Vasculitis

Pathology Department
KSU, Riyadh
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Objectives

- Know the common causes of vasculitis with special emphasis on the clinic-pathological features and mechanism of:

1. Giant cell arteritis.
2. Polyarteritis nodosa.
3. Wegener's granulomatosis.
4. Leukocytoclastic vasculitis.

Vasculitis

✖It is inflammation of vessel walls with many possible symptoms

Causes:

1. It is usually immune-mediated
   - ❖ Immune complex deposition
   - ❖ Antineutrophil cytoplasmic antibodies (ANCAs)
   - ❖ Anti-endothelial cell antibodies
   - ❖ Autoreactive T cells

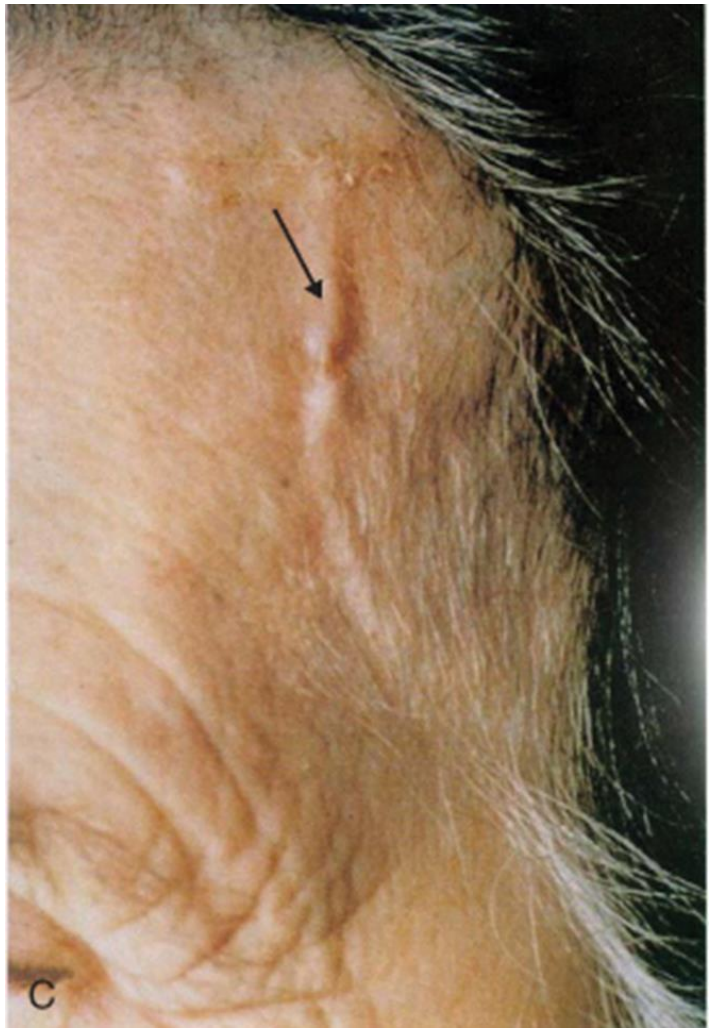
2. It can also be caused by infection, physical or chemical injury

Summary of Vasculitides

Vessel	Disease	comment
Large	Giant-cell arteritis	>50. Arteries of head.
	Takayasu arteritis	F <40. “Pulseless disease”
Medium	Polyarteritis nodosa	Young adults. Widespread.
	Kawasaki disease	<4. Coronary disease. Lymph nodes.
Small	Wegener granulomatosis	Lung, kidney. c-ANCA.
	Churg-Strauss syndrome	Lung. Eosinophils. Asthma. p-ANCA.
	Microscopic polyangiitis	Lung, kidney. p-ANCA.
	Cutaneous leukocytoclastic vasculitis	Idiopathic, infectious, drugs, chemicals, cancer and sytemic disease like HNP

