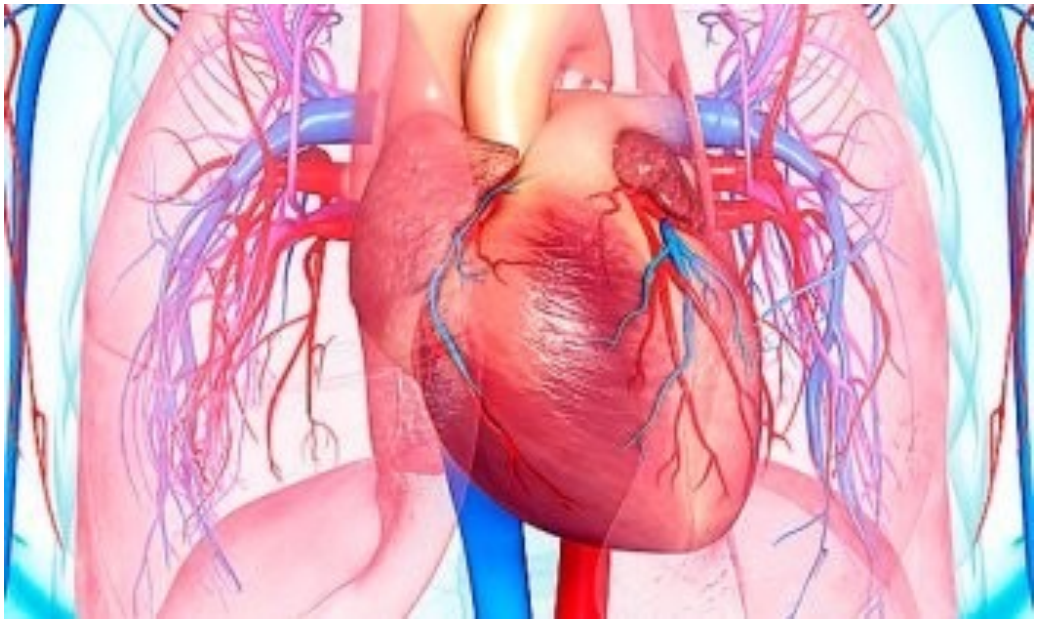
PULMONARY DISEASES OF VASCULAR ORIGIN



MARAM ABDALJALEEL, MD.
DERMATOPATHOLOGIST
UNIVERSITY OF JORDAN , SCHOOL OF MEDICINE

https://www.europeanlung.org/en/lung-disease-and-information/lung-diseases/pulmonary-vascular-disease

PULMONARY DISEASES OF VASCULAR ORIGIN

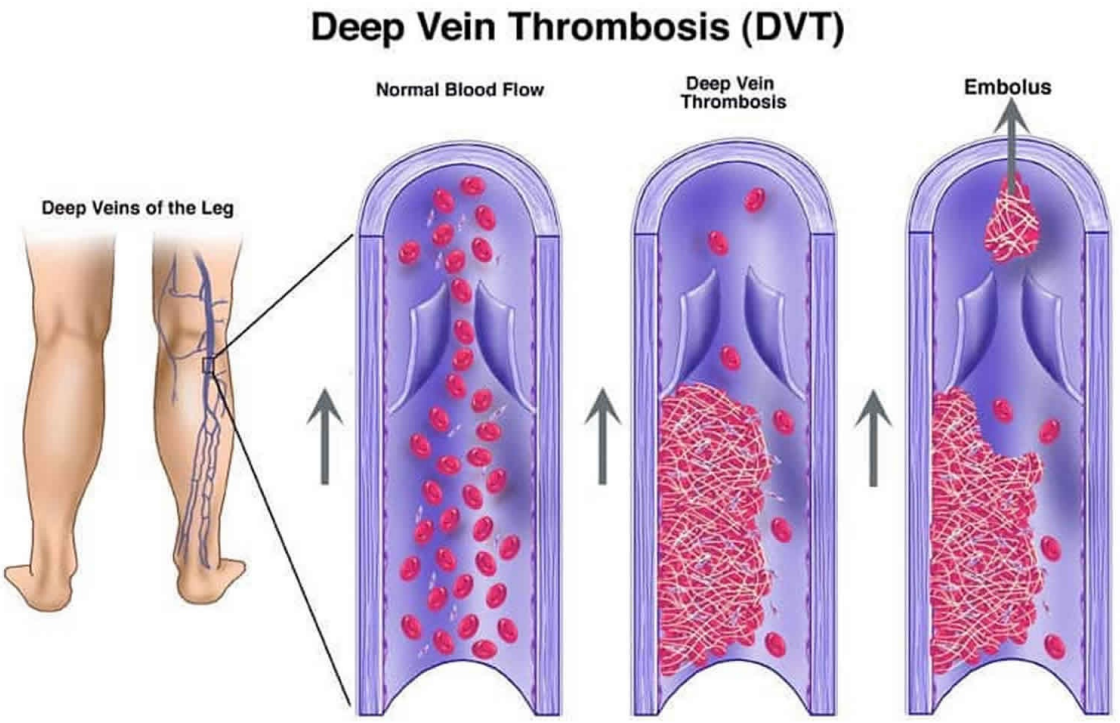
- Pulmonary Embolism, Hemorrhage, and Infarction
- Pulmonary Hypertension
- Diffuse Alveolar Hemorrhage Syndromes

PULMONARY EMBOLISM:

- **Thromboembolism**
- **Nonthrombotic pulmonary emboli**

THROMBOEMBOLISM

- >95% of PE arise from thrombi within the large deep veins of the legs, most often popliteal vein and larger veins above it.



