

AMERICAN OSTEOPATHIC SOCIETY OF RHEUMATIC DISEASES

- VASCULITIS 101
- presented by
- Robert R Speer DO,FAOSRD

VASCULITIS

- SMALL, MEDIUM, LARGE VESSELS---
- GRANULOMITIS DISEASE WITH VESSEL INFLAMMATION
- GRANULOMATIS DISEASE WITHOUT VESSEL INFLAMMATION
- NECROTIZING VASCULITIS

Symptoms in vasculitis

- Fever
- Headache
- Fatigue
- Weight loss
- General aches and pains
- Night sweats
- Rash
- Nerve problems, such as numbness or weakness

Skin changes

- Palpable purpura
- Necrotic ulcers
- Urticaria
- Nodules
- bullae

Causes of vasculitis

- Smoking
- Having chronic hepatitis B or C infections
- Having some types of autoimmune conditions, including rheumatoid arthritis, scleroderma or lupus

testing

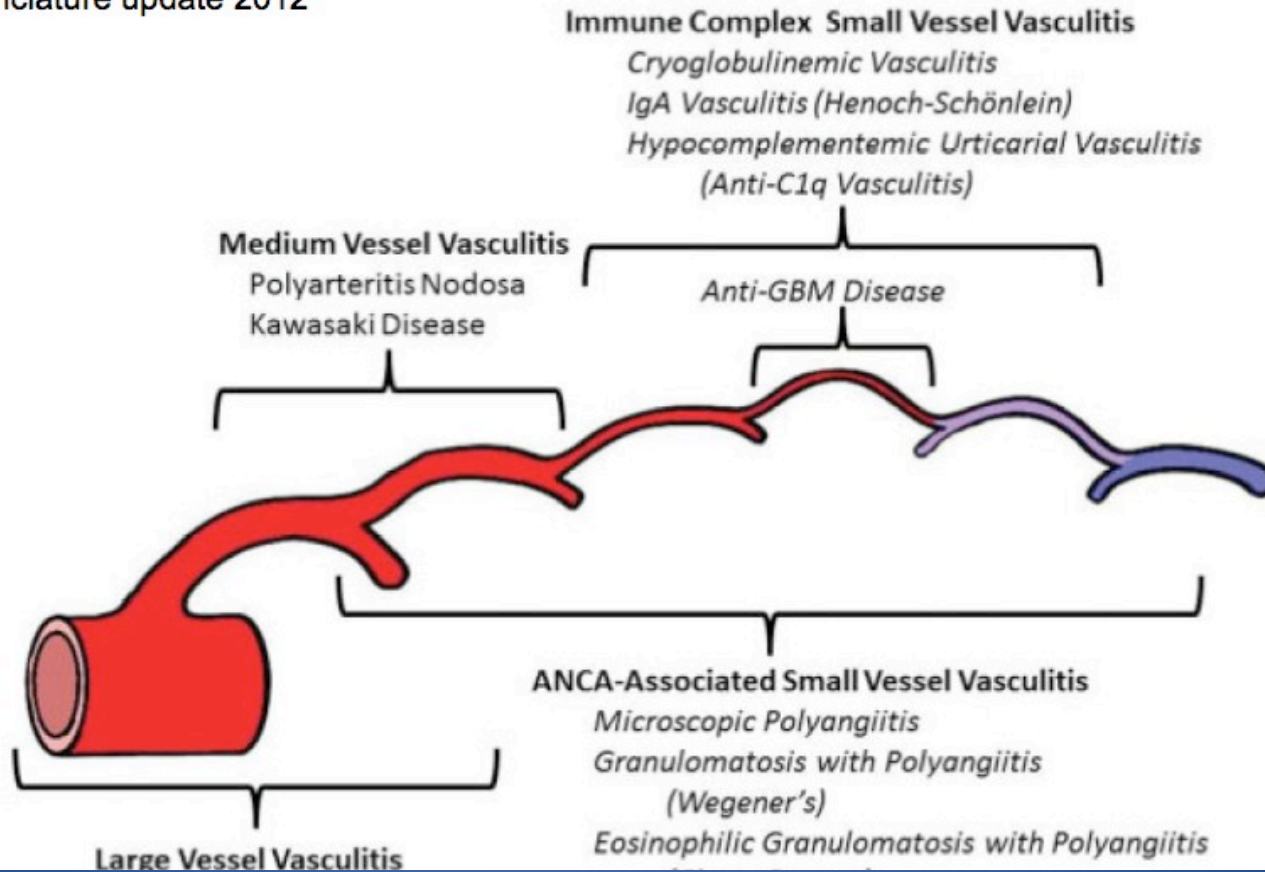
- Cbc, ANA, RA, CCP, CRP, Sedrate, Chem profile Uric Acid , ANA cascade, C3C4 compliment, hepatitis profile, urinalysis, anca (c&p), cryoglobulins, immune globulins A and E, HIV, lyme ab.
- Ultrasound, MRI, MRA, angiography, X-ray
- Biopsy

treatment

- . Corticosteroids
- . Methotrexate
- .cyclophosphomide
- . Sulfa drugs
- .biologics (tnf-inhibitors, cd-20 blockers(rituxan)
- .aspirin
- . Immune globulins

Arteritis levels

Chapel Hill Consensus Criteria
Nomenclature update 2012



SMALL VESSEL DISEASE

- Hypersensitivity angiitis-related to a specific event
- Serum sickness at one end of the spectrum
- Henoch-Schoenlein purpura at the other end
- SKIN IS MOSTLY INVOLVED
[leukocytoclastic vasculitis]

Buerger's disease

- **Buerger's disease.** This condition causes inflammation and clots in the blood vessels of your hands and feet, resulting in pain and ulcers in these areas. Rarely, Buerger's disease can affect blood vessels in the abdomen, brain and heart. It is also called thromboangiitis (throm-boe-an-jee-l-tis) obliterans.

Buerger's disease



Hypersensitivity vasculitis

- **Hypersensitivity vasculitis.** Sometimes called allergic vasculitis, the primary sign of this condition is red spots on your skin, usually on your lower legs. It can be triggered by an infection or an adverse reaction to medicine.

Palpable purpura vasculitis



HSP

- **Henoch-Schonlein purpura (IgA vasculitis)**. This condition is more common in children than in adults, and causes inflammation of the smallest blood vessels (capillaries) of your skin, joints, bowel and kidneys. Signs and symptoms include abdominal pain, blood in the urine, joint pain, and a rash on your buttocks or lower legs.

Common findings in HSP

- Abdominal pain
- Joint pain
- Palpable purpura
- Elevated ana titers, Ra factor, IgA, ASO titer
- Renal involvement
- Males 3:1 ratio
- First and second decades of life most common

Palpable purpura

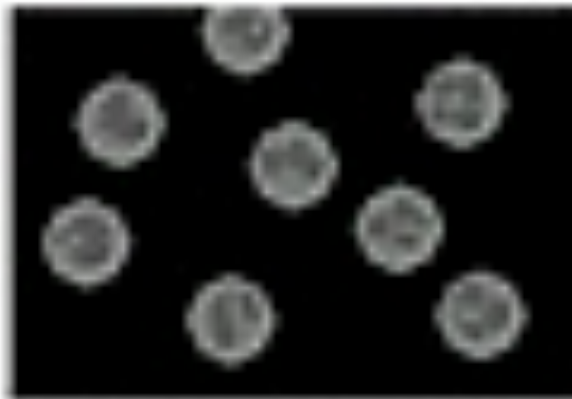


Wegener's granulomatosis

Easy to diagnose and treat – if you think of it.

Wegener's is infamous for its subtle presentation, and its lethality if (and only if) missed.

Caused by autoantibodies against proteinase 3.



Positive anti-neutrophil



Granulomas and patchy necrosis in

sore eye?
sore ear (outer?
middle? inner?)
stuffy nose?



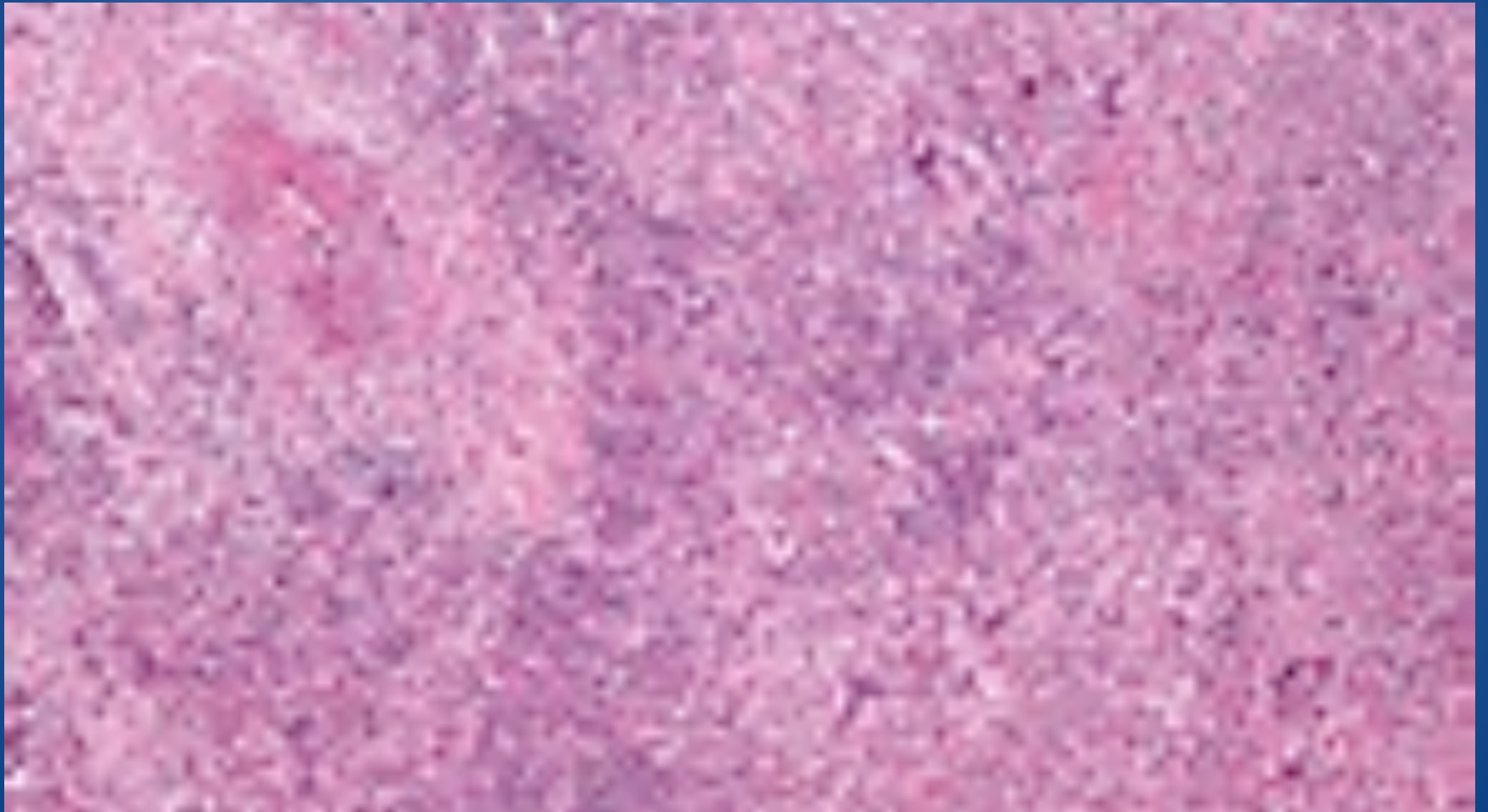
chest
x-ray
blip?

