بسم الله الرحمن الرحيم

Vasculitis

DEFINITION

- **Inflammatory condition in which there is leukocyte infiltration in vessel wall with reactive damage to mural structures.**

General consideration

- May affect one organ or multiple organs
- May cause local or systemic symptoms
- May be chronic or self limited disease
- May be primary or secondary

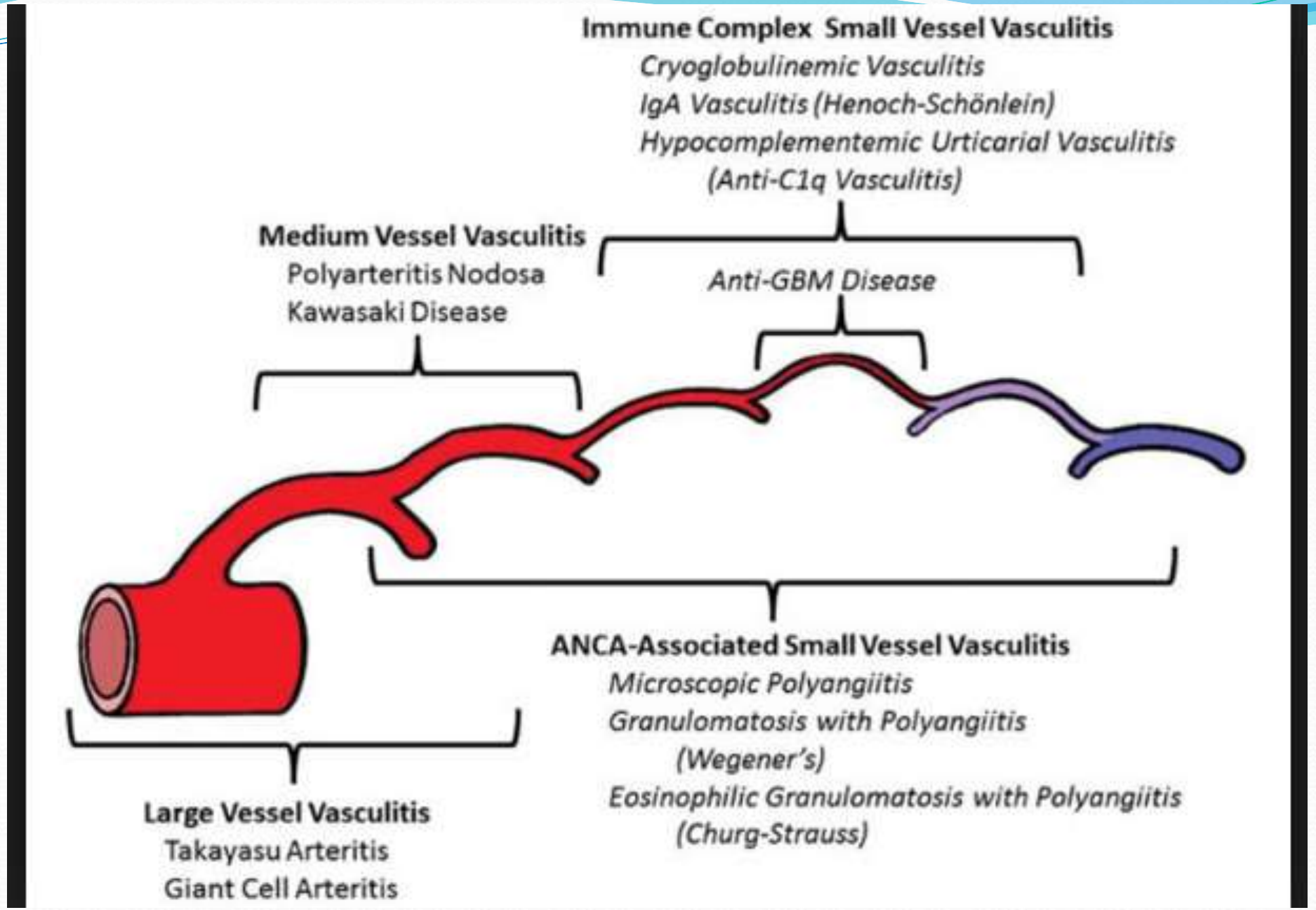
General consideration:

- Small vessels vasculitis:

  Loss of integrity → bleeding.
  Compromise in the lumen → tissue ischemia and necrosis.

- Medium and large vessels vasculitis:
  Aneurysms
  Ischemia to tissues

- Distribution of involvement depend on distribution of Ag in vessel wall.
- Patchy, but wide spread .
- Vasculitis is often serious , can be **FATAL** .
- Death in early stages disease .
  late stages treatment S/E


Immune Complex Small Vessel Vasculitis
Cryoglobulinemic Vasculitis
IgA Vasculitis (Henoch-Schönlein)
Hypocomplementemic Urticarial Vasculitis
(Anti-C1q Vasculitis)
Medium Vessel Vasculitis
Polyarteritis Nodosa
Kawasaki Disease
Anti-GBM Disease
ANCA-Associated Small Vessel Vasculitis
Microscopic Polyangiitis
Granulomatosis with Polyangiitis
(Wegener's)
Eosinophilic Granulomatosis with Polyangiitis
(Churg-Strauss)
Large Vessel Vasculitis
Takayasu Arteritis
Giant Cell Arteritis

Classification of vasculitis

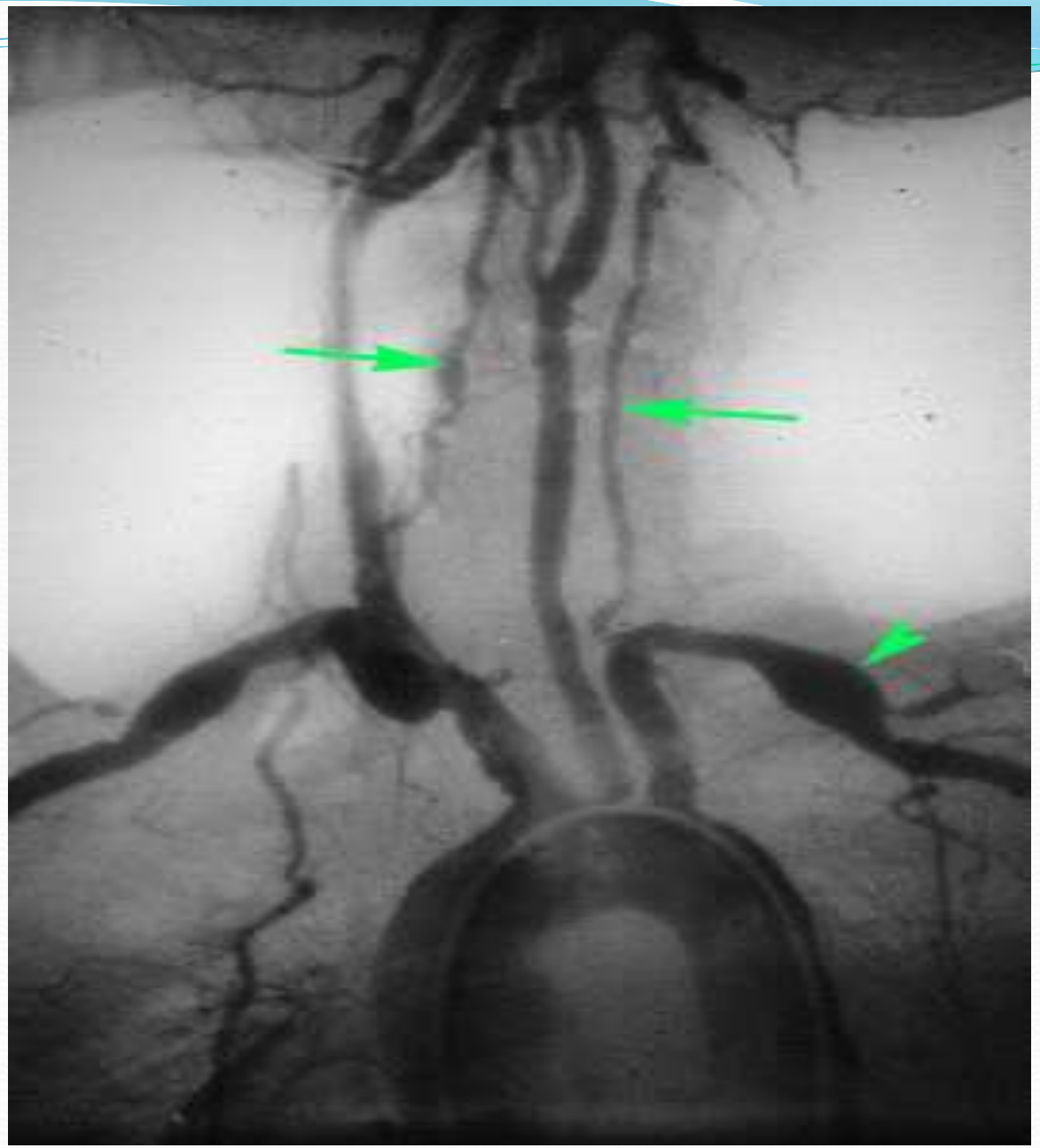
- Primary

- Secondary
  - Infections Hepatitis B , Hepatitis C
  - Malignancies.
  - Immune system disorders RA, SLE, Scleroderma
  - Reaction to drugs

- Large vessel vasculitis
  - Takayasu arteritis.
  - Giant cell arteritis (Temporal arteritis).

- Medium vessel vasculitis
  -Polyarteritis nodosa.

- Small vessel vasculitis
  - Granulomatosis and polyangitis
  - Henoch-Schonlein purpura.