- usually occurs in patients with a predisposing condition that increase the tendency to clotting (thrombophilia)

RISK FACTORS FOR VENOUS THROMBOSIS:

1. prolonged bed rest (immobilization of the legs)
2. Surgery (orthopedic surgery on the knee or hip)
3. severe trauma (burns or multiple fractures)
4. congestive heart failure
5. in women, the period around parturition or the use of OCPs (high estrogen content)
6. disseminated cancer
7. primary disorders of hypercoagulability (factor V Leiden)

CONSEQUENCES:

- **ischemia of the downstream pulmonary parenchyma** due to the nonperfused, although ventilated, segment.

- **increase in pulmonary artery pressure and vasospasm** due to the presence of embolic obstruction → blocked flow and release of mediators such as thromboxane A2, and serotonin→ vasospasm

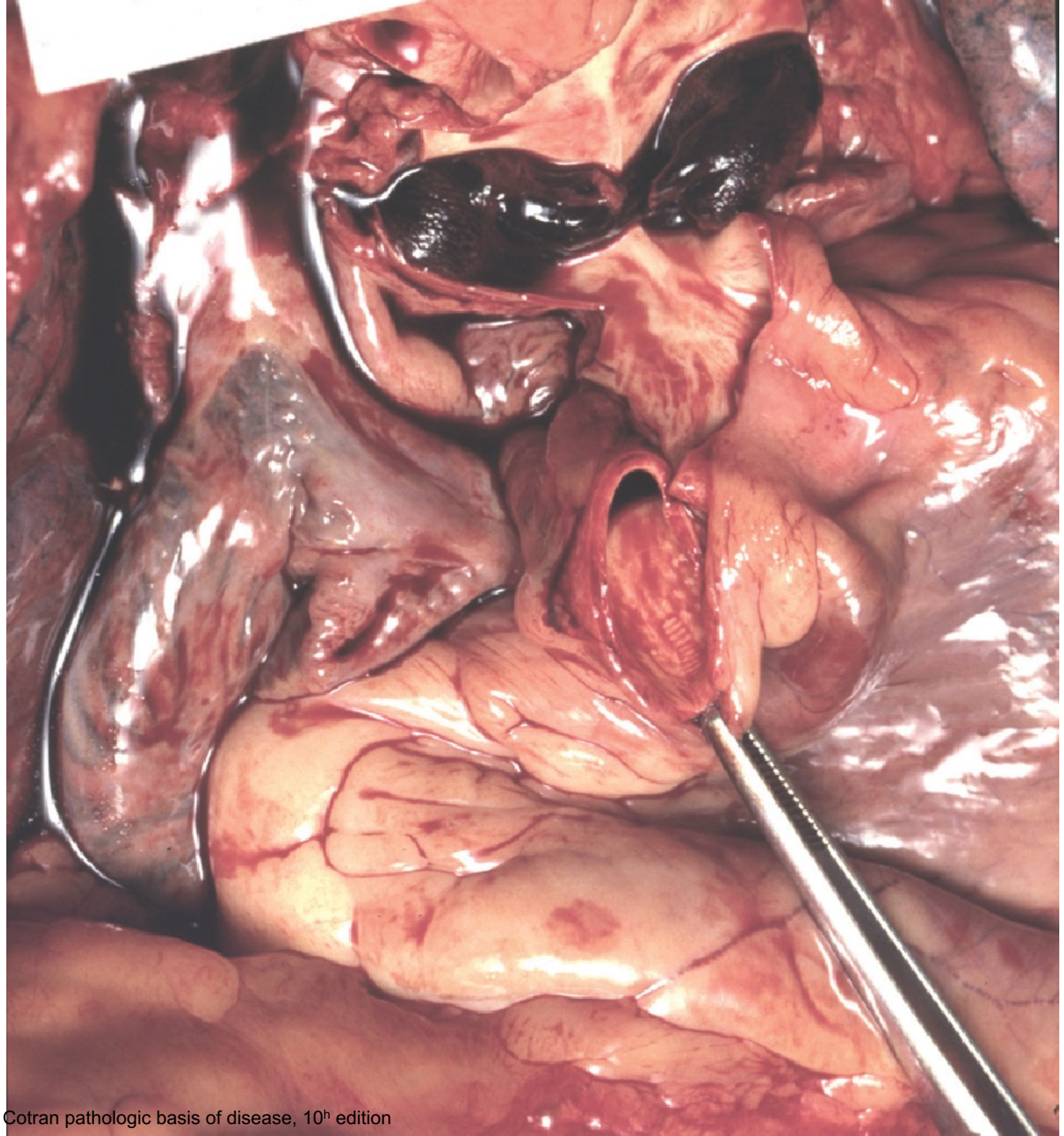
CONSEQUENCES:

- **depend mainly on:**

1- The size and number of emboli:

- large embolus may embed in the main pulmonary artery or its major branches or lodge at the bifurcation as a saddle embolus
- Smaller emboli become impacted in medium-sized and small-sized pulmonary arteries.

