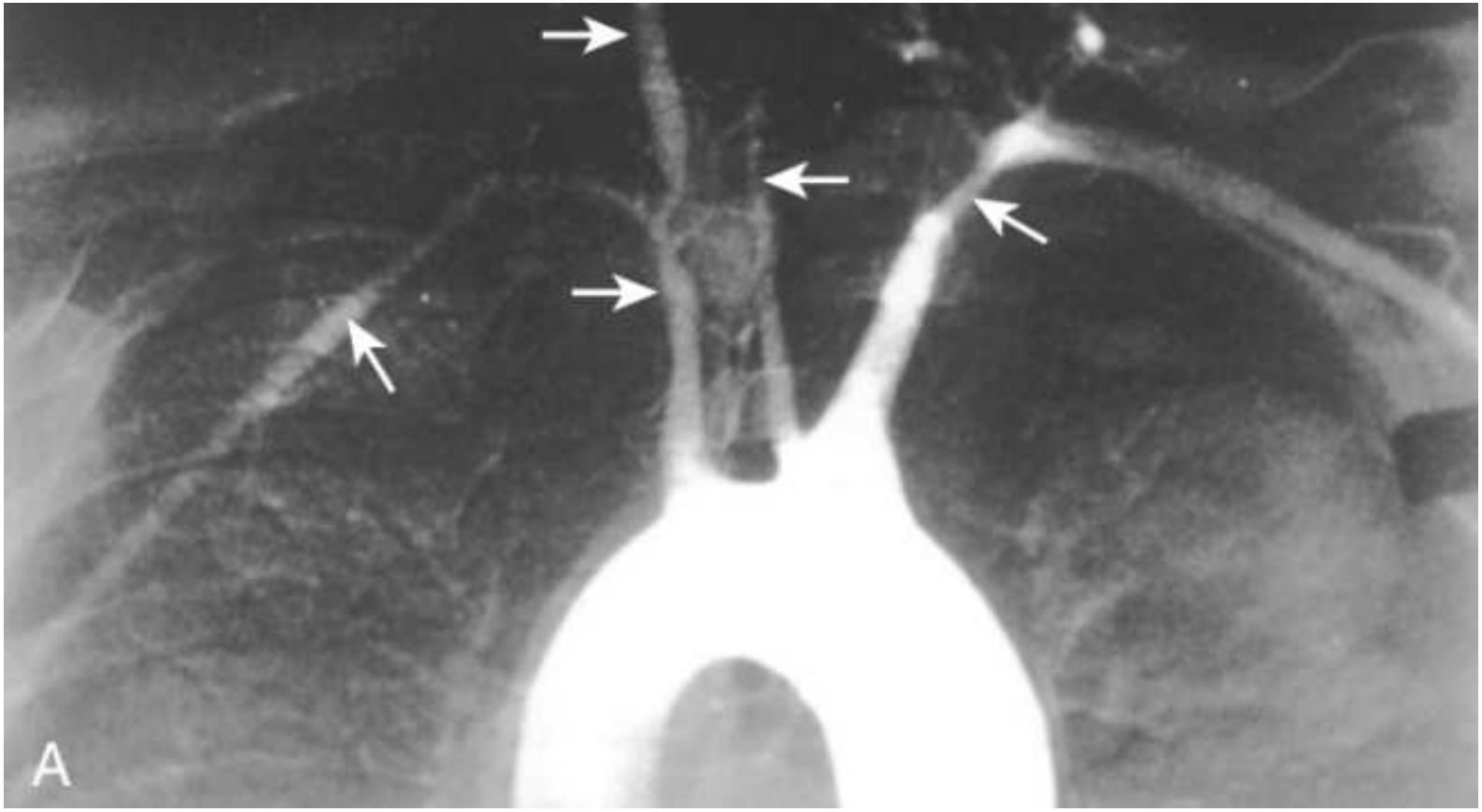
- Female : Male 4:1
- Usually between 10 and 30 years old
- Incidence about 2.6/million
- Involves aorta and its branching arteries

Takayasu Clinical Presentation

- Most common
  - upper extremity claudication (“pulseless disease”)
  - hypertension
  - carotidynia
  - dizziness
  - visual abnormalities

Diagnosis of Takayasu Arteritis

- High index of suspicion in young person presenting with:
  - CVA, MI, extremity or bowel claudication, or renovascular hypertension
  - Absent/decreased pulses in extremities
  - systemic symptoms (fatigue, weight loss)
  - elevated ESR
  - low albumin, high globulin fraction
- Angiography
  - conventional or MRI / MRA (to show inflammatory lesions)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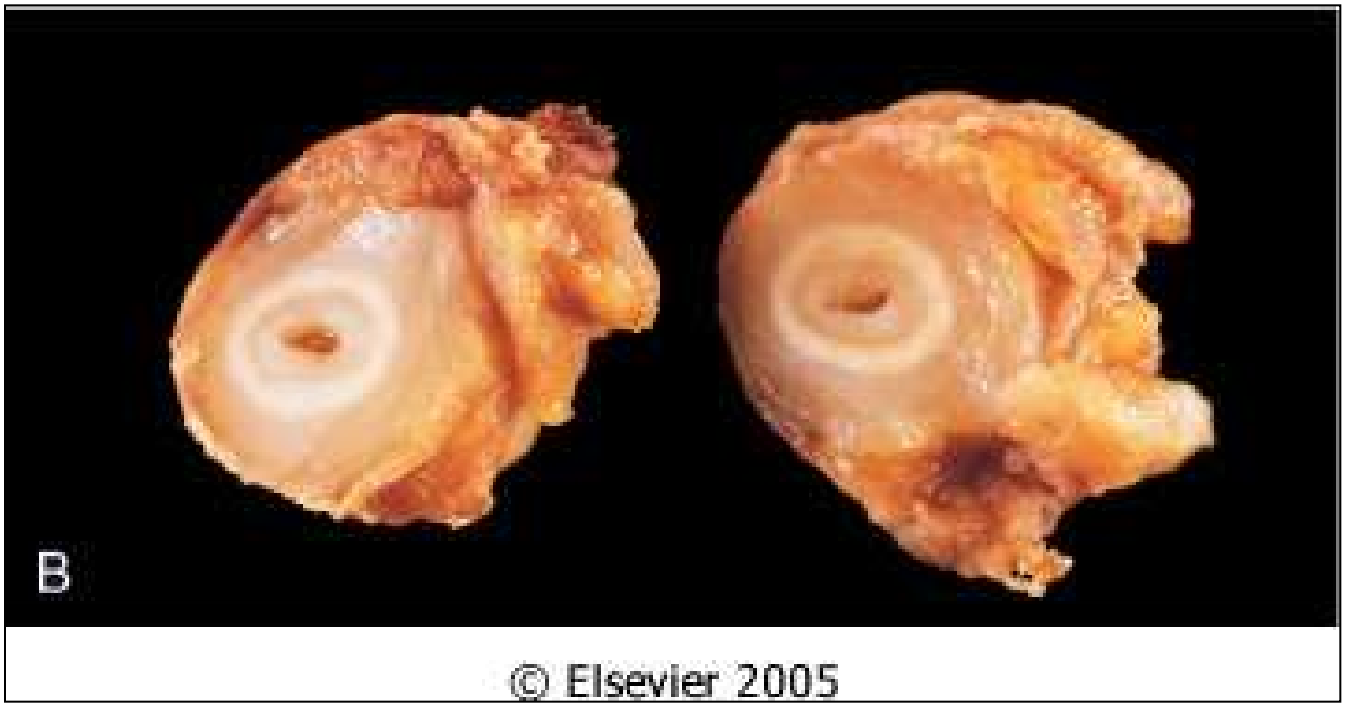
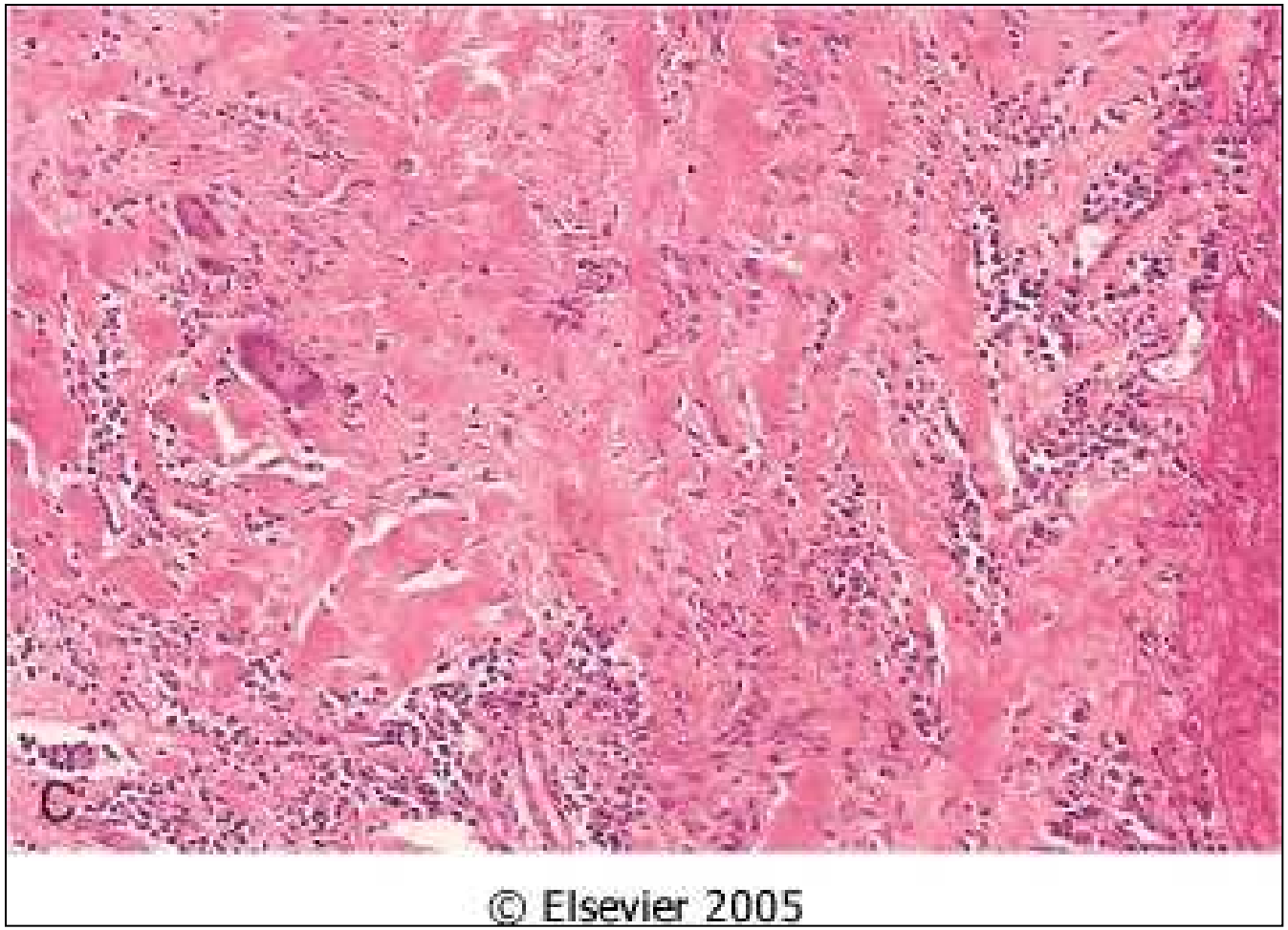


Gross photograph of two cross-sections of the right carotid artery taken at autopsy a patient, demonstrating marked intimal thickening with minimal residual lumen.



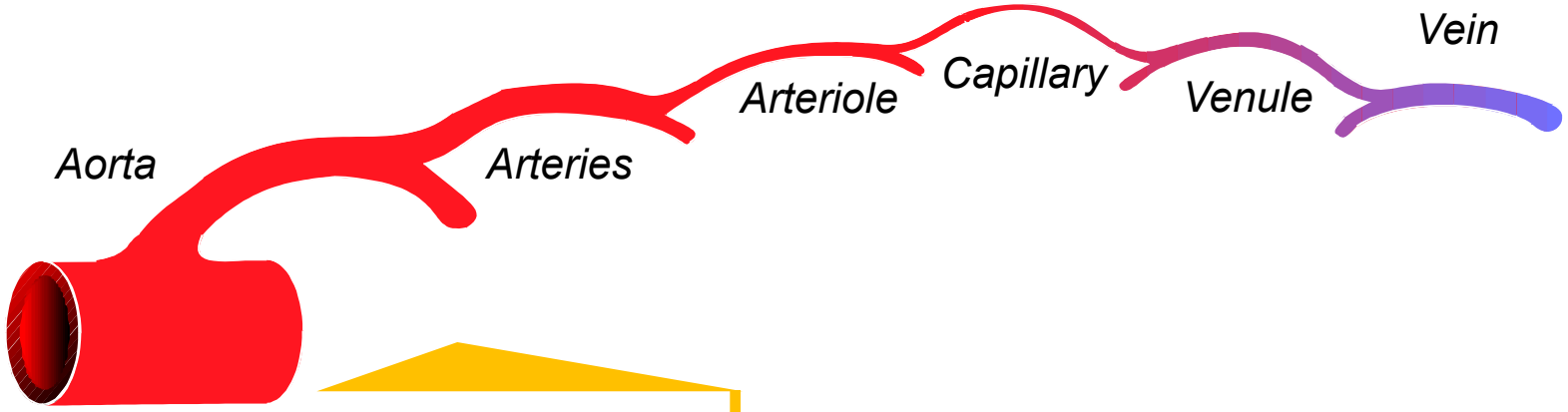
Histologic view of active Takayasu aortitis, illustrating destruction of the arterial media by mononuclear inflammation with giant cells.

Takayasu Arteritis Treatment

- 1 mg/kg/day prednisone
  - gradual taper
  - observation for flare
  - ESR not always suppressed therefore not good guide

Course of Takayasu Arteritis

- Gradual taper of steroids over months to years
- 50% have vessel stenosis despite therapy
  - surgical bypass
  - aortic valve repair
- 90% overall survival



Medium Vessel Vasculitis

- Necrotizing arteritis <u>without</u> mucocutaneous lymph node (MCLN) syndrome
  - Polyarteritis Nodosa (PAN)
- Necrotizing arteritis <u>with</u> MCLN syndrome
  - Kawasaki Disease

Polyarteritis Nodosa (PAN)

- Any age (including children)
- Peak in 50-60 year olds
- Male : Female 2:1
- **Association with hepatitis B** but usually idiopathic
- Prevalence estimated to range from 2 to 33 per million
  - The reduction in hepatitis B infection has been associated with a reduction in PAN prevalence

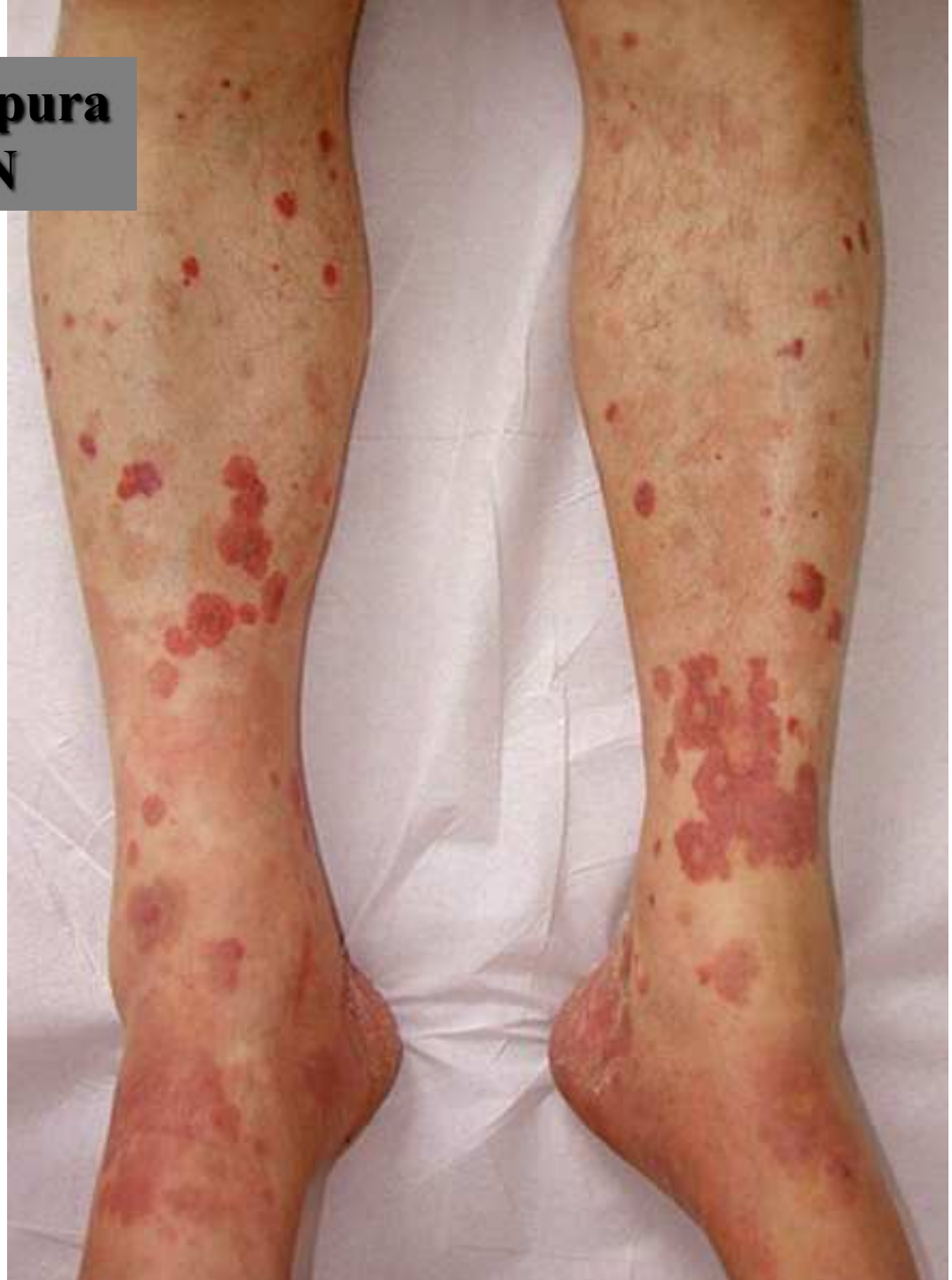
PAN Clinical Presentation

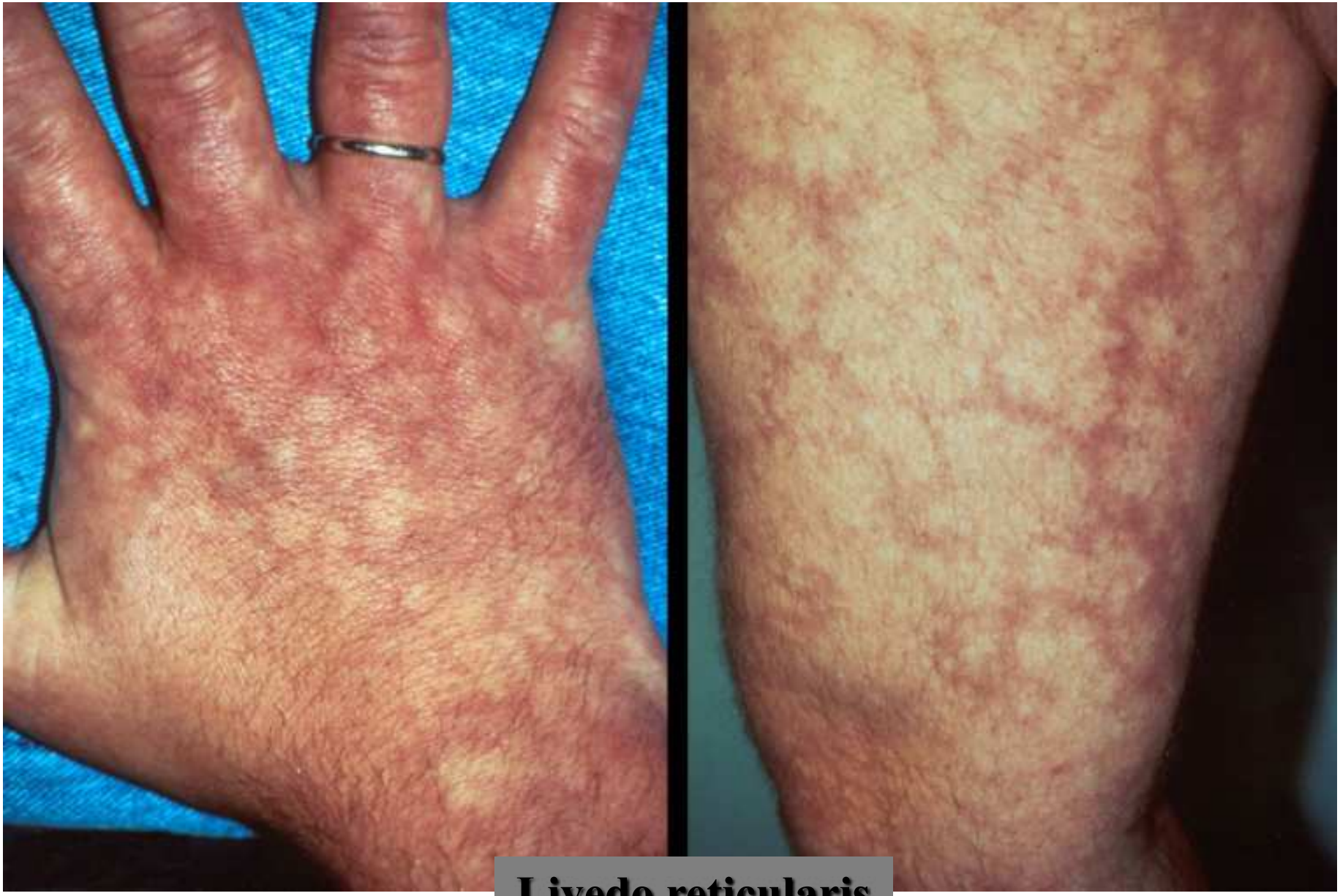
- Insidious onset systemic symptoms
  - weight loss
  - fever
  - myalgias
  - arthralgias
  - peripheral sensory neuropathy