Takayasu arteritis

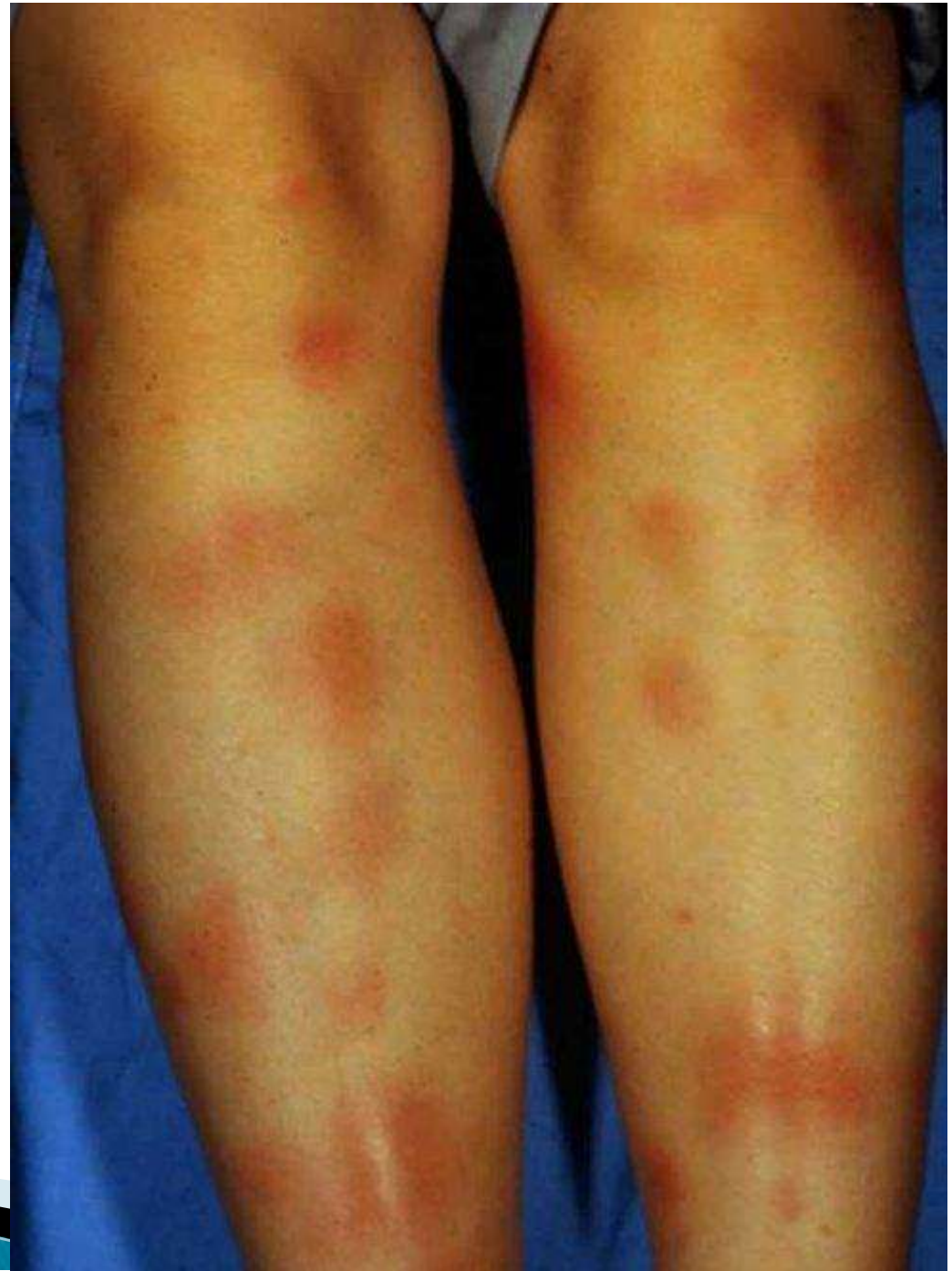
- Affects aorta and its branches.
- Females are 80-90% F:M 10:1
- Age 10-40 years.
- Japan 150 new cases / year .
- Affects abdominal aorta and pulmonary vessels in 50% of cases.
- Symptoms :

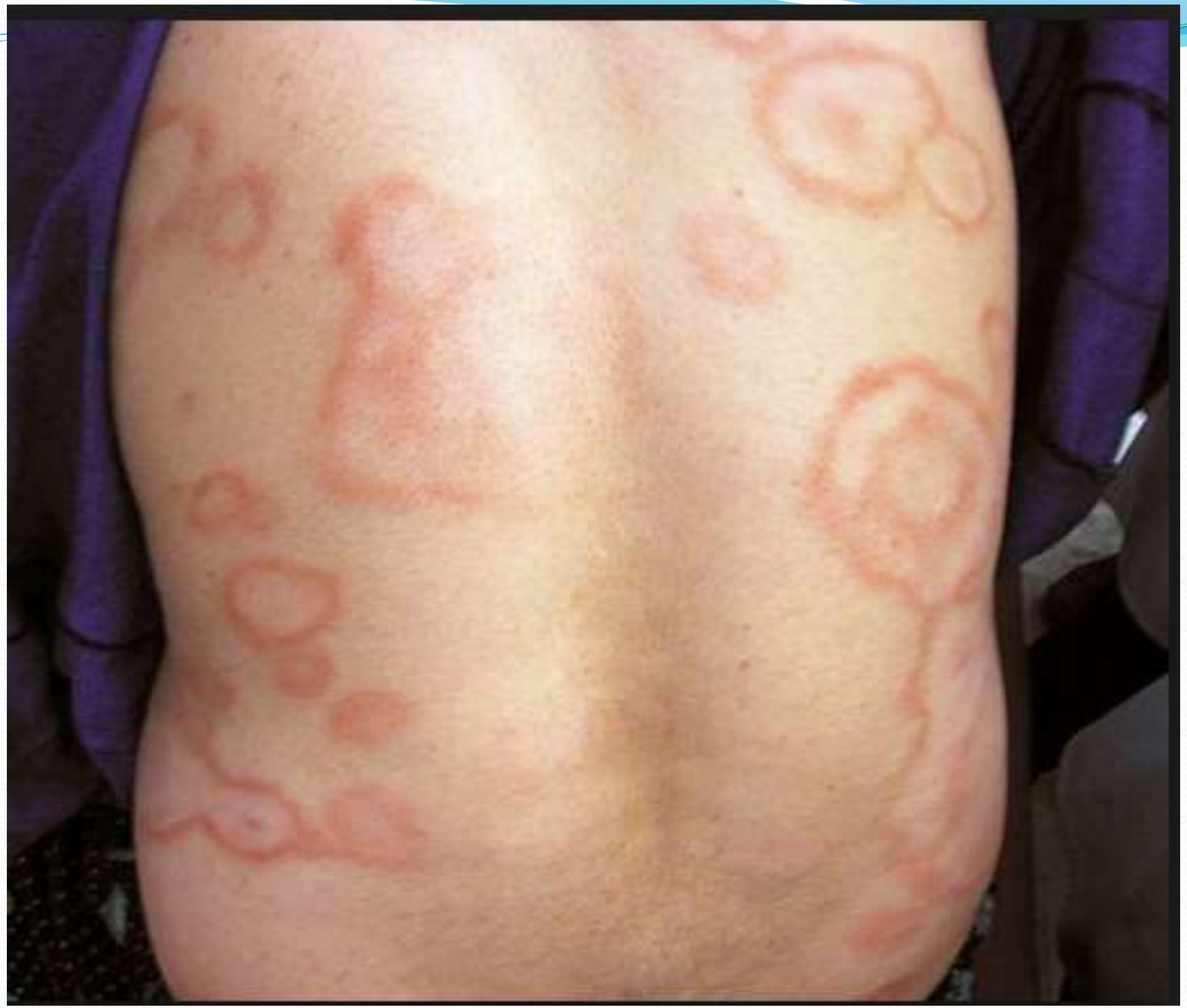
  general: fatigue ,weight loss ,fever

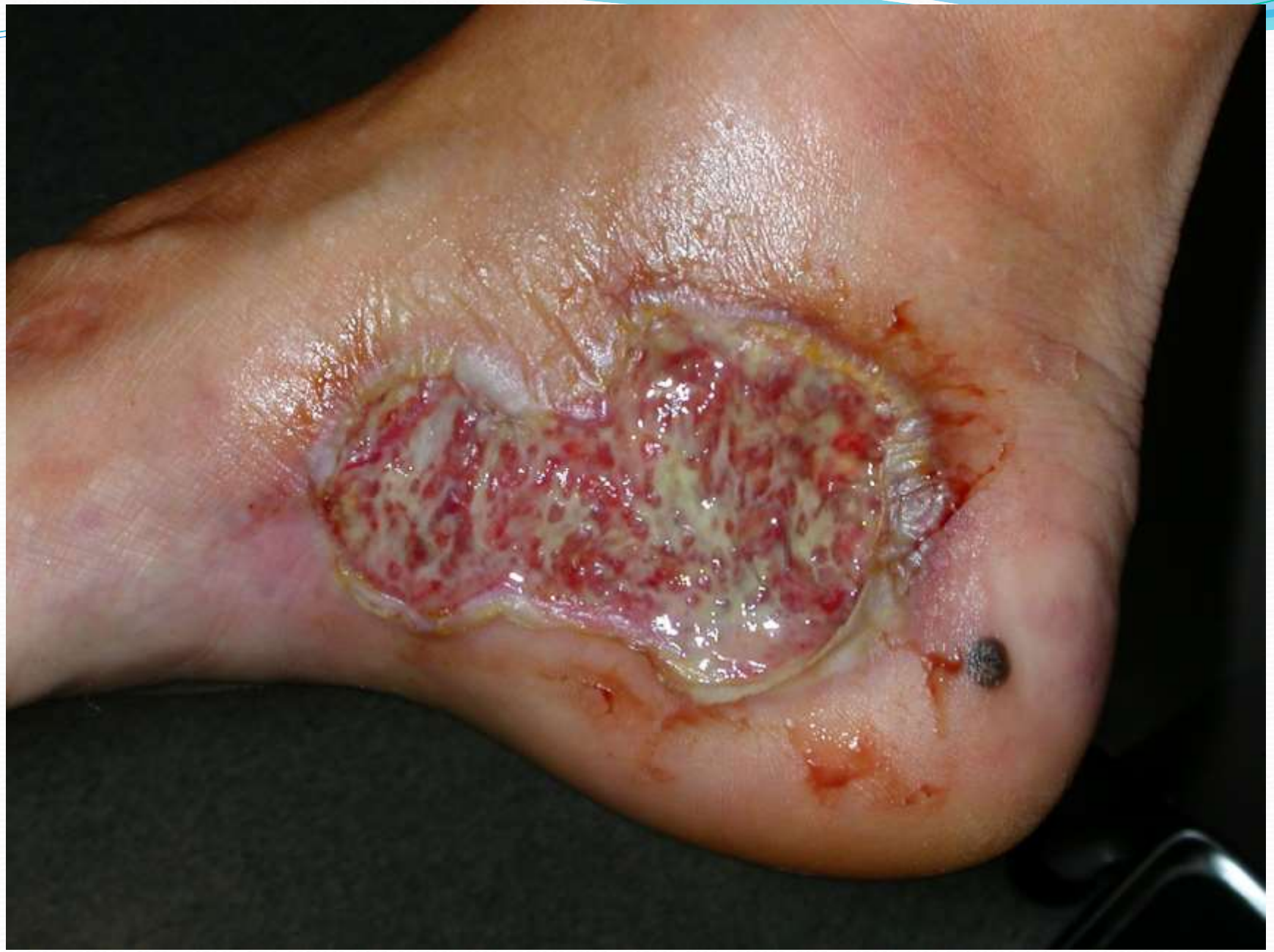
  vascular: leg and arm claudication.

Takayasu arteritis(cont.)

- Symptoms:
  - subclavian steal syndrome →vertebral ischemia→ syncope.
  -gangrene and ischemic ulcers.
  -arthralgias ,myalgia.
  -skin lesions :erythema nodosum
  pyoderma gangrenosum
  -SOB, chest pain ,hemoptysis
  pulmonary hypertension.
  - angina pectoris







Takayasu (cont.)

- Physical exam:
  - looks ill, febrile .
  -low B.P in one or both limbs.
  -difference of >10mm is typical.
  -a symmetrical pulses , diminished.
  -bruits over subclavian, brachial, or abdominal vessels.
  -hypertension in 50% (renal artery involvement).

Takayasu (cont.)

- Investigations:
  - -high ESR , CRP .
  - -low albumin.