sore joint?

trace of
blood?

destruct
of the
lung cavities
& sinuses
kidney failure

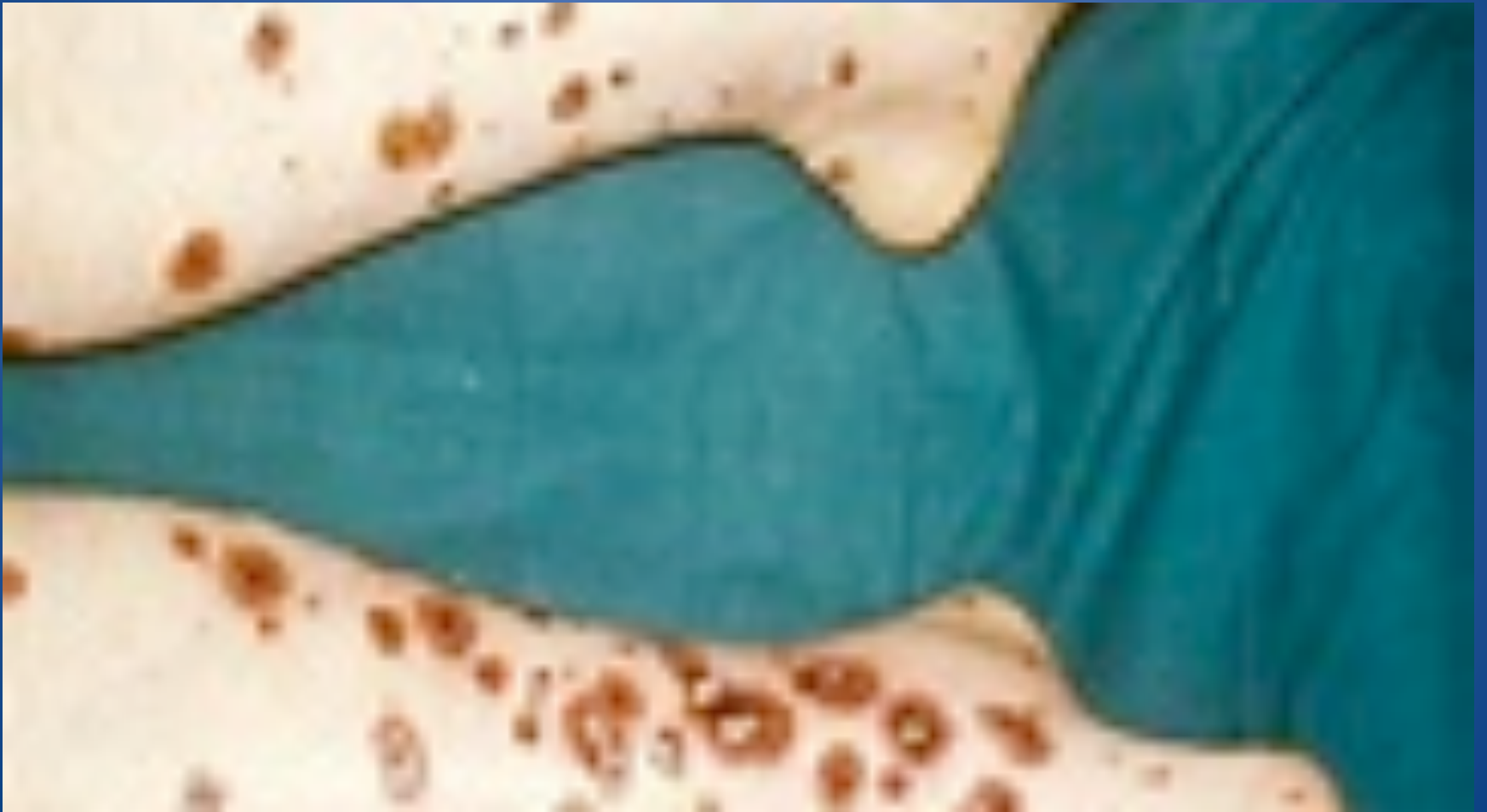
Blood vessel occlusion



Granulomatous eye involvement



Granulomatitis vasculitis



Pulseless disease

- **Takayasu's (tah-kah-YAH-sooz) arteritis.** This form of vasculitis affects the larger arteries in the body, including the aorta. Signs and symptoms include joint pain, loss of pulse, high blood pressure, night sweats, fever, general malaise, appetite loss, headaches and visual changes.

Cardiac involvement



Polyarteritis nodosa

- Small and medium sized arteries affecting all three layers [common at the bifurcation]
- Affects all ages birth to old age with a male 3:1 ratio
- Onset can be acute or insidious with fever, htn, renal disease, abdominal pain
- Lung is notably spared
- eosinophilia

PAN

- Associated with Hep B antigen and antibody
- Anuerysmal dilation of medium sized arteries {mesenteric}
- Major mortality associated with renal failure, GI bleeding, cerebral bleeding

Poly arteritis nodosa subgroup in children

- KAWASAKI DISEASE [Mucocutaneous lymph node syndrome]
- Presents as high fever, conjunctival congestion, dry mouth with fissures, red strawberry tongue
- Rash—erythema multiforme, purpura, morbilliform in nature
- Elevated IgE, CRP, decrease complement
- Large joint arthritis in 25% of children
- Mortality from coronary artery disease

MCLNS

- **Kawasaki disease.** This condition most often affects children younger than age 5. Signs and symptoms include fever, rash and redness of the eyes. It is also called mucocutaneous lymph node syndrome.

MCLNS



MCLNS



MCLNS



Giant Cell Arteritis

- First- temporal arteritis with or without poly myalgia rhuematica
- Second- takayasu arteritis [pulseless disease, aortic arch arteritis]
- Female 8:1 ratio
- Renal vascular hypertension, cns manifestations, aortic insufficiency,
- Migratory arthritis in 25% of cases

GCA

- **Giant cell arteritis.** This condition is an inflammation of the arteries in your head, especially at the temples. Giant cell arteritis can cause headaches, scalp tenderness, jaw pain, blurred or double vision, and even blindness. It is also called temporal arteritis.

temporal arteritis



cryoglobulinemia

- **Cryoglobulinemia.** This condition results from abnormal proteins in the blood. Signs and symptoms include rash, joint pain, weakness, and numbness or tingling.

Cryoglobulinemia

• How Is Cryoglobulinemia Defined?

• Types Of Cryoglobulinemia?

• Symptoms Of Cryoglobulinemia?



Serum cryoglobulins



Necrotic rash in cryoglobulemia



Occlusion in distal vessels



Cryoglobulin rash and early toe occlusion



Increased cells and
crystalline deposits
is generated by
light microscopy

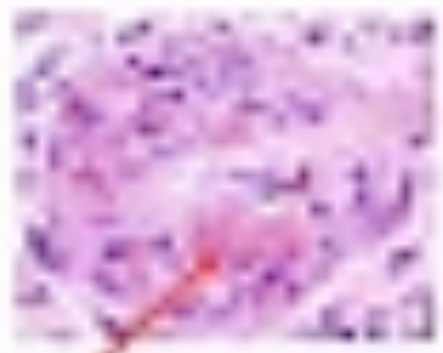


Normal appearance

Crystalline deposits are
generated by
immunofluorescence
microscopy



Crystalline deposits in
renal artery vasculitis



Crystalline deposits

End stage occlusion



- **Microscopic polyangiitis.** This form of vasculitis affects small blood vessels, usually those in the kidneys, lungs or nerves. You may develop abdominal pain and a rash, fever, muscle pain and weight loss. If the lungs are affected, you may cough up blood.