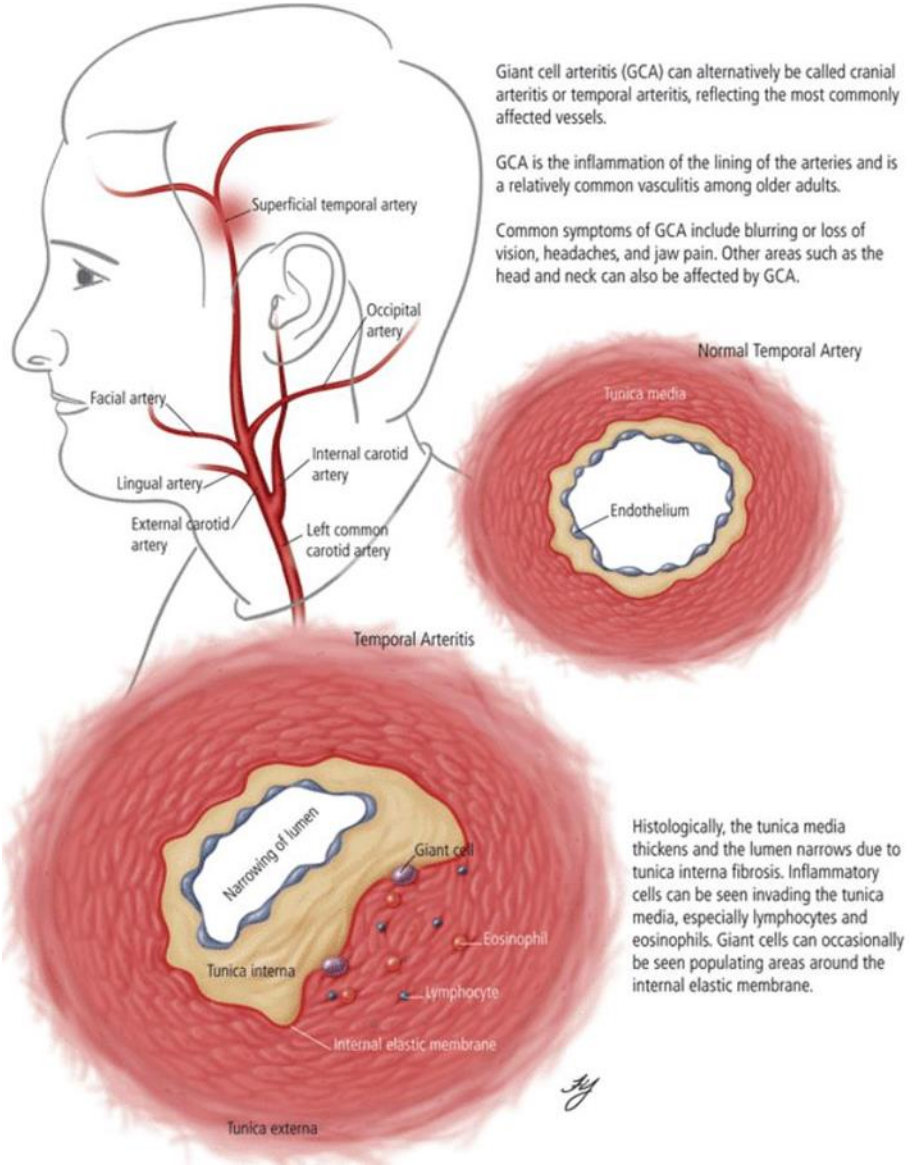
Giant-Cell (Temporal) Arteritis

- Most common type of vasculitis
- Patients >50, F:M = 2:1.
- Chronic, granulomatous inflammation of large to small arteries, especially in head particularly the branches of the carotid artery (temporal a. and branches of the ophthalmic a.)
- Involvment is segmental, acute and chronic.



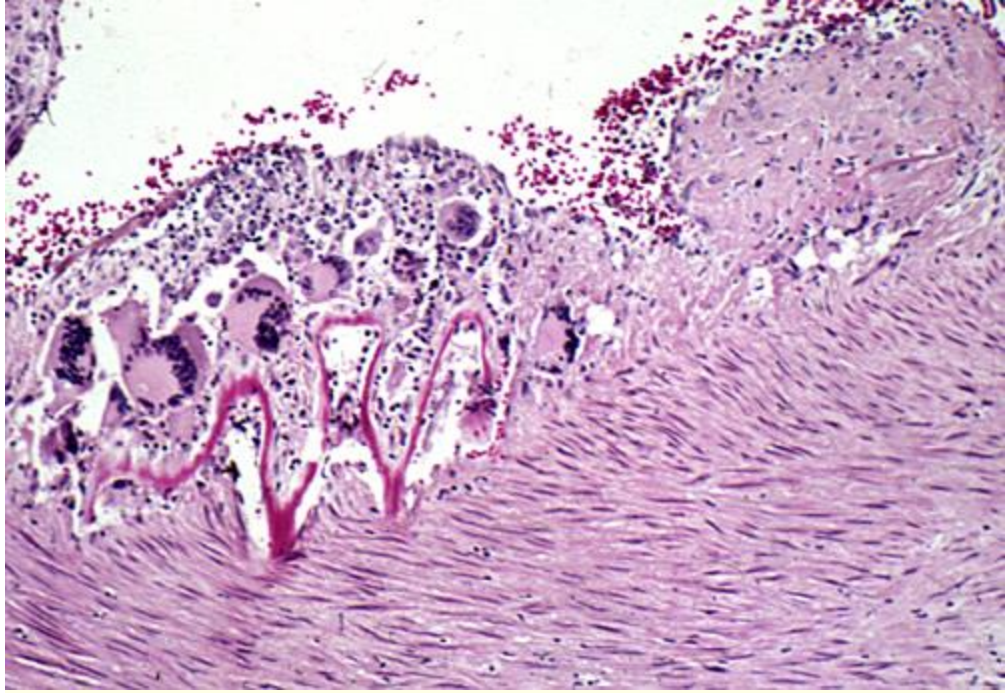
Clinical Features

- ✖Symptoms :
  - ✚fever, facial pain or headache, often most intense along the course of the superficial temporal artery,
  - ✚Thickened and painful temporal artery
  - ✚Jaw pain
  - ✚Visual problems and acute vision loss
- ✖The diagnosis depends on biopsy and histologic confirmation.
- ✖Treatment: corticosteroids