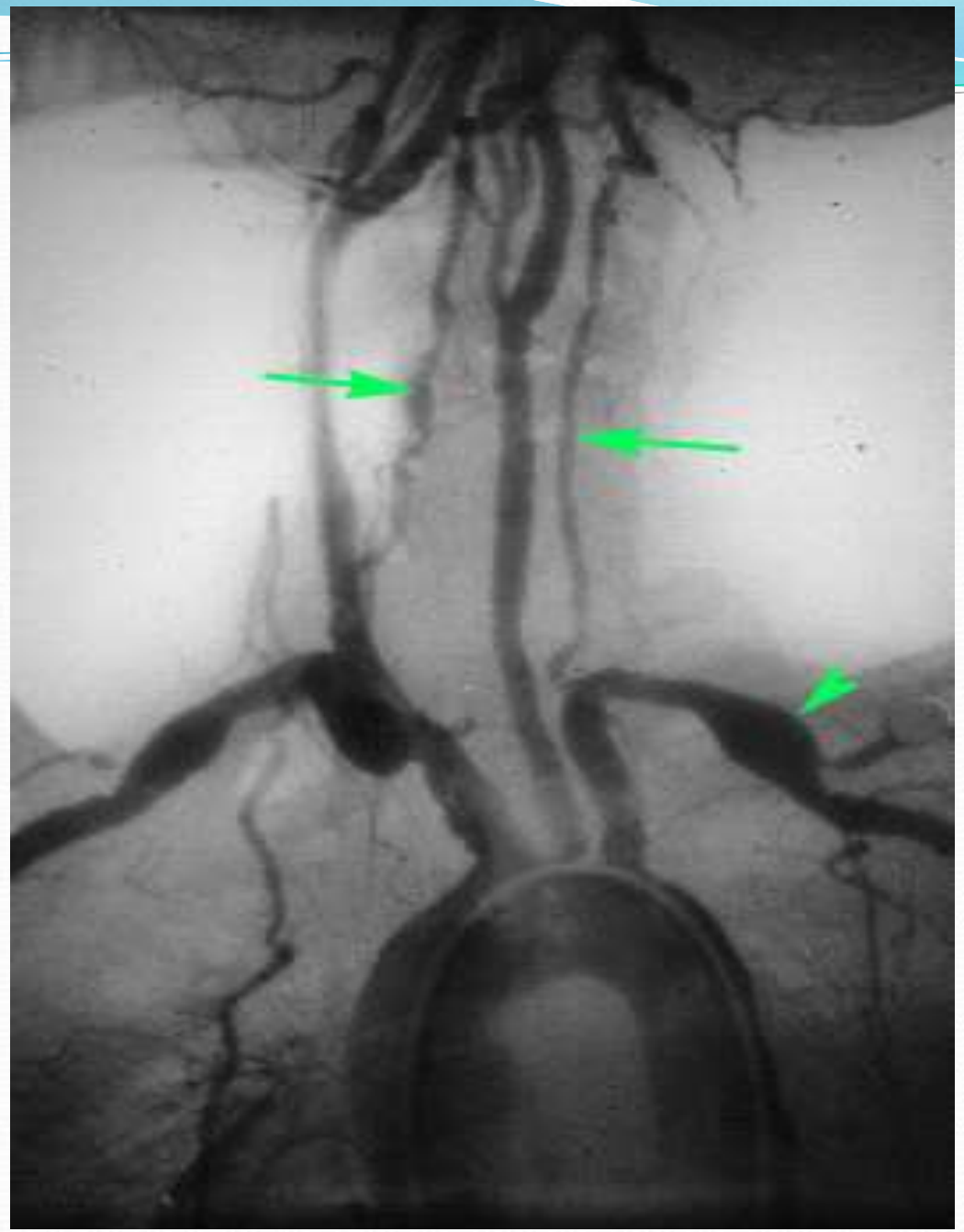
CXR : mediastinal widening (aneurismal dilatation).

Angiography :

- -narrowing , dilatations , beading.
- -patchy involvement.
- -collateral vessels.

BIOPSY : NOT AN OPTION.

CT , MRI ,PET scan.

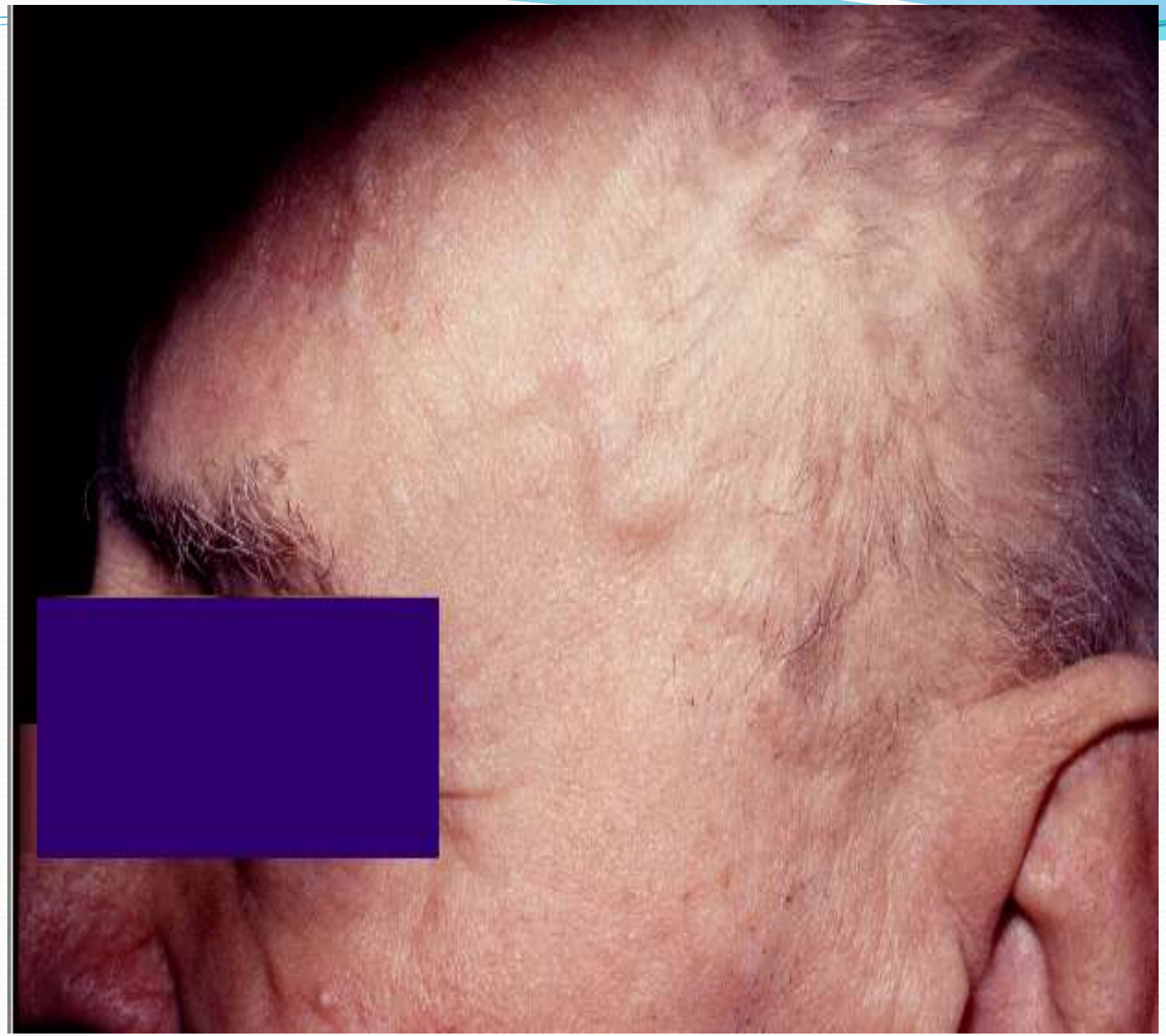


Treatment:

- Corticosteroids
  -prednisolone 1 mg/kg initially .

- Immunosuppressant :
  Methotrexate, AZA , Mycophenolate, Rituximab

- REVASCULARIZATION.



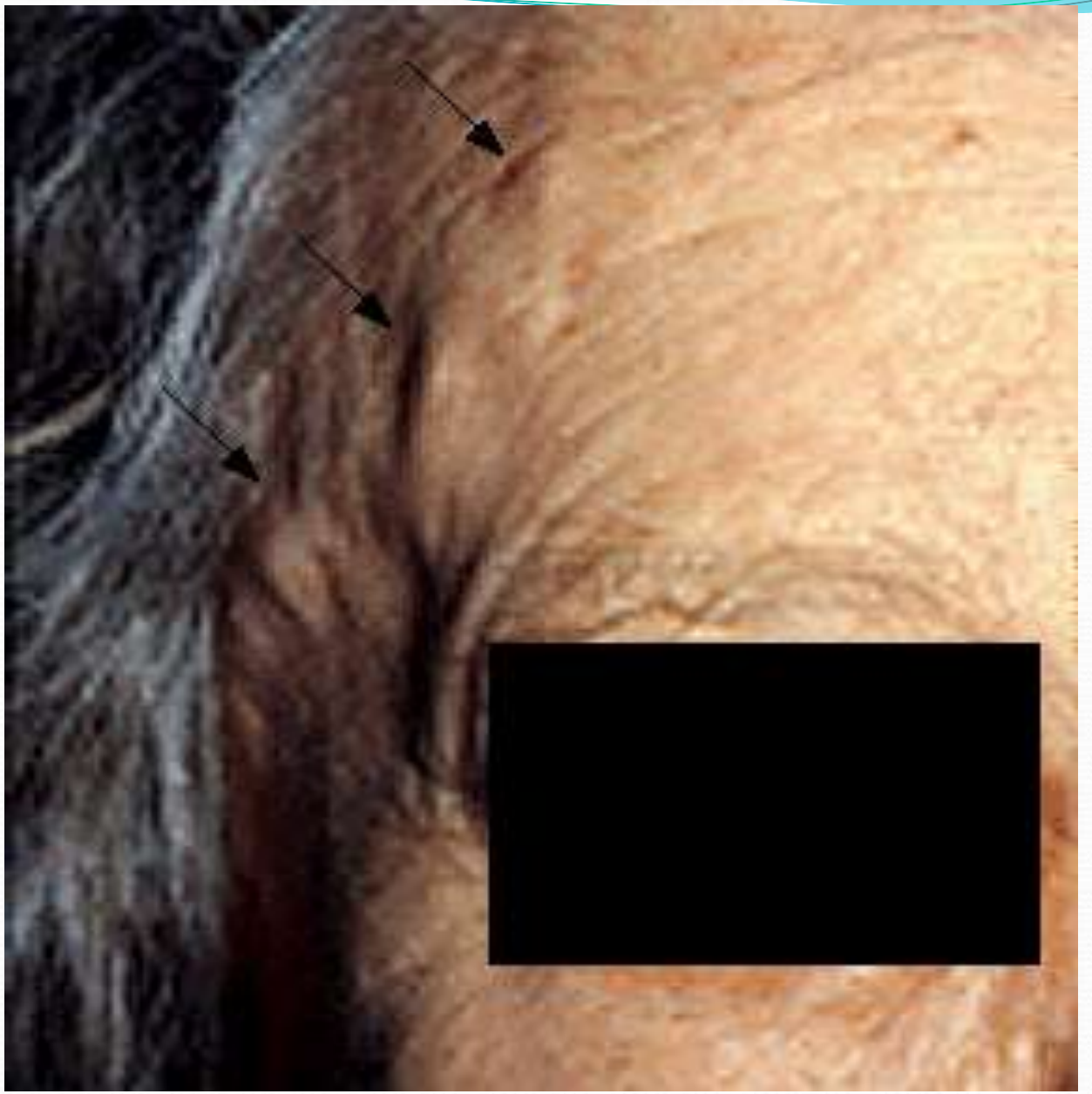
GIANT CELL ARTERITIS

- AGE > 50 YEARS.
- FEMALE:MALE 3:2
- Affects mainly the cranial branches of the aortic arch.

