Vasculitis

Note:

The important notes with YELLOW colour

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Vasculitis

Vascular inflammatory injury, often with necrosis

Vasculitis

Causes

- immune-mediated :
  - Immune complex deposition
  - Antineutrophil cytoplasmic antibodies (ANCAs)
  - Anti-endothelial cell antibodies

- invasion of vascular walls by infectious pathogens
- Physical and chemical injury
- NOTE: primary vasculitis is more serious than secondery

NOTES

- Immun complex: Hepatitib-B & AB in PAN, Drugs like penicillin (foreign material or durg modified self protiens)
- Circulating immune (antigen-antibody) complexes may also be seen-for example, DNA-anti-DNA complexes in systemic lupus erythematosus (SLE)-associated vasculitis

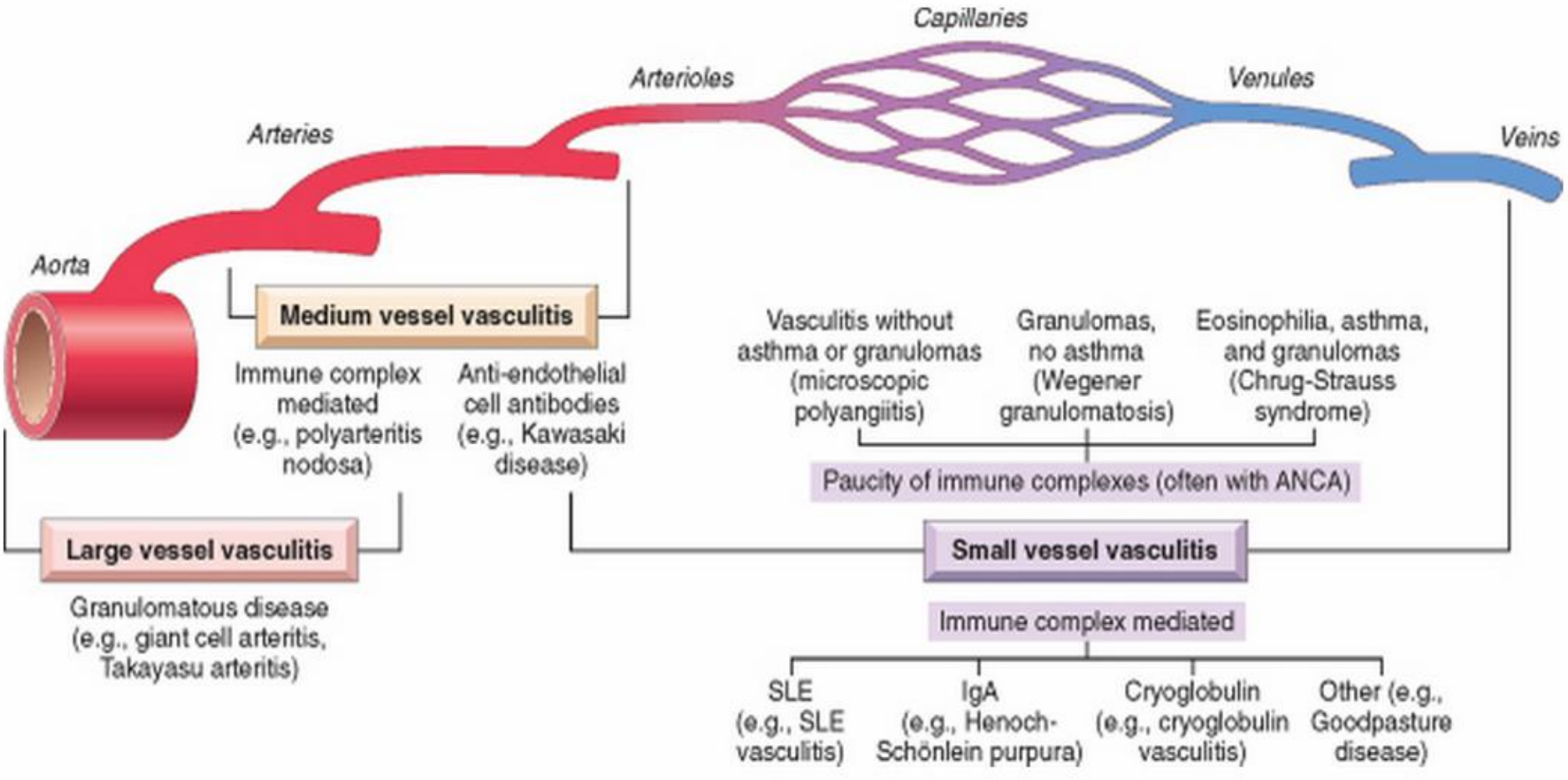
- *Infections can also indirectly induce a noninfectious vasculitis,* for example, by generating immune complexes or triggering cross-reactivity

- Antiendothelial : Kawasaki

Summary of Vasculitides

Vessel	Disease	Notes
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.