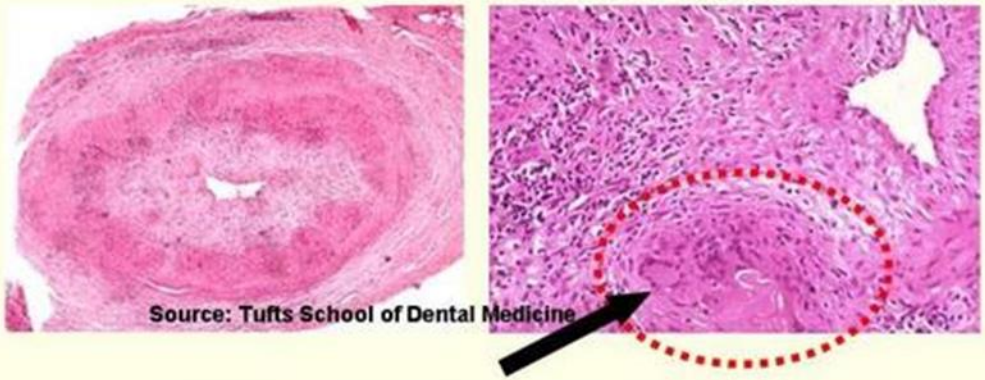


Giant-Cell (Temporal) Arteritis: morphology

- ✖Granulomatous inflammation of the blood vessel wall
- ✖Giant cells
- ✖Disruption and fragmentation of internal elastic lamina
- ✖The healed stage reveals collagenous thickening of the vessel wall and the artery is transformed into a fibrous cord



Temporal Arteritis



Giant cells (arrow) within a granuloma (circle) of granulomatous inflammation

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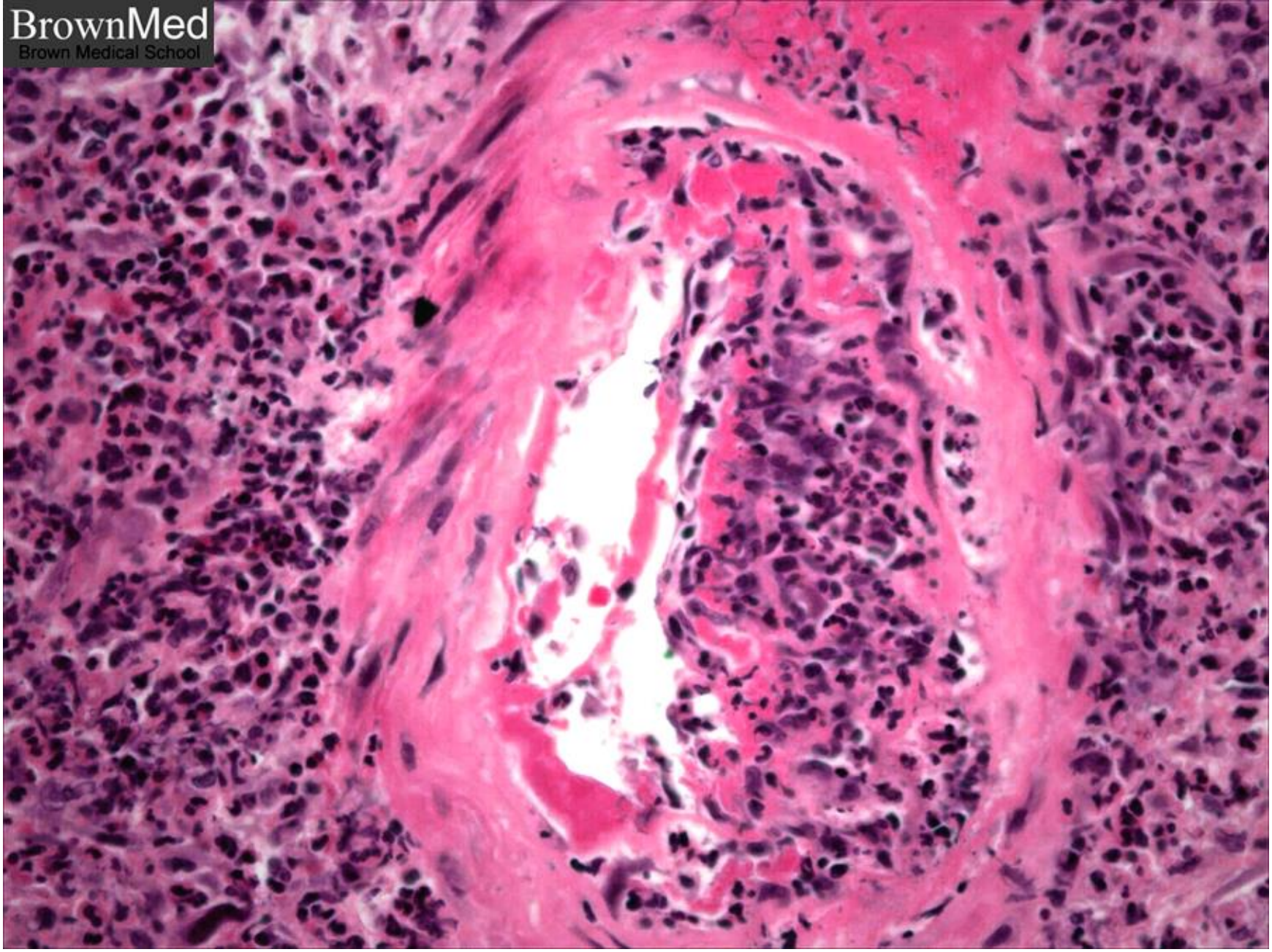
Polyarteritis Nodosa