Symptoms

-fever ,weight loss , fatigue.

-headache : 2/3 of cases temporal artery tenderness.



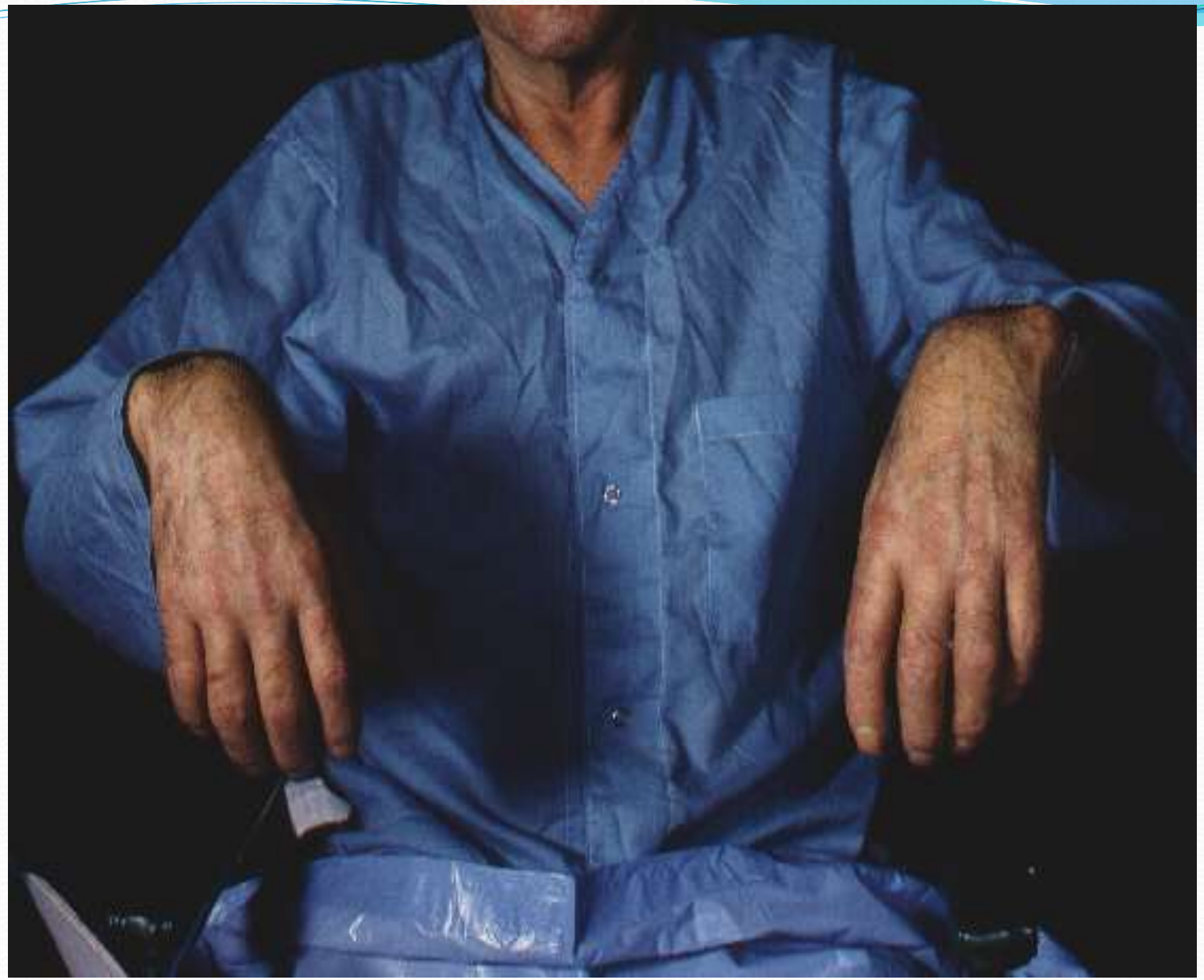
GCA (CONT.)

- Jaw claudication : 50%
  -tongue or throat pain.
- Visual loss 15-20%
  -early manifestation.
  -abrupt visual field defect in one eye → blindness.
  -next eye may follow in 1-2 weeks.
- Arm claudication.

GCA (DIAGNOSIS)

- ANY PATIENT > 50 YEARS WITH :
  - Headache
  -Abrupt loss of vision
  -Symptoms of polymyalgia rheumatica
  -Unexplained fever or anemia
  -high ESR, high CRP

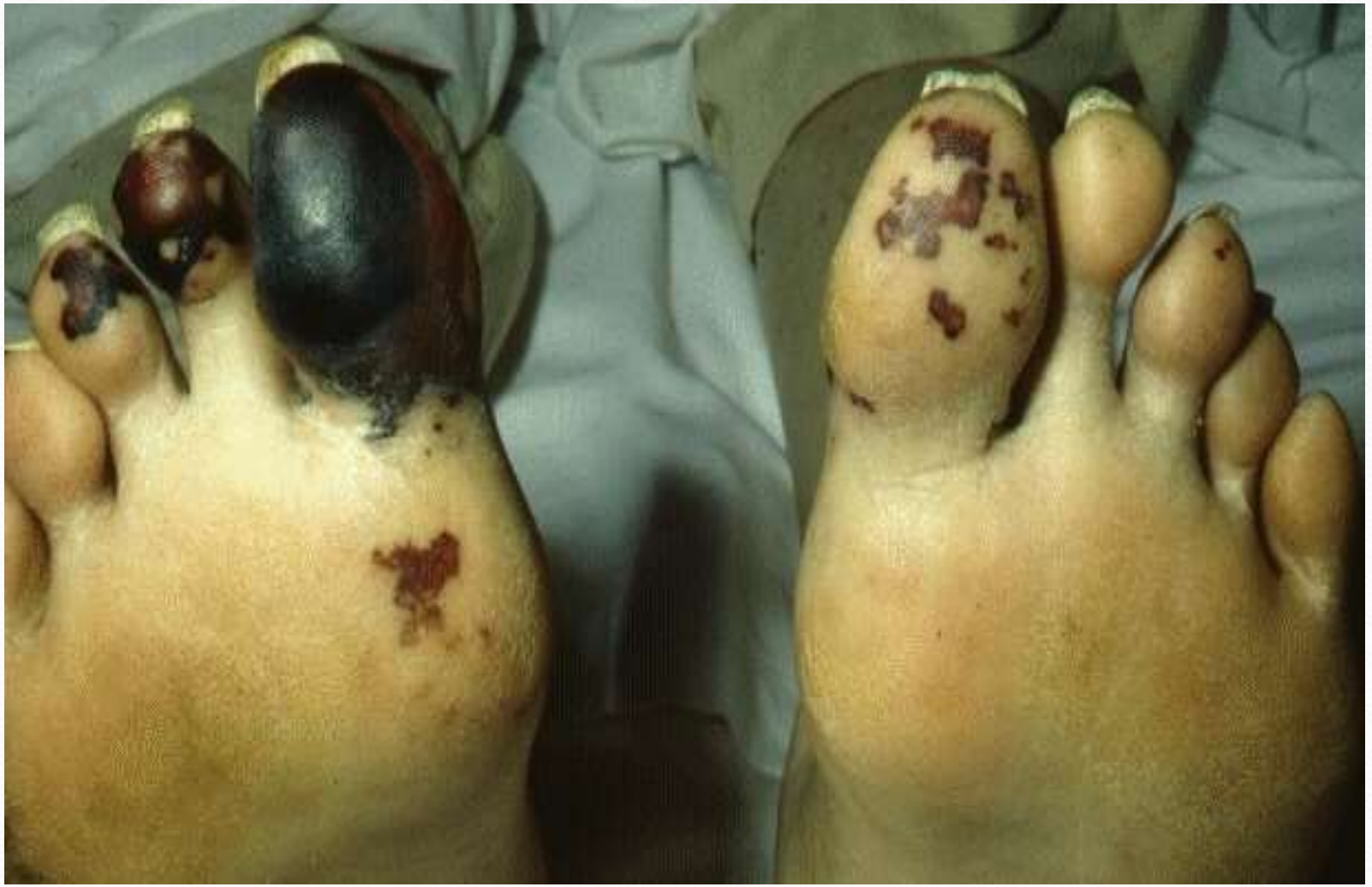
Diagnosis : temporal artery biopsy.



Polyarteritis nodosa

A systemic necrotizing vasculitis.

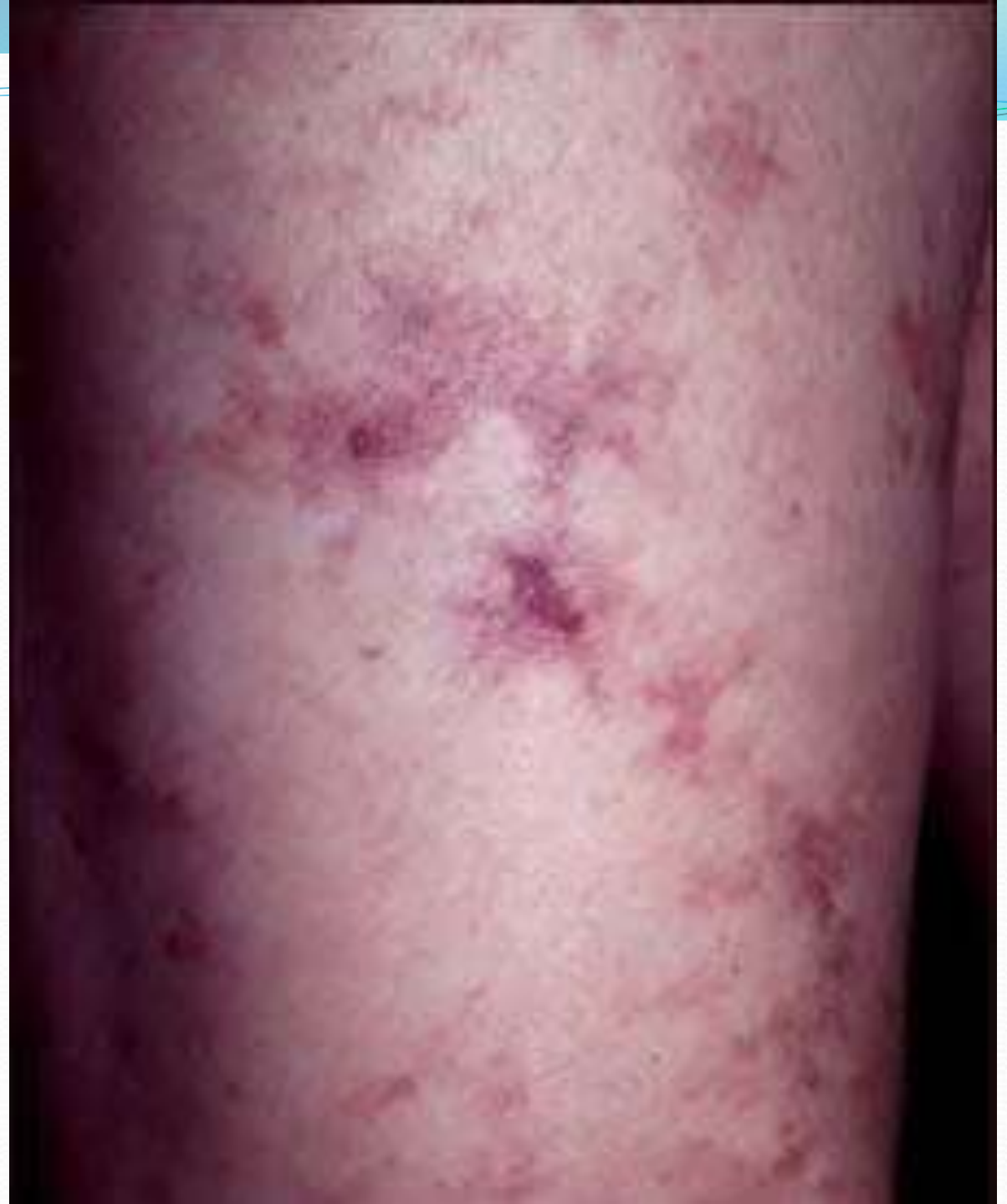
- **Affects small and medium sized arteries.**