PAN Clinical Presentation

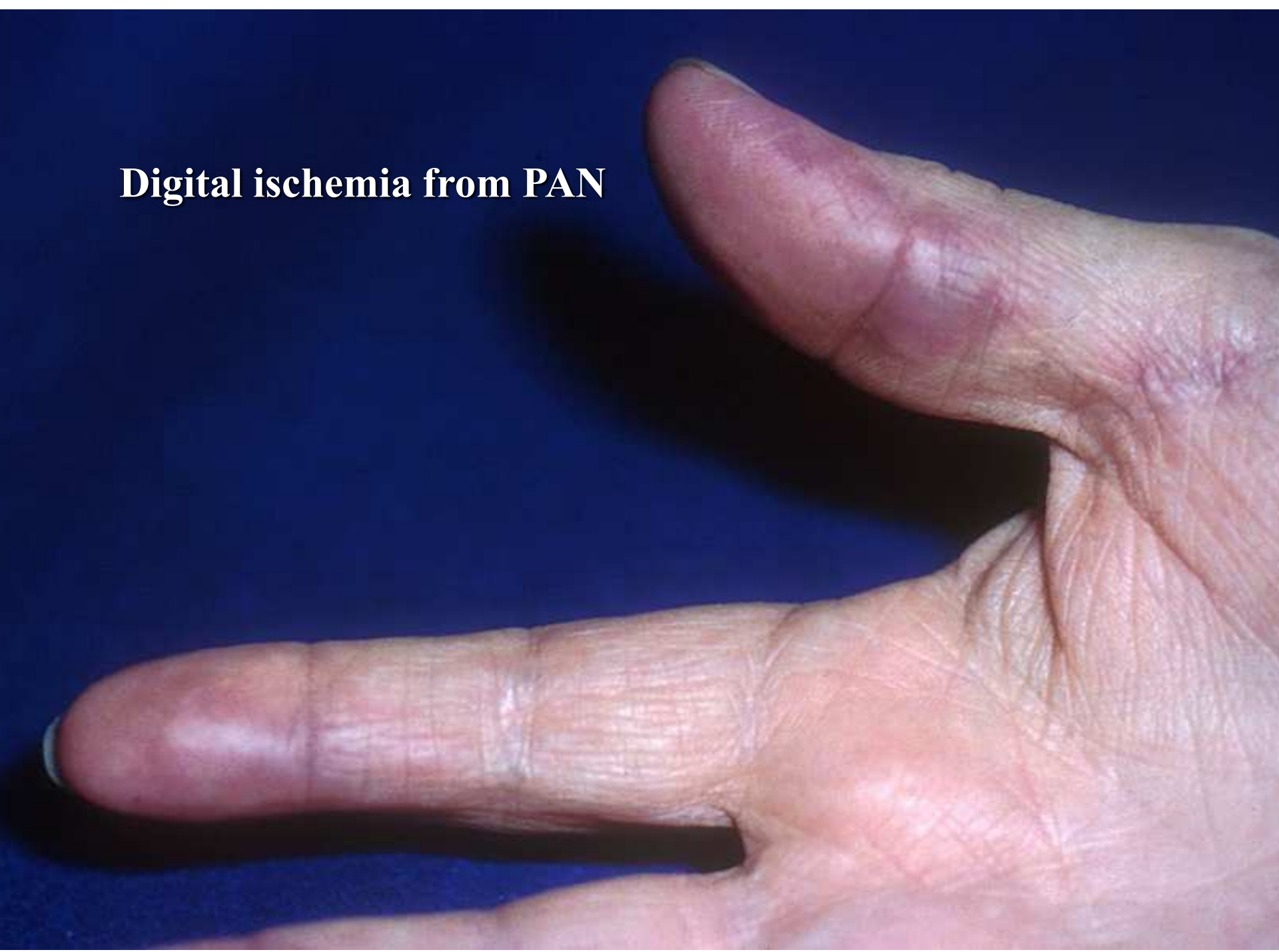
- Abrupt onset of
  - palpable purpura (1/3 of cases)
  - new onset hypertension (renal artery involvement)
  - intestinal ischemia: abdominal pain and bloody stools
  - infarction of a kidney or other major organ
  - loss of multiple nerves (mononeuritis multiplex)
    - Classic presentation is a wrist drop
  - livedo reticularis, splinter hemorrhages, digital gangrene
  - testicular pain
- Striking tendency to spare the lungs

Palpable purpura from PAN



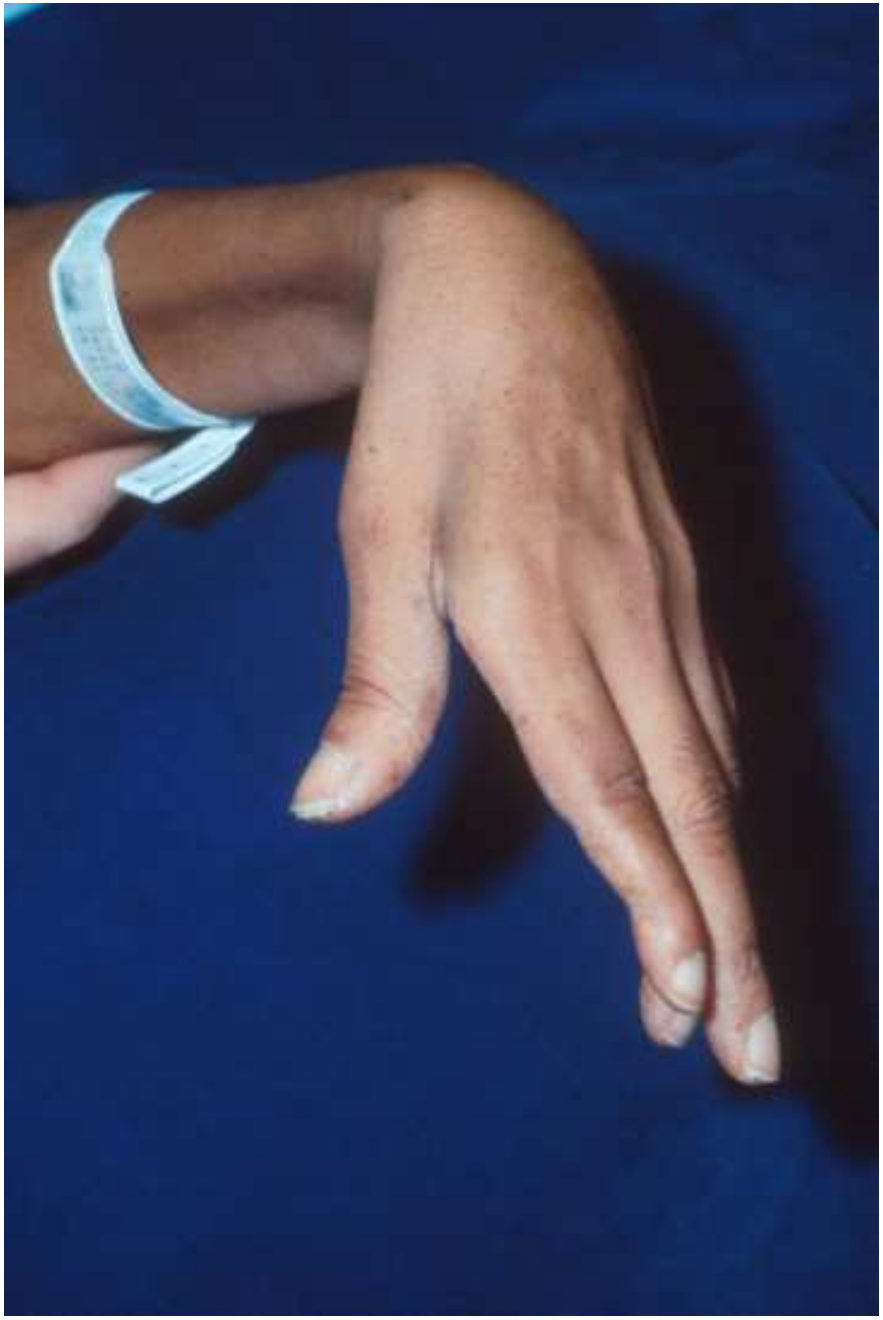


Livedo reticularis

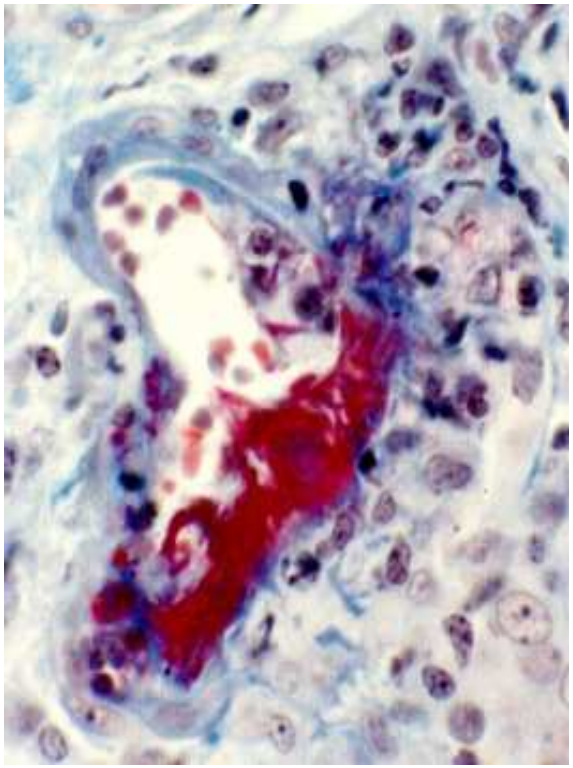

Digital ischemia from PAN

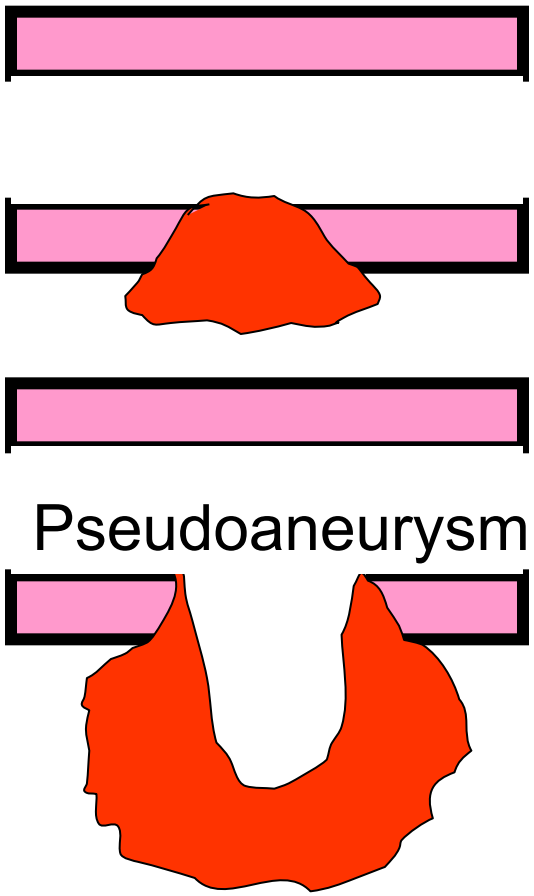

Digital ischemia from PAN

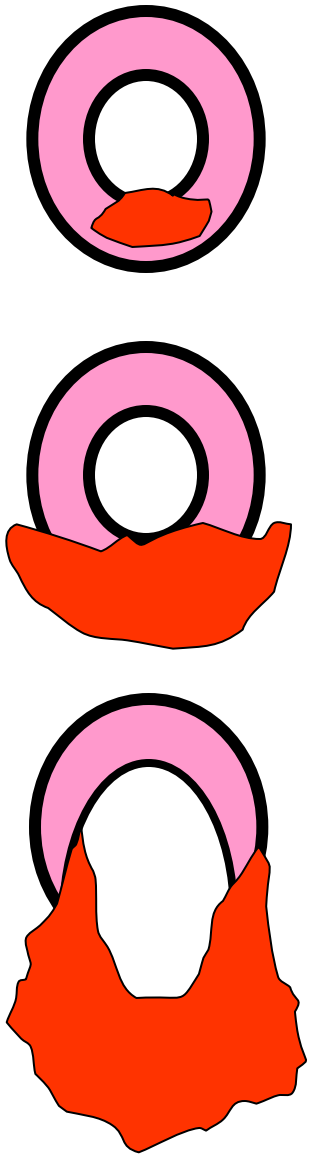
Wrist drop from mononeuritis multiplex associated with PAN



If the necrotizing inflammation extends through the vessel wall into the perivascular tissue, inflammatory aneurysms (pseudoaneurysms) will develop.

