

Vasculitis

- Auto immune inflammation and necrosis of blood vessels.(ischemia.And bleeding)
- any blood vessel involved

Vasculitis aetiology

- **primary** :an idiopathic condition
- **secondary**:
manifestation of
 - **Infectious disease**(Hepatitis B surface antigen (HBsAg) and HCV)
 - **Malignant** disease
 - **rheumatic** disease.e.g SLE

Vasculities presentation

3 main concepts responsible for cl features

1-multisystem disease inflammatory disease

Constitutional symptoms (fever, weight loss)

High ESR, severe anemia, thrombocytosis(increase platelets)

2-symptoms- obstructive or narrowing end-organ
ischemia or hemoohage

3-Multisystem.

Necrotic and bloody

VASCULITIS

Nervous system

- stroke

Heart

- myocardial infarction
- hypertension

Digestive system

- bloody stool 🩸
- abdominal pain

Joints

- pain
- arthritis

Skin

- palpable purpura
- livedo reticularis

Eye

- reduced visual acuity

Nose

- bleeds 🩸

Lungs

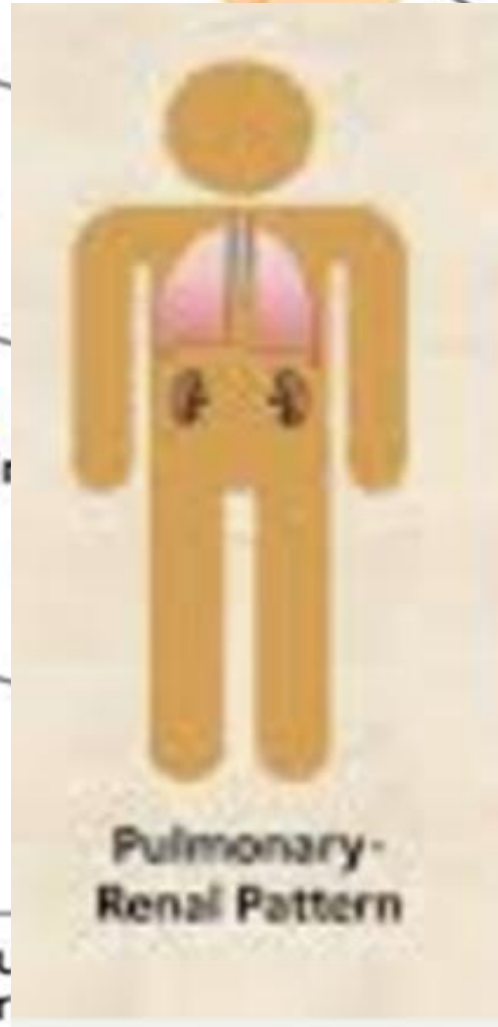
- bloody cough 🩸
- lung infiltrates

Kidneys

- glomerular nephritis

Muscle

- pain



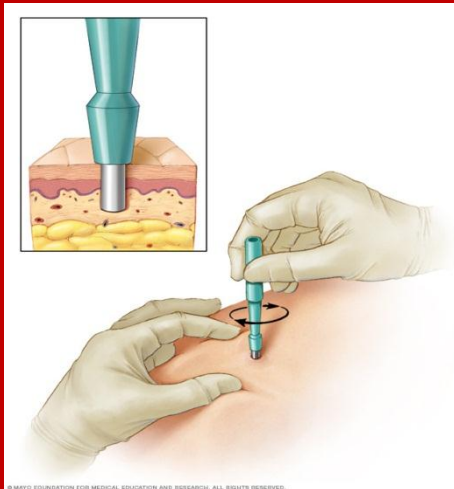
Pulmonary-
Renal Pattern

General symptoms:

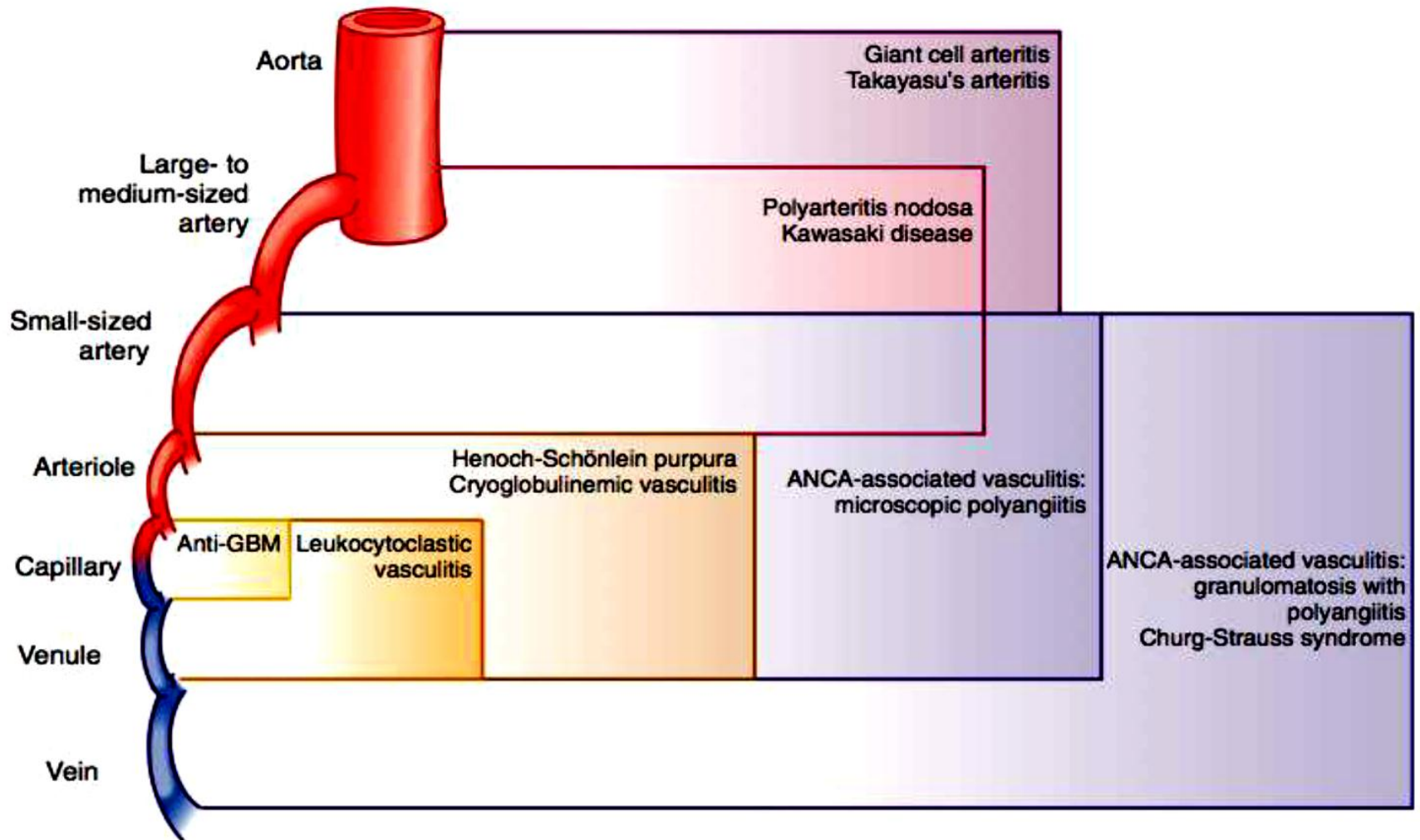
- fever
- headache
- weight loss

2 helpful diagnostic tests

1. serologic testing help in diagnosis in minority
2. biopsy often necessary



Classification of vasculitis depend of mostly involved vessel size



Classification

- by vessel size :
- a. Large vessel:
 - (Giant cell arteritis)aorta and temporal artery.
 - takayasu 's arteritis. Begingingin of branches out of aorta.
- b. Medium vessel:
 - Polyarteritis nodosa (PAN)

Small vessel:

(1) Immune-complex mediated:

(a) anti-glomerular basement membrane (anti-GBM) disease.