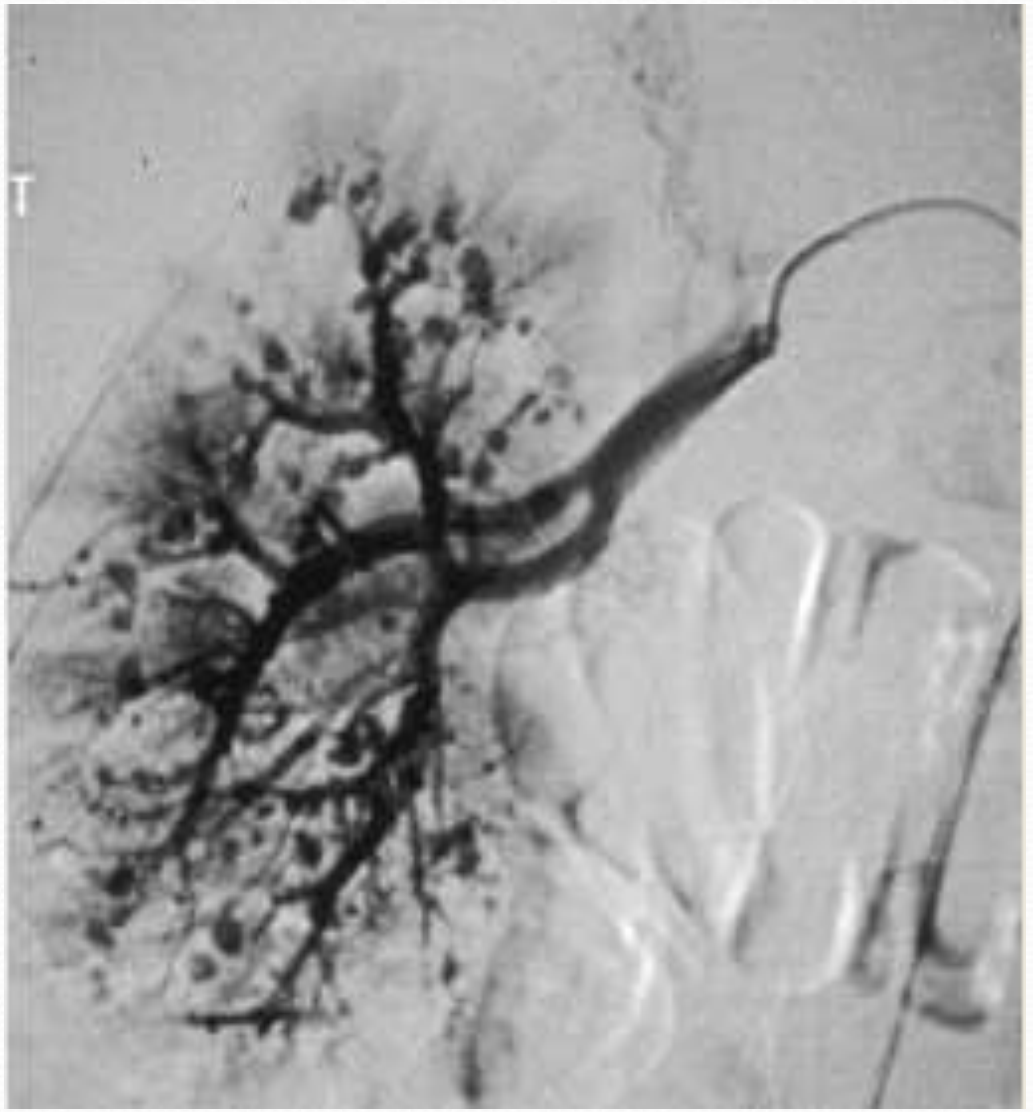


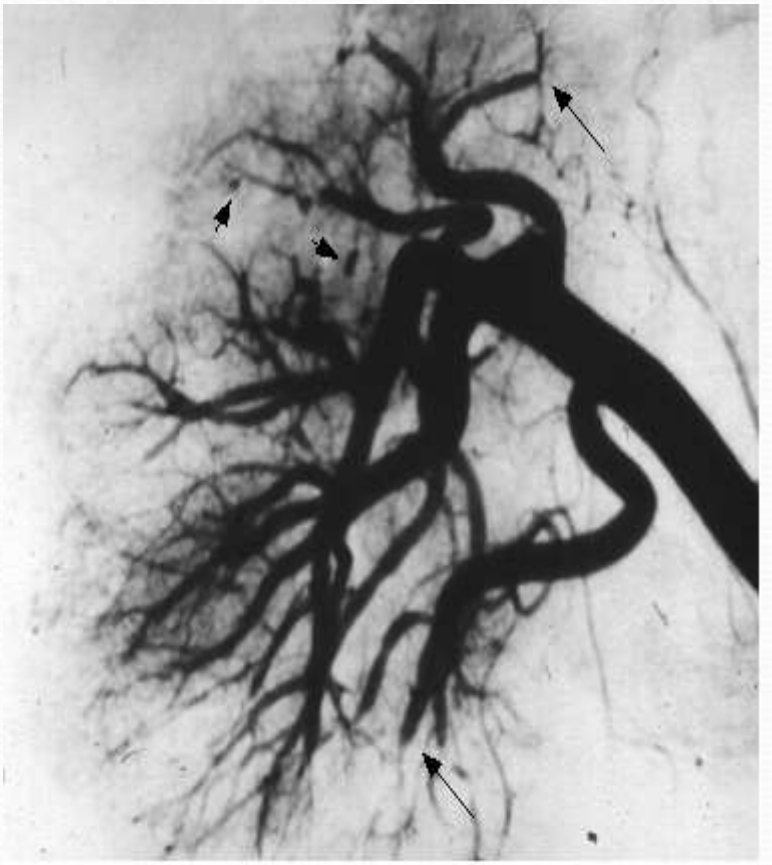
Classification criteria PAN

- **Livedo reticularis**.
- **Mono-neuropathy or poly neuropathy**.
- **New onset of diastolic B.P > 90 mm Hg**.
- **High urea>40 creat.>132 umol/l**
- **HBsAg +ve**.
- Characteristic **arteriographic abnormalities**.

- **NOTE : NEGATIVE ANCA**





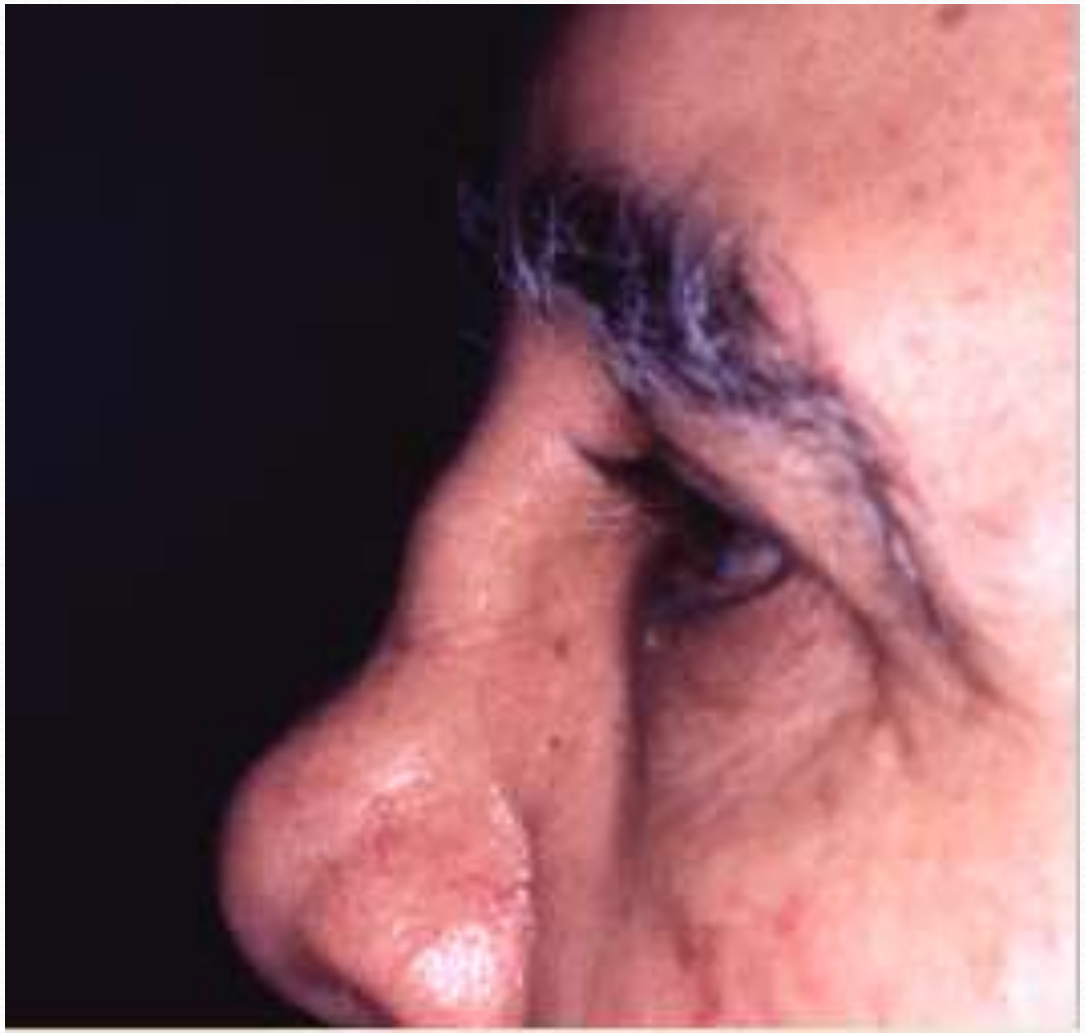


Polyarteritis nodosa Renal arteriogram in large vessel polyarteritis nodosa showing characteristic microaneurysms (small arrows) and abrupt cutoffs of small arteries (large arrows). (From Rose, BD, Pathophysiology of Renal Disease, 2d ed, McGraw-Hill, New York, 1987.)