- Cutaneous only or systemic
- Disease of young adults.
- there is segmental necrotizing inflammation of **arteries of medium to small size, in any organ** (esp kidney and skin) except the lung.
- Polyarteritis nodosa has been associated with hepatitis B or hepatitis C

Polyarteritis Nodosa

- ✖Clinical manifestations result from ischemia and infarction of affected tissues and organs.
- ✖Fever, weight loss, abdominal pain and melena (bloody stool), muscular pain and neuritis.
- ✖Renal arterial involvement is often prominent and is a major cause of death.
- ✖Particularly characteristic of PAN is that all the different stages of activity (i.e. active and chronic stages) may coexist in same artery or in different vessels at the same time.
- ✖Fatal if untreated, but steroids and cyclophosphamide are curative.



Polyarteritis nodosa with segmental inflammation and fibrinoid necrosis and occlusion of the lumen of this artery. Note that part of the vessel wall at the left side is uninvolved.

Wegener granulomatosis

is a necrotizing vasculitis characterized by the **triad** of

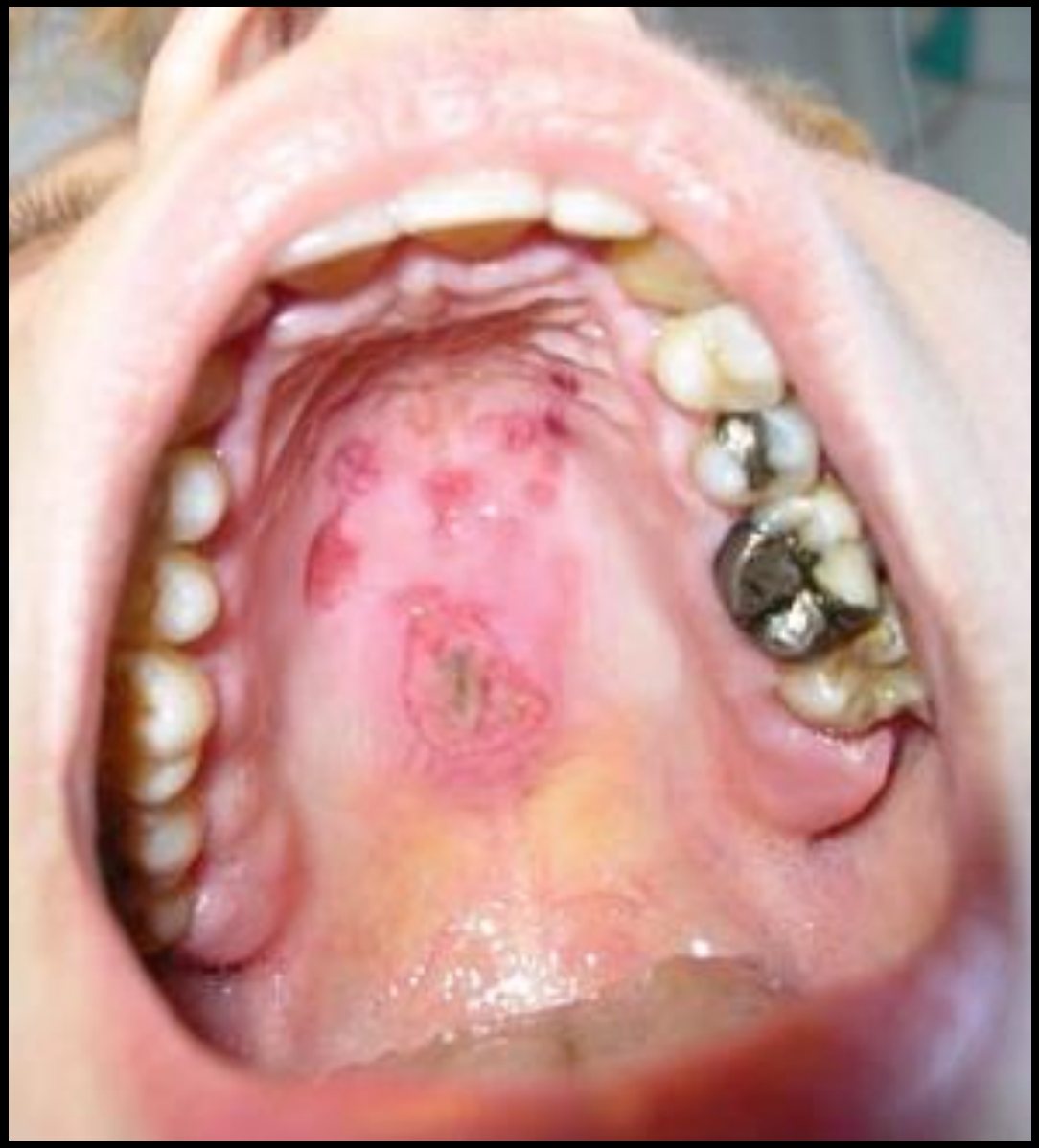
1) **necrotizing granulomas** of the upper and lower respiratory tract