(b) Henoch-Schonlein purpura (HSP).

(2) ANCA-associated

(a) granulomatosis with polyangiitis.

(b) eosinophilic granulomatosis with polyangiitis.

Henoch-Schonlein purpura (HSP)



Henoch Schonlein purpura (HSP)

- **Henoch Schonlein purpura (HSP) is an immune mediated disease associated Immunoglobulin A (IgA) deposition within the affected organs**
- **common in children between the ages of 2 and 6 years.**
- **Skin The rash develops mainly on the buttocks, legs and feet.**
- **Arthritis mainly ankles and knee**
- **intestines:abdominal pain or bloody stools May lead to intussusception**
- **kidneys. Ig A GLOMERULONEPHRITIS.**

Polyarteritis Nodosa PAN

- General
 - hepatitis B antigen in 30% of pt with PAN
 - PAN in 1-5% of hepatitis B cases.
 - medium arteries, aneurysms
 - Hypertension
 - Proteinuria and neuropathy
 - Skin necrotic vasculitis

Skin



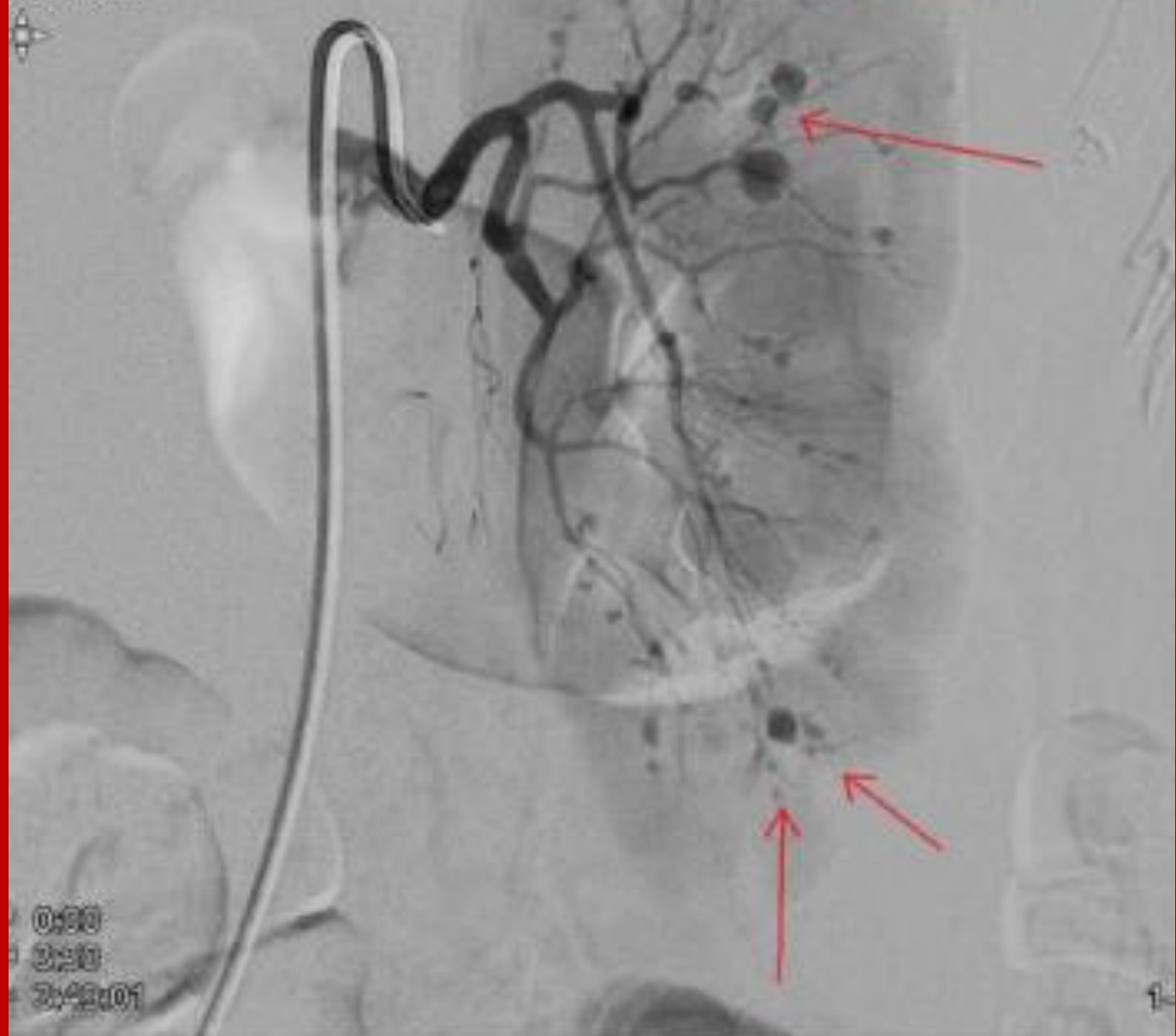
Renal angiogram



47-year-old man with abdominal pain, weight loss, and an elevated ESR

Right renal arteriogram reveals multiple microaneurysms within the upper pole of the kidney on this selective right renal artery injection (upper pole branch)

mi 0°
rg 0°
D 27 cm



0:00
3:20
3:42:01

Polyarteritis Nodosa

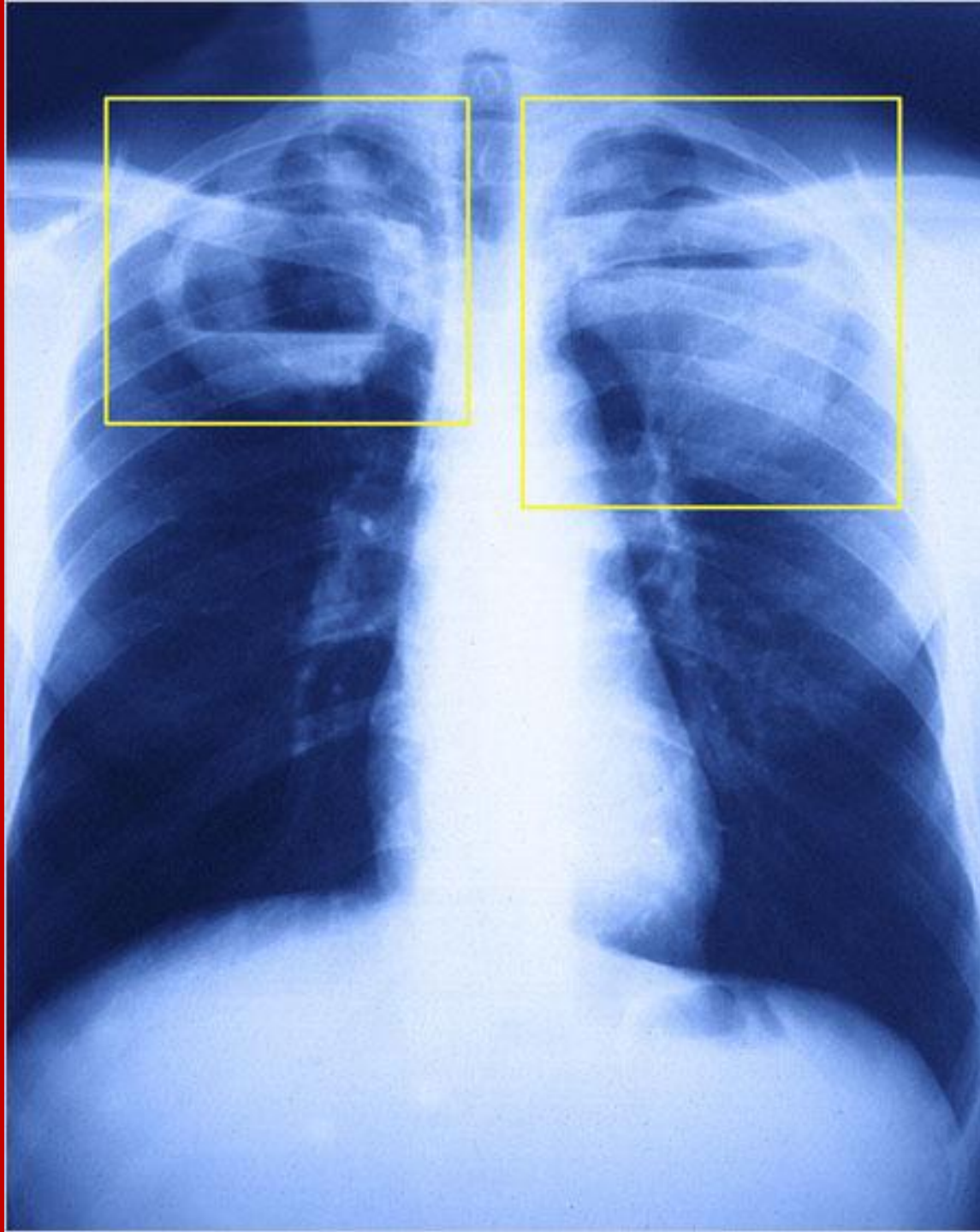
- **Biopsy of skin or peripheral nerve**
Visceral angiography
- RX:Corticosteroids; cytotoxic drugs