You should know the type of vessel which will be affected and the type of immunity



Giant-Cell (Temporal) Arteritis

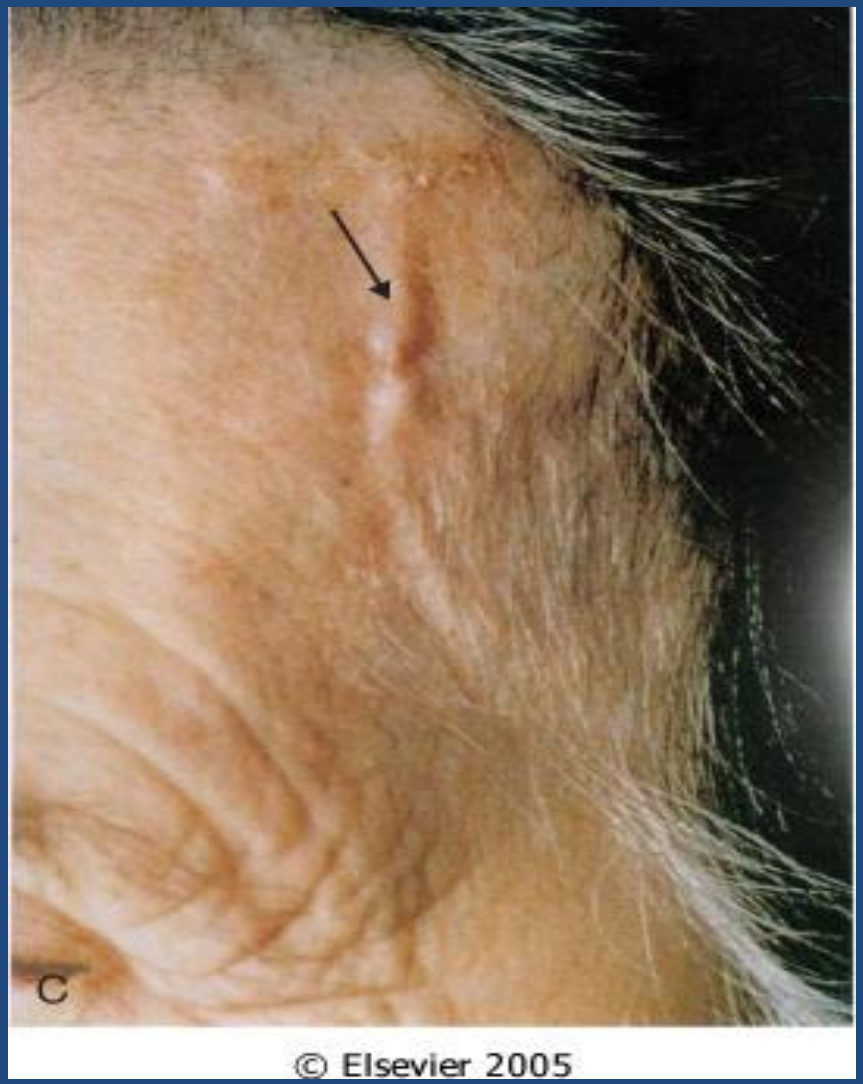
- The most common
- Chronic, typically granulomatous inflammation of large to small-sized arteries
- Principally affects the arteries in the head- especially the temporal arteries
- Rarely the aorta (*giant-cell aortitis*)