2) necrotizing or granulomatous **vasculitis** of small to medium-sized vessels

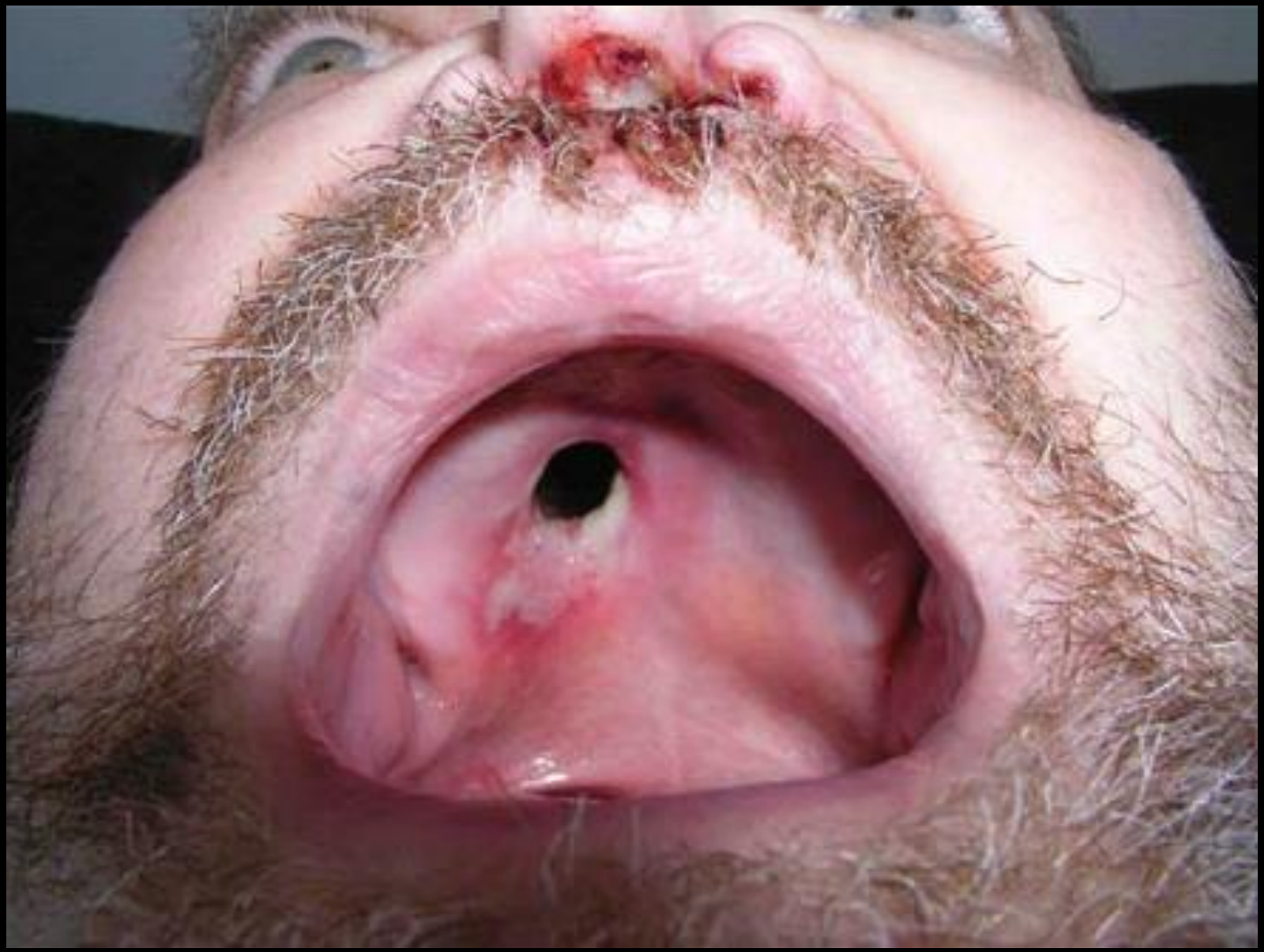
3) **renal disease** in the form of necrotizing, crescentic, glomerulonephritis.

Wegener Granulomatosis

- Males are affected more often than females, at an average age of about 40 years
- C-ANCAs(antineutrophilic cytoplasmic antibodies) is positive in serum of more than 95% of patients.
- Persistent pneumonitis , chronic sinusitis , mucosal ulcerations of the nasopharynx , and evidence of renal disease.
- Untreated: fatal - may lead to death within 2 years if not treated.