granulomatosis with polyangiitis(Wegener's Granulomatosis)

- Upper airway
 - epistaxis, **NECROTIC(BLOODY)** recurrent sinusitis
- Lower airways
- (lung nodules or cavitation)
- Glomerulonephritis.

Dx of granulomatosis with polyangiitis.

- antineutrophil cytoplasmic antibody (c-ANCA)
- and
- **biopsy** OF INVOLVED ORGAN
 - Treatment
 - cyclophosphamide 2 mg/kg plus prednisone 1 mg/kg



eosinophilic granulomatosis with polyangiitis Churg-Strauss Syndrome

- triad of systemic vasculitis, asthma, and eosinophilia
- peripheral eosinophilia up to 74%, lung infiltrate and high ESR.
- RX STEROIDS

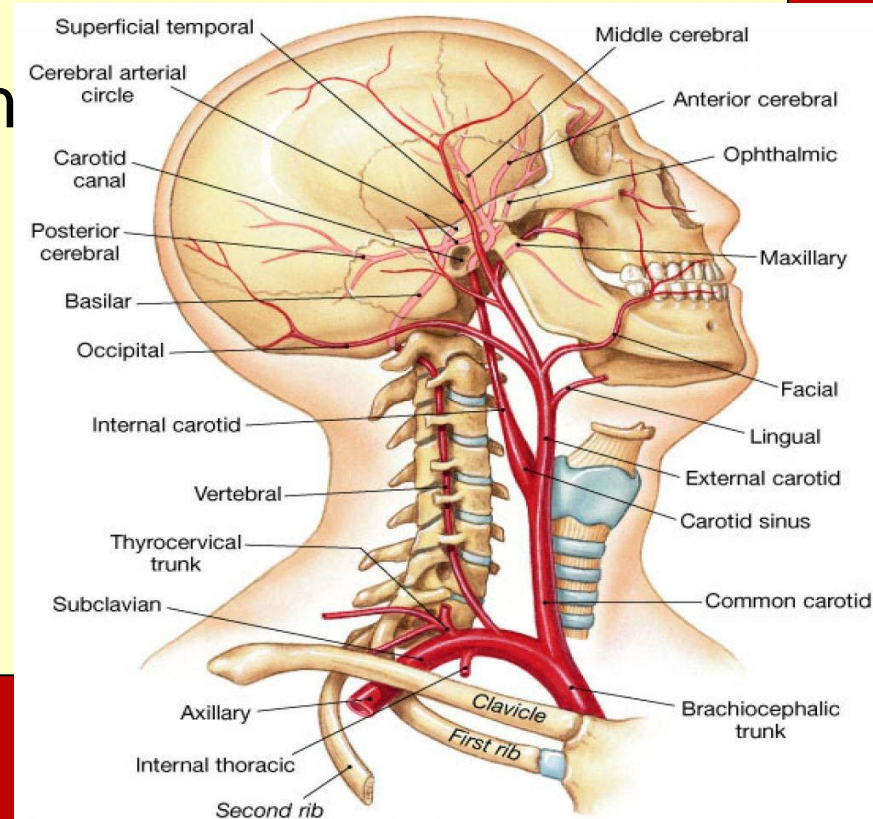
Giant Cell Arteritis

- headache (old patient with constant, boring)
 - temporal artery tender, erythematous 50%
 - scalp tender
 - jaw claudication 50% pain with chewing

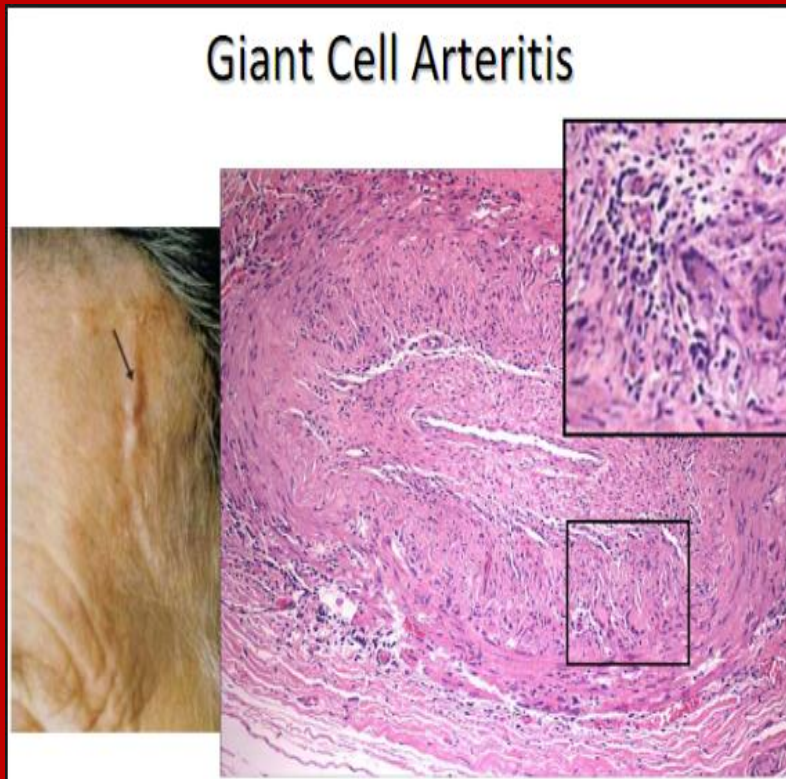


Giant Cell Arteritis

- **blindness** > 1/3 untreated patients
- ESR usually > 50 mm/h
- Dx confirmation
- **temporal artery biopsy**