Notes : -but also the vertebral and ophthalmic arteries

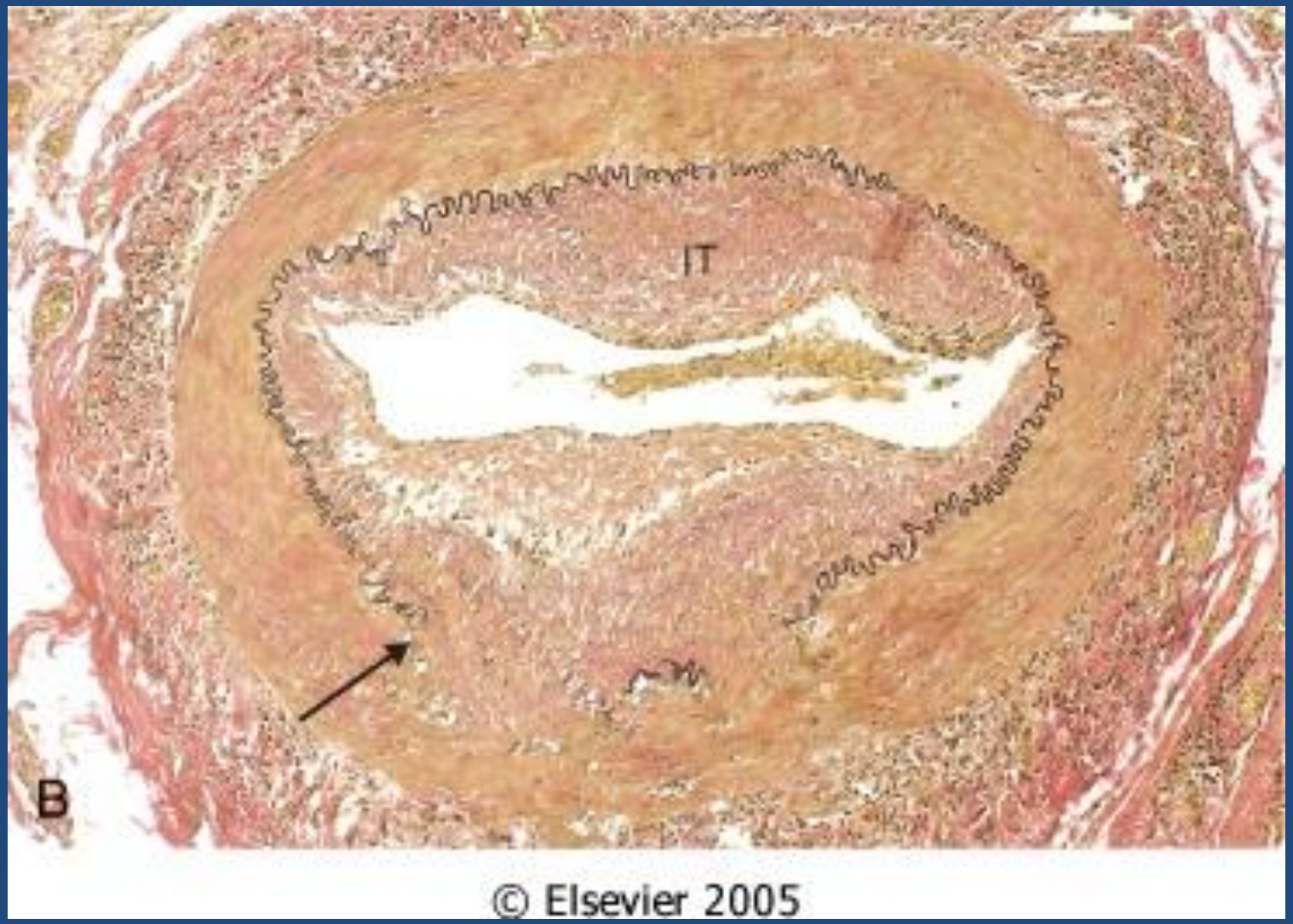
Giant-Cell (Temporal) Arteritis

- Unknown cause
- Likely immune origin, T cell-mediated

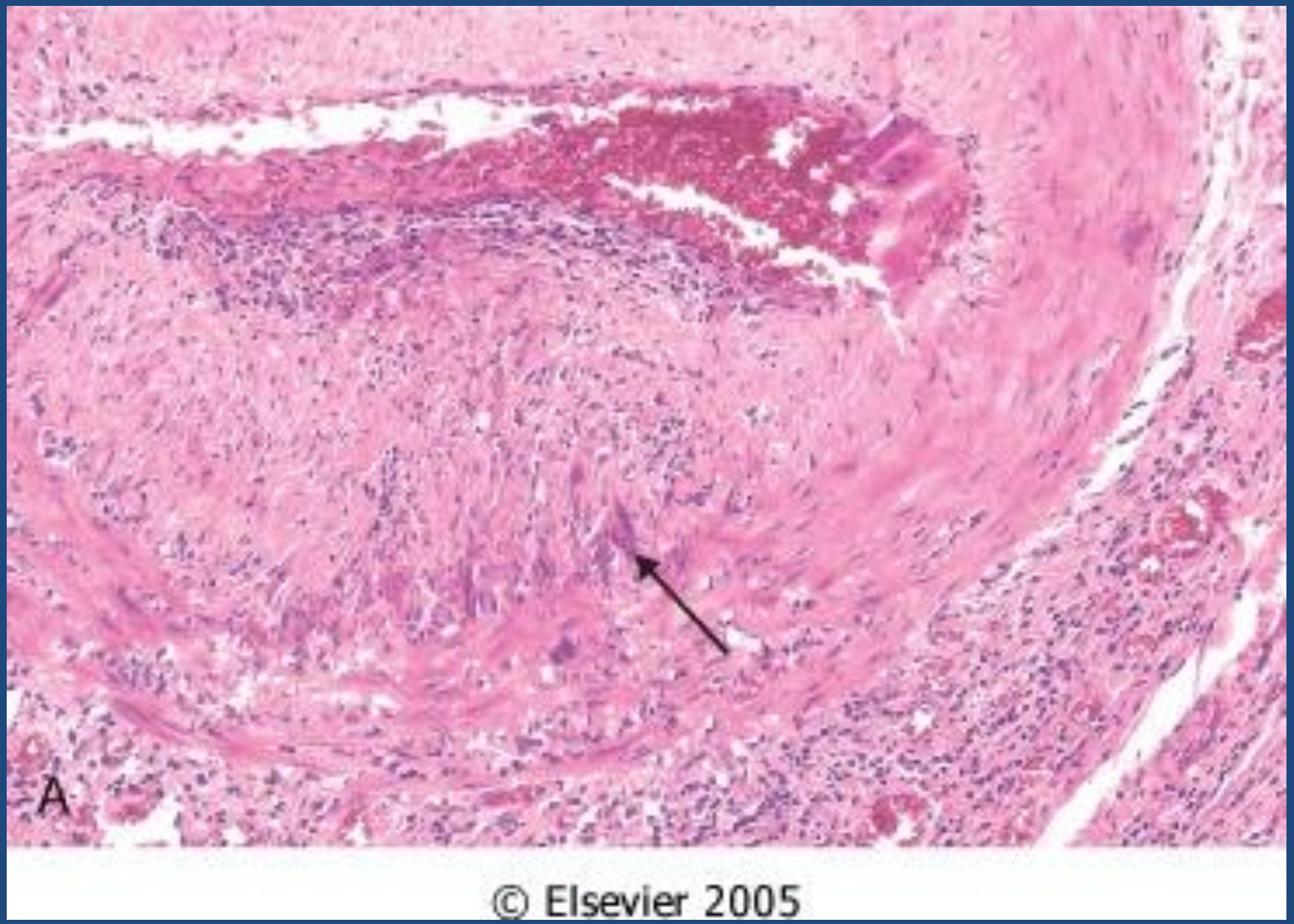
Notes: Nodularity, thickness and firm vessel
Can be segmental process



Notes: The granuloma is centered on the interanl elastic lamina



Nodular intimal thickness
Can be healed (remember ATCHscle) , you don’t have to see granuloma



Giant-Cell (Temporal) Arteritis
Clinical features

- > 50 years of age
- Vague symptoms:
  - Fever, fatigue and weight loss
- May involve facial pain or headache
- Most intense along the course of the superficial temporal artery, which is painful to palpation “tenderness”

Notes

- Ocular symptoms (associated with involvement of the ophthalmic artery) abruptly appear in about 50% of patients
- Female predominance
- these range from diplopia to complete vision loss.