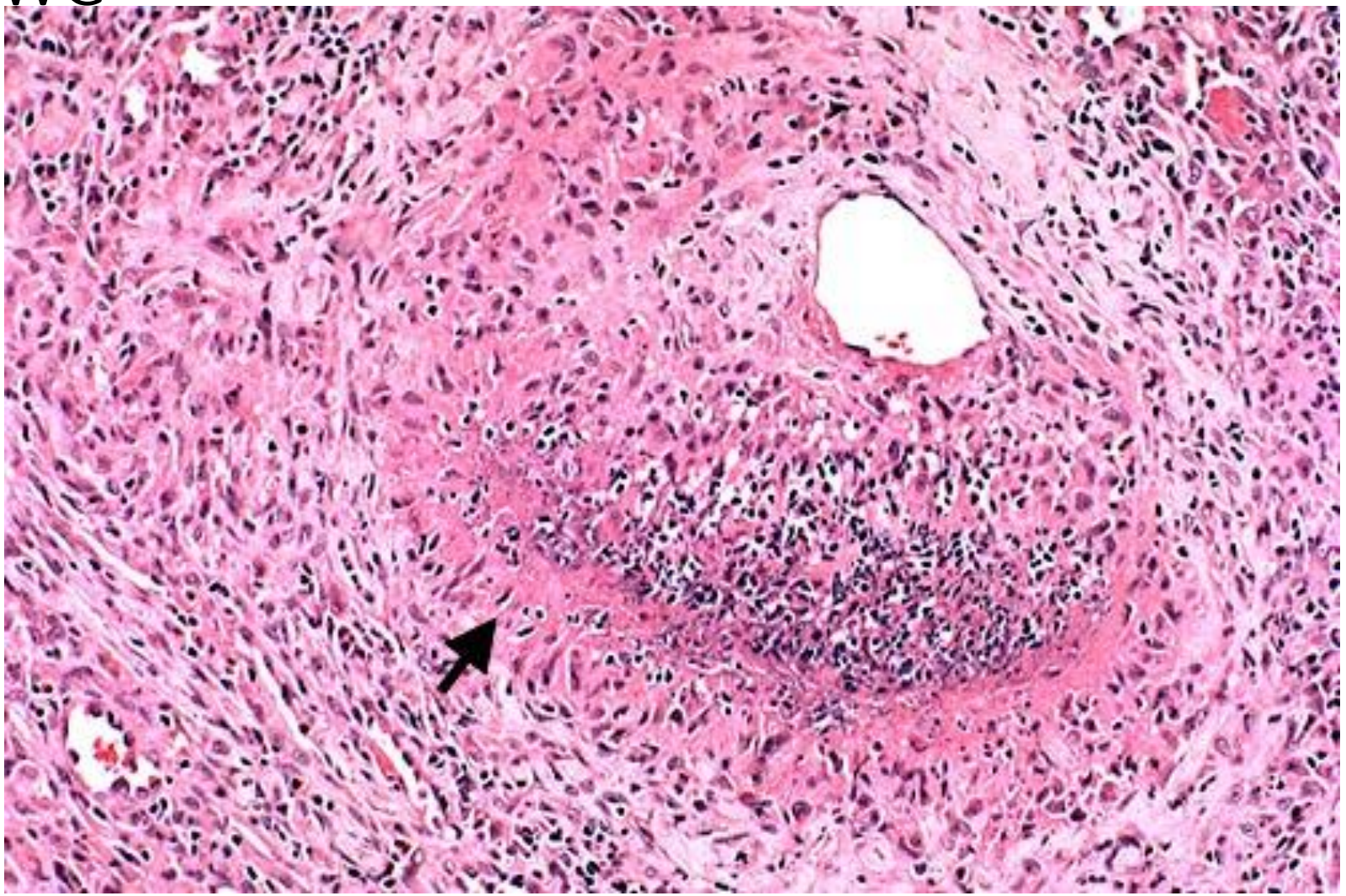


Wegener granulomatosis: palatal ulceration



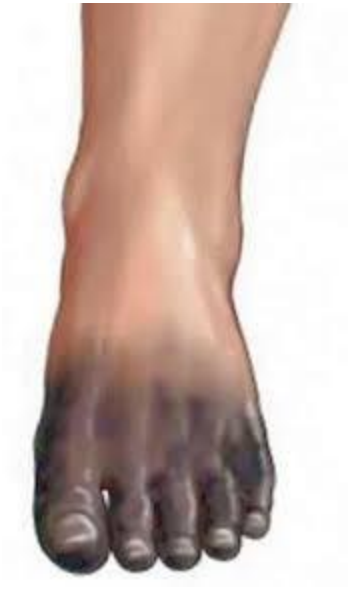
Wegener granulomatosis: palatal destruction

WG

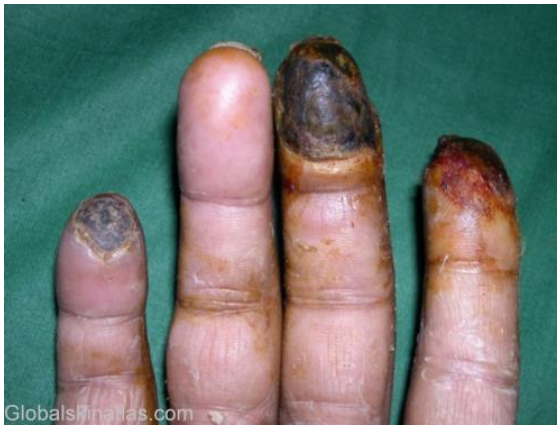


Thromboangiitis obliterans (Buerger disease)

- medium-sized and small arteries,
- Leg and hands.
- heavy smokers of cigarettes, before age 35.