Next in line- arteritis associated with granulomatous disease

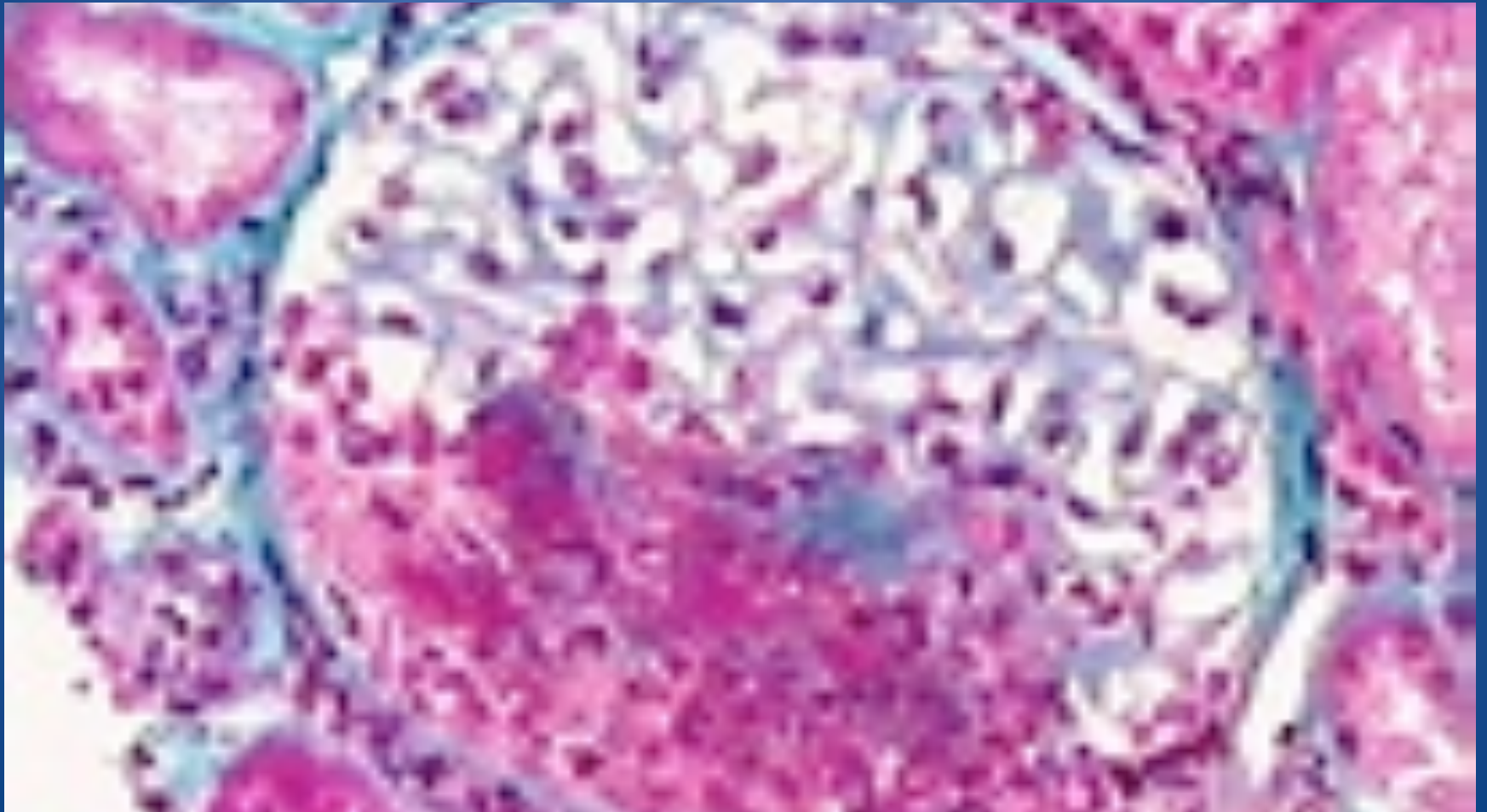
- WEGENERS GRANULOMATOSIS
- the triade of diagnosis
- 1-necrotizing granulomatous lesions of the respiratory tract, sinus-saddle nose deformity-pulmonary infiltrates
- 2-focal necrotizing glomerulonephritis with crescent formation
- 3- necrotizing vasculitis of medium and small vessels, arteriols and venules [separate in space from the granulomas]

Aka wegeners granulomatosis

- **Granulomatosis with polyangiitis.** This condition causes inflammation of the blood vessels in your nose, sinuses, throat, lungs and kidneys. Signs and symptoms include nasal stuffiness, sinus infections, nosebleeds and possibly coughing up blood. But most people don't have noticeable symptoms until the damage is more advanced.

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Vessel in wegeners granulomatosis



Vascular rash in wegeners



Saddle nose in GPA



Necrotic rash in wegeners



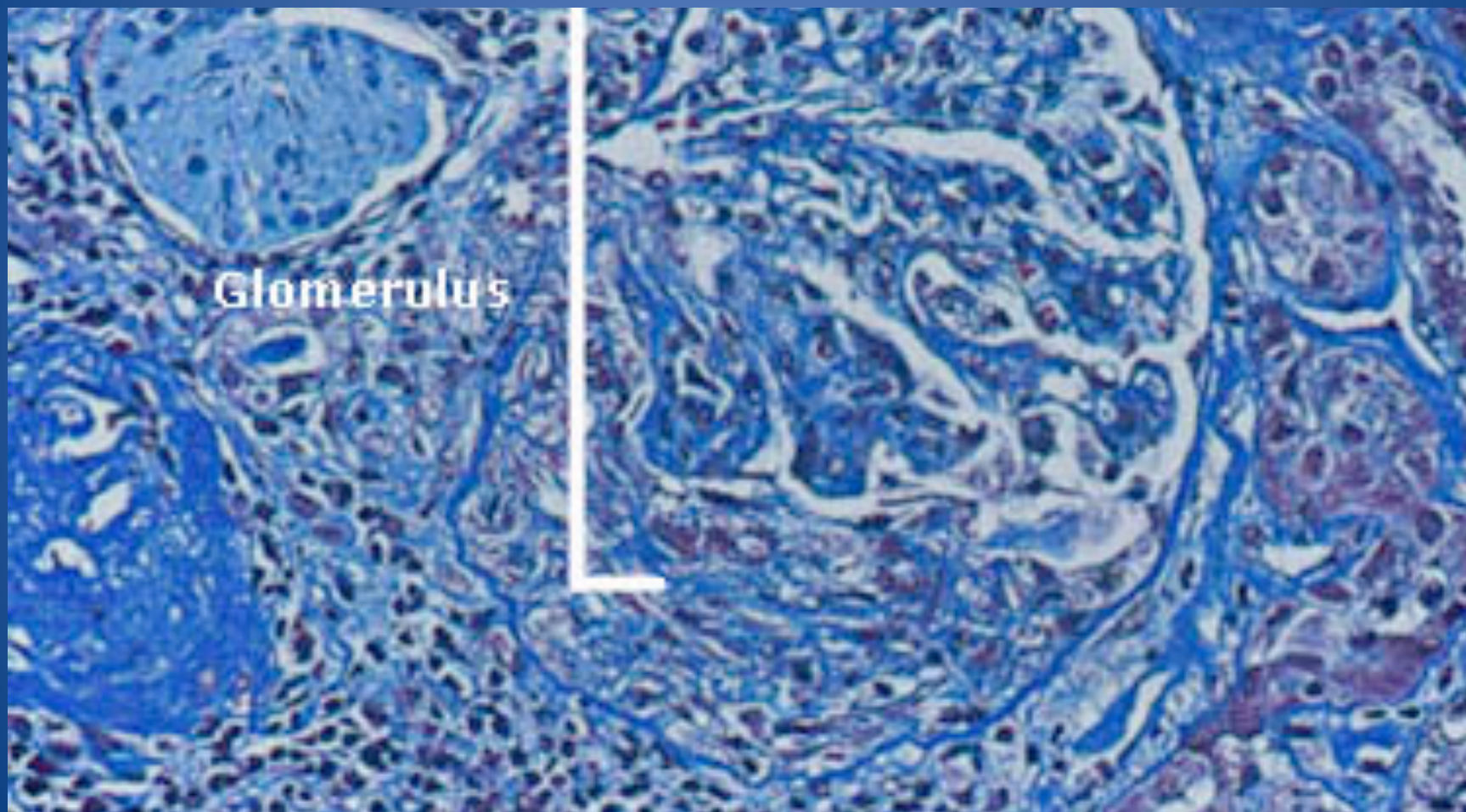
EGP

- **Churg-Strauss syndrome (Eosinophilic granulomatosis with polyangiitis)**. This condition is very rare. It mainly affects the lungs, skin, kidneys, heart and nerves in your limbs. Signs and symptoms vary greatly and include asthma, skin changes, nerve pain and nasal allergies.