PAN (treatment)

- Steroids .
- Cyclophosphamide.
- Hypertension control:

  ACE inhibitor – may worsen renal function
  Calcium channel blocker.
- Renal transplantation.



GPA (Granulomatosus and polyangitis)

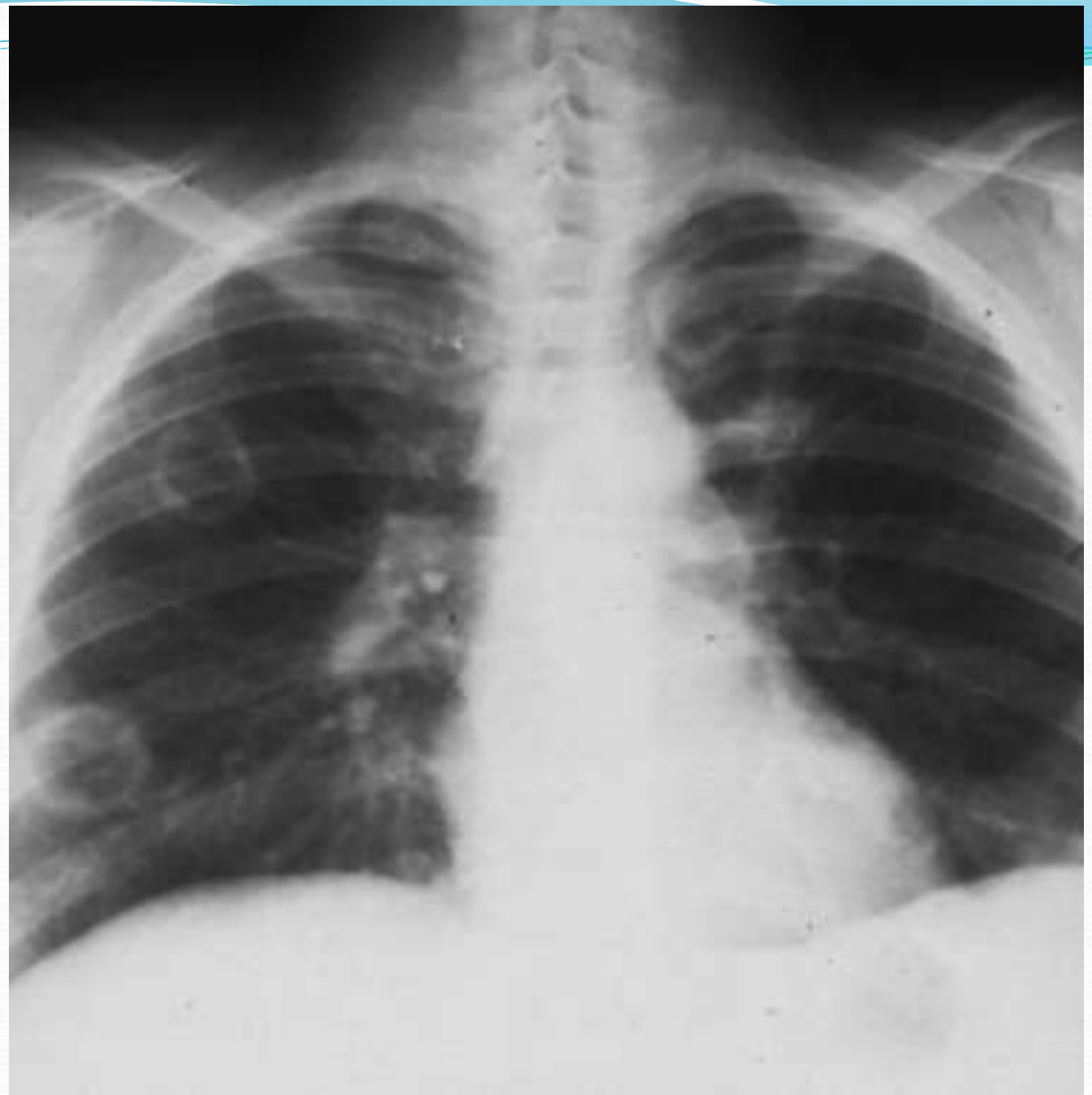
- Formerly known as Wegeners' granulomatosus.
- Systemic granulomatous vasculitis of the medium and small arteries, as well as the venules and arterioles.
- Typically causes granulomatous inflammation of upper and lower respiratory tract and pauci-immune glomerulonephritis in the kidneys.
- Positive C-ANCA

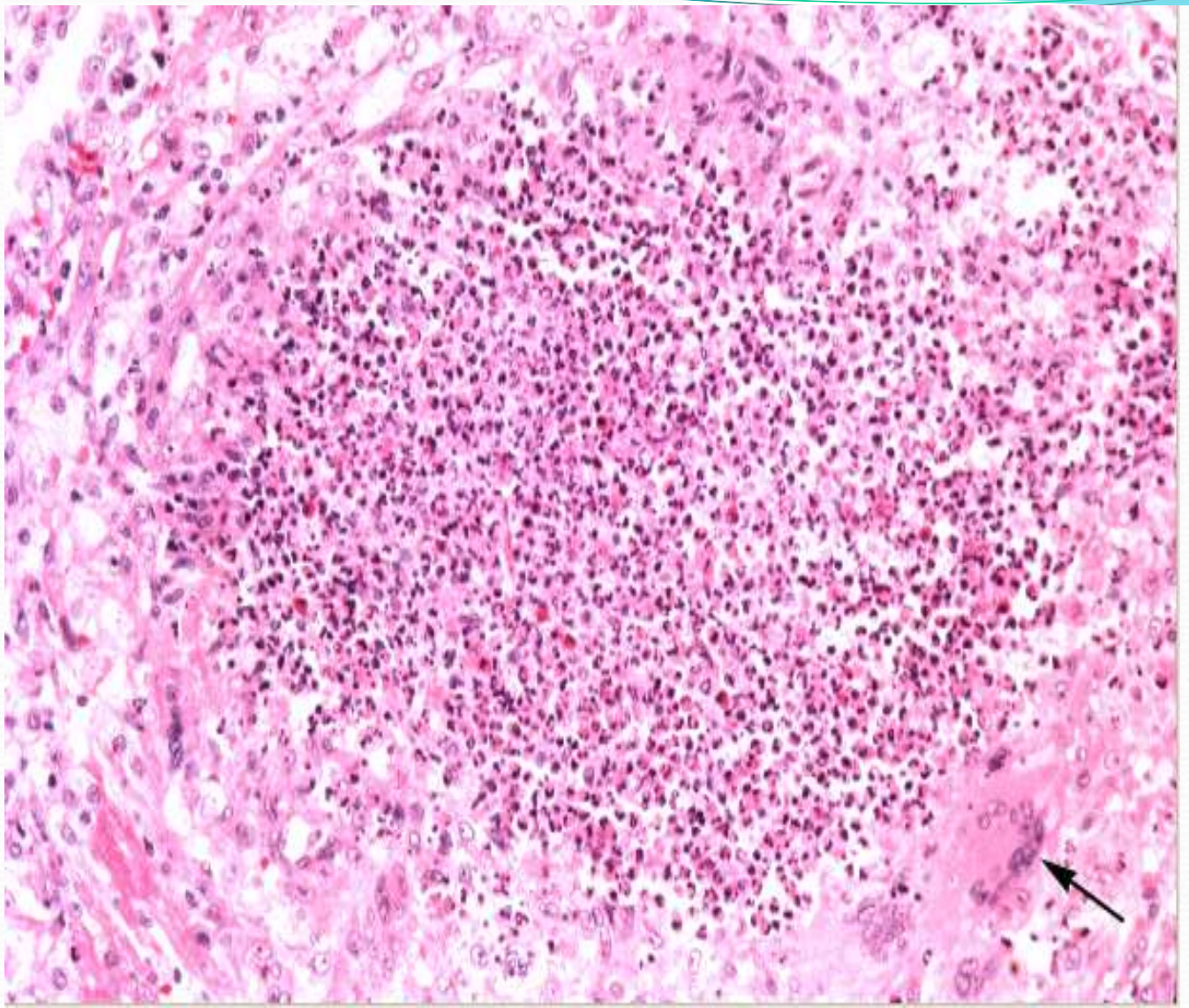
Clinical picture

- Upper respiratory
  - Rhinorrhea
  -nasal discharge/ulcers
  -sinusitis
- Lower respiratory
  -dyspnea
  -cough
  -hemoptysis

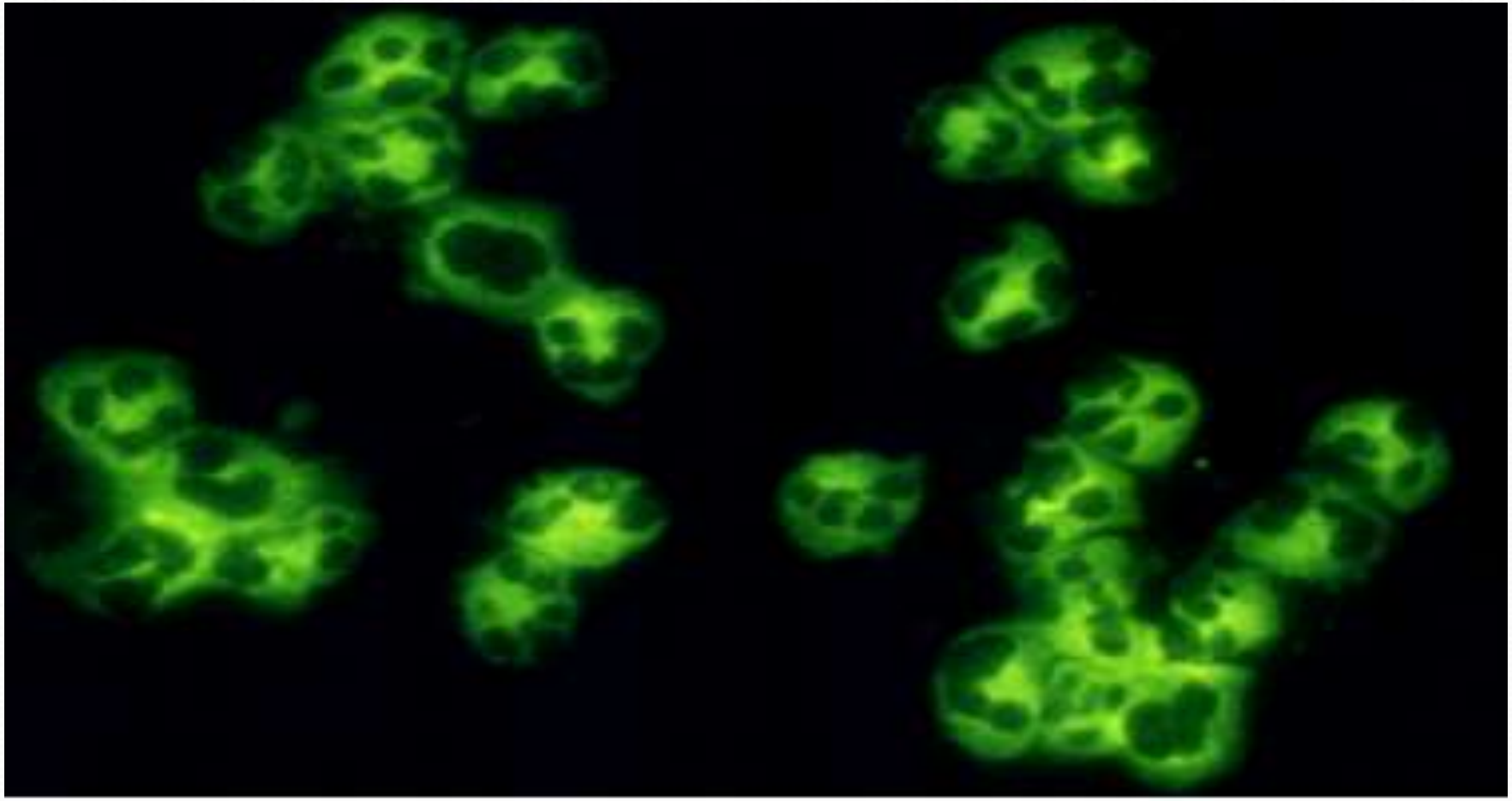
CXR : nodules, alveolar opacities, pleural opacities , alveolar hemorrhage, cavities.