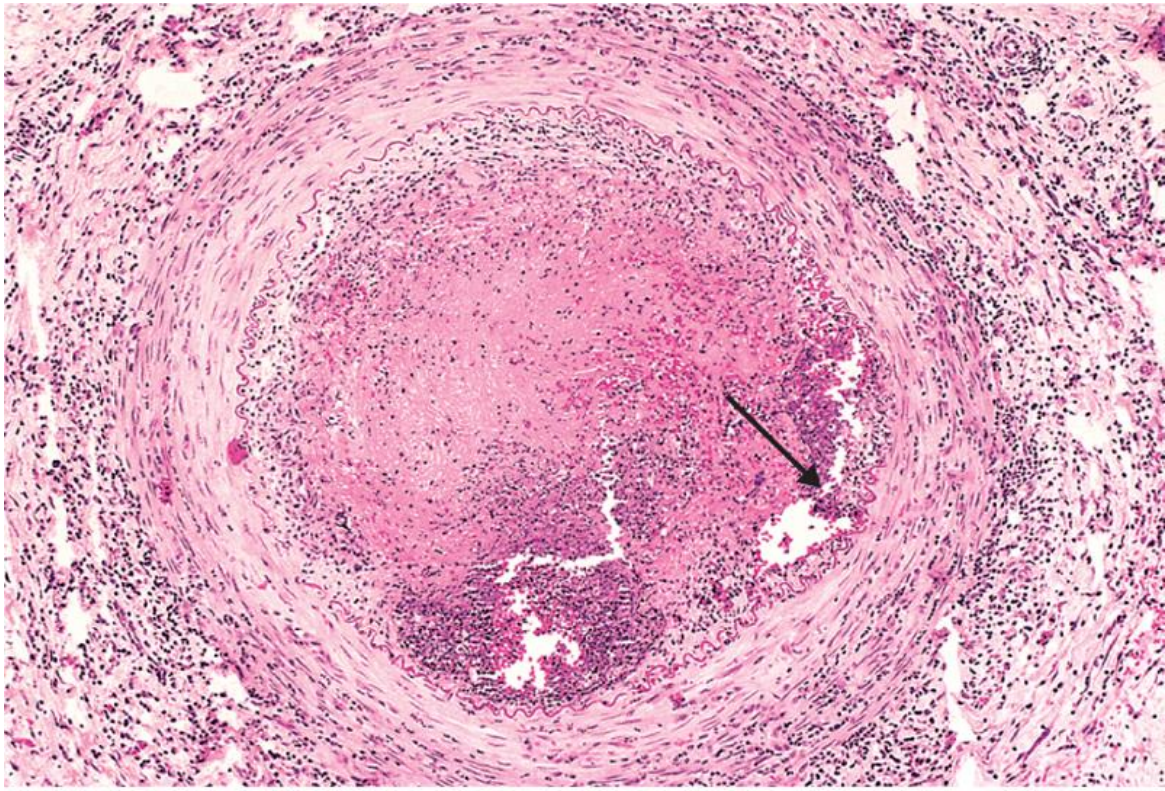


- pain in the affect hand or foot induced by exercise (called *instep claudication*).
- Patients tend to have pain even at rest, due to the neural involvement. Chronic ulcerations of the toes, or fingers may appear, followed in time by gangrene.
- Abstinence from cigarette smoking in the early stages of the disease brings relief from further attacks



Thromboangiitis obliterans (Buerger disease)

- Microscopically, there is acute and chronic inflammation, accompanied by luminal thrombosis.



Cutaneous leukocytoclastic or hypersensitivity vasculitis (angiitis)

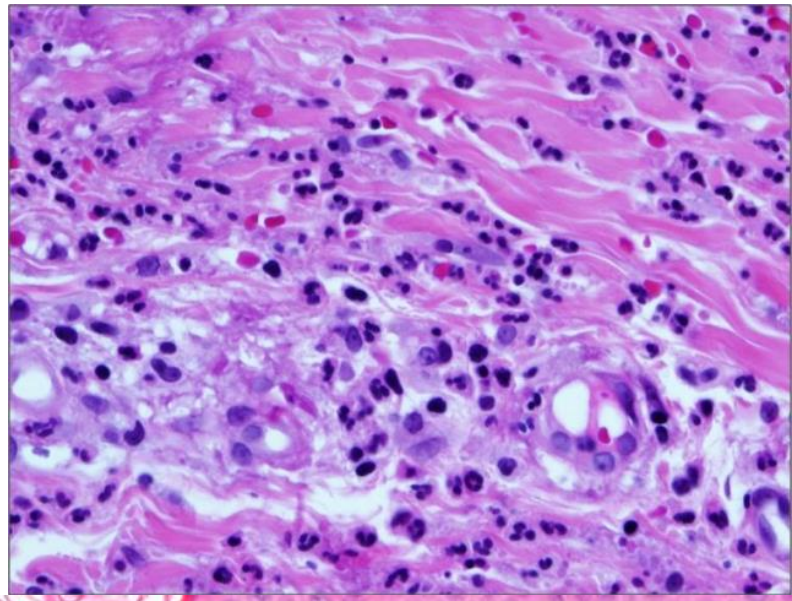
- Can be cutaneous only or systemic
- is inflammation of small blood vessels (usually post-capillary venules in the dermis),
- palpable purpura.
- the most common vasculitis seen in clinical practice.
- Leukocytoclasis = (karyorrhexis of neutrophils) in and around the vessels.
- It affects many organs e.g. skin (most common), mucous membranes, lungs , brain, heart, GI , kidneys and muscle.