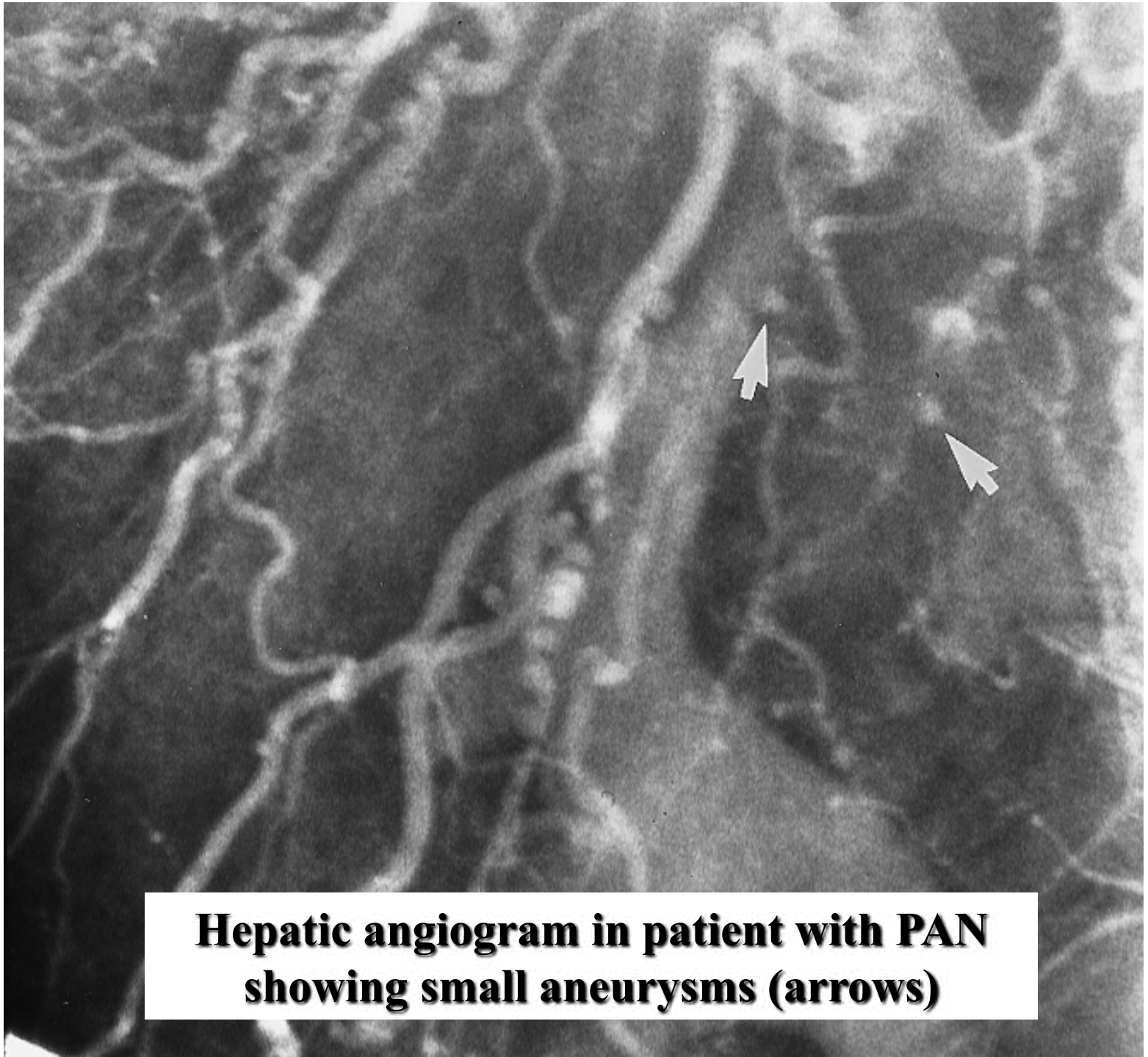






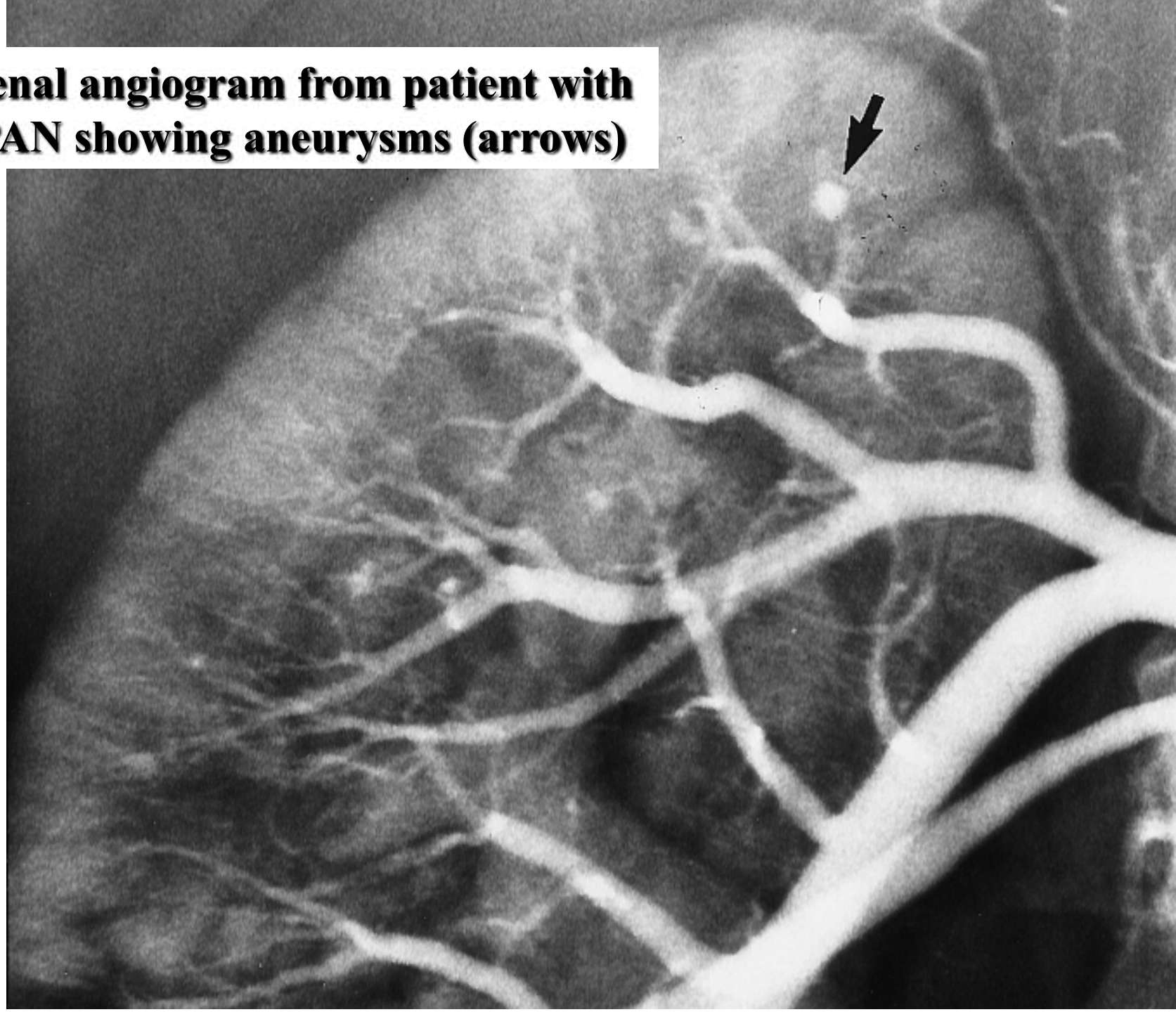
Diagnostic Studies for PAN

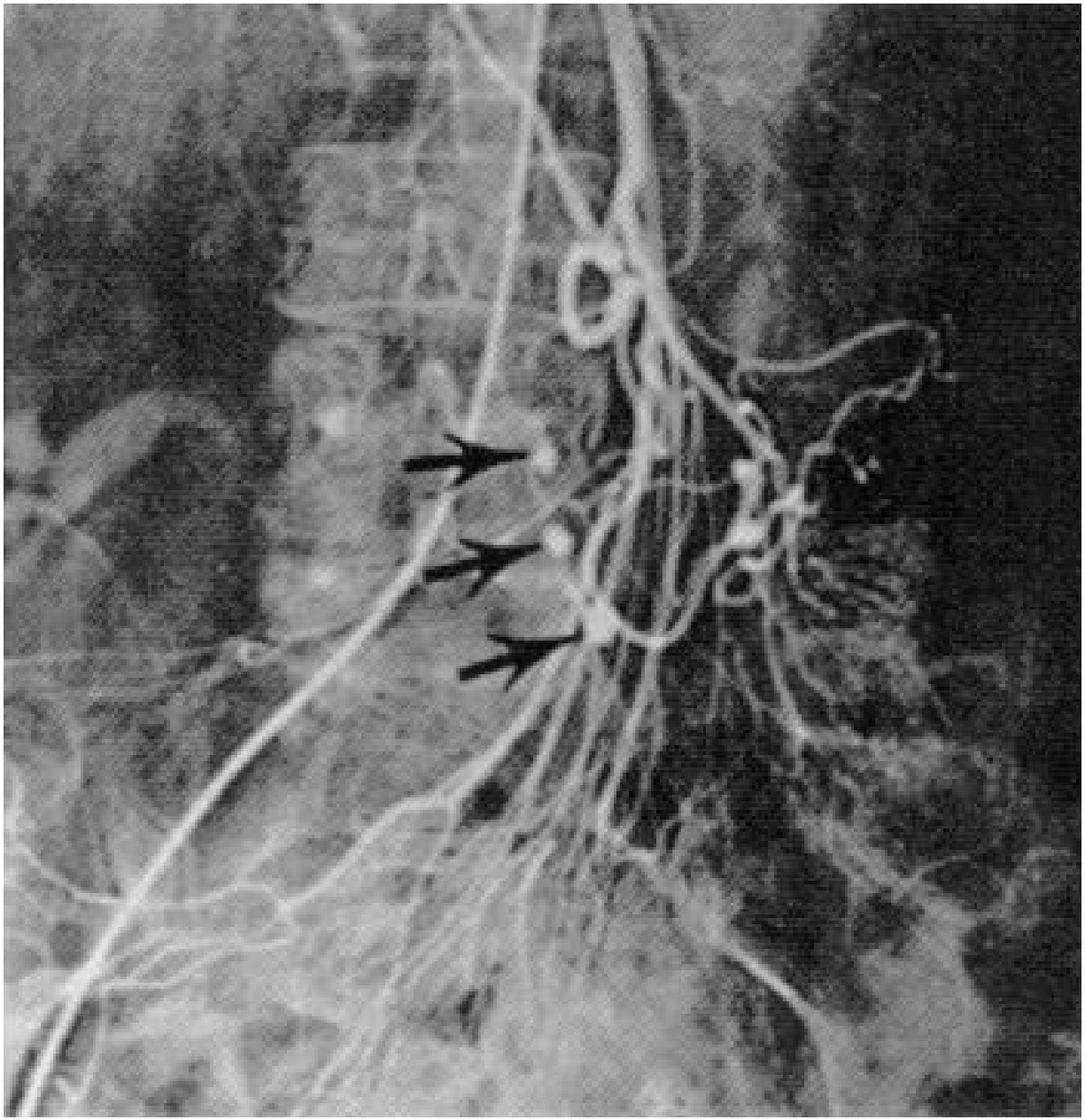
- Biopsy
  - if skin involvement, biopsy may be diagnostic
  - target organ (but avoid kidney biopsy if possible)
- Laboratory evaluation
  - ESR
  - hepatitis B serology
- Angiography
  - guided by clinical organ involvement



Hepatic angiogram in patient with PAN showing small aneurysms (arrows)

Renal angiogram from patient with PAN showing aneurysms (arrows)





Mesenteric arteriogram in patient with PAN and abdominal pain

ACR Classification Criteria of PAN

Classified as PAN in patients with a documented vasculitis in whom at least 3 of the following are present:

- Otherwise unexplained weight loss greater than 4 kg
- Livedo reticularis
- Testicular pain or tenderness
- Myalgias (excluding that of the shoulder and hip girdle), weakness of muscles, tenderness of leg muscles, or polyneuropathy
- Mononeuropathy or polyneuropathy
- New-onset diastolic blood pressure greater than 90 mmHg
- Elevated levels of serum blood urea nitrogen (>40 mg/dL or 14.3 mmol/L) or creatinine (>1.5 mg/dL or 132 micromol/L)
- Evidence of hepatitis B virus infection via serum antibody or antigen serology
- Characteristic arteriographic abnormalities not resulting from noninflammatory disease processes
- A biopsy of small- or medium-sized artery containing polymorphonuclear cells

Sensitivity of 82% and specificity of 87%

PAN Treatment

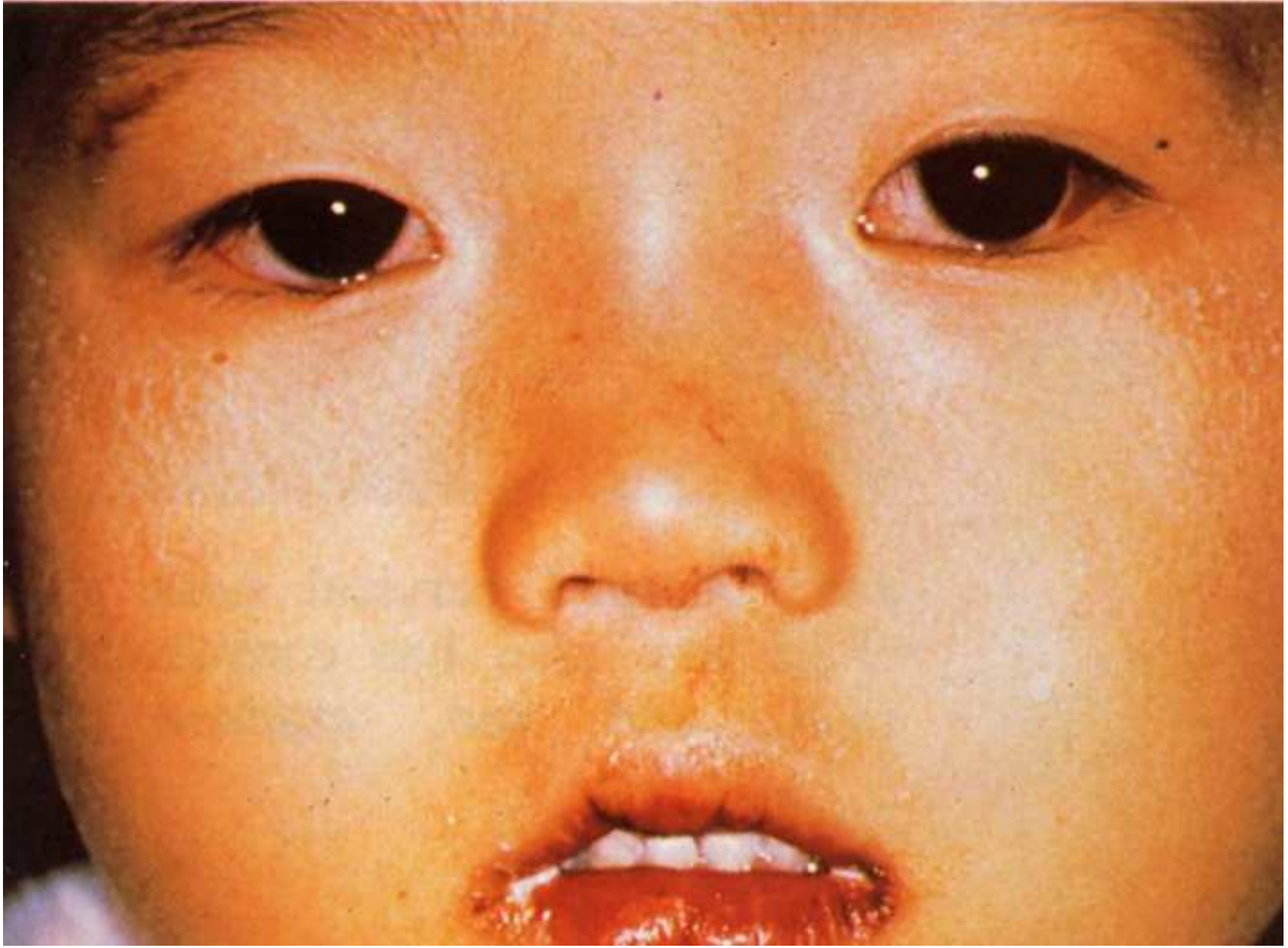
- Universally fatal in 1-2 years if not treated

- Prednisone
  - 1 mg/kg/day in divided doses initially
  - pulse of methylprednisolone 1 g IV if fulminant disease
- Cytotoxic drugs in addition
  - oral cyclophosphamide up to 2-4 mg/kg/day

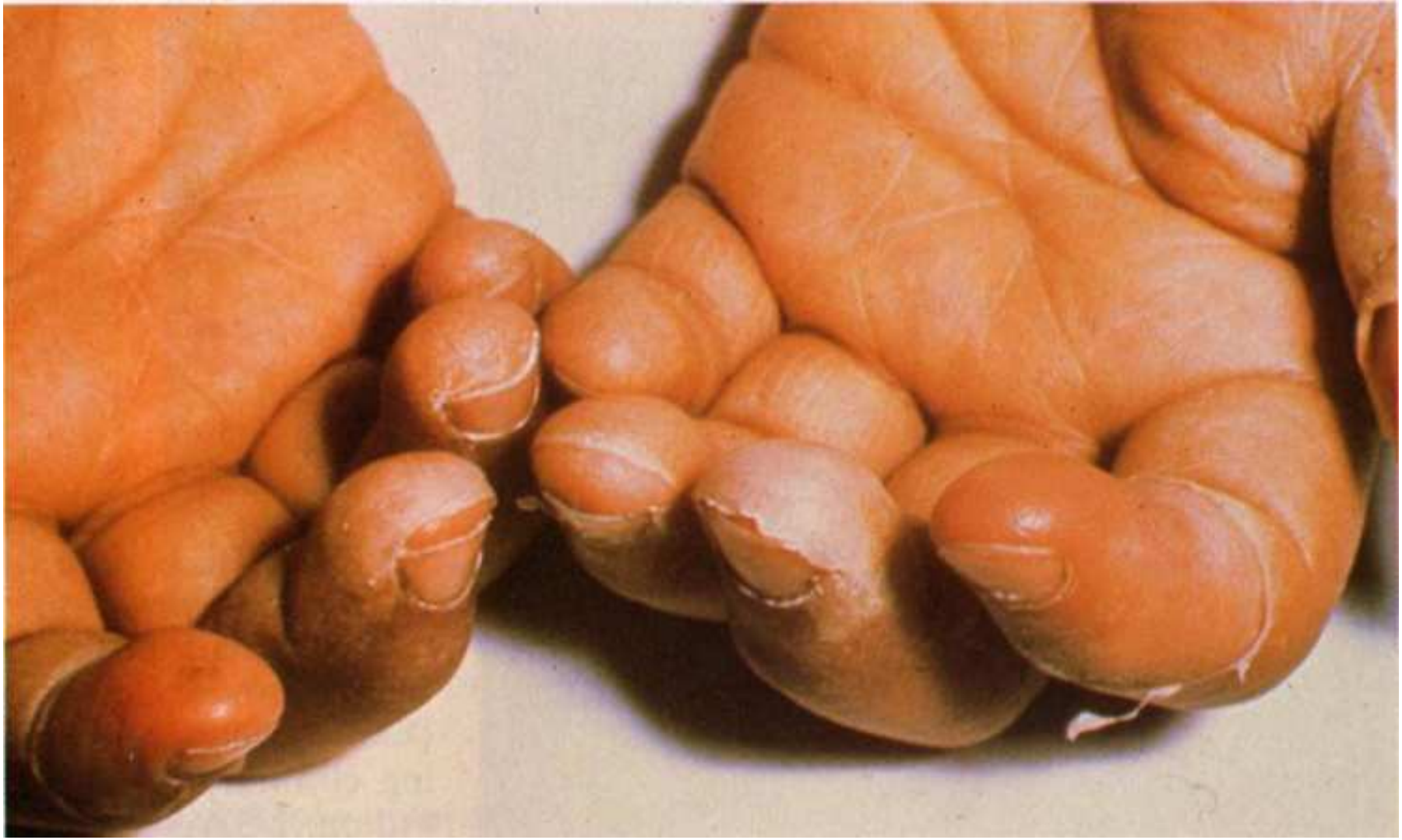

川崎病
Kawasaki
Disease
川崎富作

Kawasaki Disease

- AKA Mucocutaneous Lymph Node Syndrome
- Acute febrile illness of infancy and early childhood
- Antiendothelial antibodies
- Usually self-limited, but may cause vasculitis of coronary artery and lead to myocardial infarct
- Fever, conjunctivitis, enlarged cervical lymph nodes
- Edema, erythema, desquamation of hands and feet
- Treated with IV immunoglobulins and aspirin – reduces cardiovascular sequelae from 20% to 4%



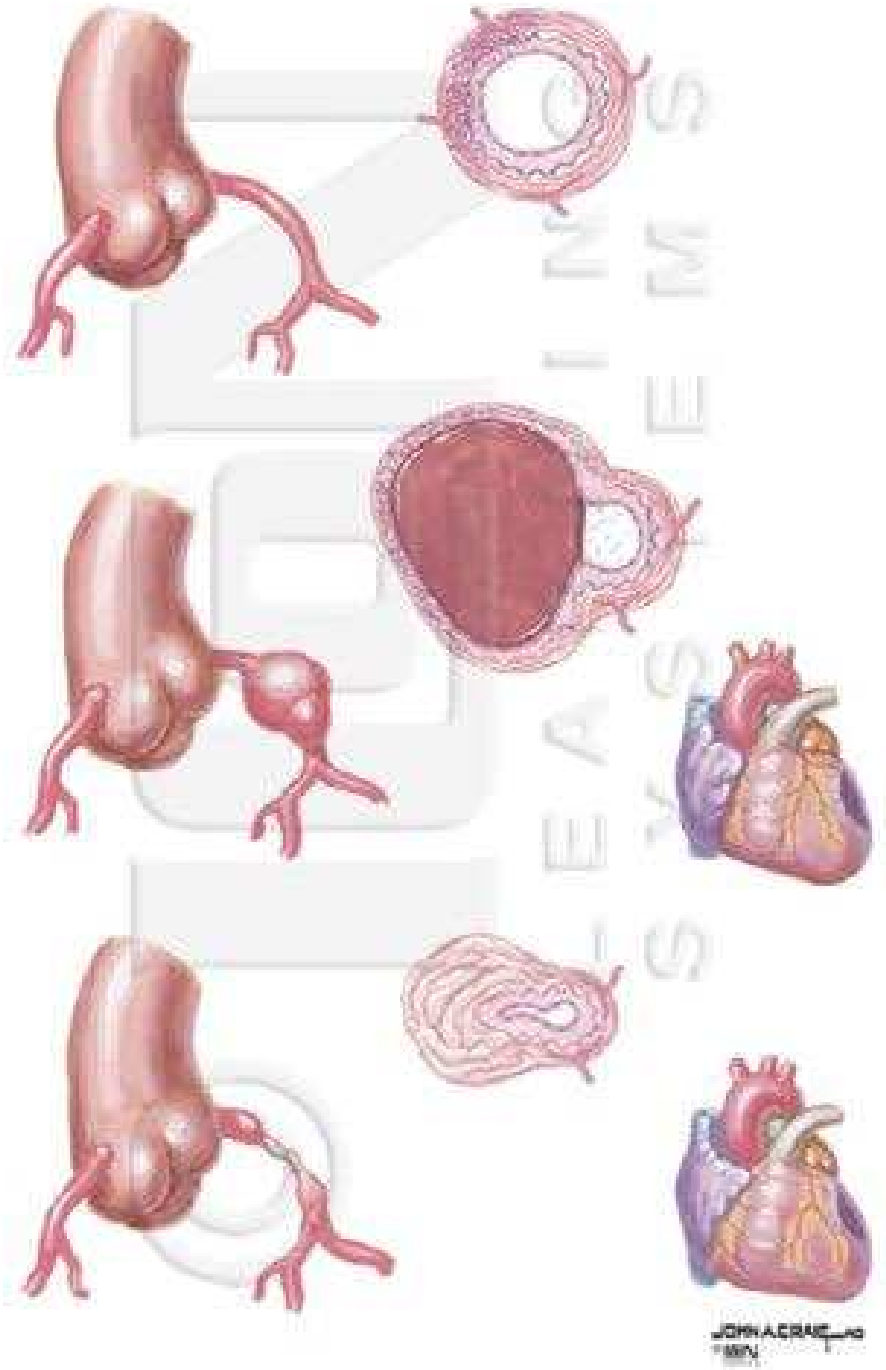
Child with Kawasaki Disease: Conjuntivitis and inflammation of mucous membranes of the mouth.