Temporal artery biopsy



Polymyalgia Rheumatica

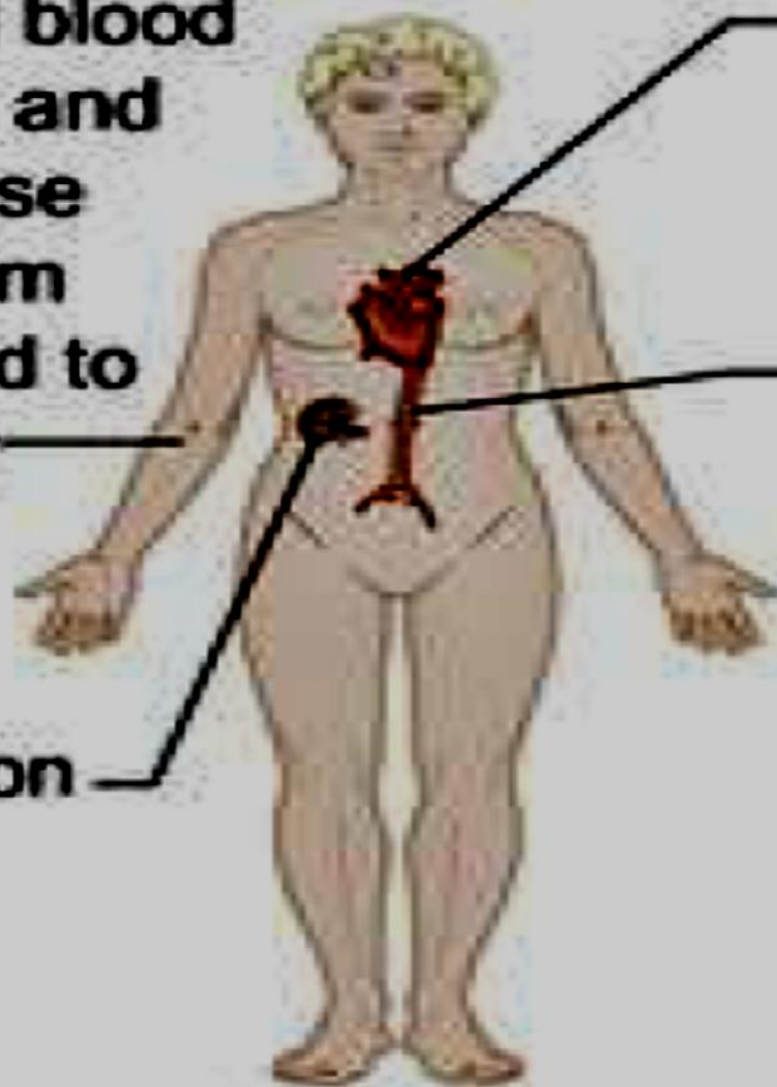
- accompanying syndrome in 50% temporal arteritis
- clinical syndrome
 - Age more than 50 y
 - morning stiffness proximal muscles pain
 - increased ESR
 - no inflammatory joint or muscle disease*****
 - low grade fever, weight loss, malaise

○ Treatment

- Polymyalgia rheumatica
 - Prednisolone 10-15mg daily tapered over 9-12 months
- Temporal arteritis
 - Prednisolone 40-60mg daily tapering over 12-18 months

Reduced blood pressure and weak pulse in one arm compared to the other

Renal dysfunction



Aortic insufficiency

Inflammation of aorta and large branches

Takayasu's arteritis

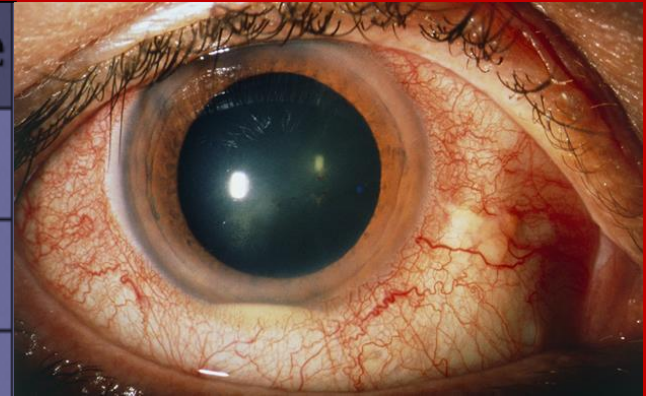
Behcet's Disease

- Vasculitis with triad
 - oral, genital ulcers, ocular (inflammation uveitis or iritis)
 - oral
 - aphthous-like
 - painful, clusters on lips, gingiva, buccal, tongue