- Kidneys active sediment , high creatinine



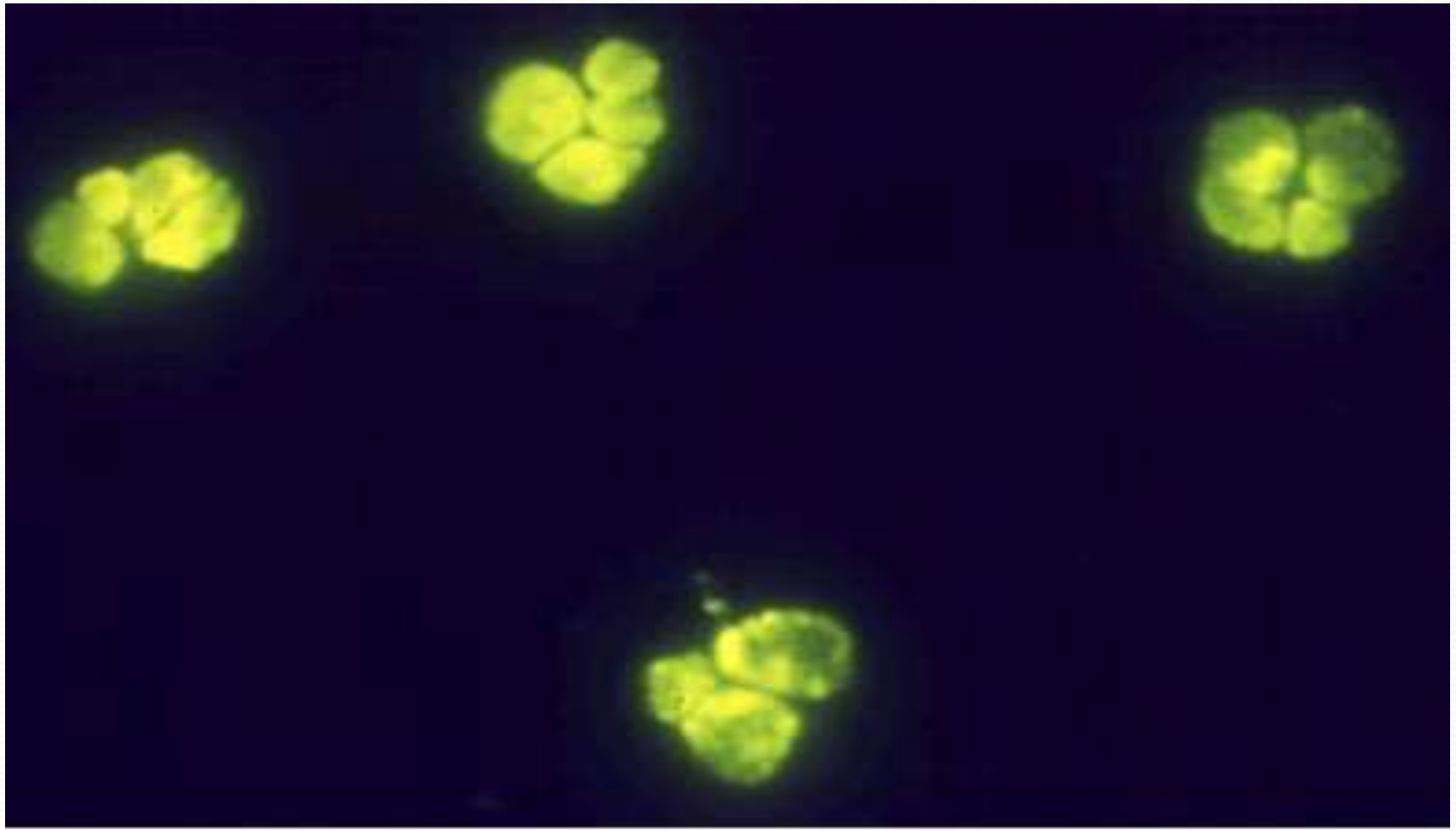


C- ANCA (PROTEINASE 3)

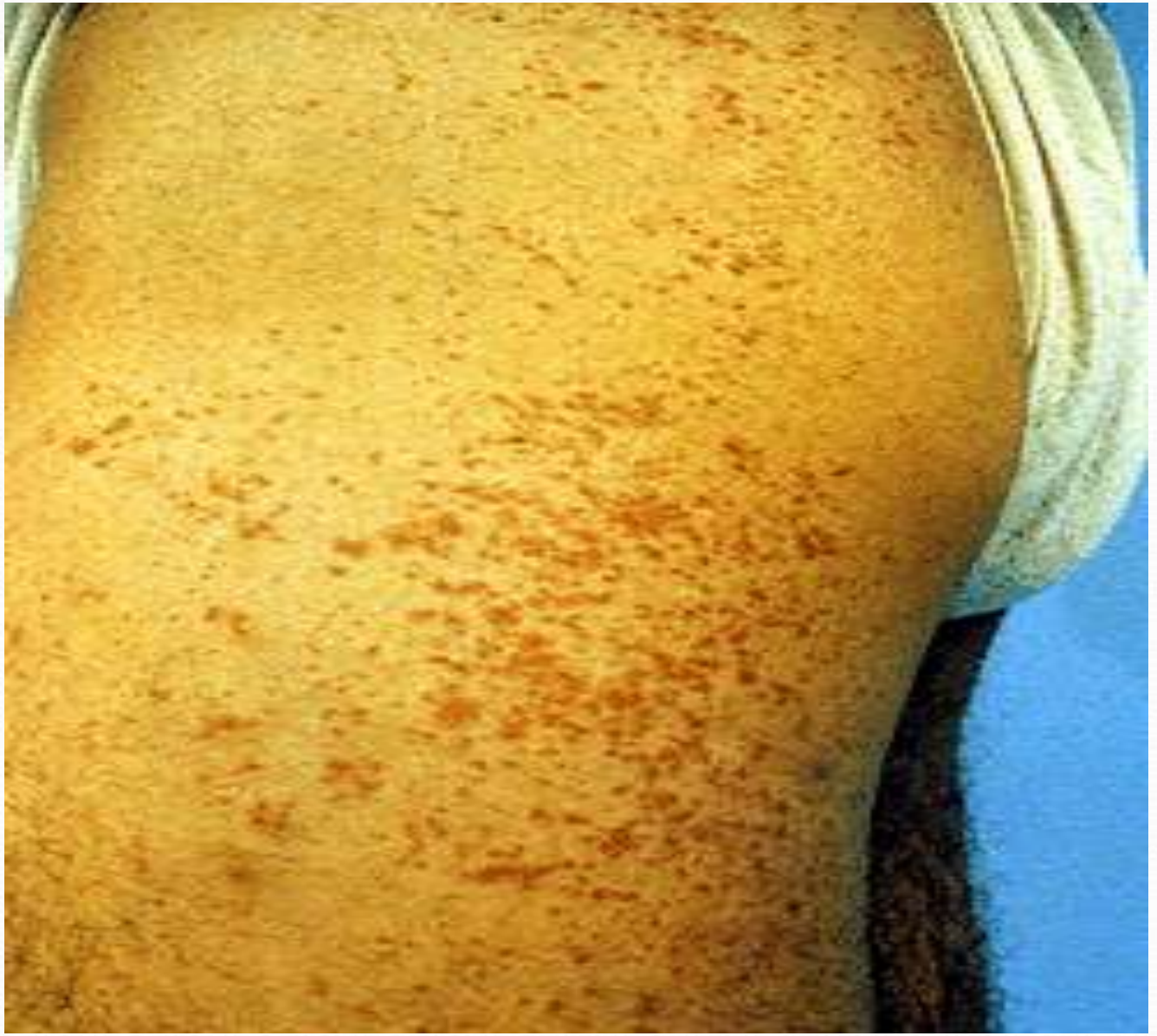


C-ANCA pattern Demonstration of cytoplasmic

P-ANCA (Myeloperoxidae- MPO)