Desquamation in Kawasaki Disease

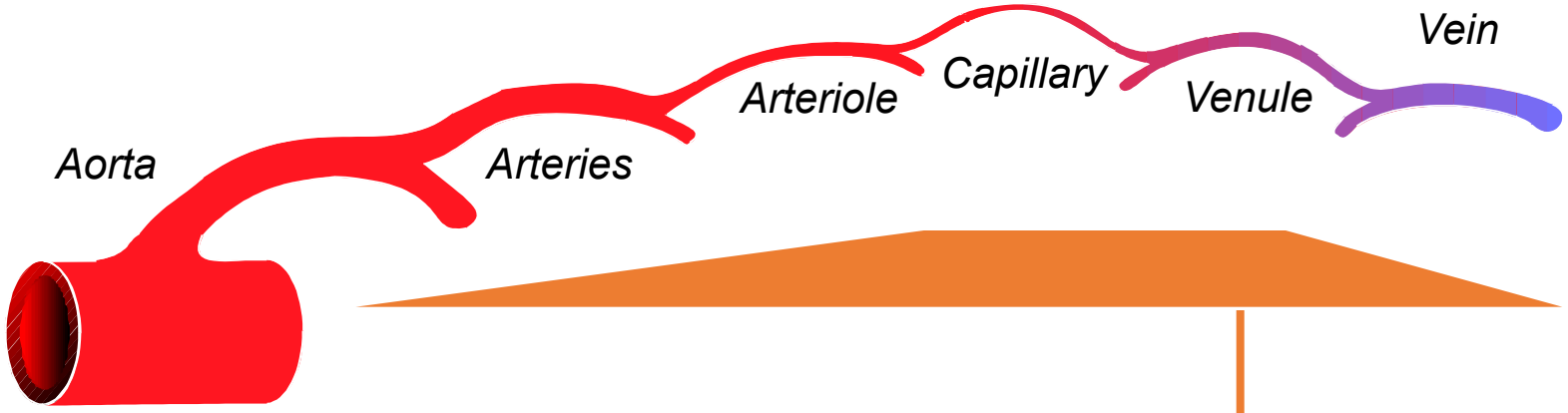
Coronary Artery Manifestations

- Coronary artery aneurysms are the major complication of Kawasaki disease
  - May result in myocardial ischemia / infarction and sudden death
  - Treatment to reduce these complications includes IVIG (intravascular immunoglobulin) and aspirin



Types of Small Vessel Vasculitis

- ANCA-associated
  - Granulomatosis with polyangiitis (GPA)
  - Microscopic polyangiitis (MPA)
  - Eosinophilic granulomatosis with polyangiitis (Churg-Strauss)
- Non-ANCA-associated
  - Primary:
    - IgA vasculitis (Henoch-Schonlein)
    - Cryoglobulinemic vasculitis
    - Idiopathic cutaneous vasculitis
  - Secondary:
    - Drug reactions
    - Connective tissue diseases (SLE, RA, Sjogren syndrome)
    - Infection (hepatitis C, HIV)
    - Neoplasia (hematologic or solid)

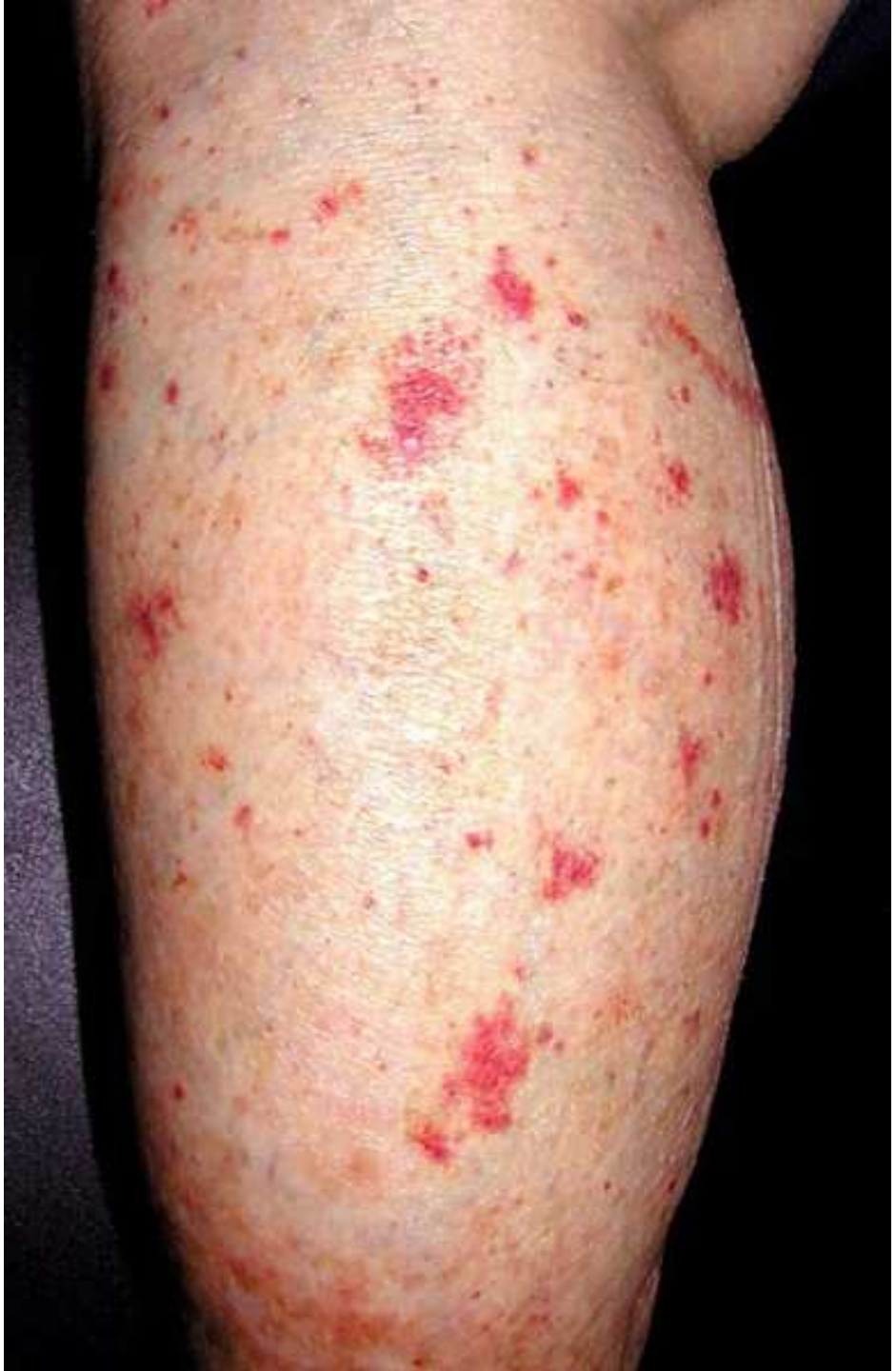


Small Vessel Vasculitis

- immune complexes in vessels
  - other sources for immune complexes → Other Im Cx Vasculitis
  - cryoglobulins in blood and vessels → Cryoglobulinemic Vasculitis
  - IgA-dominant vessel wall deposits → H-S Purpura
  - SLE or rheumatoid arthritis → Lupus/Rheumatoid Vasculitis
- paucity of vascular Ig (often ANCA positive)
  - vasculitis with no asthma or granulomas → MPA
  - Granulomas → GPA
  - eosinophilia, asthma and granulomas → EGPA

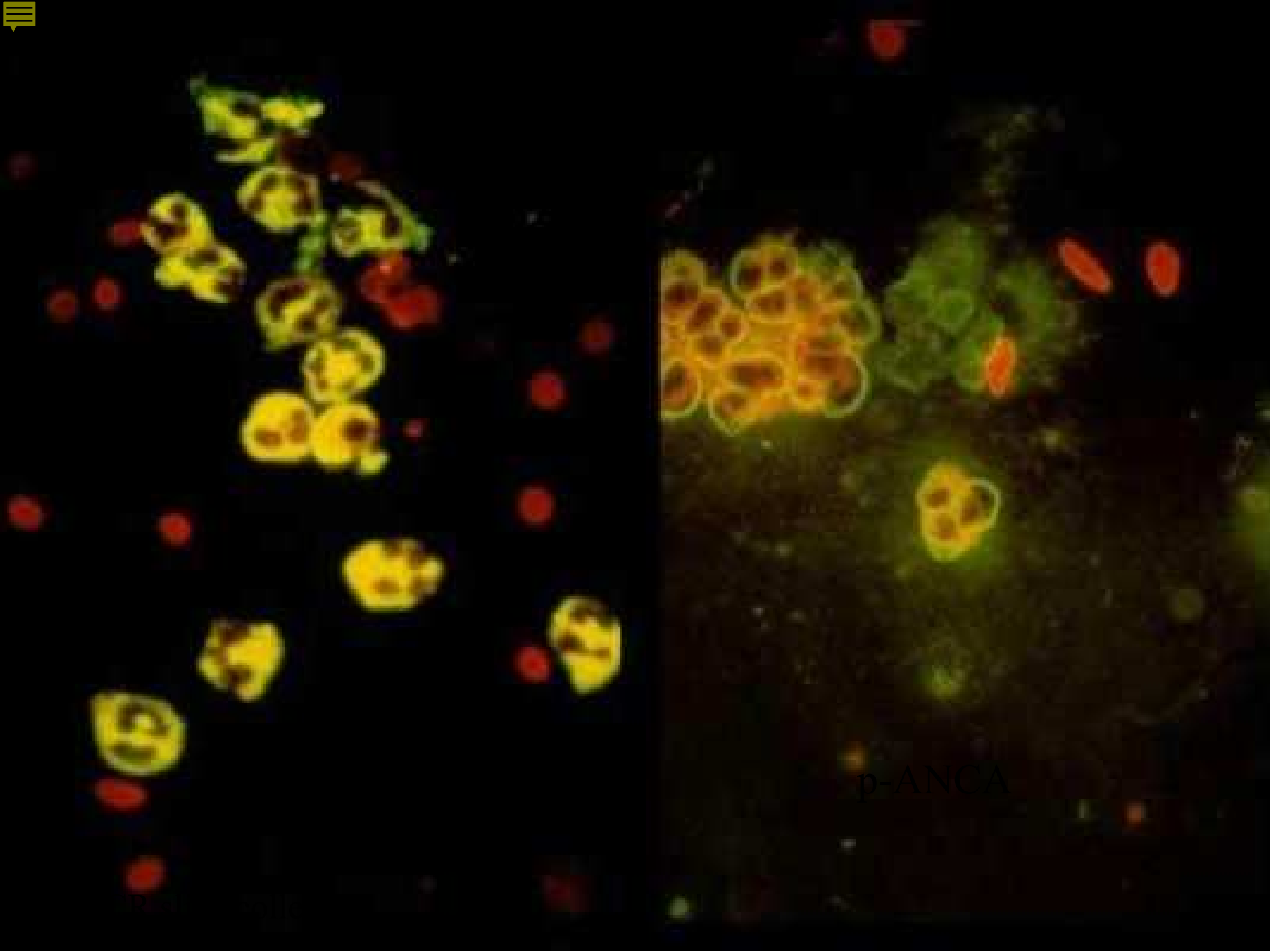

Small vessel vasculitis

Palpable purpura in cryoglobulinemic vasculitis



ANCA

- Anti-neutrophil cytoplasmic antibodies
  - reported by distribution of antibody staining in cytoplasm
    - P-ANCA = perinuclear, associated with myeloperoxidase antigen (MPO)
    - C-ANCA = cytoplasmic, 90% due to proteinase 3 (PR3)
- Must be confirmed with antigen-specific testing for PR3 and MPO to be specific

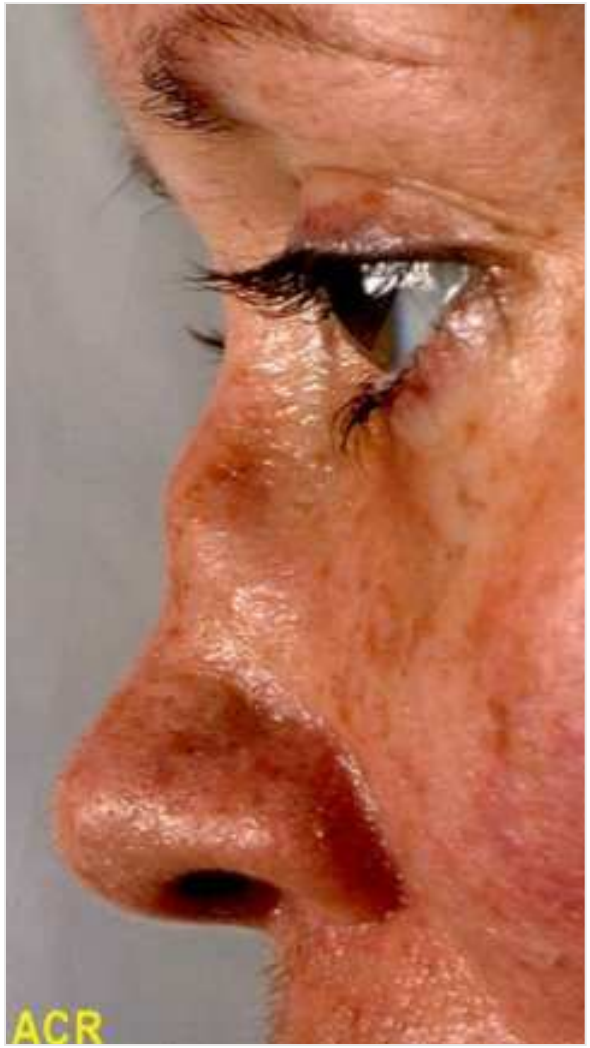

p-ANCA

Granulomatosis with Polyangiitis

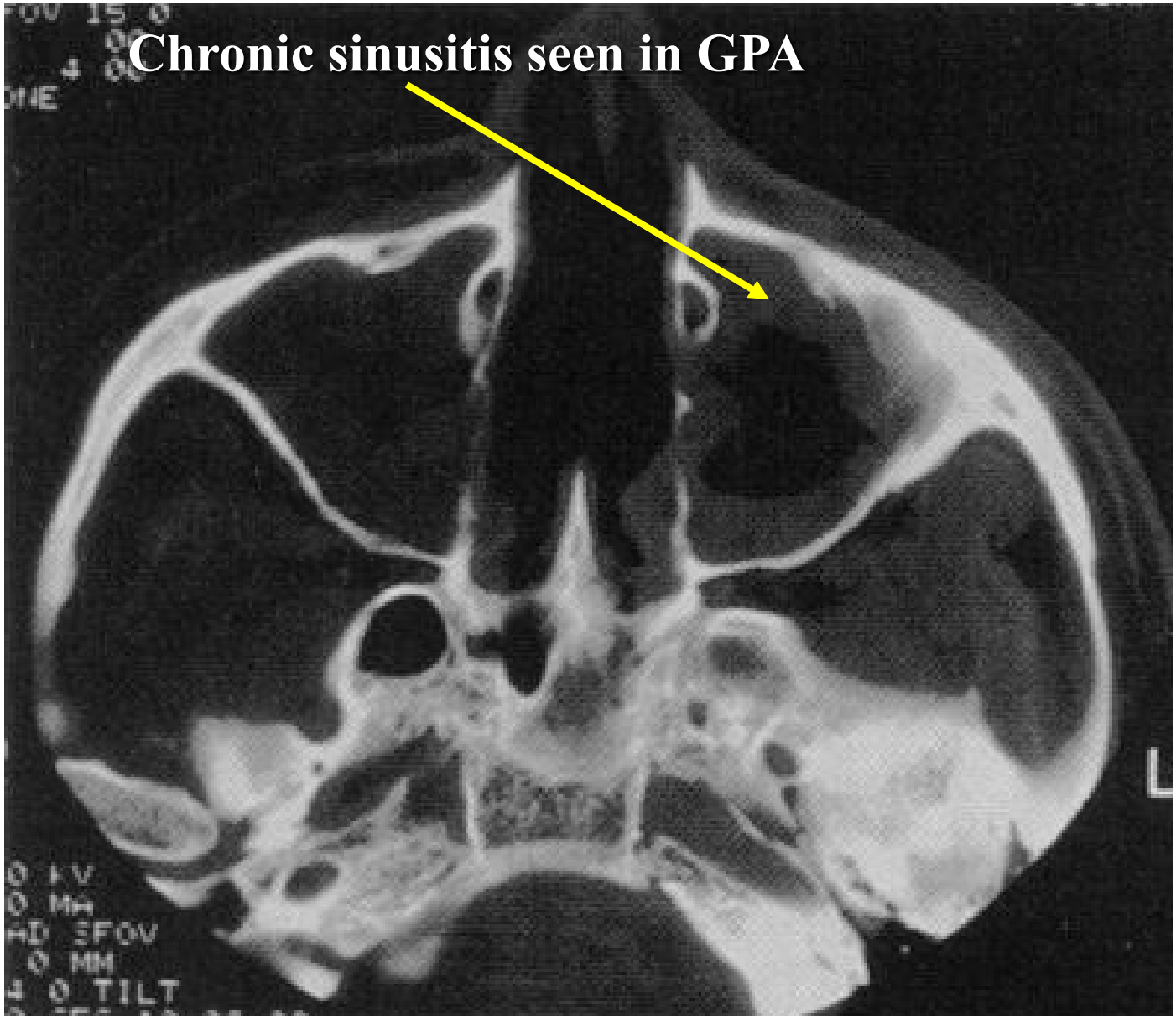
- 30-50 year olds; Male : Female 1:1
- More common in Caucasian groups
- c-ANCA and PR3 positive in 85%
  - p-ANCA and MPO positive in 10-15%