- Plymyalgaia rheumatica association: myalgia, arhtaralgia and fever

Giant-Cell (Temporal) Arteritis

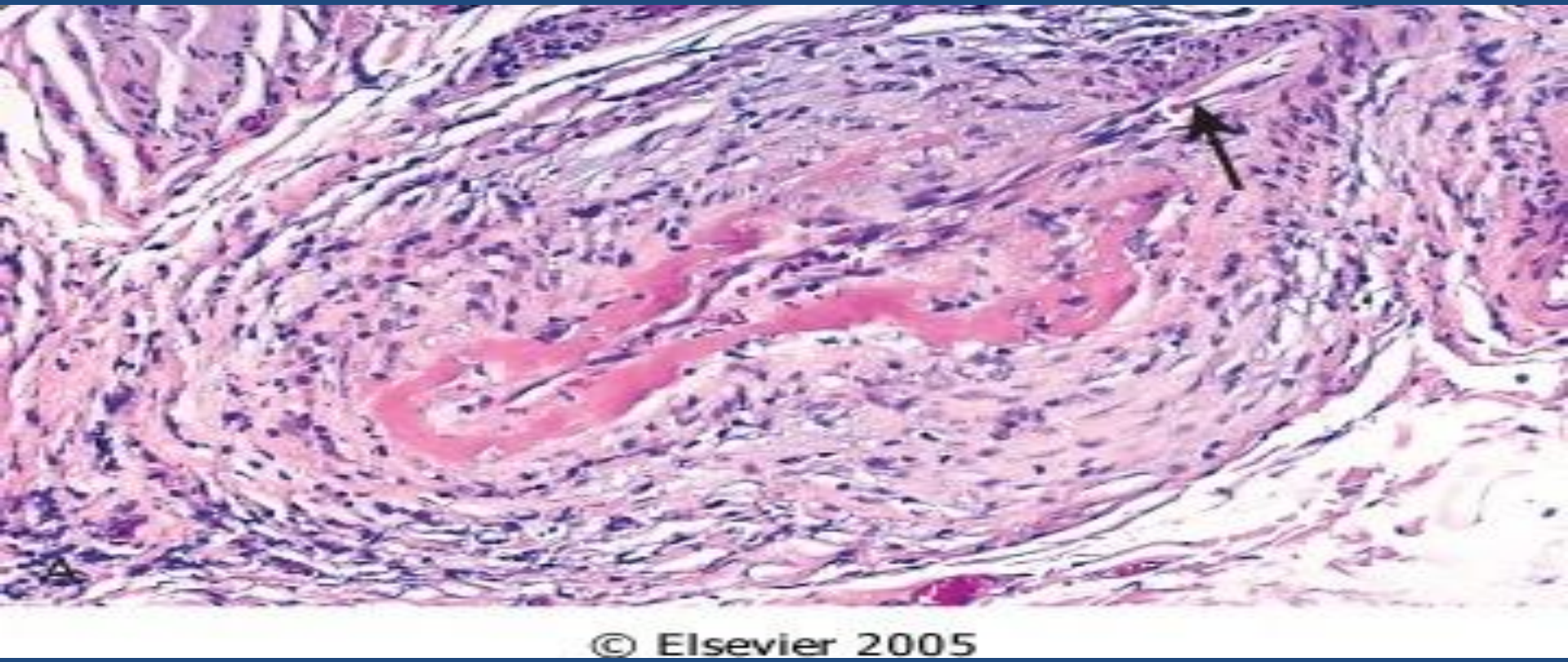
- Definite diagnosis depends on:

biopsy of an adequate segment and histological confirmation

- Treatment: corticosteroids
- Notes : However, because temporal arteritis is extremely segmental, adequate biopsy requires at least a 2- to 3-cm length of artery; even then, a negative biopsy result does not exclude the diagnosis

Polyarteritis Nodosa
PAN

- Systemic except lung
- Small or medium-sized muscular arteries
- **BUT NOT** arterioles, capillaries, or venules
- Typically involving renal and visceral vessels but **sparing** the pulmonary circulation



- Notes: Biopsy is needed for Dx
- with a mixed infiltrate of neutrophils, eosinophils, and mononuclear cells, frequently accompanied by **fibrinoid necrosis**
- Thrombosis, transmural
- arrow NOT involved
- Sharply demarcated

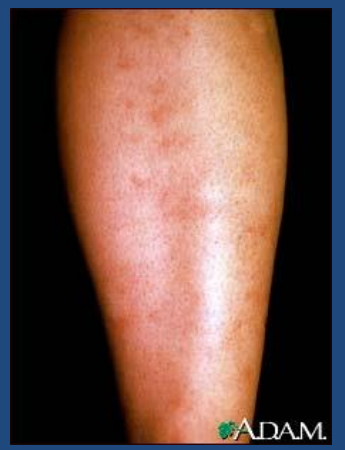
Polyarteritis Nodosa

- The most important feature of this vasculitis:
- all stages of activity (from early to late) may coexist in different vessels or even within the same vessel

Polyarteritis Nodosa
Clinical picture

- Largely young adults
- Typically episodic, with long symptom-free intervals
- Because the vascular involvement is widely scattered, the clinical findings may be varied and puzzling