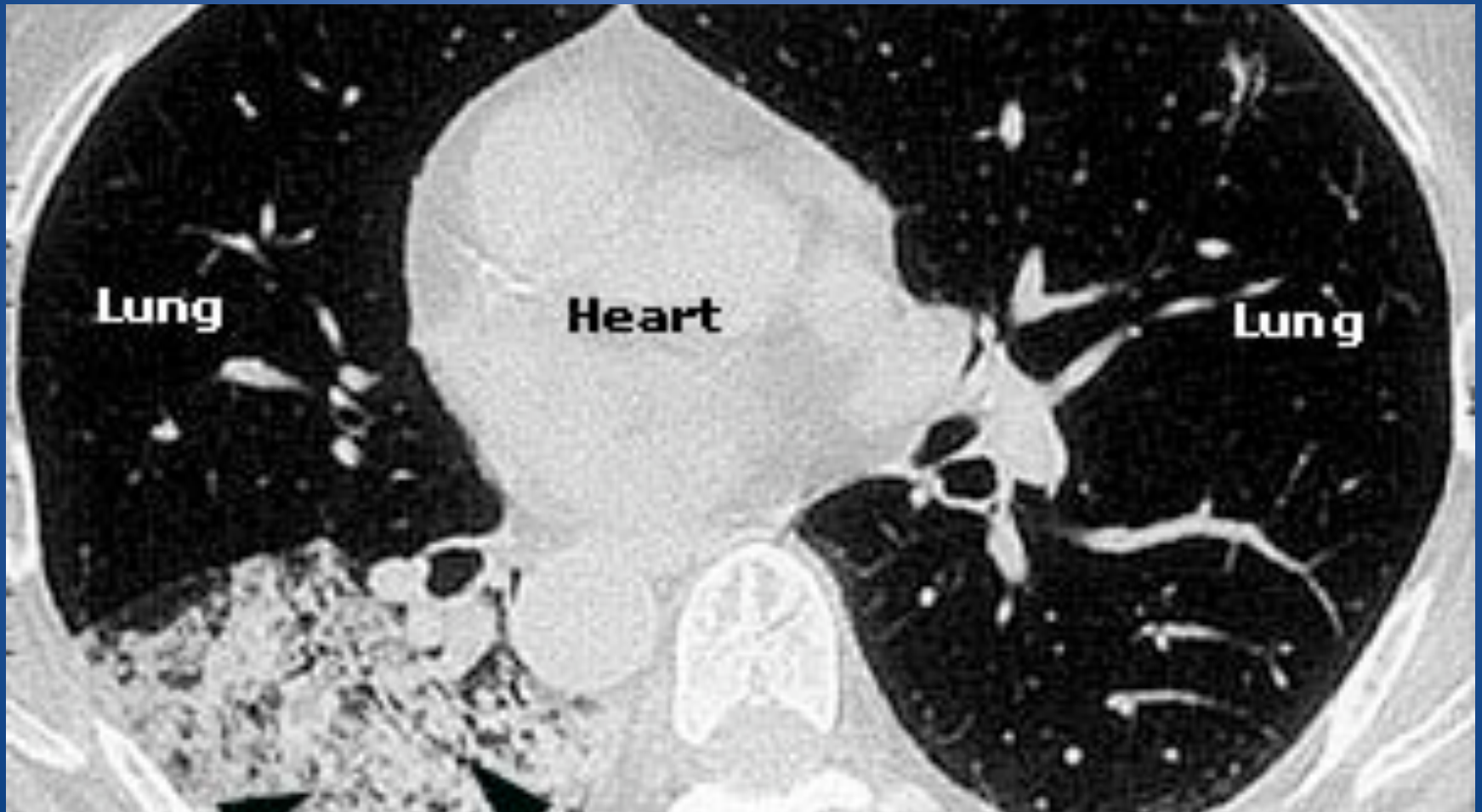
Churg-straus syndrome

- is a *small vessel necrotizing vasculitis*
- classically associated with asthma, allergic rhinitis, lung infiltrates, peripheral eosinophilia, necrotizing granulomas, infiltration by eosinophils.
- extremely rare disorder.
- purpura, GIT bleeding, and renal disease are the major associations.
- Cardiomyopathy (60% of patients) → a major cause of morbidity and death.
- Pathogenesis: p-ANCA associated