GPA Clinical Presentation

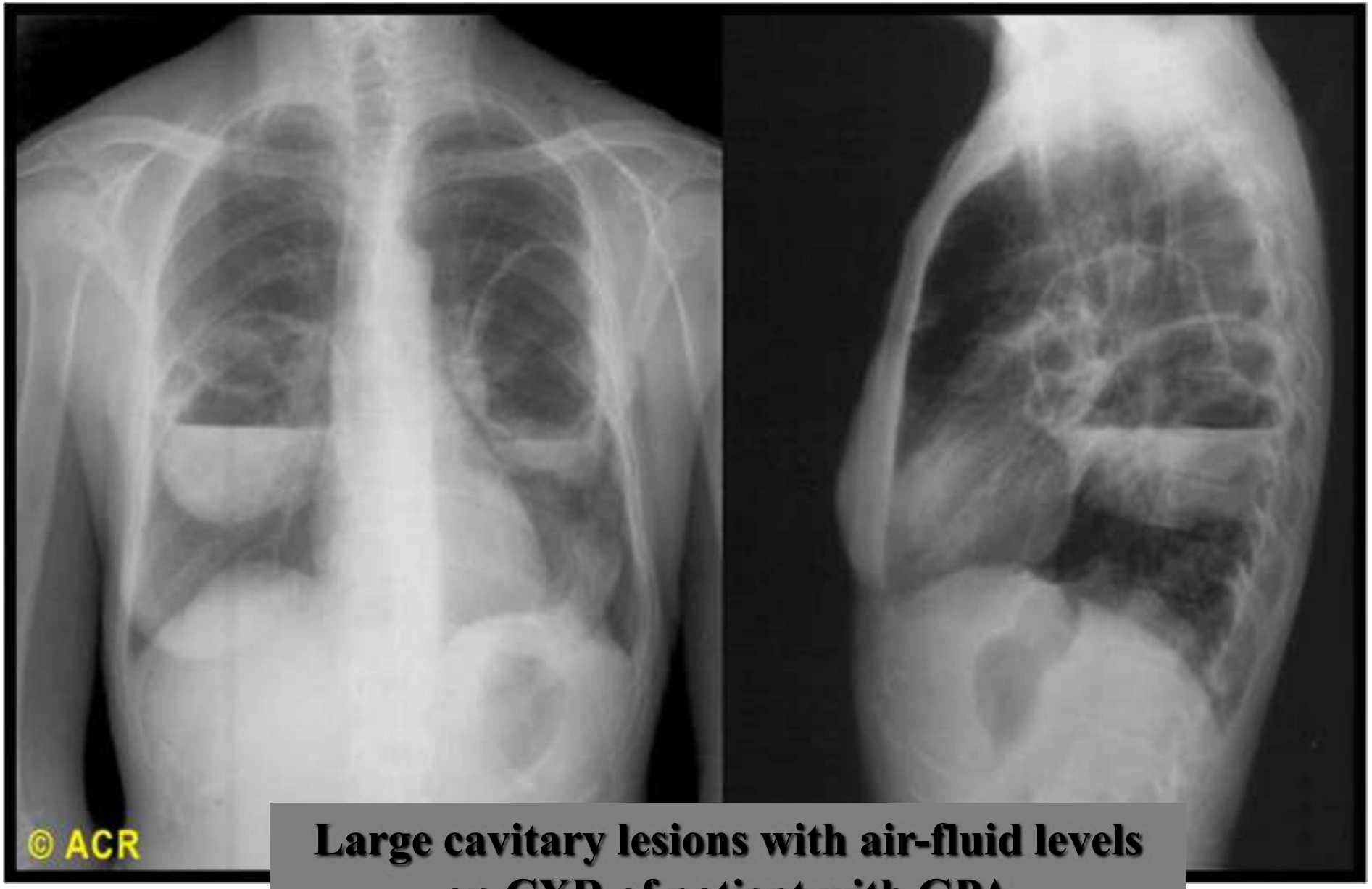
- Involves
  - upper respiratory tract
    - tracheal stenosis
    - mass lesions, locally invasive
      - sinusitis
      - otitis media
      - proptosis
      - saddle nose deformity
  - lower respiratory tract
    - nodular lesions
    - progression to cavitary lesions
  - kidneys
    - pauci-immune crescentic glomerulonephritis
  - potentially skin and ocular manifestations



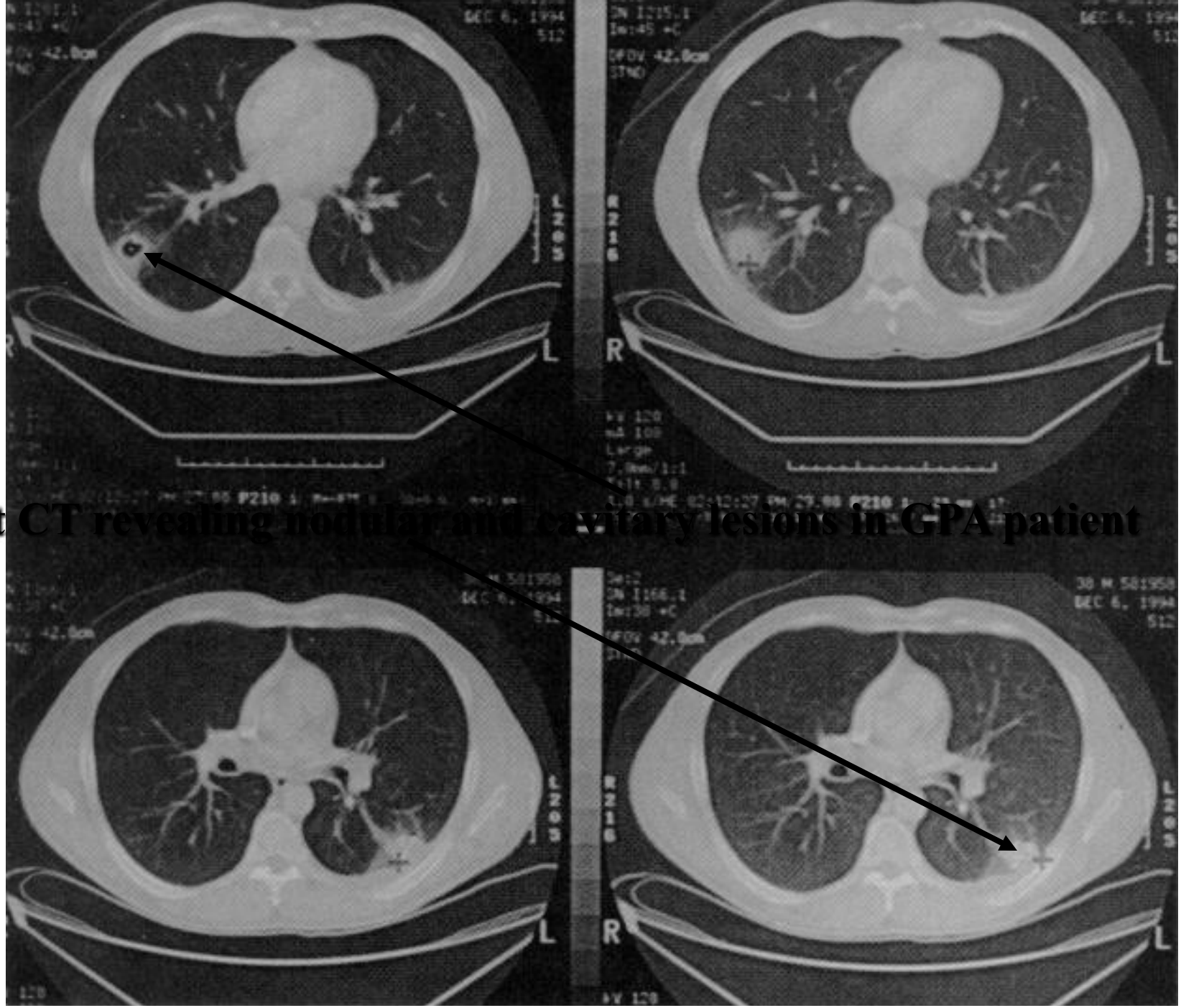
Saddle-nose deformity in a patient with GPA



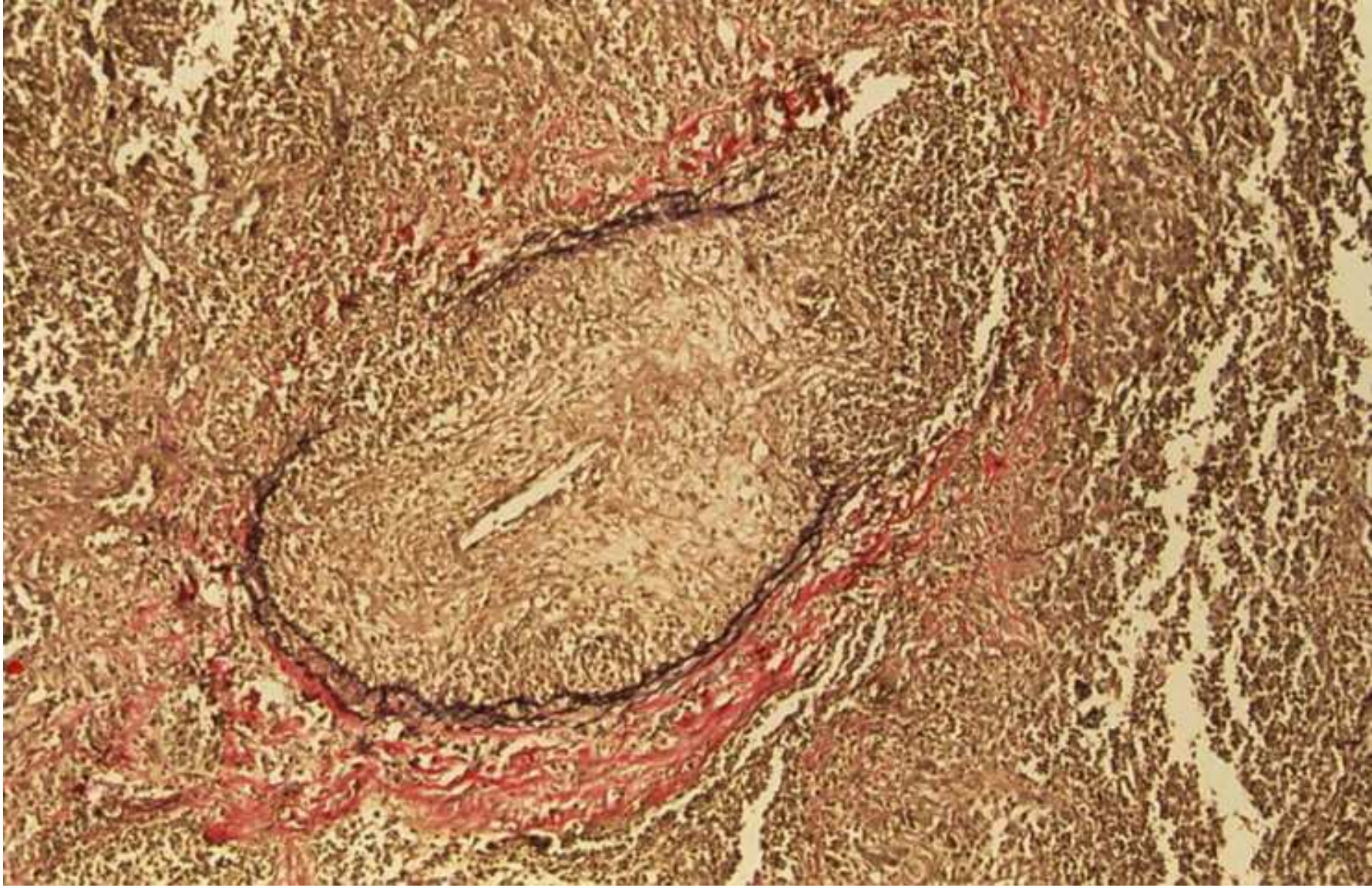
Ruddy: Kelley's Textbook of Rheumatology, 6th ed., 2001



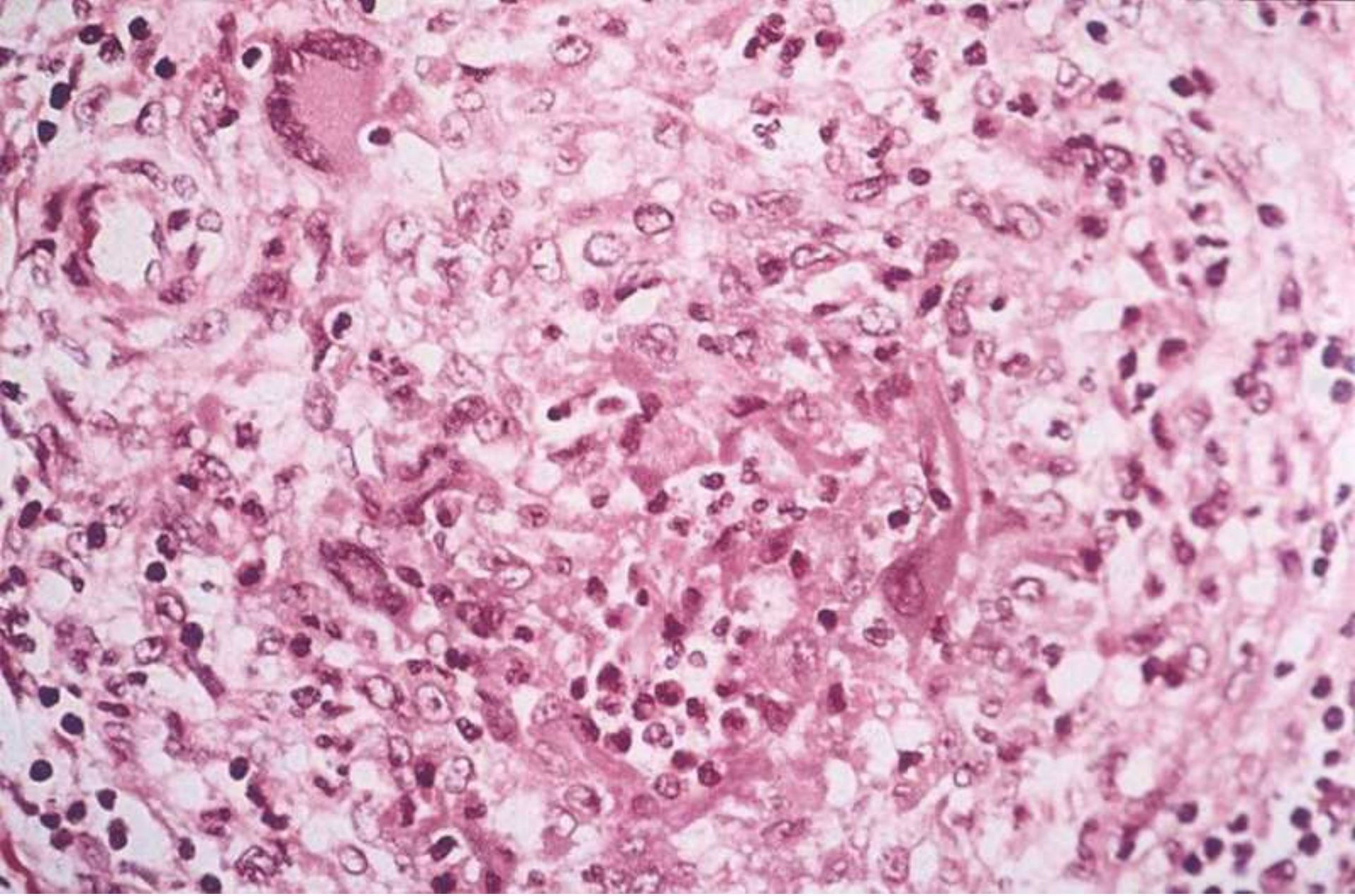
Large cavitary lesions with air-fluid levels on CXR of patient with GPA



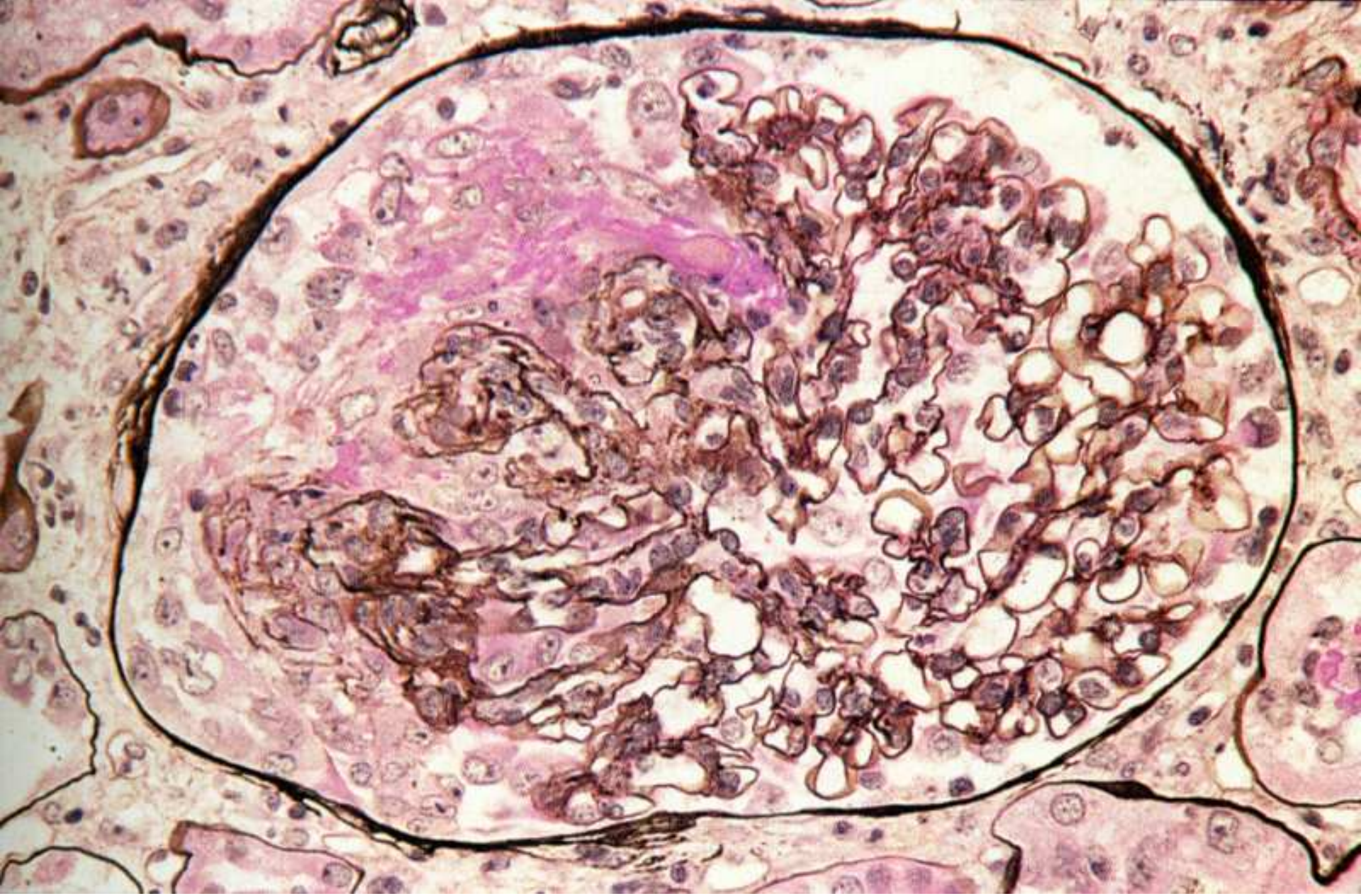
Chest CT revealing nodular and cavitary lesions in GPA patient