Notes: The course can vary from acute to chronic



Polyarteritis Nodosa
Clinical picture

- Fever and weight loss
- Examples on systemic involvement:
  - Renal (arterial) involvement is common and a major cause of death
    - Hypertension, usually developing rapidly
    - Abdominal pain and melena (bloody stool)
    - Diffuse muscular aches and pains
    - Peripheral neuritis
- Biopsy is often necessary to confirm the diagnosis

Polyarteritis Nodosa

- **No** association with ANCA
- Some 30% of patients with PAN have hepatitis B antigenemia
- If untreated, the disease is fatal in most cases
- Therapy with corticosteroids and other immunosuppressive therapy results in remissions or cures in 90%
- PAN is an example of **heterogeneicity** disease

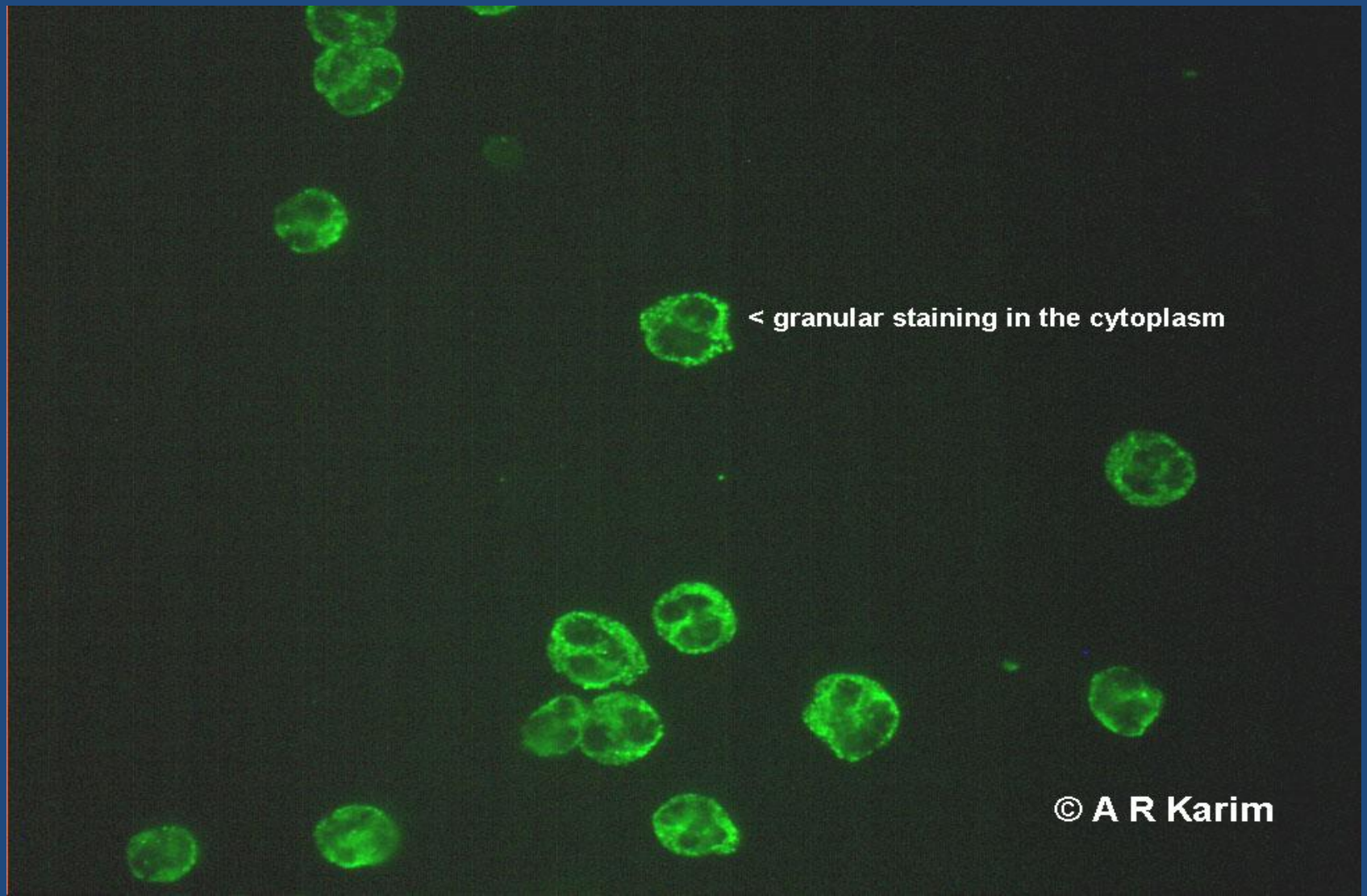
Polyarteritis Nodosa
Complications

- Vessel rupture
- Impaired perfusion:
  - Ulcerations
  - Infarcts
  - Ischemic atrophy *(**not infarction**)*
  - Haemorrhages in the distribution of affected vessels may be the first sign of disease