Cutaneous leukocytoclastic or hypersensitivity vasculitis (angiitis)

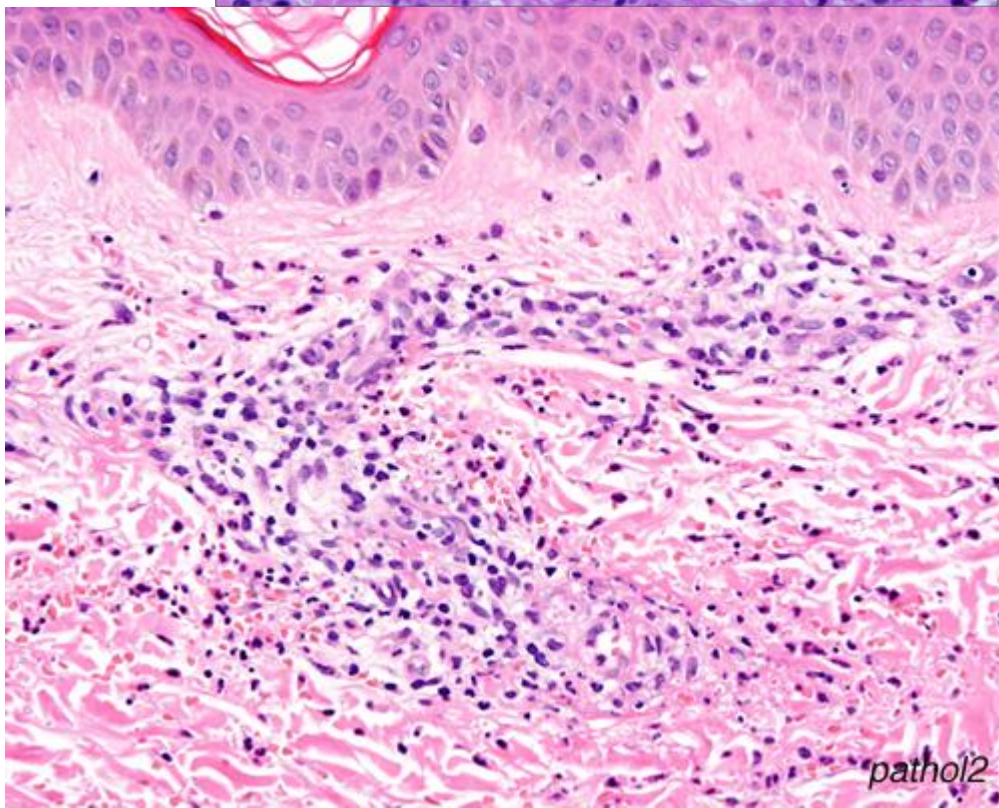
Causes:

- Idiopathic
- immunologic reaction to an antigen that may present as
  - Drugs e.g penicillin
  - Infectious microrganisms e.g. strept. and other infections,
  - food products and toxic chemicals
  - tumor antigens in various cancers.
- It may be a part of a systemic diseases.
  a) **collagen vascular diseases** (lupus erythematosus, rheumatoid arthritis),
  b) **Henoch-Schonlein purpura**

Cutaneous leukocytoclastic or hypersensitivity vasculitis (angiitis)



- Skin biopsy is often diagnostic.
- Histologically there is infiltration of vessel wall with neutrophils, which become fragmented called as leukocytoclasia or nuclear dust.



Henoch-Schonlein purpura(HSP)

- ✖HSP is an IgA-mediated, autoimmune systemic small vessel hypersensitivity vasculitis of childhood.It causes skin purpura, arthritis, abdominal pain, gastrointestinal bleeding, orchitis and nephritis.
- ✖The aetiology remains unknown.
- ✖Immunoglobulin A (IgA) and complement component 3 (C3) are deposited on arterioles, capillaries, and venules.
- ✖Serum levels of IgA are high in HSP

Objectives

1. **Giant cell arteritis.**
   - granulomatous inflammation
   - Temporal artery …pain in its caurse
   - Fever, facial pain or headache
   - Visual problems and acute vision loss

2. **Polyarteritis nodosa.**
   - Cutaneous only or systemic
   - Medium to small size, in any organ (esp kidney and skin)
   - hepatitis B or hepatitis C
   - ischemia of affected tissues and organs

3. **Wegener's granulomatosis.**
   - necrotizing granulomas respiratory tract
   - necrotizing or granulomatous vasculitis (small to medium vessels)
   - renal disease …crescentic, glomerulonephritis.
   - C-ANCA

4. **Leukocytoclastic vasculitis.**
   - small blood vessels
   - Skin biopsy is often diagnostic ..neutrophiles, karyorrhexis
   - Idiopathic , antigenic reaction, systemic disease
   - Henoch-Schonlein purpura

5. ***Thromboangiitis obliterans***
   - medium-sized and small arteries
   - Young …smokers
   - Leg and hands
   - Thrombosis….. gangrene

Ahmed Alhumaidi
Associate professor