Granulomatous vasculitis with destruction of the elastica in GPA, elastic stain



Granulomatous vasculitis in GPA



Necrotizing glomerulitis in GPA

GPA Treatment

- High (~80%) mortality if untreated
- Initial immunosuppressive therapy for induction of remission
  - Prednisone 1mg/kg/day or higher initially
  - ADD: Cyclophosphamide (usually oral daily)

    or Rituximab IV
  - Methotrexate for milder disease
- Maintenance immunosuppression after achieving remission
- Pneumocystis carinii (jiroveci) pneumonia prophylaxis

Microscopic Polyangiitis (MPA)

- Originally described as variant of PAN
- Chapel Hill consensus definition
  - "necrotizing vasculitis affecting small vessels (i.e., capillaries, venules, or arterioles) with few or no immune deposits"
- 40-60 year olds; Male : Female 1.8 : 1
- Frequently associated with necrotizing glomerulonephritis & pulmonary capillaritis
  - thus one of the "pulmonary renal" syndromes

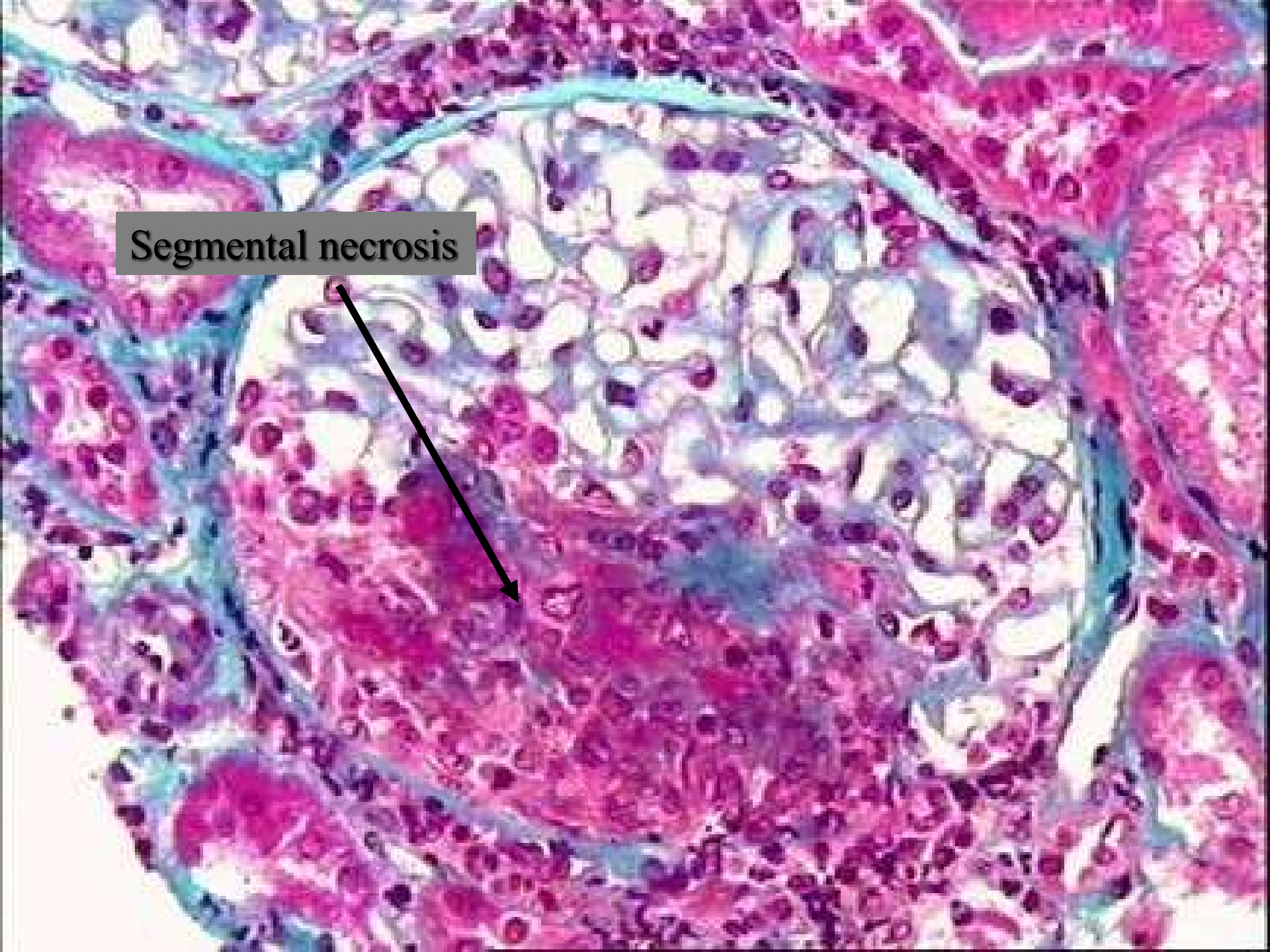
MPA Presentation

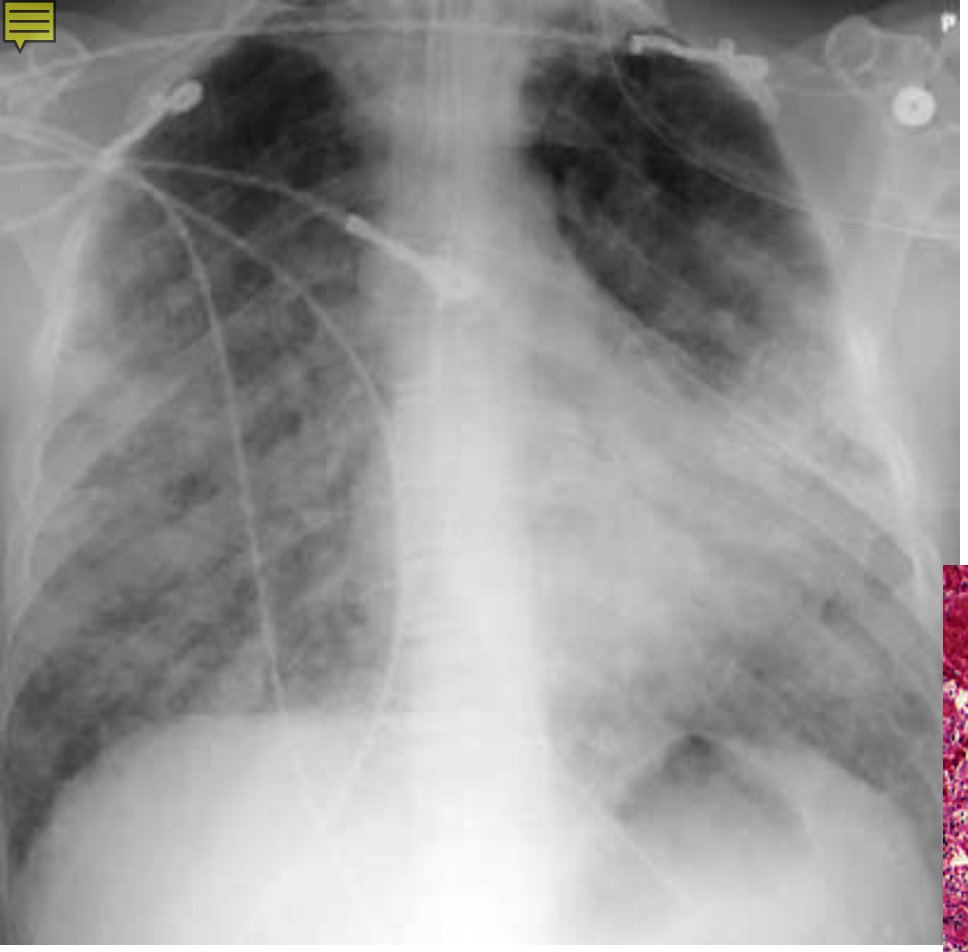
- Hyperacute
  - rapidly progressive glomerulonephritis
  - pulmonary hemorrhage
- Insidious
  - intermittent constitutional symptoms
  - purpura
  - mild renal disease
  - periodic hemoptysis

MPA Clinical Features

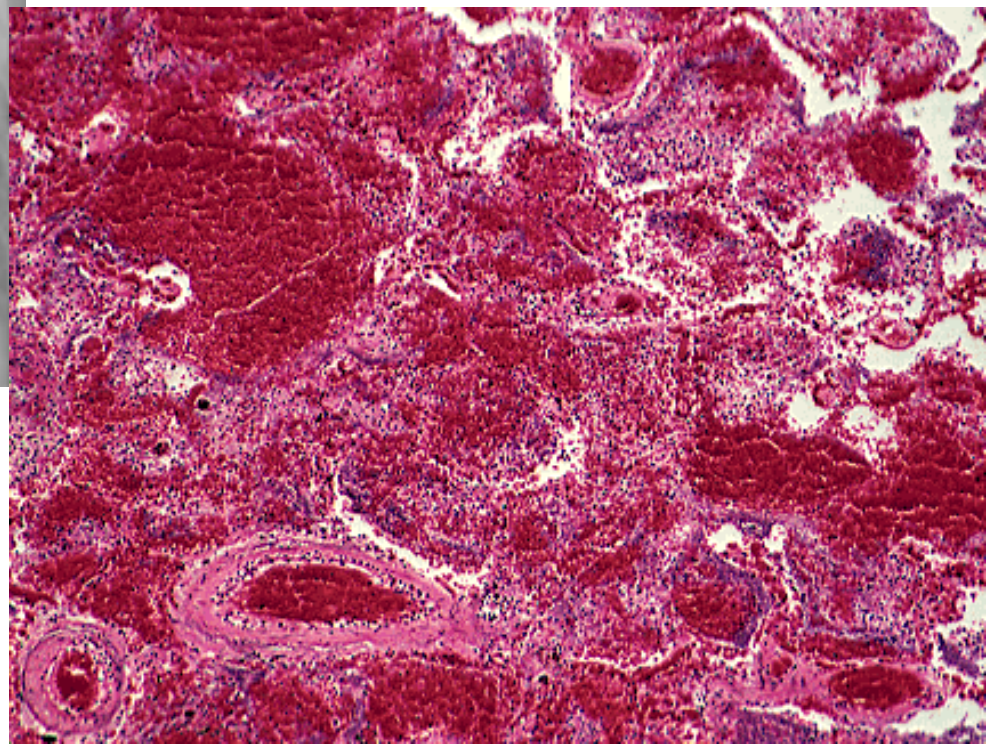
Clinical Feature	Percentage
Constitutional symptoms	76-79
Fever	50-72
Renal disease	100
Arthralgias	28-65
Purpura	40-44
Pulmonary disease (hemorrhage, infiltrates, effusion)	50
Neurologic disease (central, peripheral)	28
Ear, nose, throat	30

- P-ANCA positive with +MPO confirmation in 60 to 85 %
- Occasional C-ANCA PR3+


Segmental necrosis



MPA pulmonary infiltrates



Alveolar hemorrhage from pulmonary capillaritis

MPA Treatment

- Prednisone 1mg/kg/day or higher initially
- ADD: Cyclophosphamide (usually oral daily)

  or Rituximab IV
- 5 yr survival on this regimen is 80%
- Alternatives
  - Azathioprine or methotrexate

Eosinophilic Granulomatosis with Polyangiitis (EGPA) Clinical Presentation

- AKA Churg-Strauss syndrome
- Insidious prodrome
  - allergic prodrome (3-7 years)
    - asthma (adult onset)
    - allergic rhinitis with polyposis
    - eosinophilic pneumonia
    - eosinophilic gastroenteritis
  - prodrome may abate with vasculitis onset
- ANCA+ (MPO, P-ANCA) in up to 70%

EGPA Clinical Presentation

- Vasculitis phase
  - Pulmonary disease
    - during vasculitis or prodromal phase
    - fleeting but lobar, interstitial, and nodular patterns on x-ray
    - occasionally pulmonary hemorrhage without renal disease (to distinguish from GPA and MPA)
  - Neurologic disease
    - peripheral neuropathy (mononeuritis multiplex, symmetric polyneuropathy)

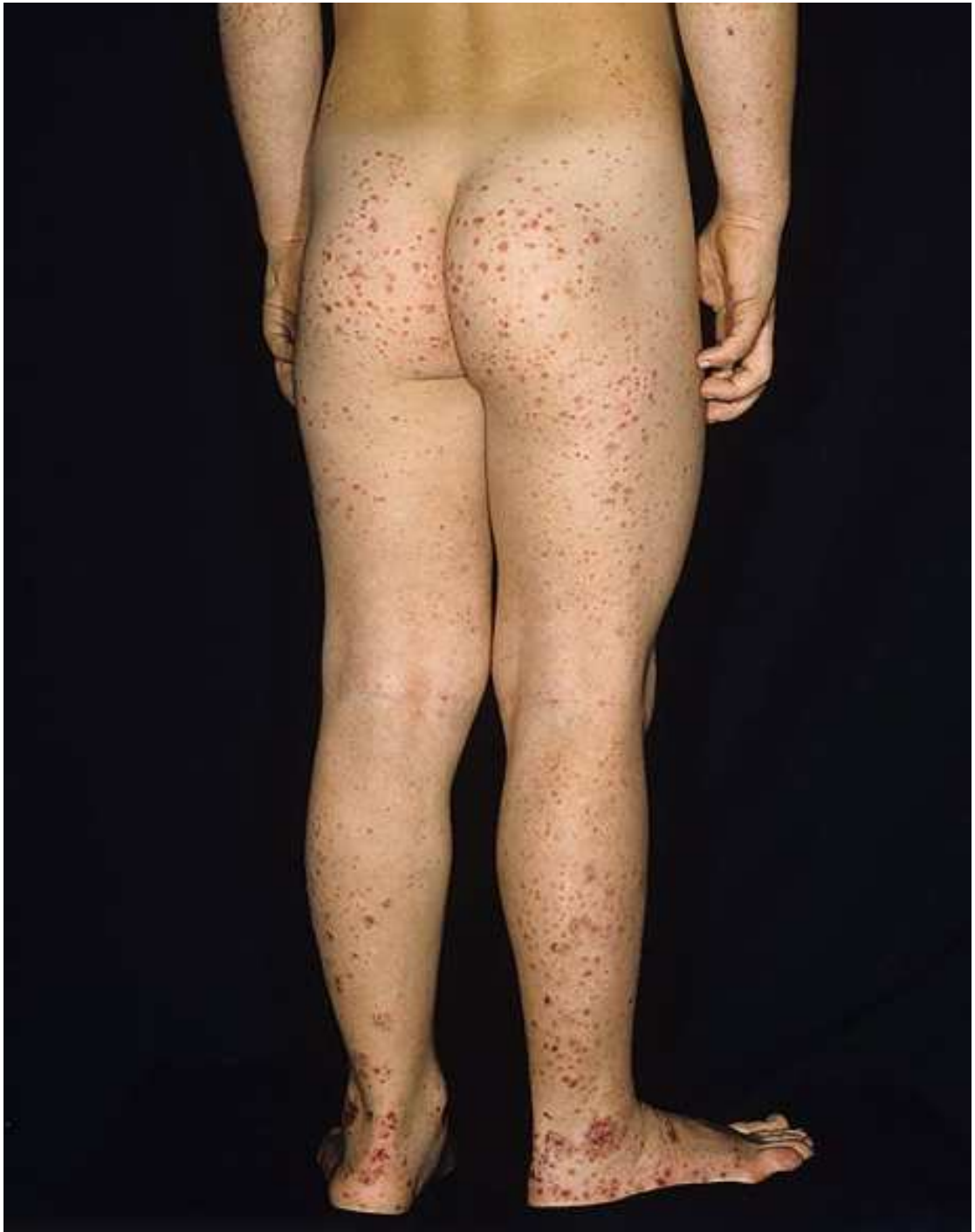
EGPA Prognosis

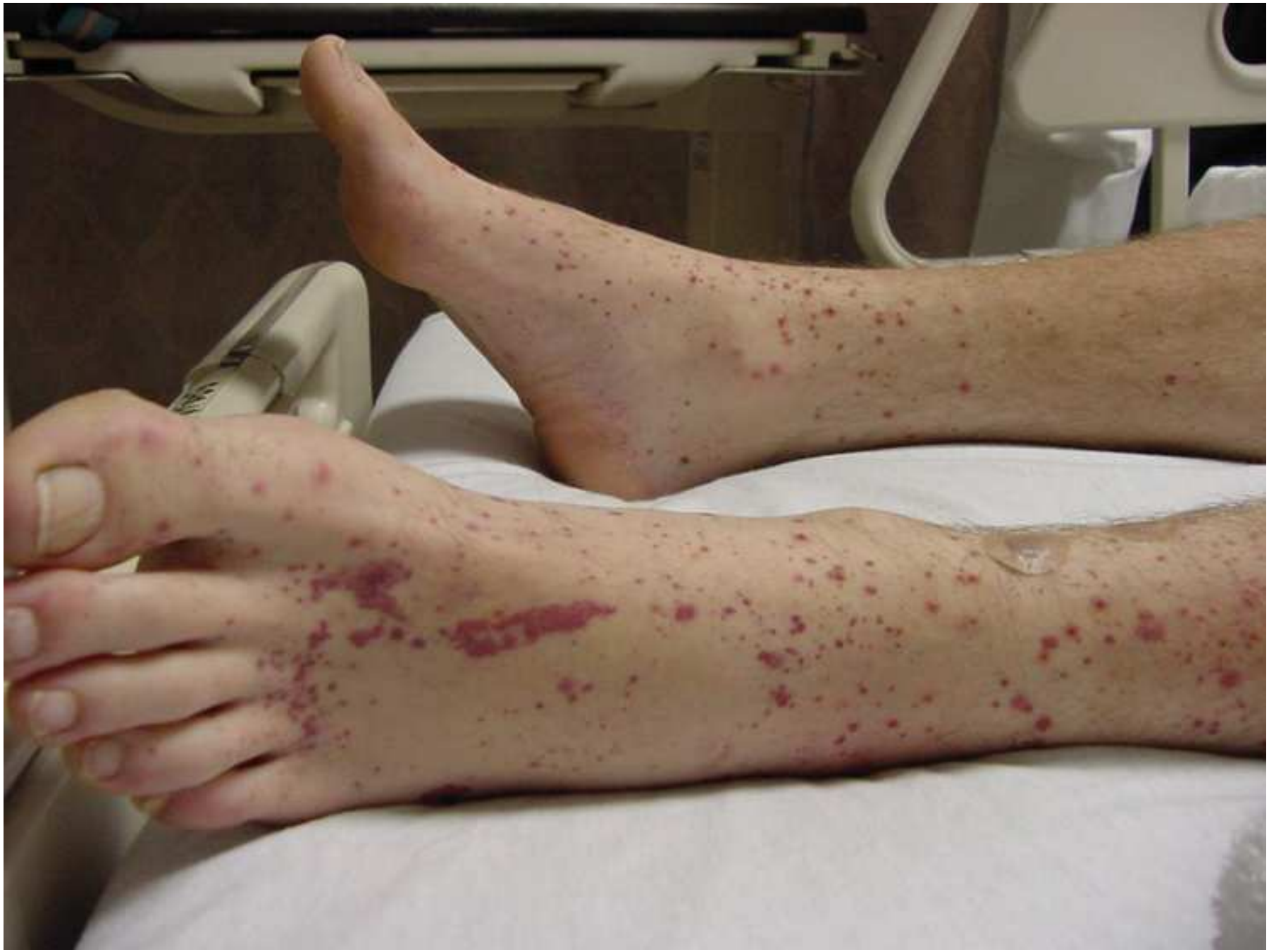
- 5-year survival 80%
- Poor outcome with
  - azotemia (creatinine concentration >1.58 mg/dl)
  - proteinuria (>1 g/d)
  - gastrointestinal tract involvement
  - cardiomyopathy
  - CNS involvement