Notes: Like other vasculitis

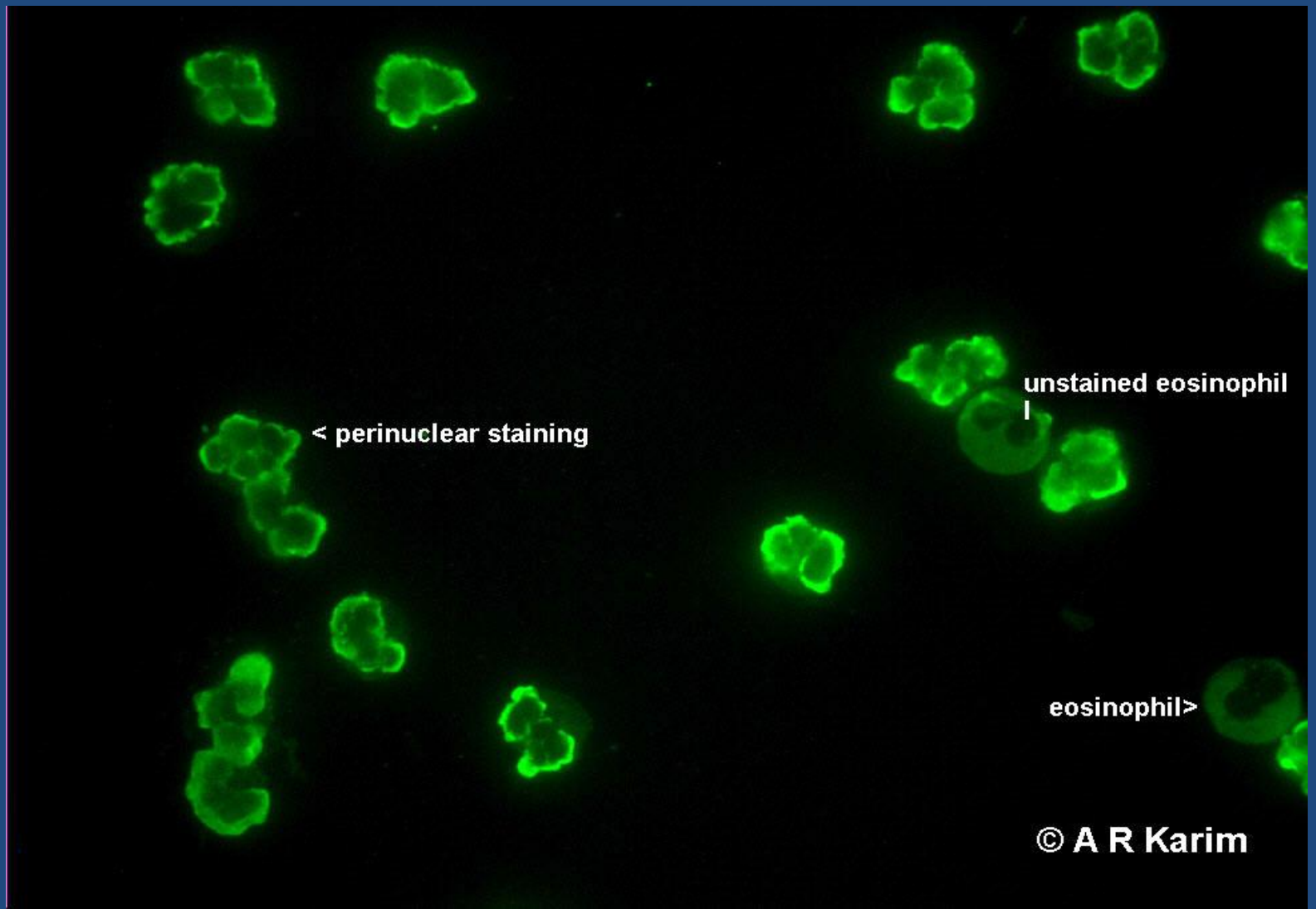
c-ANCA

"cytoplasm anti-neutrophil cytoplsmic antibody"



p-ANCA

“ perinuclues anti-neutrophil cytoplsmic antibody “



Antineutrophil Cytoplasmic Antibodies

- Cytoplasmic localization (c-ANCA) -> the most common target antigen is proteinase-3 (PR3)
  - typical of Wegener granulomatosis
- Perinuclear localization (p-ANCA) -> most of the autoantibodies are specific for myeloperoxidase (MPO)
  - microscopic polyangiitis and Churg-Strauss syndrome
- ANCAs serve as useful diagnostic markers for the ANCA-associated vasculitides
- Their levels can reflect the degree of inflammatory activity