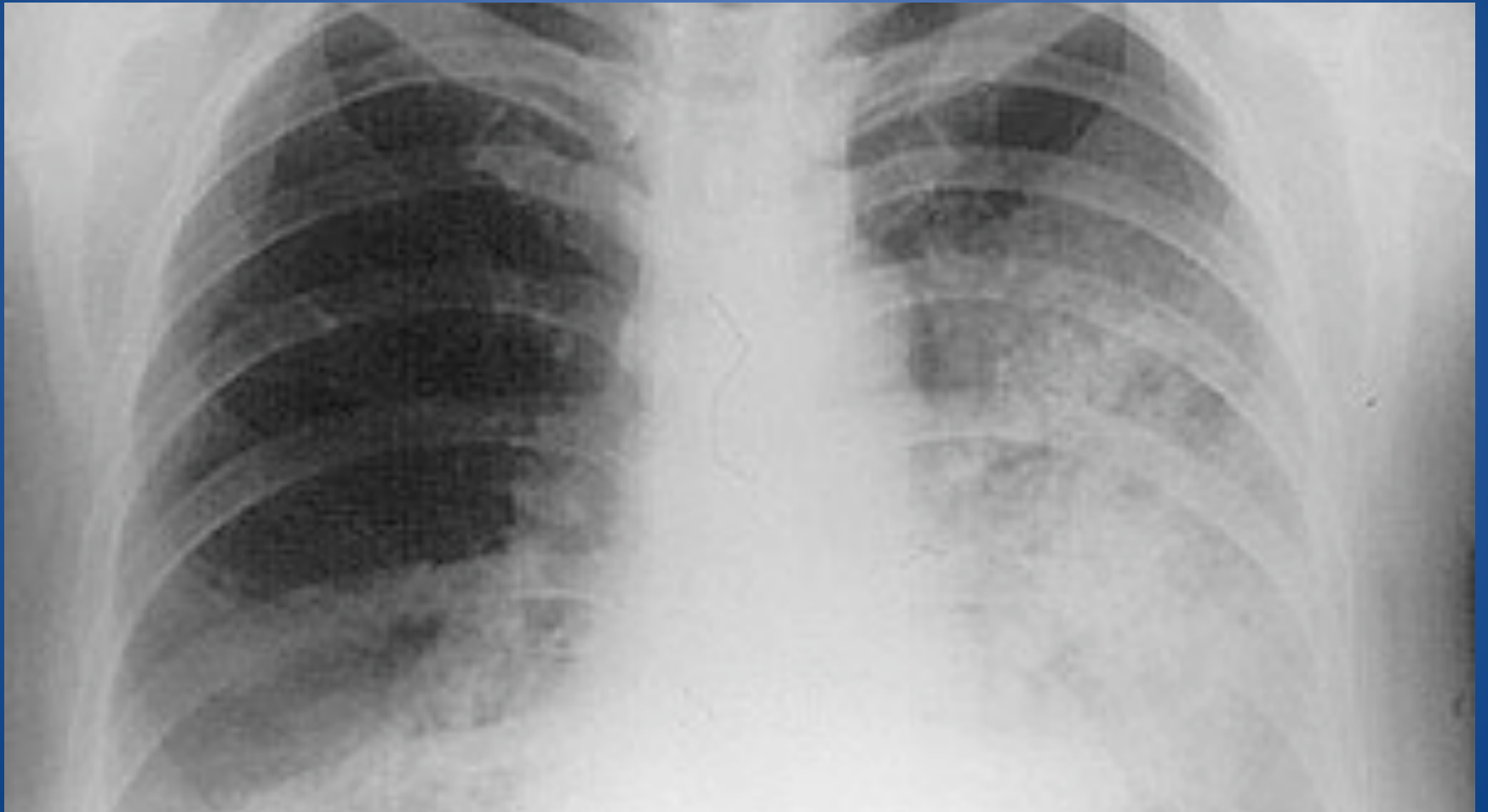
Renal biopsy



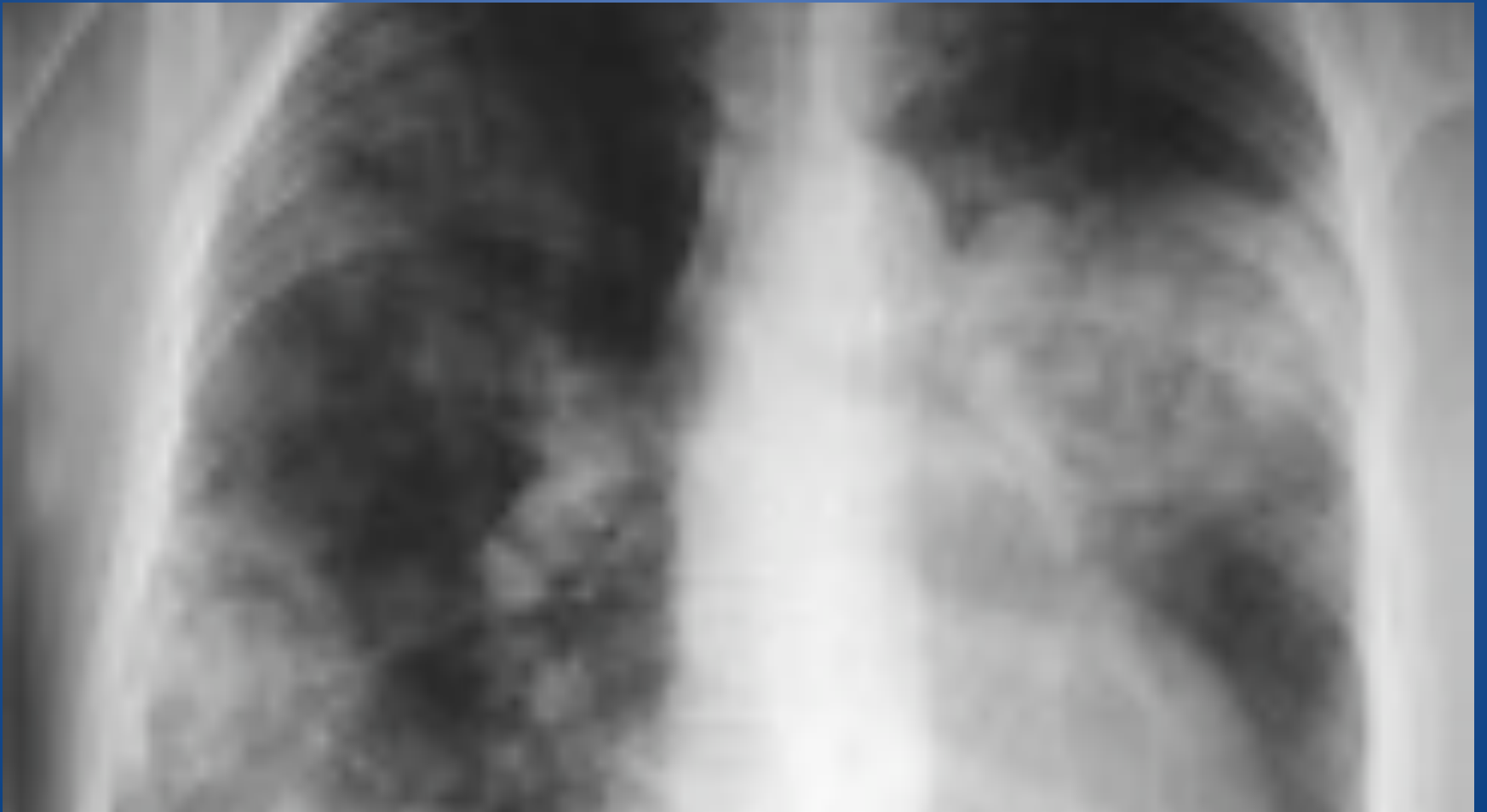
Pulmonary bleeding



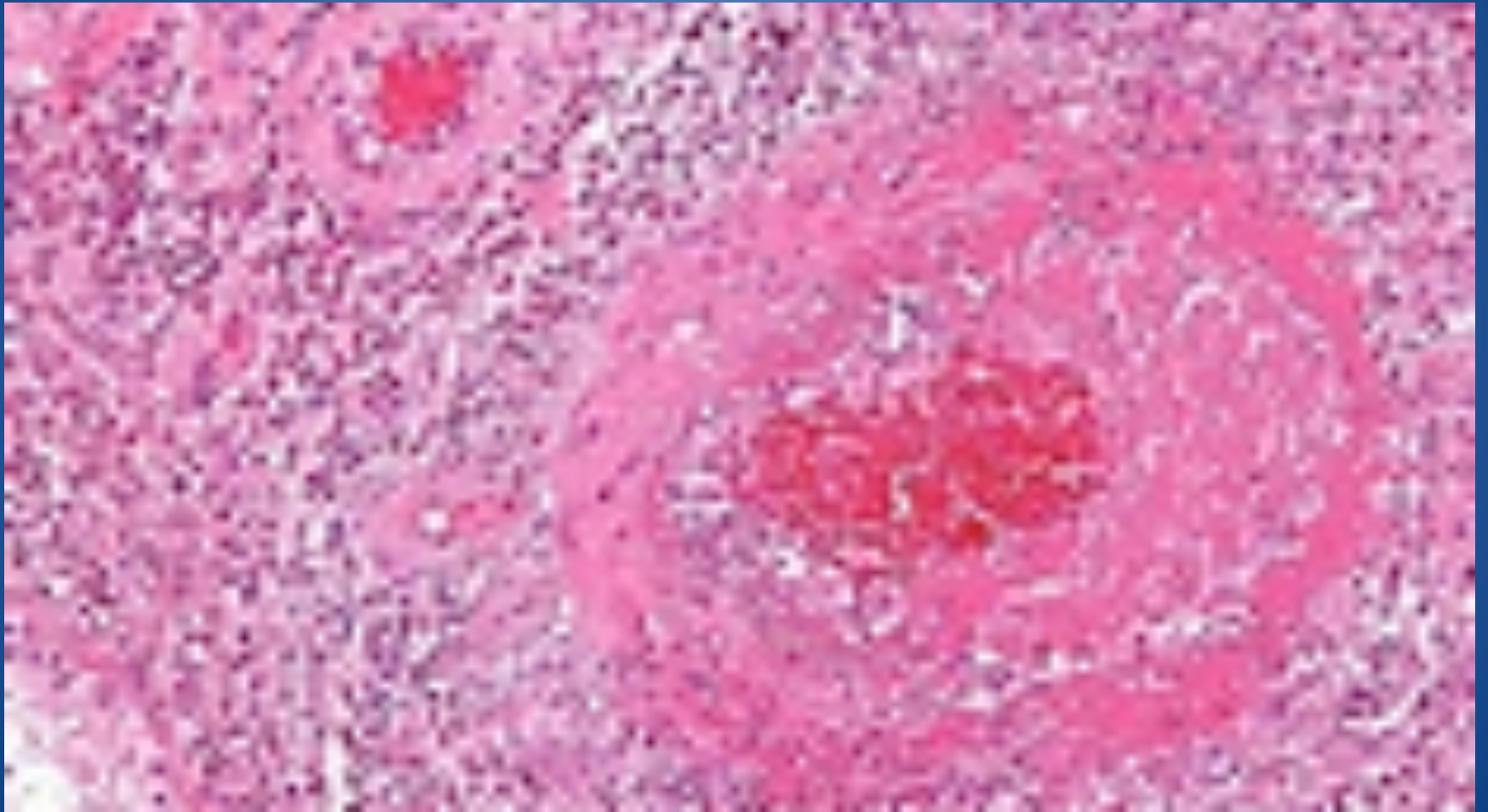
Pulmonary bleed



Lung in churg-strauss



Eosinophilic vessel occlusion



PAN

- **Polyarteritis nodosa.** This form of vasculitis usually affects the kidneys, the digestive tract, the nerves and the skin. Signs and symptoms include a rash, general malaise, weight loss, muscle and joint pain, abdominal pain after eating, high blood pressure, muscle pain and weakness, and kidney problems.

Necrotic rash in PAN

What is Polyarteritis Nodosa?

It is a pathological condition in which the medium and small sized arteries in the body become swollen and are significantly damaged when attacked by immune cells.