Small Vessel Vasculitis
Immune-Complex-Mediated

- IgA Vasculitis
  - AKA Henoch-Schönlein Purpura (HSP)
  - palpable purpura in children; 50% < 5 years old
  - IgA deposition on histopathology
  - leukocytoclastic vasculitis
  - associated arthritis, abdominal pain

- Hypersensitivity vasculitis (Small-Vessel Cutaneous Vasculitis)
  - similar picture in association with serum sickness and drug reactions
  - distinguished by IgG deposition





IgA Vasculitis Outcomes

- Better prognosis in children
  - spontaneous resolution is the norm
- Older children and adults
  - 10 to 30 percent progress to chronic renal disease and end-stage renal failure

Ruddy: Kelley's Textbook of Rheumatology, 6th ed., 2001

IgA Vasculitis Therapy

- No controlled studies
- Success with early corticosteroids if renal involvement
- If nephrotic range proteinuria, 15mg/kg/day x 3 days can be used
- Immunosuppressive agents if renal disease continues or recurs with steroid taper
  - usually after renal biopsy obtained

Hypersensitivity Vasculitis
Clinical Presentation

- Often abrupt onset of rash
  - Arthus reaction or type III hypersensitivity
  - In association with exposure to:
    - drug
    - infectious agent
- Papular eruption (inflammatory)
  - more in dependent areas
  - discrete or coalescent lesions
- Associated with fever, arthralgia, malaise

Hypersensitivity Vasculitis Treatment

- Remove offending agent
  - drug
  - infection
- If major organ involvement, 1/2-1 mg/kg/day prednisone tapered as disease resolution allows
- No need for cytotoxic agents if isolated cutaneous disease

Behçet Disease – Outside the Box

- A systemic vasculitis which can involve vessels of all sizes – on both arterial & venous sides of the circulation
- Seen more frequently in
  - Japan
  - Middle East
  - Mediterranean region
- Usually young adults 20-40 years old
- Males > Females

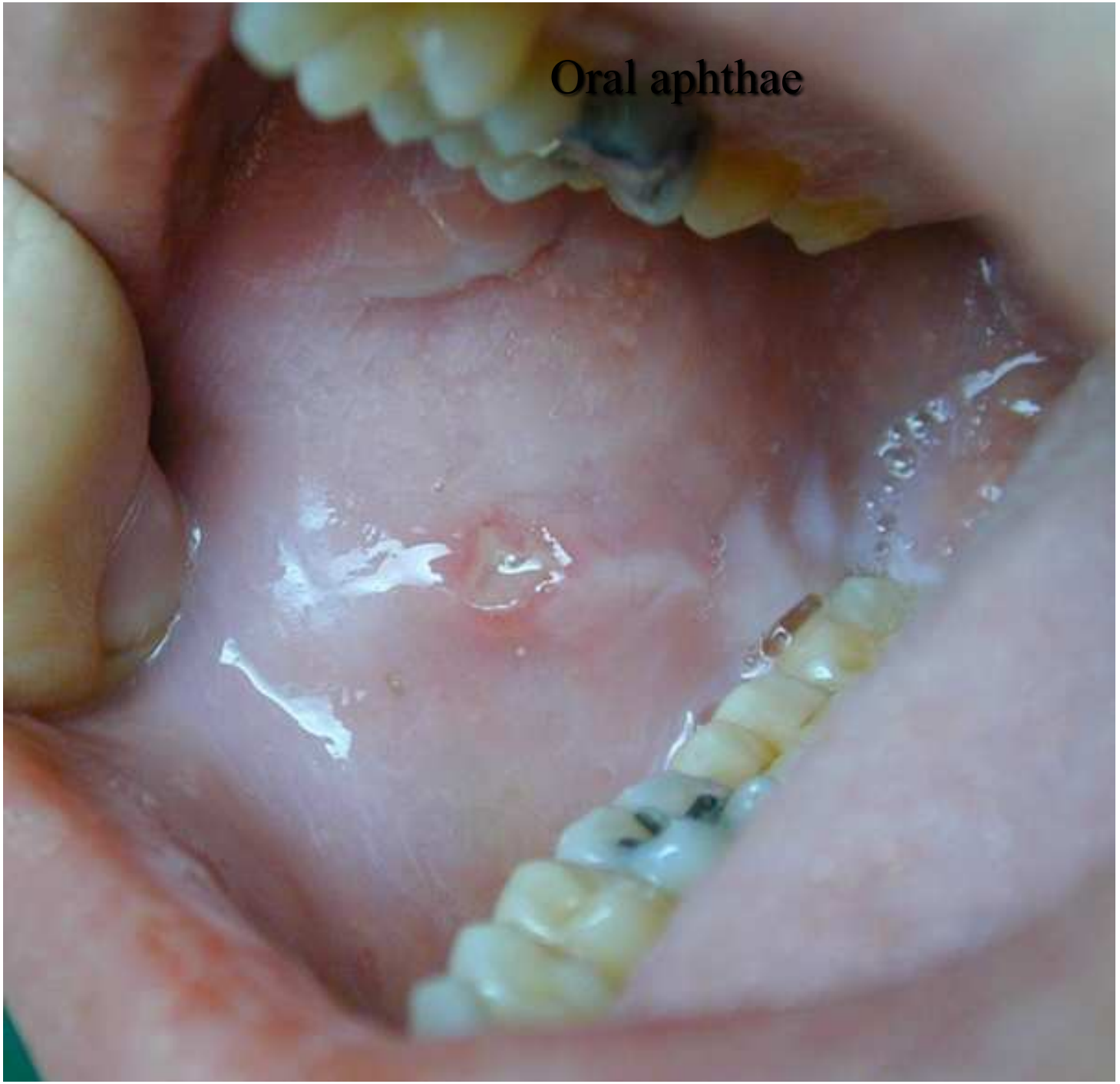
Ruddy: Kelley's Textbook of Rheumatology, 6th ed., 2001

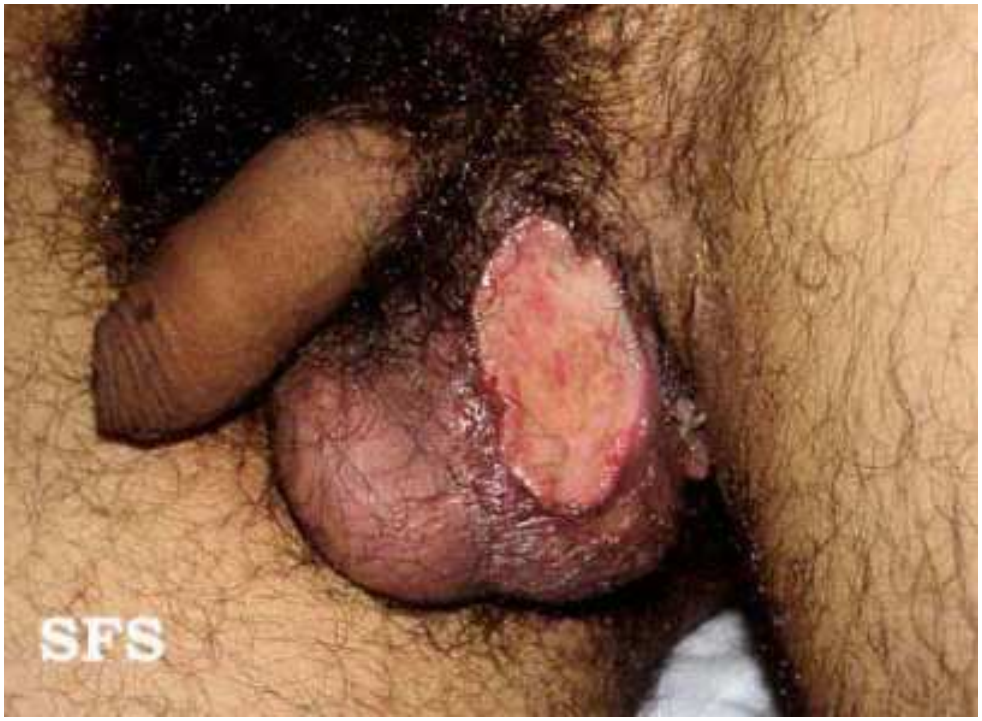
Behçet Disease Clinical Aspects

- Usual presentation with oral and genital ulcers for years
  - oral aphthae, recurrent, painful - sine qua non of disease
  - buccal mucosa, gingiva, lips, and tongue
  - males on scrotum or penis
  - females on labia or vagina

- Followed by ocular and neurologic manifestations

Ruddy: Kelley's Textbook of Rheumatology, 6th ed., 2001


Oral aphthae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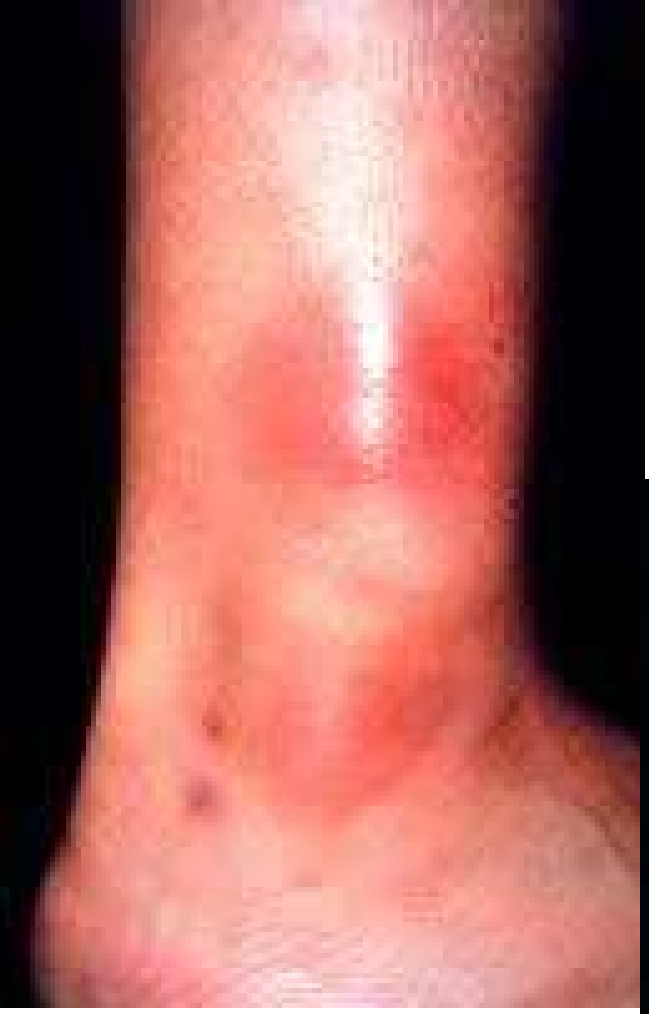
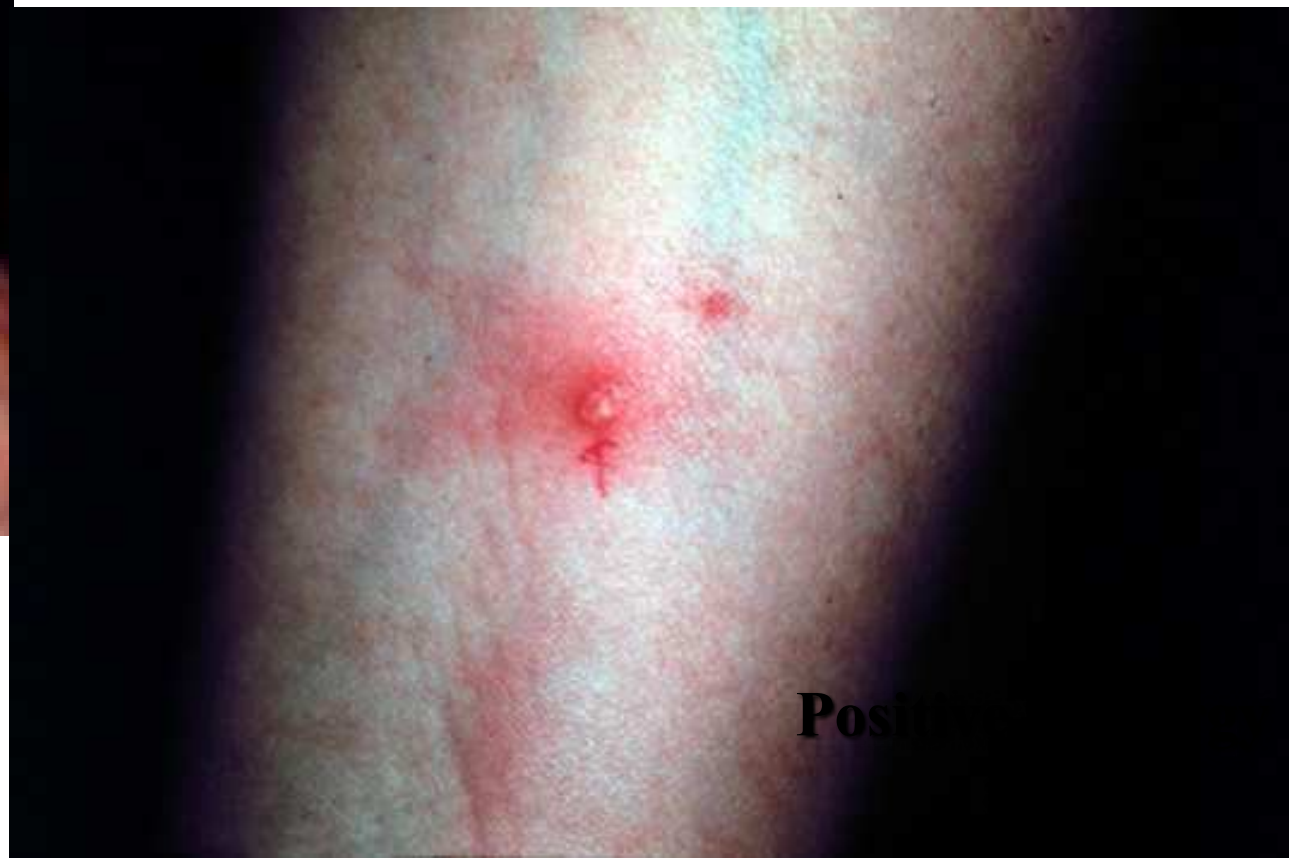
Behçet Disease Clinical Aspects

- Cutaneous lesions
  - erythema nodosum
  - pyoderma gangrenosum
  - acneiform lesions
  - pustular lesions including pathergy lesion (tested by sterile puncture of the skin)
    - despite sterile needle puncture, you still get a pustular lesion

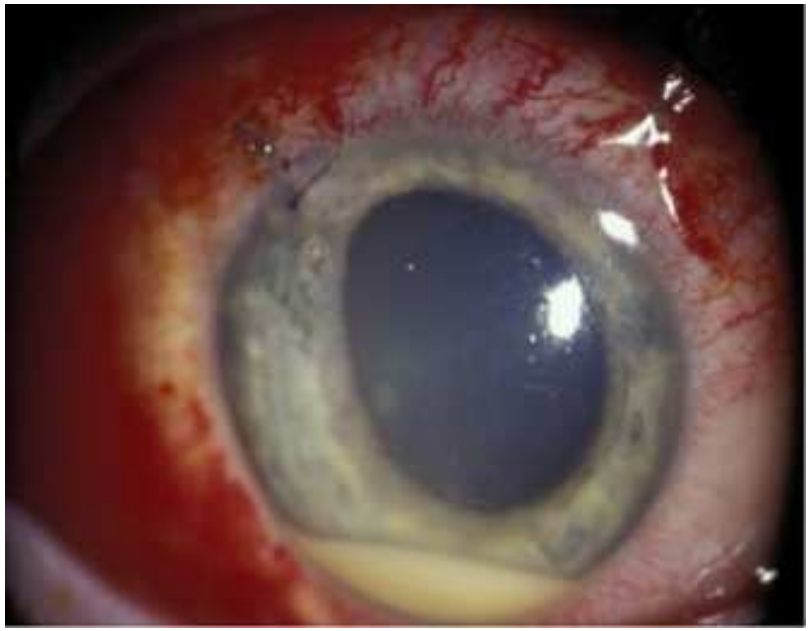
Erythema nodosum





Behçet Disease Clinical Aspects

- Ocular involvement (70-85% of patients)
  - anterior uveitis
  - posterior uveitis (major cause of blindness)
  - conjunctivitis
  - corneal ulceration
  - hypopyon iritis
  - retinal vasculitis



Behçet Disease Clinical Aspects

- Arthritis – knees, wrists, ankles
  - 40-50% of cases
- CNS involvement – presenting with HA, seizures
  - a significant source of morbidity and mortality
- GI involvement
  - can result in GI hemorrhage
  - large aphthae can lead to GI perforation
- Generally follows relapsing/remitting course with more aggressive course in men
- Association with HLA-B51
- Pulmonary and CNS involvement typically have high mortality

Behçet Differential Diagnosis

- Inflammatory bowel disease (IBD)
- Systemic lupus erythematosus
- Reactive arthritis (Reiter's syndrome)
- Herpetic infections (PCR testing recommended)

Ruddy: Kelley's Textbook of Rheumatology, 6th ed., 2001

Behçet Disease Therapy

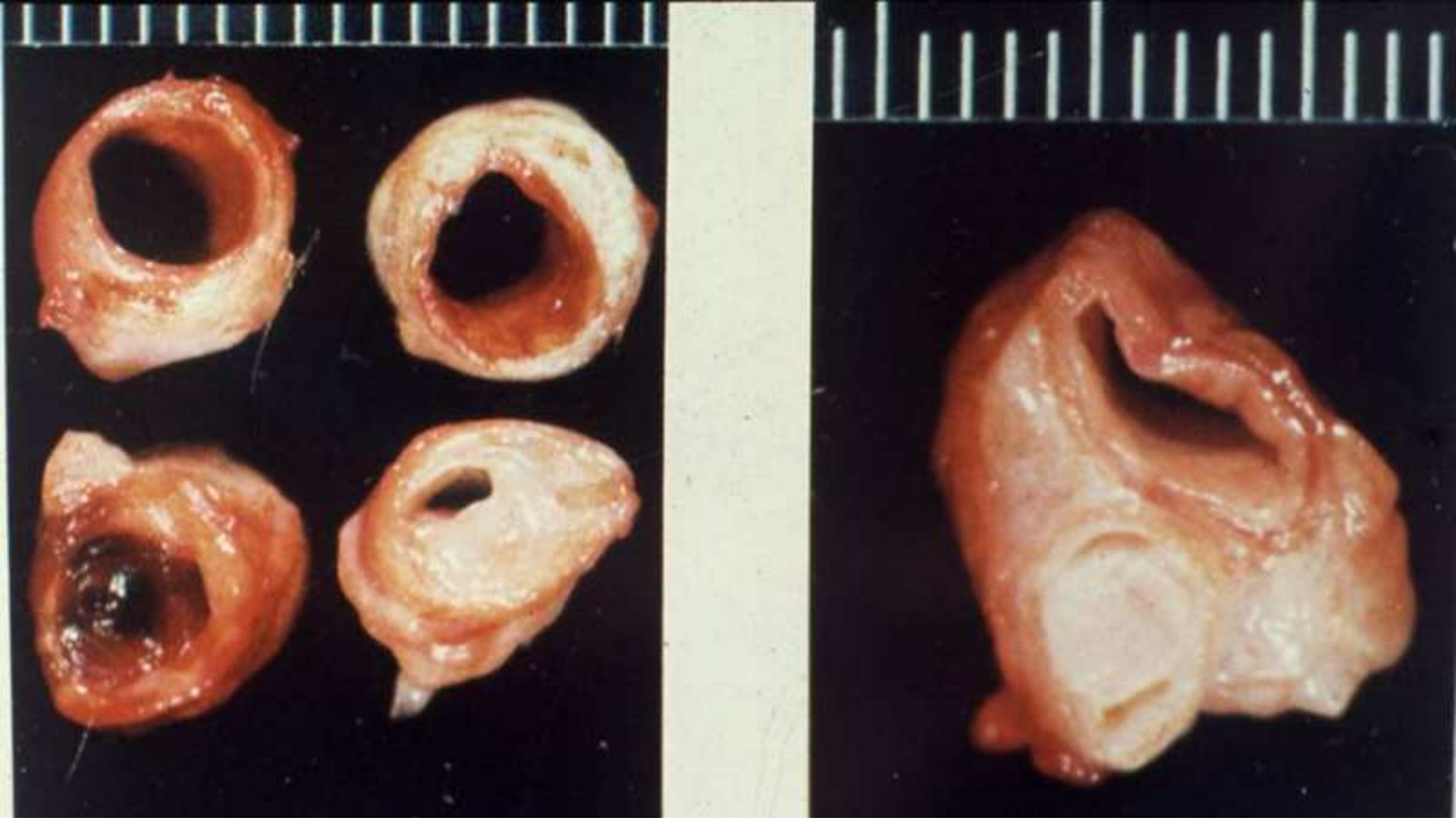
- Mucocutaneous lesions
  - Topical or intralesional corticosteroids
  - Colchicine 0.6-1.8 mg/d
  - Dapsone 50-100 mg/d
  - If severe or refractory
    - Thalidomide 100 mg/d
    - Methotrexate 7.5-20 mg/wk
    - Prednisone