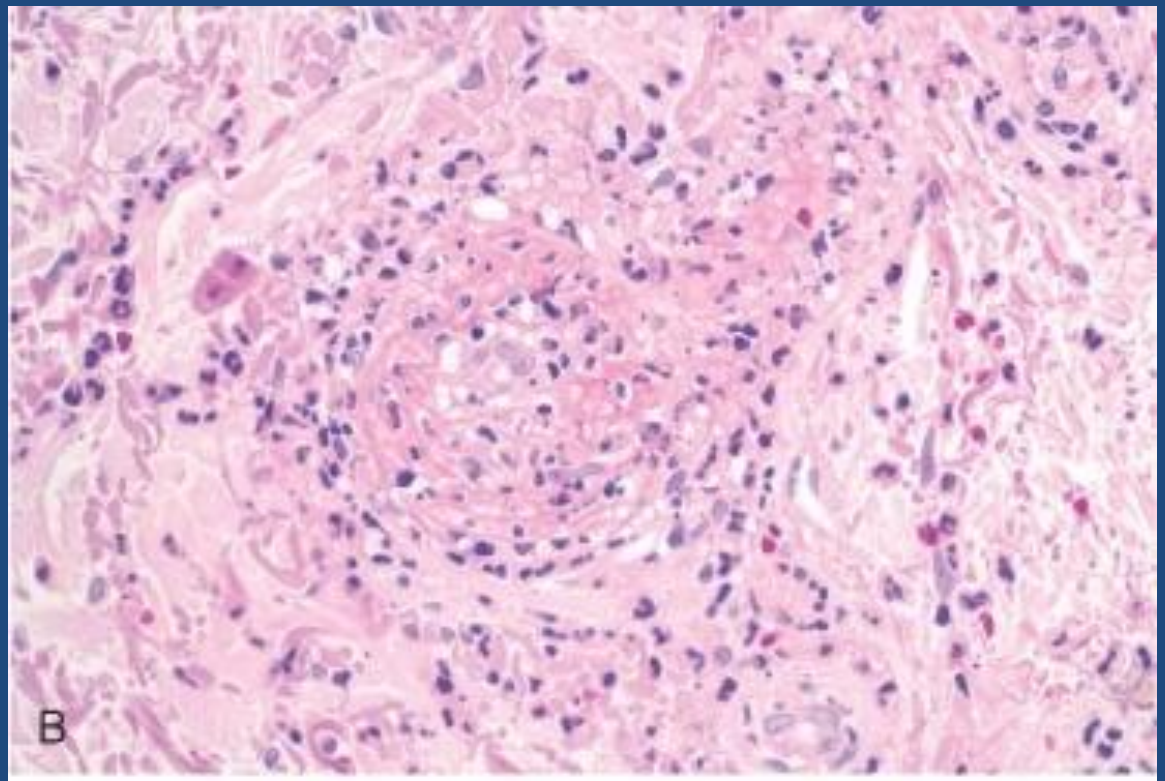
Microscopic Polyangiitis

- Necrotizing vasculitis that generally affects capillaries as well as arterioles and venules of a size smaller than those involved in PAN
- Rarely, larger arteries may be involved
- *All lesions of microscopic polyangiitis tend to be of the **same age** in any given patient*
- *Ex: Necrotizing glomerulonephritis (90% of patients) and pulmonary capillaritis are particularly common*

Microscopic Polyangiitis

Pathogenesis

- In many cases, an antibody response to antigens such as drugs (e.g., penicillin), microorganisms (e.g., streptococci), heterologous proteins, or tumor proteins is the presumed cause
- This can result in immune complex deposition, or it may trigger secondary immune responses
- p-ANCAs are present in more than 70% of patients



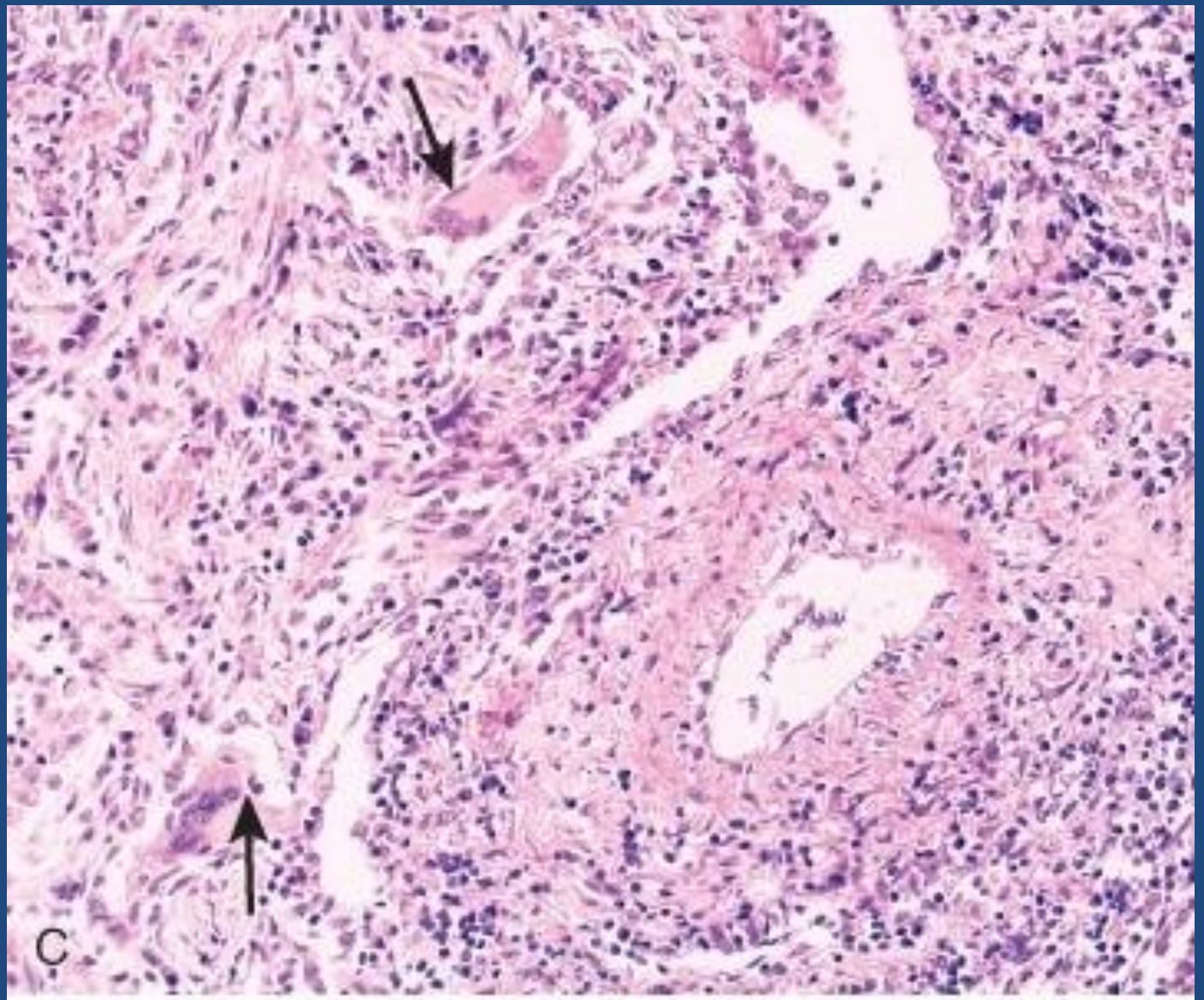
Microscopic Polyangiitis

- Depending on the organ involved, major clinical features include:
  - Hemoptysis
  - *H*ematuria and proteinuria
  - Bowel pain or bleeding
  - Muscle pain or weakness
  - Palpable cutaneous purpura