For More Information:
Visit: www.epainassist.com



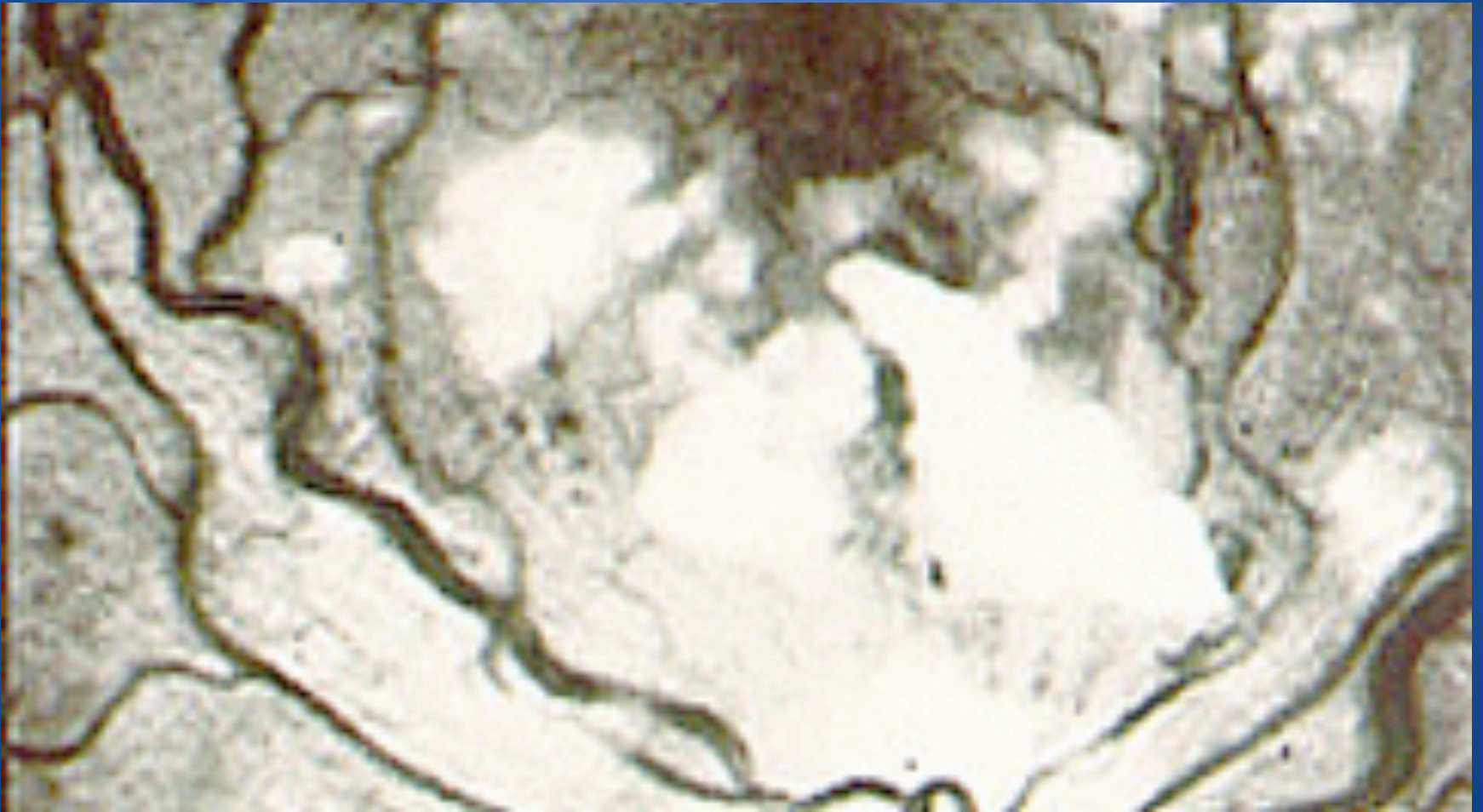
Livedoraticularis rash



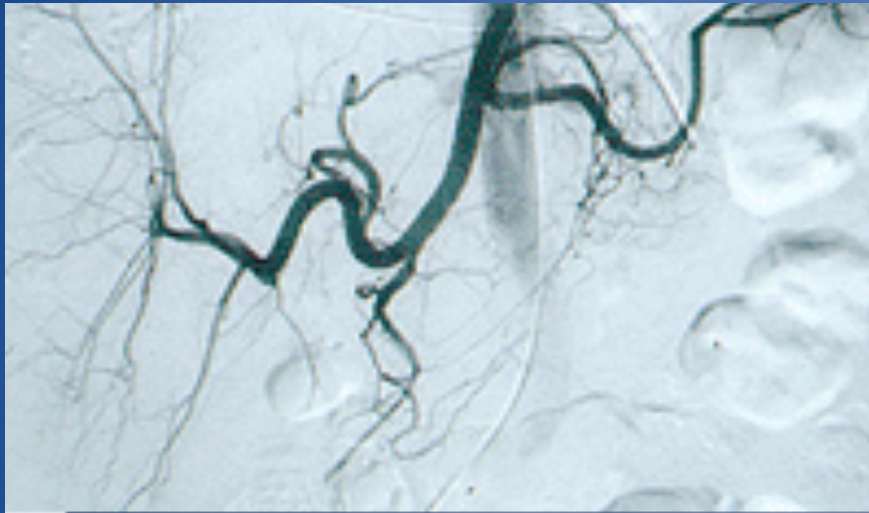
MPA

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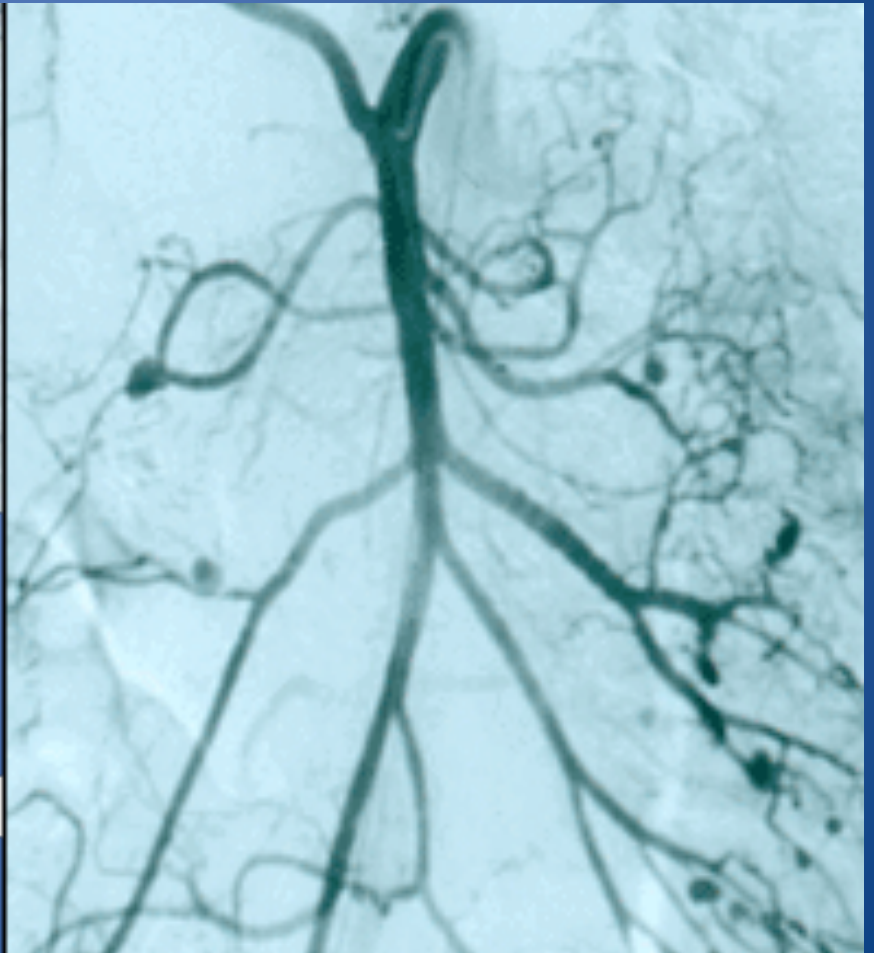
Retinal infarcts



PAN microaneurysm



▲ A normal mesenteric angiogram, demonstrating arteries of normal caliber coming off the aorta.

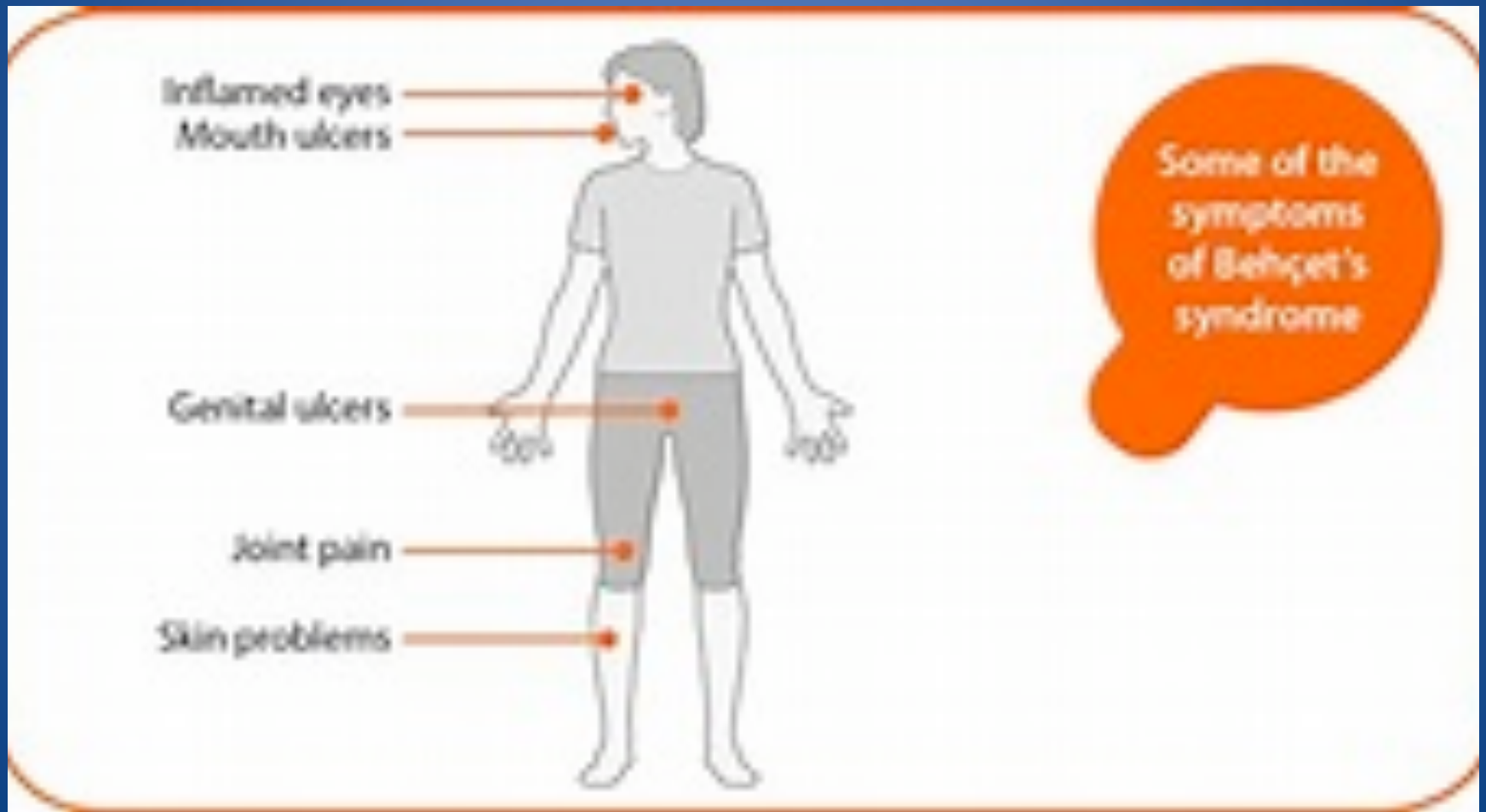


A flagrant abnormal angiogram from a patient with polyarteritis nodosa (PAN). Note the large
▶

Behchets disease

- **Behcet's (beh-CHETS) disease.** This condition causes inflammation of your arteries and veins. Signs and symptoms include mouth and genital ulcers, eye inflammation, and acne-like skin lesions.

Behçet's syndrome







behcets



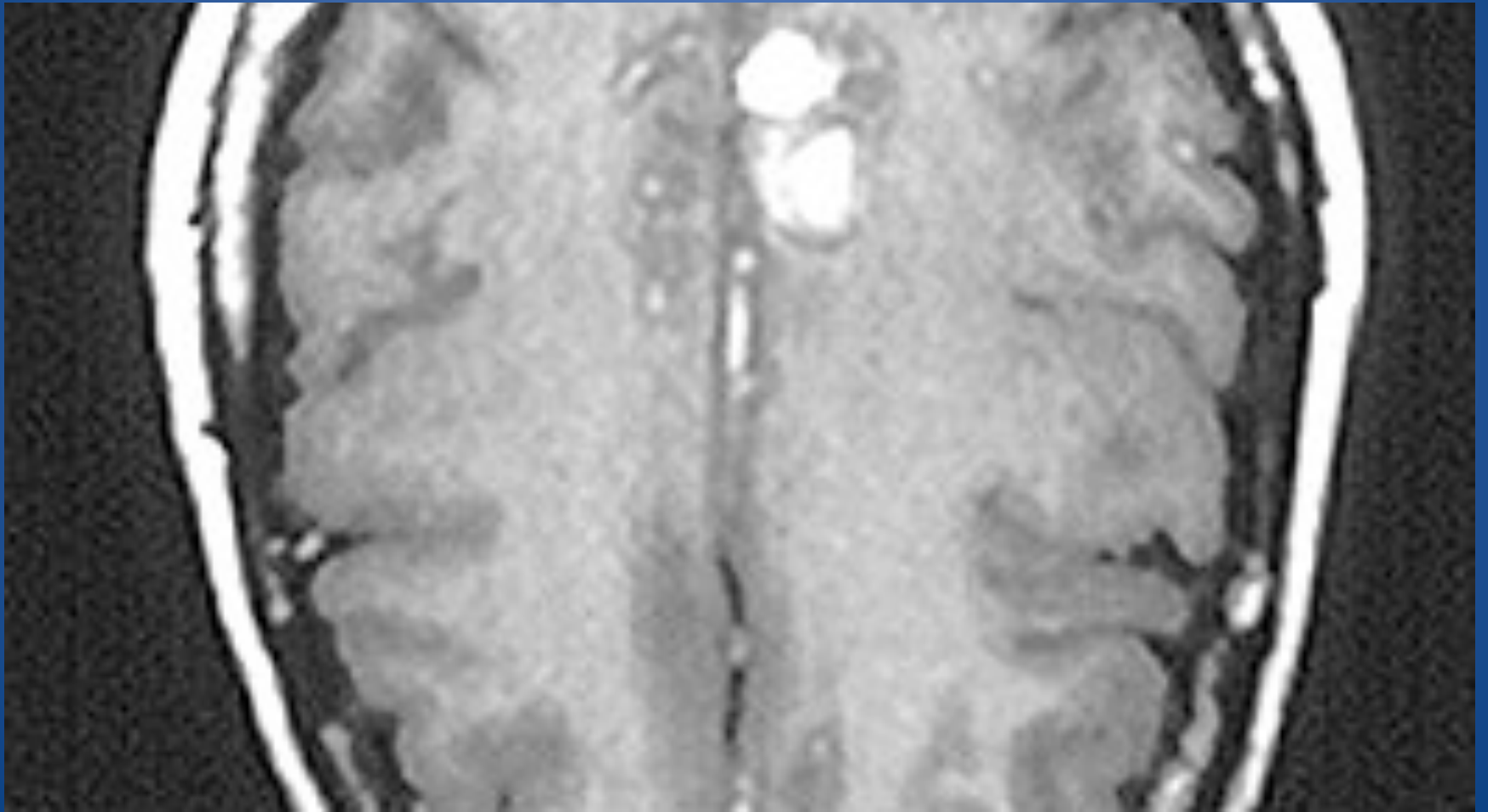
Granulomatous angitis

- Mimics GCA but is intra cranial
- Produces severe neurologic symptoms

Behcets syndrome



Intracranial bleed



Allergic granulomatosis of Churg and Strauss

- More generalized granulomatous angitis
- Spares the upper respiratory tract and kidney
- Positive for chronic allergy
- Positive high titer rheumatoid factor
- Pan and churg-strauss have very similar pathogenic changes except pan is neutrophilic and css is eosinophilic