Table 39–1. Frequency of Clinical Manifestations of Behçet Disease

Feature	Frequency (%)
Oral ulcers	100
Genital ulcers	75
Skin lesions	60–90
Arthritis	50
Gastrointestinal disease	25
Thrombophlebitis	20
Central nervous system disease	10–20

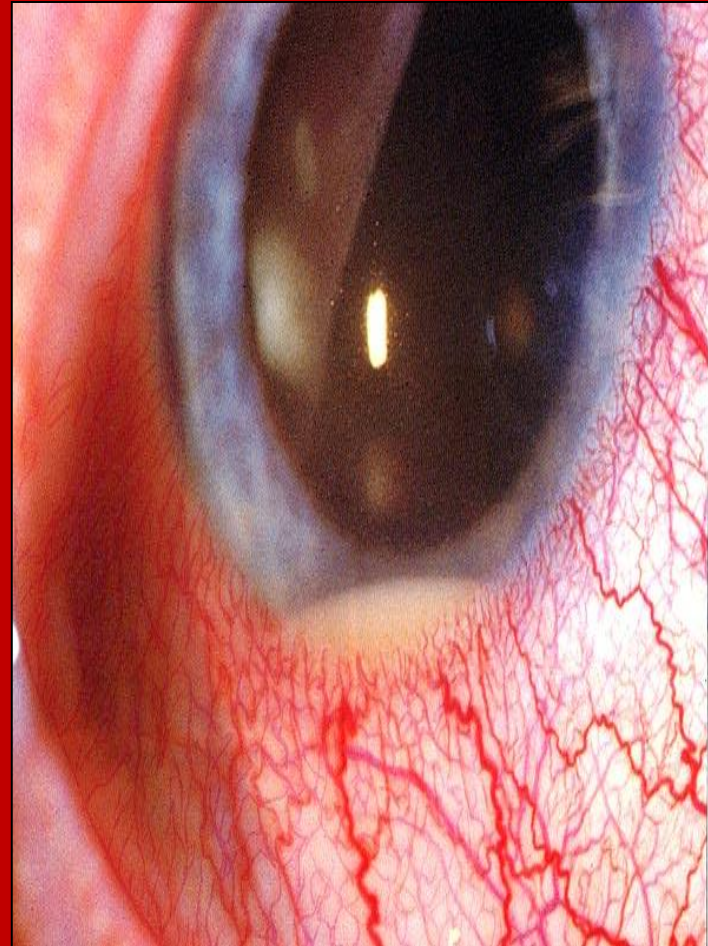


Increase incidence of dvt

Behcet's Disease

eye

- uveitis, iritis
- Hypopyon=
- Accumulation of exudate in anterior chamber.



Behcet's Disease

- other findings
 - associated with HLA-B51
- Treatment:
 - _local steroids,colchicine,systemic steroids
 - azothioprine, cyclosporin,cyclophosphamide.



Pathergy test

exaggerated skin injury occurring after minor trauma such as bump, bruise, needle stick injury.

Complications

- Blindness
- Paralysis
- Embolism/thrombosis - pulmonary, vena cava, peripheral DVT
- Aneurysms



Hemorrhagic purpuric lesions



- Blood dis - DIC –anti phospholipid lanti body syndome
- infection sabe-septic emboli-
- Inflammn vasculitis
- tumors –
- Metabolic cholestrol emboli etc

-
- Erythrocyte sedimentation rate and C-reactive protein are increased
 - Complete blood count with white cell differential demonstrates anaemia, leucocytosis, eosinophilia and thrombocytosis
 - X-ray of the sinuses demonstrates shadowing or fluid levels
 - Chest X-ray demonstrates pulmonary infiltrates
 - Urinary sample demonstrates haematuria
 - ANCA suggest small artery vasculitis
 - Tests are ordered based on the clinical picture and they can also be used to demonstrate the triggering or perpetuating factor in secondary vasculitis and to exclude pseudovasculitis

- Thank u