Wegener Granulomatosis

- Triad:
  - *Acute necrotizing granulomas* of the 1-upper and 2-lower respiratory tract (lung), or both
  - *Necrotizing or granulomatous vasculitis* affecting small to medium-sized vessels (most prominent in the lungs and upper airways)
  - *Focal necrotizing, often crescentic, glomerulitis*

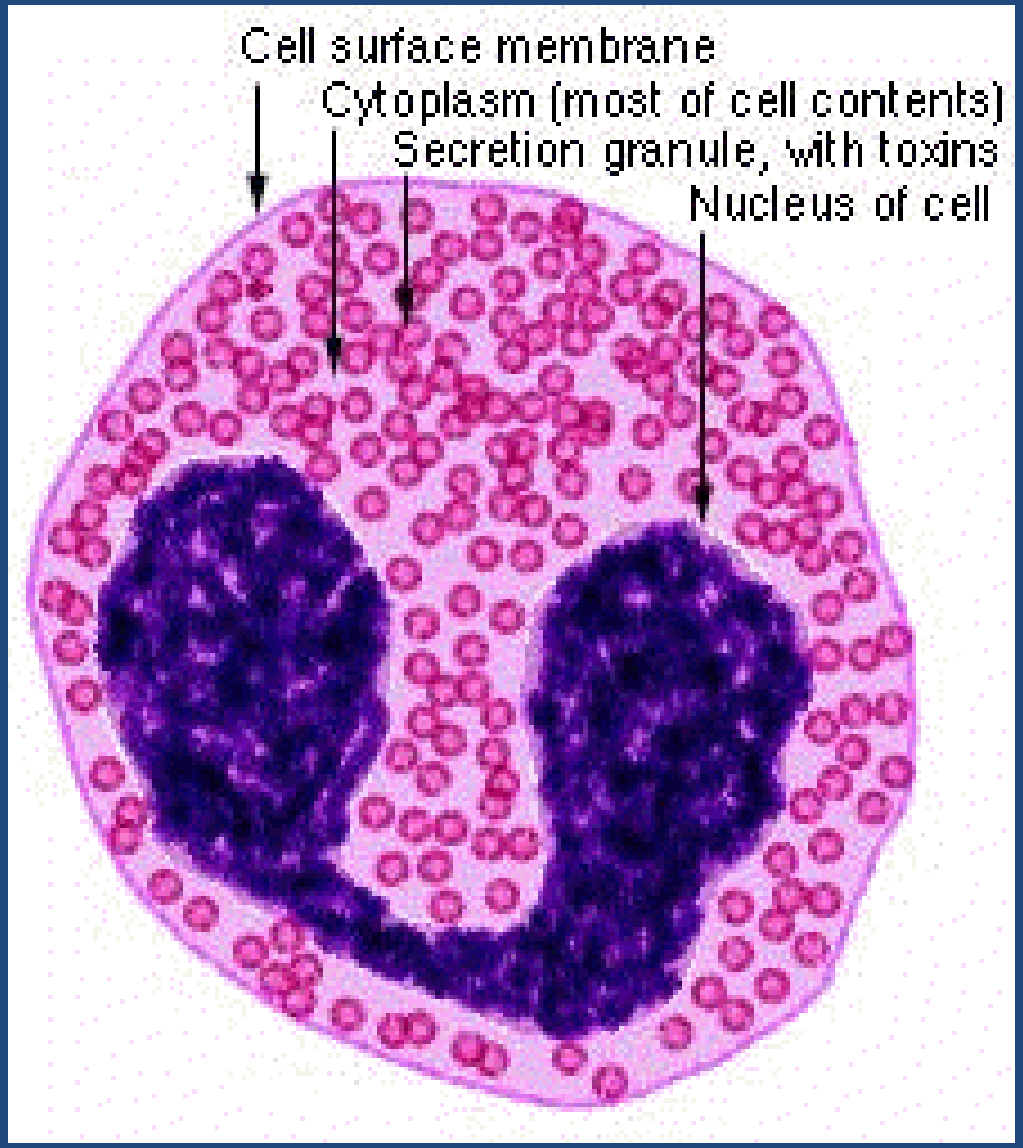
- *Notes:* No causative agent, although immune complexs occasionally seen
- clinically, this resembles PAN except that there is also respiratory involvement.
- Kidney severity is variable



Wegener Granulomatosis

- 40-50 years
- Without Rx -> 80% die
- With Rx -> 90% live (not cured)
- The Rx -> immunosuppression

Notes: M>F


Cell surface membrane
Cytoplasm (most of cell contents)
Secretion granule, with toxins
Nucleus of cell

Churg-Strauss syndrome

- **Eosinophil**-rich and granulomatous inflammation involving the respiratory tract and necrotizing vasculitis affecting small vessels
- Associated with **asthma** and blood eosinophilia
- Associated with p-ANCAs.