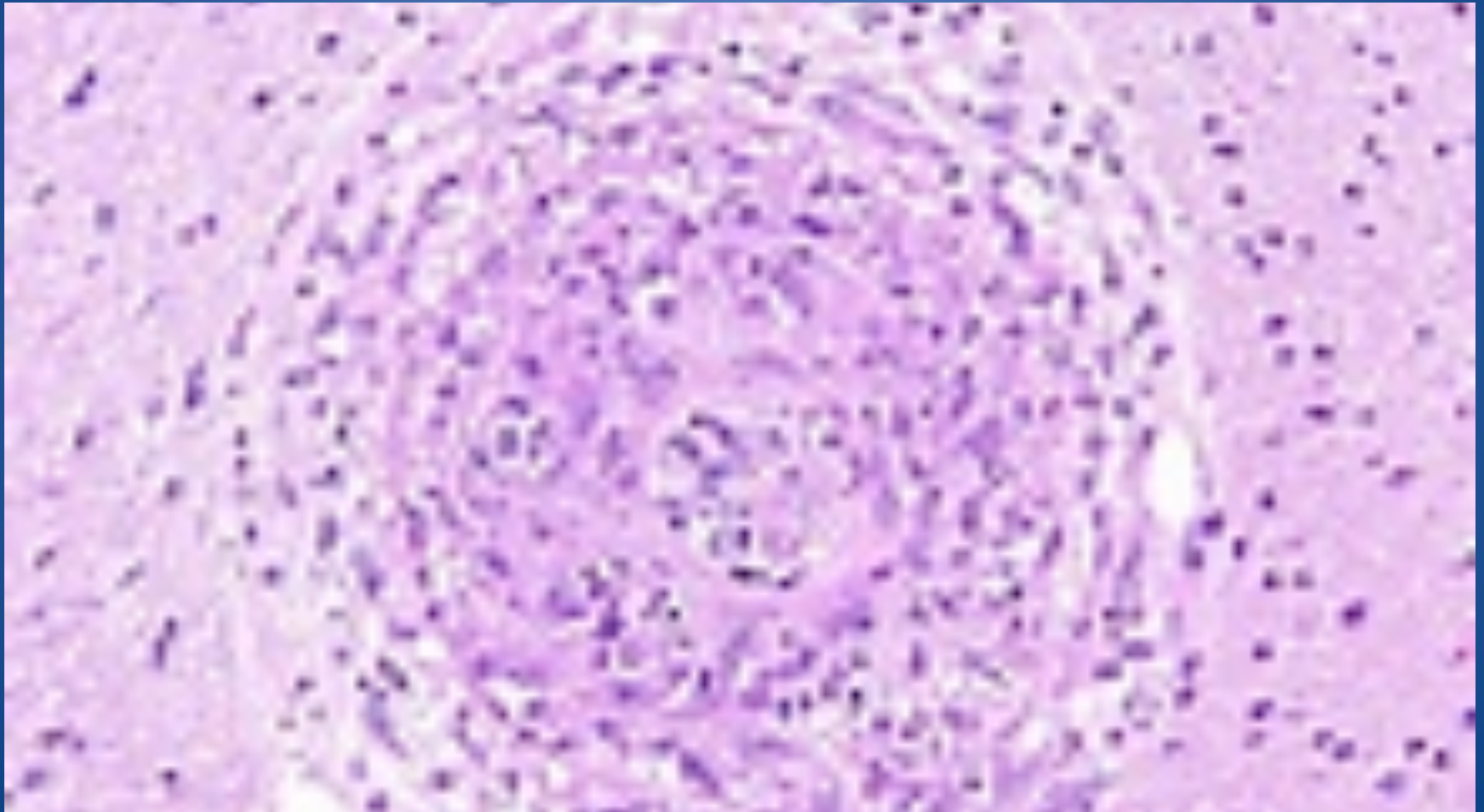
Lymphomatoid granulomatosis

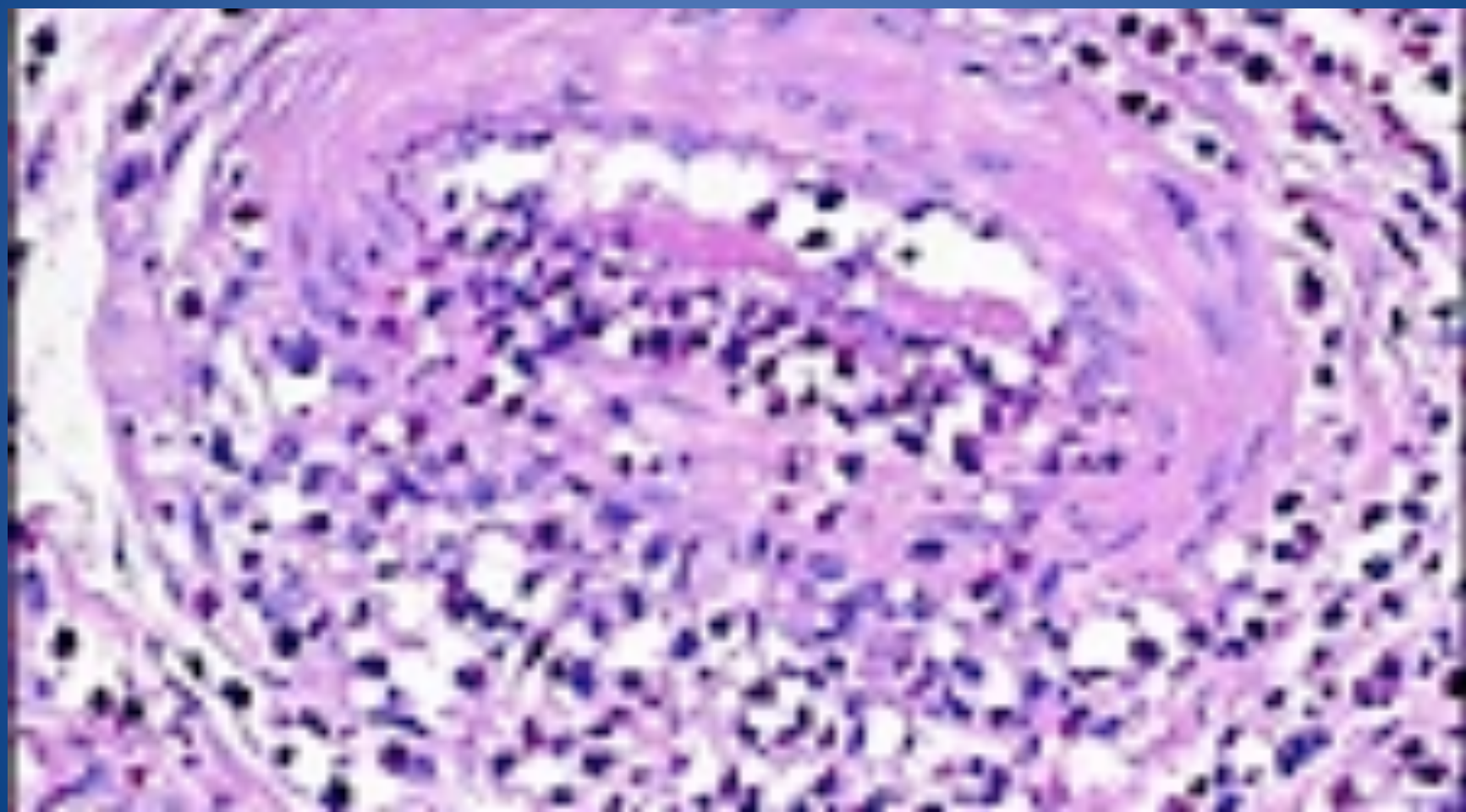
- Vascular necrosis produced in the absence of inflammatory vasculitis
- Can affect lungs ,skin,cns,and the kidney but glomerulonephritis does not occur