Behçet Disease Therapy

- Systemic disease (severe ocular or CNS disease)
  - Prednisone 1 mg/kg/d
  - Azathioprine
  - Cyclophosphamide
  - TNF-α antagonists

Thromboangiitis Obliterans
Buerger's Disease

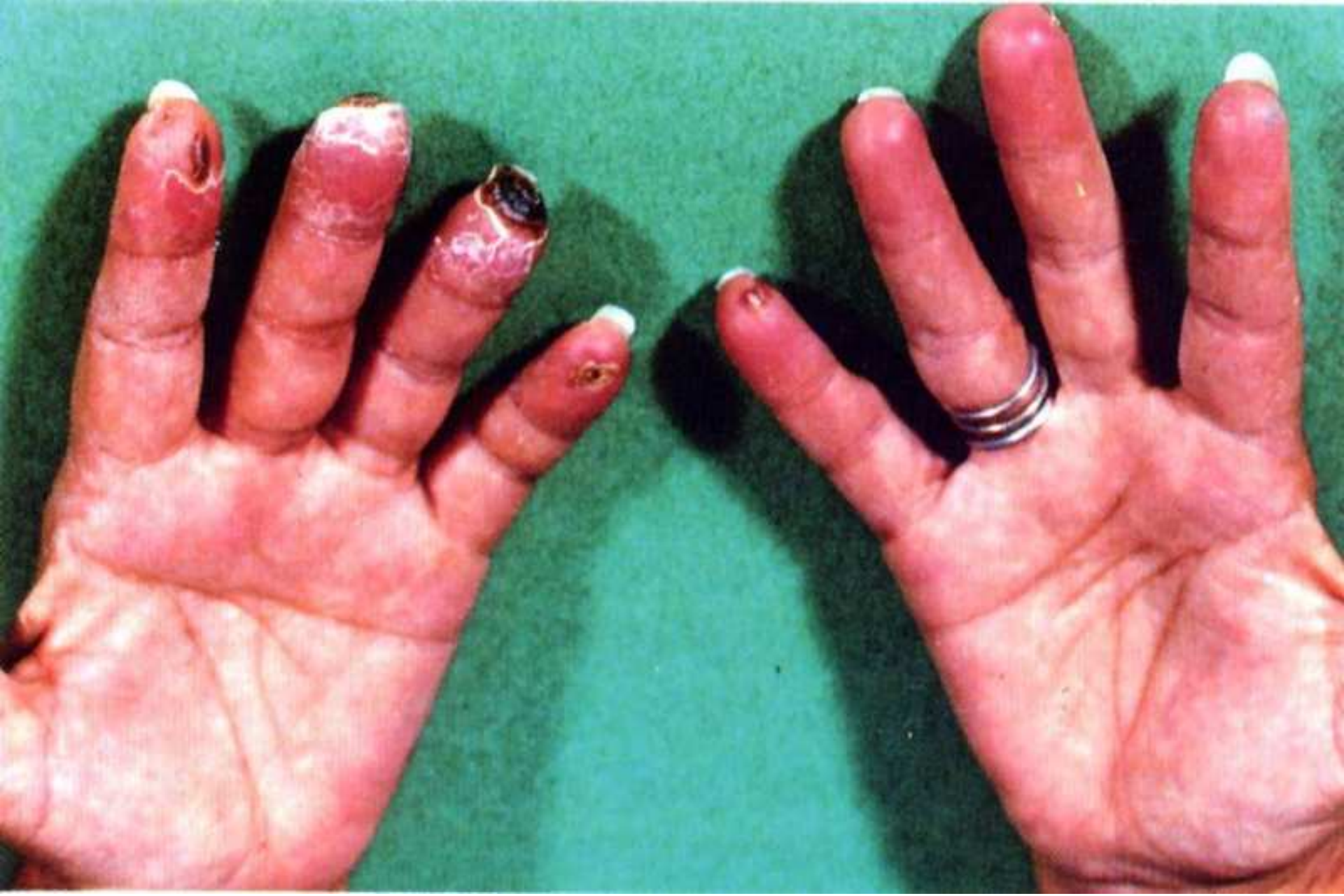
- Male smokers, age 25-50
- Vasculitis and thrombosis of medium sized vessels
- HLA association
- Israel, Japan, India
- Raynaud's, vascular insufficiency, pain, chronic ulcerations.



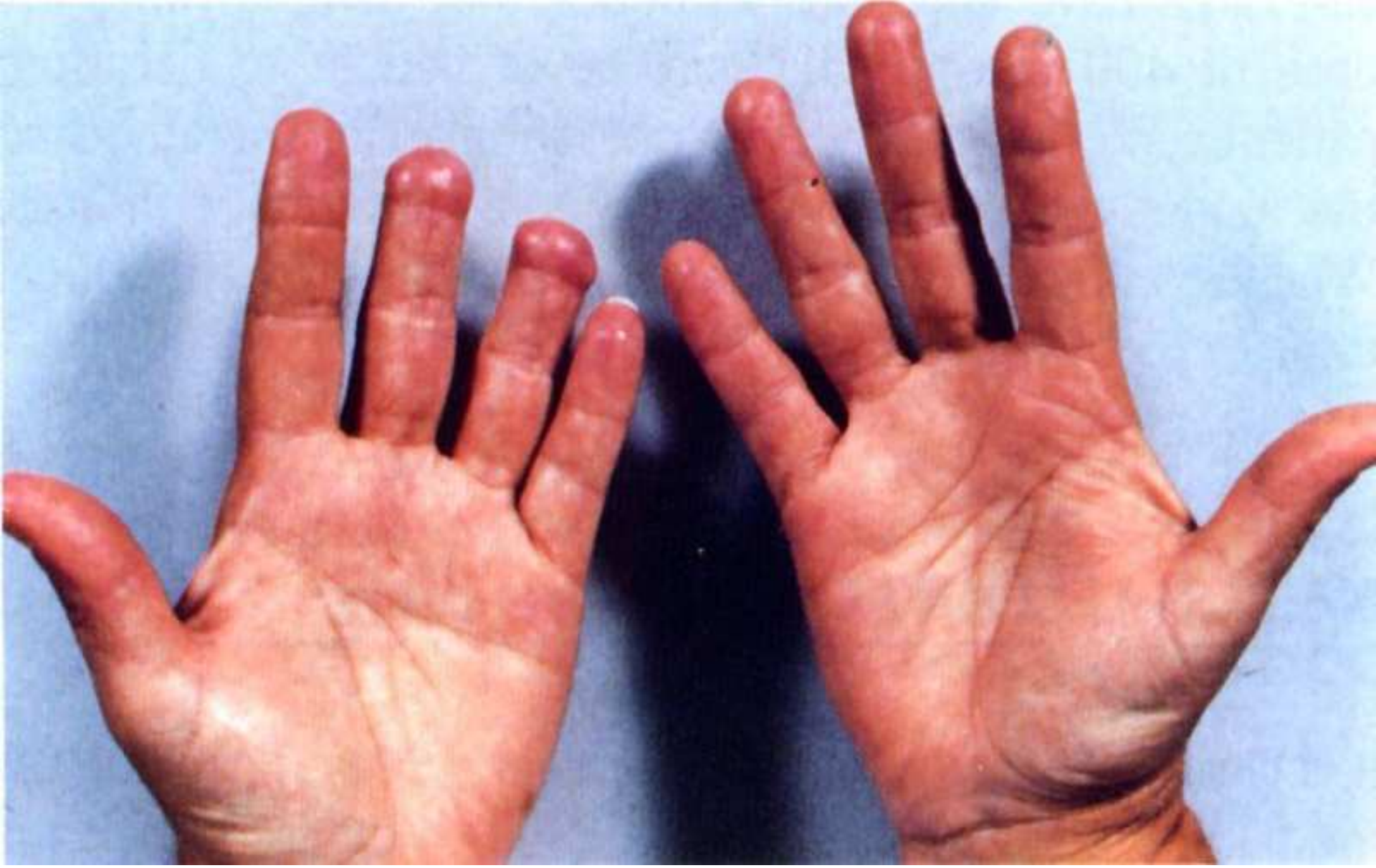
Vascular occlusions in Buerger Disease

(Thromboangiitis Obliterans)

Thromboangiitis Obliterans (Buerger Disease). The lumen is occluded by a thrombus containing abscesses (arrow), and the vessel wall is infiltrated by leukocytes.



Autoamputation in Buerger disease



Healed autoamputation in Buerger disease after patient quit smoking.

A 30 year old Israeli male smoker develops instep claudication as well pain, ulceration, and necrosis of his fingers and toes. The most effective way to give relief from attacks is:

- Aggressive immunosuppression.
- Arterial grafts.
- Endarterectomy.
- Smoking cessations.

A 65 year old male develops fever, malaise, weight loss, and an elevated erythrocyte sedimentation rate. He has pain over the right temporal artery and sudden right sided blindness. A biopsy of the temporal artery would show:

Atheromatous emboli.

Dissection.

Giant cell inflammation.

Thromboarteritis obliterans.

The most likely clinical course is:

Effective response to steroids.

Gangrenous necrosis of the right side of the face.

Permanent bilateral blindness.

Transformation to angiosarcoma.

A thirty year old female develops fatigue, weight loss, and fiver. She has reduced blood pressure in the upper extremities and no radial pulse is palpable. An aortic arch angiogram shows narrowing of the brachiocephalic, carotid, and subclavial arteries. The most likely diagnosis is:

Giant cell arteritis.

Kawasaki disease.

Polyarteritis nodosa

Takayasu arteritis.

A 30 year old male with chronic hepatitis B develops a rapid onset of hypertension, testicular pain, mononeuritis multiplex, and pancreatitis. Microscopic examination of his arteries would show:

Atherosclerosis.

Granulomatois inflammation.

Thrombosis

Transmural necrotizing inflammation with pseudoaneurysm formation.

A two year old male develops an acute febrile illness, cervical lymph node enlargement, conjunctivitis, oral erythema, and a desquamative skin rash of the hands. The importance of recognizing this syndrome lies in the prevention of

- Cerebral hemorrhage
- Coronary artery disease
- Disseminated intravascular coagulopathy
- Meningitis

40 year old male develops hemoptysis with nodular lung lesions, epitaxis, sinusitis, and acute renal failure. C-ANCA with Pr-3 specificity is present. If untreated, the prognosis is:

80% mortality in one year.

Chronic debilitating lung disease.

Likely dialysis dependent, but remission of respiratory symptoms.

Prompt recovery after a few weeks.

A 50 year old male develops hemoptysis, hematuria, and acute renal failure. He has antibodies to myeloperoxidase. The diagnosis is most likely:

- Giant cell arteritis.
- Kawasaki disease.
- Microscopic polyangiitis
- Myelodysplasia.
- Takayasu disease

Additional Slides for Review

Types of Vasculitis

Type of vasculitis	Affected body part(s)	People often affected	Symptoms can include
Takayasu arteritis	Aorta (the main artery that gets blood from the heart)	Women between 10 and 40. Not common in the United States.	Pain and weakness in arms or legs, belly pain after eating
Giant cell arteritis	Aorta, arteries on face and head	Older people and people with a condition called "polymyalgia rheumatica"	Headache, pain in the jaw, trouble seeing clearly or seeing from 1 eye
Polyarteritis nodosa	Kidneys, skin, other body parts	People with lupus, rheumatoid arthritis, hepatitis B or C, HIV (human immunodeficiency virus, the virus that causes AIDS), or a cancer called "hairy cell leukemia"	Tiredness, weight loss, fever, red bumps or purple bumps on skin, numbness or tingling in hands or feet, belly pain
Kawasaki disease	Heart, other body parts	Children under 5 years old	Fever, swelling, and redness in areas with inflammation
Primary central nervous system vasculitis	Brain and spinal cord	Middle-aged adults	Headache, confusion, stroke
Churg-Strauss vasculitis	Lungs, nose and sinuses, skin, other body parts	People with asthma	Asthma, allergies, sores on the skin
Granulomatosis with polyangiitis (Wegener's)	Nose, sinuses, lungs, kidneys	Older adults	Fever, joint pain, weight loss, runny or "crusty" nose, ear infections, cough, brown or dark urine
Henoch-Schönlein purpura	Skin, kidneys	Children between age 3 and 15, but can affect adults	Belly and joint pain, skin rash, brown or dark urine
Cryoglobulinemia	Blood	People infected with hepatitis C	Red or purple skin bumps, tiredness, joint and muscle pain, swollen glands
Hypersensitivity vasculitis	Skin	People taking medicines that cause a reaction	Red or purple skin bumps or other rash
Behçet's disease	Mouth, skin, genitals, eyes, other body parts	Adults age 20 to 40	Sores in mouth or on skin or genitals, eye problems, joint pain and swelling

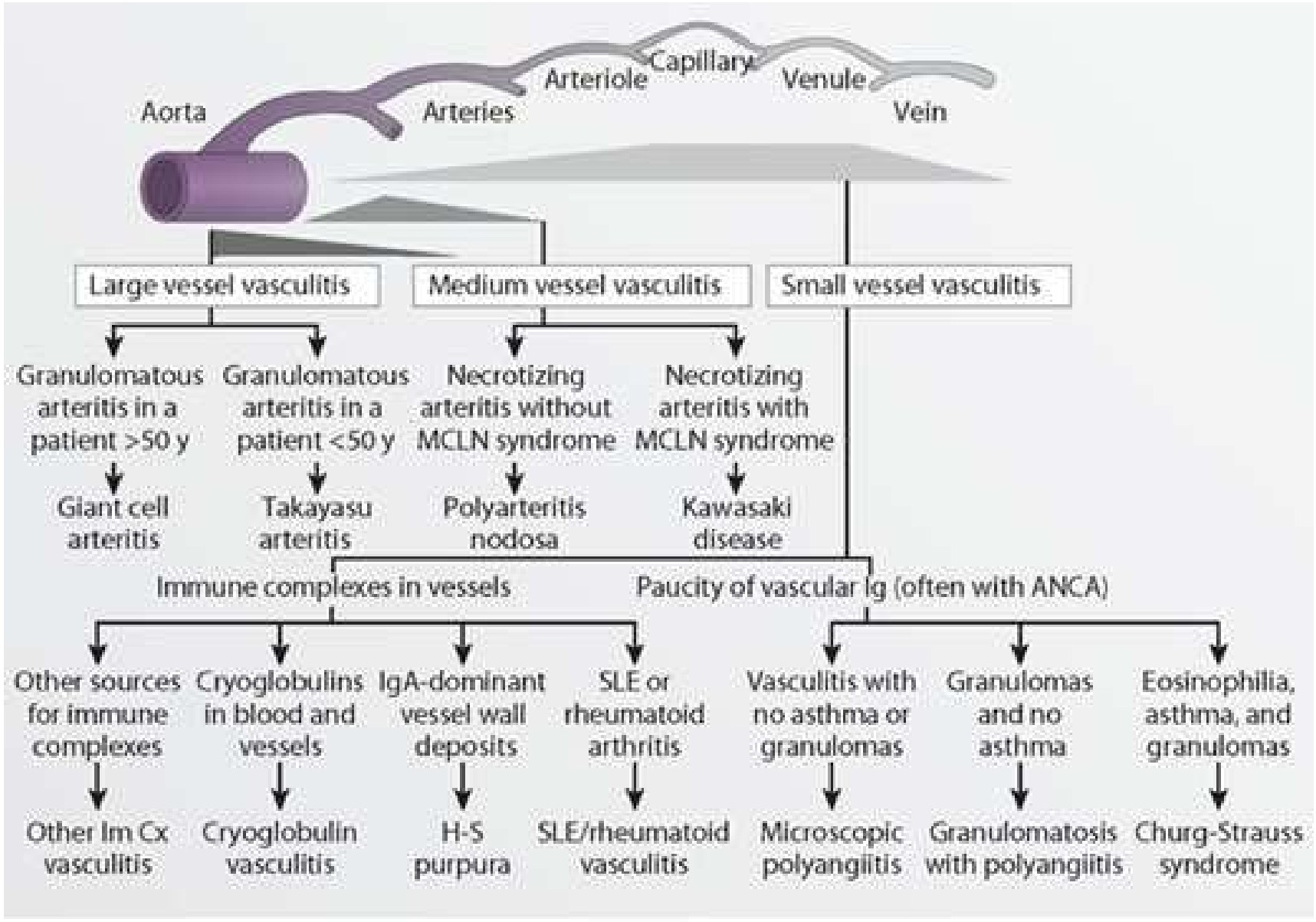
Mimics of Vasculitis

Major categories of mimics of vasculitis

Infectious causes (eg, endocarditis, HBV, HCV, HIV)
Atherosclerosis
Thromboembolic disease
Congenital causes (eg, aortic coarctation, middle aortic syndrome)
Hereditary disorders (eg, Marfan syndrome, Ehlers-Danlos syndrome)
Fibromuscular dysplasia
Hypercoagulable states (eg, APS, TTP)
Vasospastic disorders (eg, RCVS, drug exposures)
Other multisystem inflammatory disorders (eg, sarcoidosis, Susac syndrome)
Malignancy (eg, lymphoma, leukemia)
Iatrogenic (eg, postradiation therapy)
IgG4-related disease

HBV: hepatitis B virus; HCV: hepatitis C virus; HIV: human immunodeficiency virus; APS: antiphospholipid syndrome; TTP: thrombotic thrombocytopenic purpura; RCVS: reversible cerebral vasoconstriction syndrome; IgG4: immunoglobulin G4.

Vascular Distribution of Major Types of Vasculitis



Source: Imboden JB, Hellmann DB, Stone JH: *Current Diagnosis & Treatment: Rheumatology*, 3rd Edition: www.accessmedicine.com