P-ANCA pattern Demonstration of perinuclear



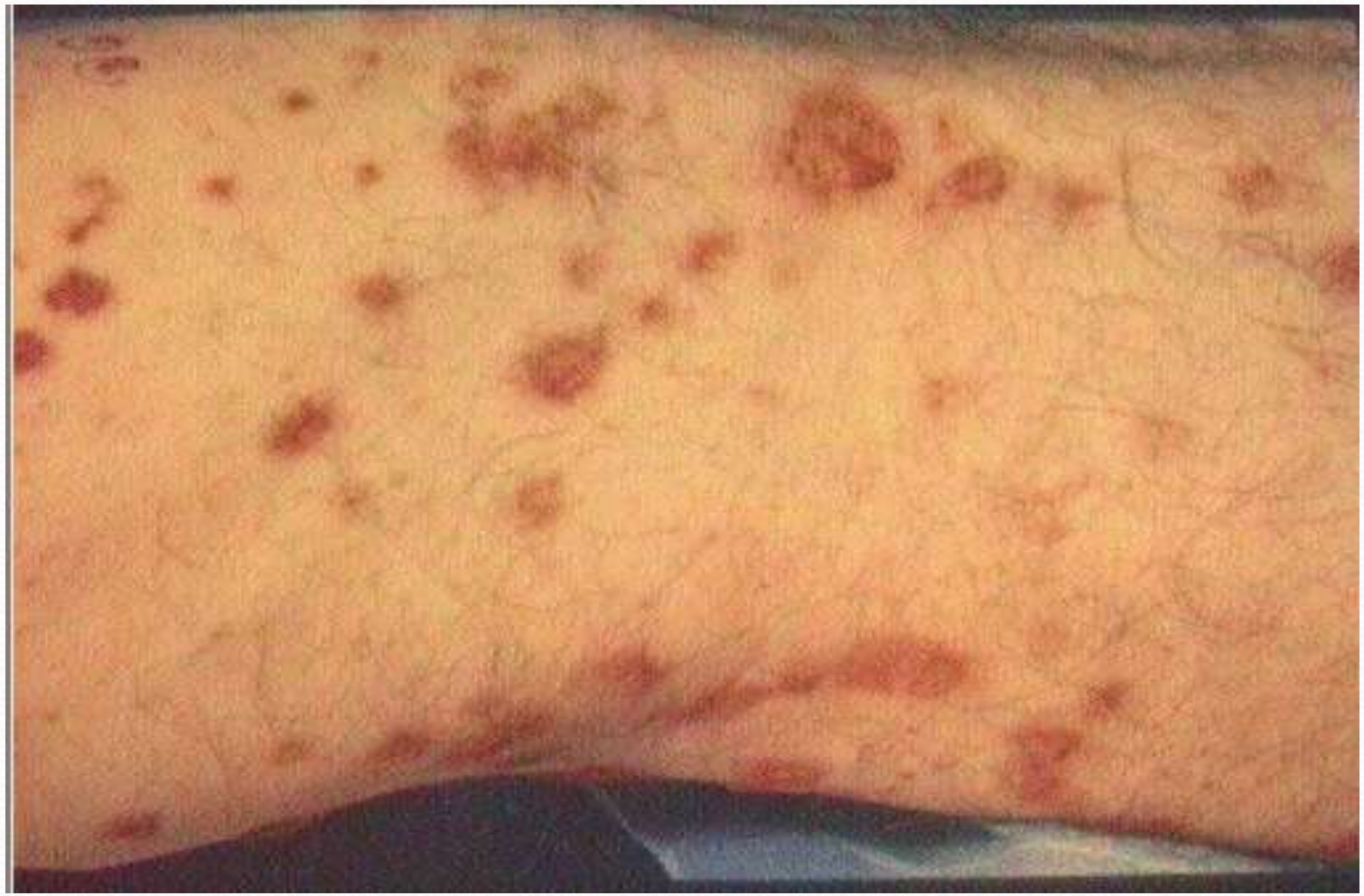
Henoch-schonlein purpura

- A systemic vasculitis with prominent cutaneous component.
- It is a form of hypersensitivity vasculitis.
- Characterized by deposition of IgA containing immune complexes.
- Occurs more in children esp. < 5 years.
- Usually follows URTI.
- Renal involvement is more severe in older children.

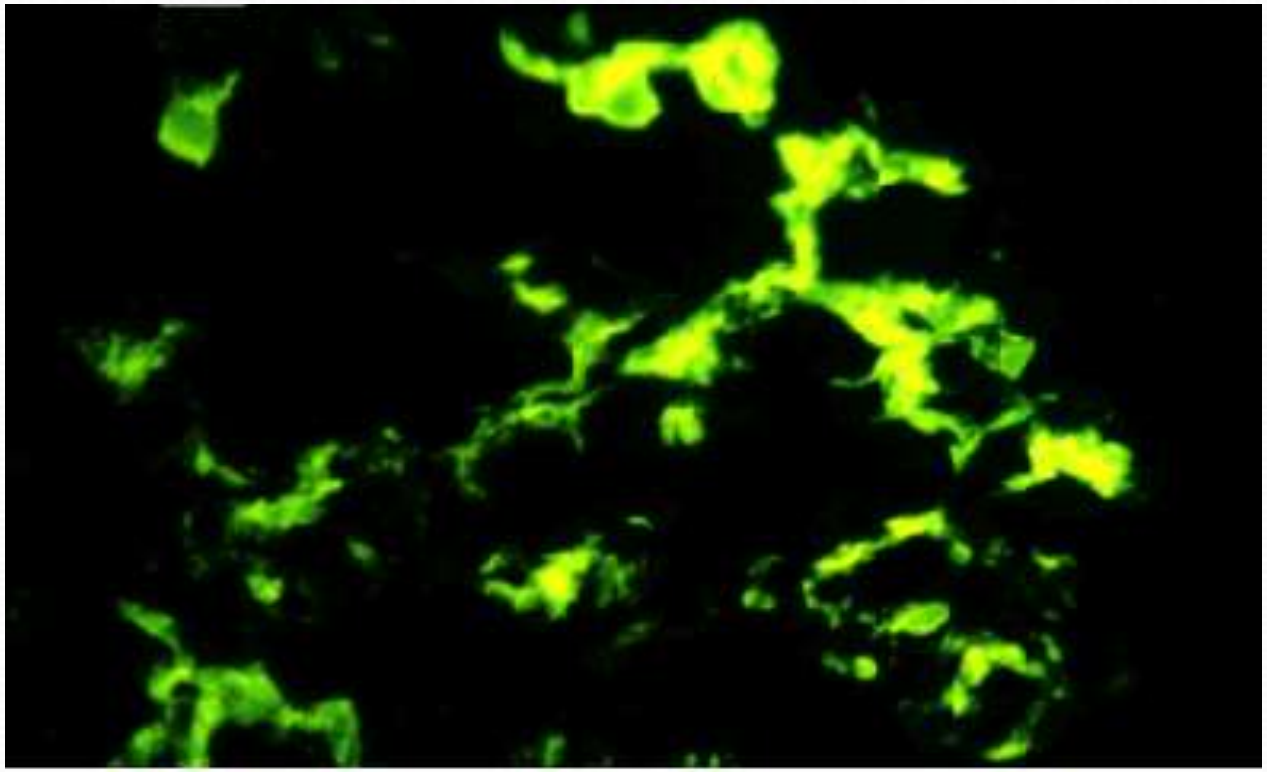
HSP (cont.)

- Clinically : develops over days-weeks

-purpura 100%
-arthralgia 82%
-abdominal pain 63%
-renal disease 40%
-GI bleeding 33%

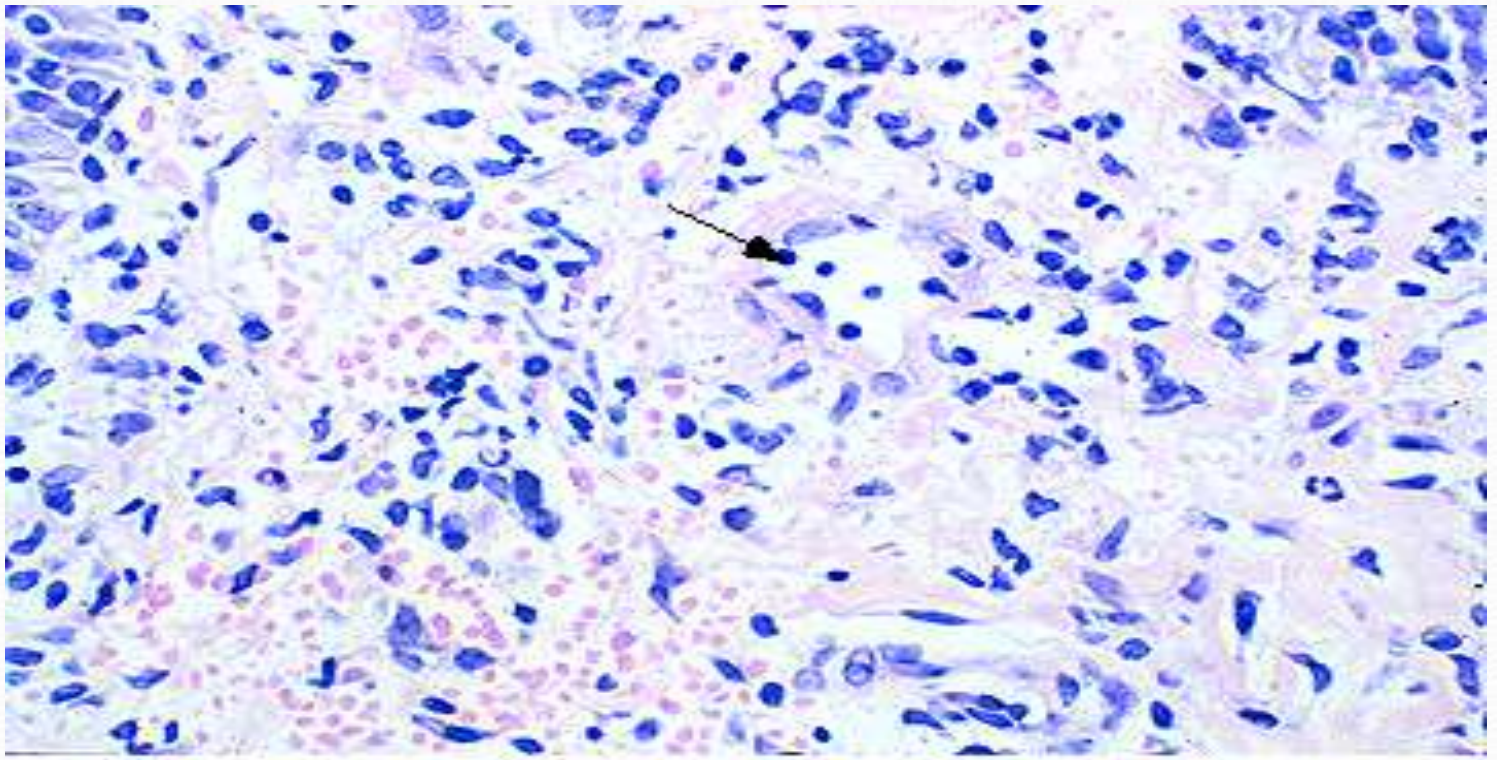


HSP (kidney biopsy)



Mesangial IgA deposits Immunofluorescence microscopy demonstrating large, globular mesangial IgA deposits that are diagnostic of IgA nephropathy or Henoch-Schönlein purpura. Note that the capillary walls are not outlined, since the deposits are primarily limited to the mesangium. Courtesy of Helmut Rennke, MD.

HSP (skin biopsy)



Leukocytoclastic vasculitis Leukocytoclastic vasculitis involving the dermal papillae capillaries and venules, a finding that probably reflects an Arthus type III immune complex reaction. Courtesy of Cynthia Magro, MD.

HSP (TREATMENT)

- Most patients recover with no specific treatment. 94% children
  89% adults.
- Steroids are indicated in:
  -crescentic glomerulonephritis.
  -GI vasculitis (abdominal pain).
- Occasionally cyclophosphamide may be needed
- Plasmapharesis, IV immunoglobulin-rarely

Approach to vasculitis in adults

- **Clues** :

- Mononeuritis multiplex (a symmetric poly – neuropathy) highly suggestive of vasculitis esp PAN (if we exclude DM).
- Palpable purpura like HSP.
- Pulmonary-renal syndrome.
- Pre-existing CT disease. i.e SLE

Approach to vasculitis

- History:

  -drugs - hepatitis C , B

  -SLE

  -sex

Demographic Characteristics of 807 Patients with Vasculitis†

Disease category	Percent with disorder	Mean age at disease onset	Percent female
Polyarteritis nodosa	15	48	38
Churg-Strauss syndrome	3	50	37
Wegener's granulomatosis	10	45	37
Hypersensitivity vasculitis	12	47	54
Henoch-Schonlein purpura	10	17	46
Giant cell arteritis	26	69	75
Takayasu's arteritis	8	26	86
Other vasculitis, type unspecified	16	44	55

†Data from from Hunder, GG, Arend, WP, Bloch, DA, et al. Arthritis Rheum 1990; 33:1065.

Approach to vasculitis

- Lab. Tests

-CBC,ESR

- MSU, 24-hr. urine collection for cr cl., proteinuria

-urea, creatinine

- Complements

-ANA

-ANCA PR3, MPO

-EMG and NCS (mononeuritis multiplex).

-Tissue biopsy .

-Arteriography.