Granulomatous angitis rash

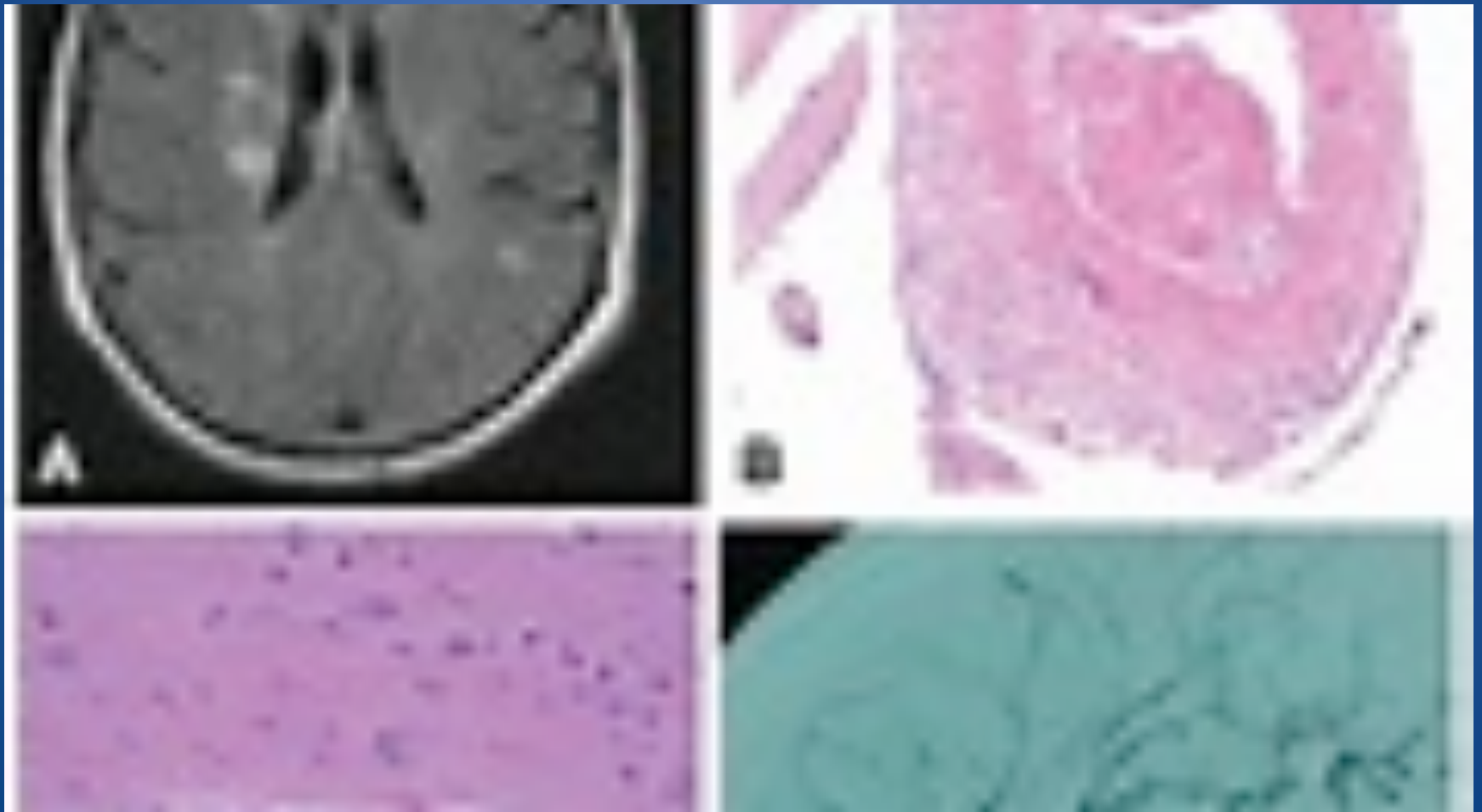


Blood vessel in granulomatous angitis





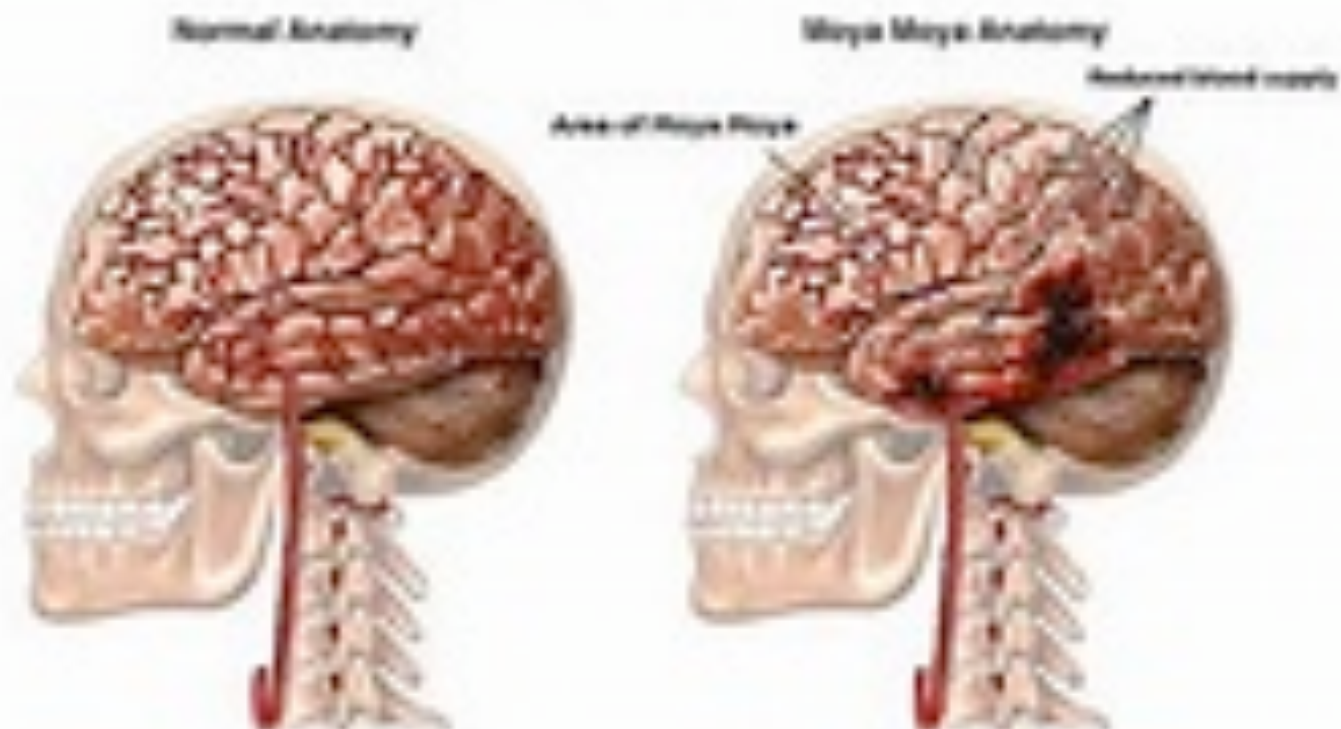
Vessel occlusion brain



Moyamoya disease

- Intimal proliferation with narrowing of the vessel without vasculitis,
- Causes headaches, neurologic complications, leading to hemiplegia
- Main causes are oral contraceptives and ergot derivatives

Moya Moya Syndrome



Moya moya vessel constriction



Moya moya angiogram

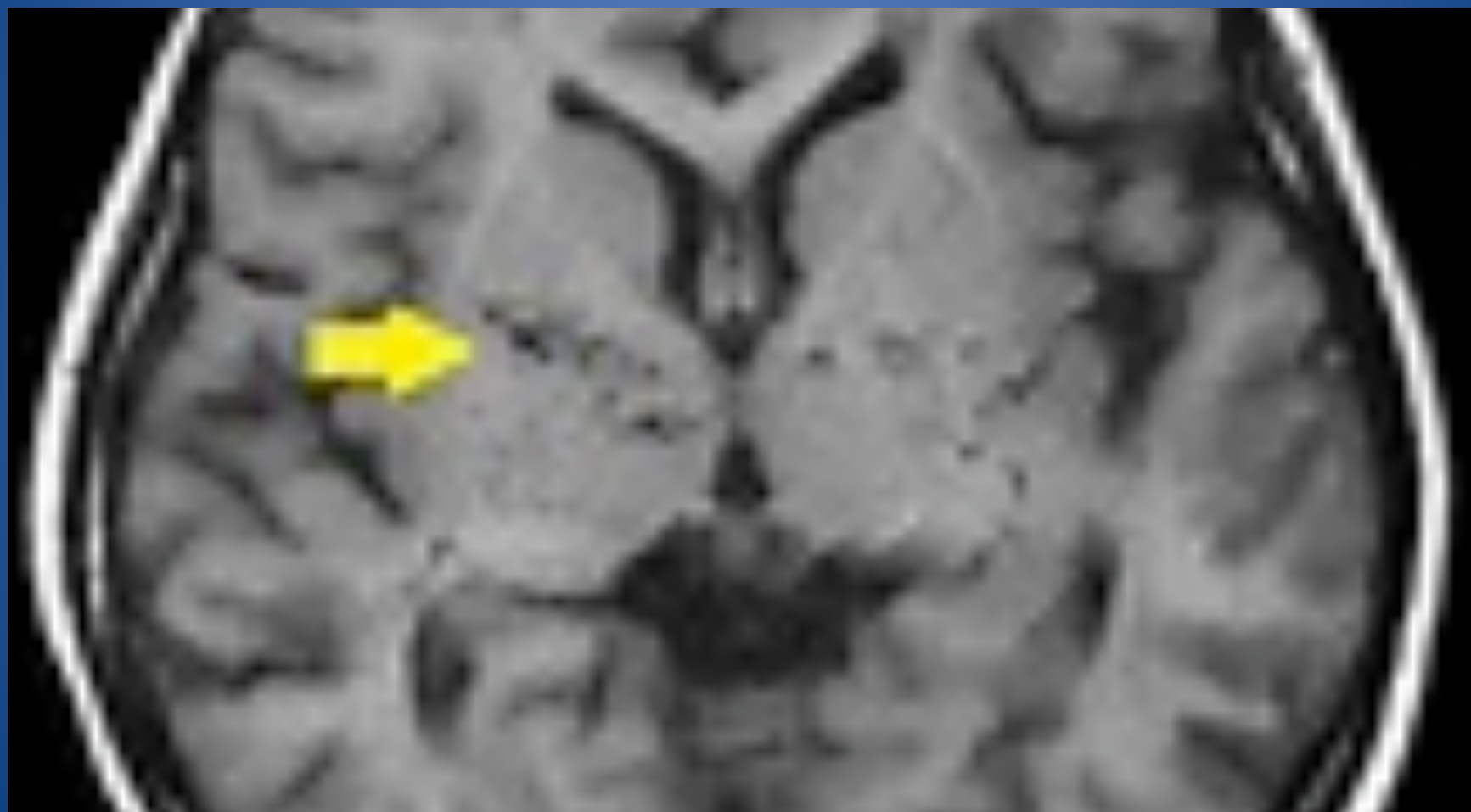


eP eom

Moyamoya Disease

For information,
Visit www.epistemon.com





Summary

- LARGE VESSEL [AORTA,cranial arteries
- examples- GCA, Takayasu, aortitis of ankylosing spondylitis

- MEDIUM SIZED Muscular arteries
- examples- PAN, Kawasaki disease, cogans syndrom

summary

- SMALL muscular arteries and veins
- examples– Churg-Strauss, Wegeners, livedo reticularis, thrombo angitis obliterans, cns angitis

- VENULES and ARTERIOLES
- examples- leukocytoclastic vasculitis, urticarial vasculitis, henoch-schonlein purpura,

THE END