2- the cardiopulmonary status of the patient.



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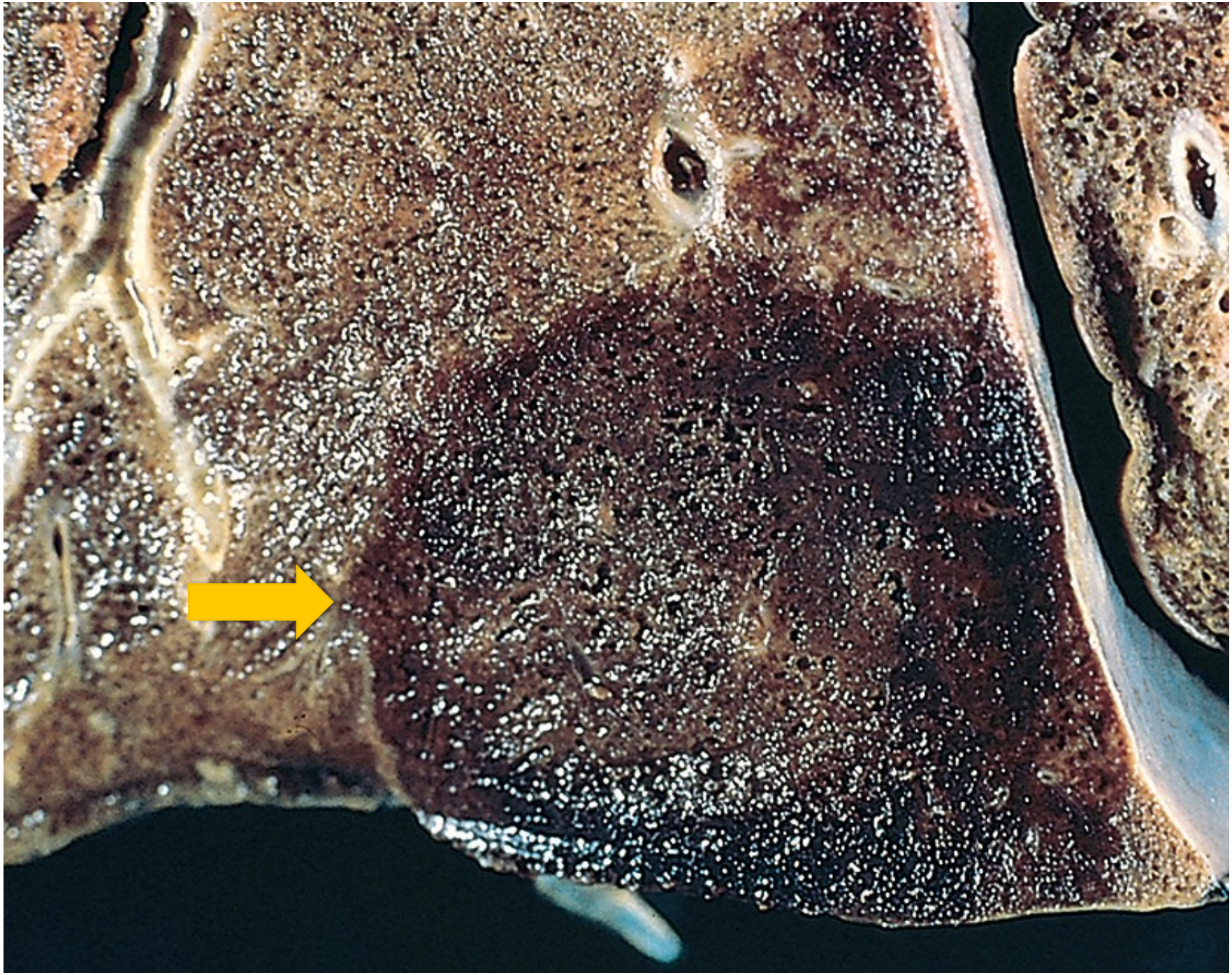
MORPHOLOGY:

- **No morphologic alternations**: large emboli (causing sudden death)
- **alveolar hemorrhage**: Smaller emboli
- **infarction :**
  - compromised cardiovascular status (congestive heart failure)
  - The more peripheral the embolic occlusion, the higher the risk for infarction.
    - ¾ lower lobes & >50% multiple.
  - wedge-shaped, with their base at the pleural surface and the apex pointing toward the hilus of the lung.

PULMONARY INFARCTS

- Typically, hemorrhagic with red-blue areas of coagulative necrosis
- The adjacent pleura surface is covered by fibrinous exudate
- The occluded vessel is located near the apex of the infarcted area.



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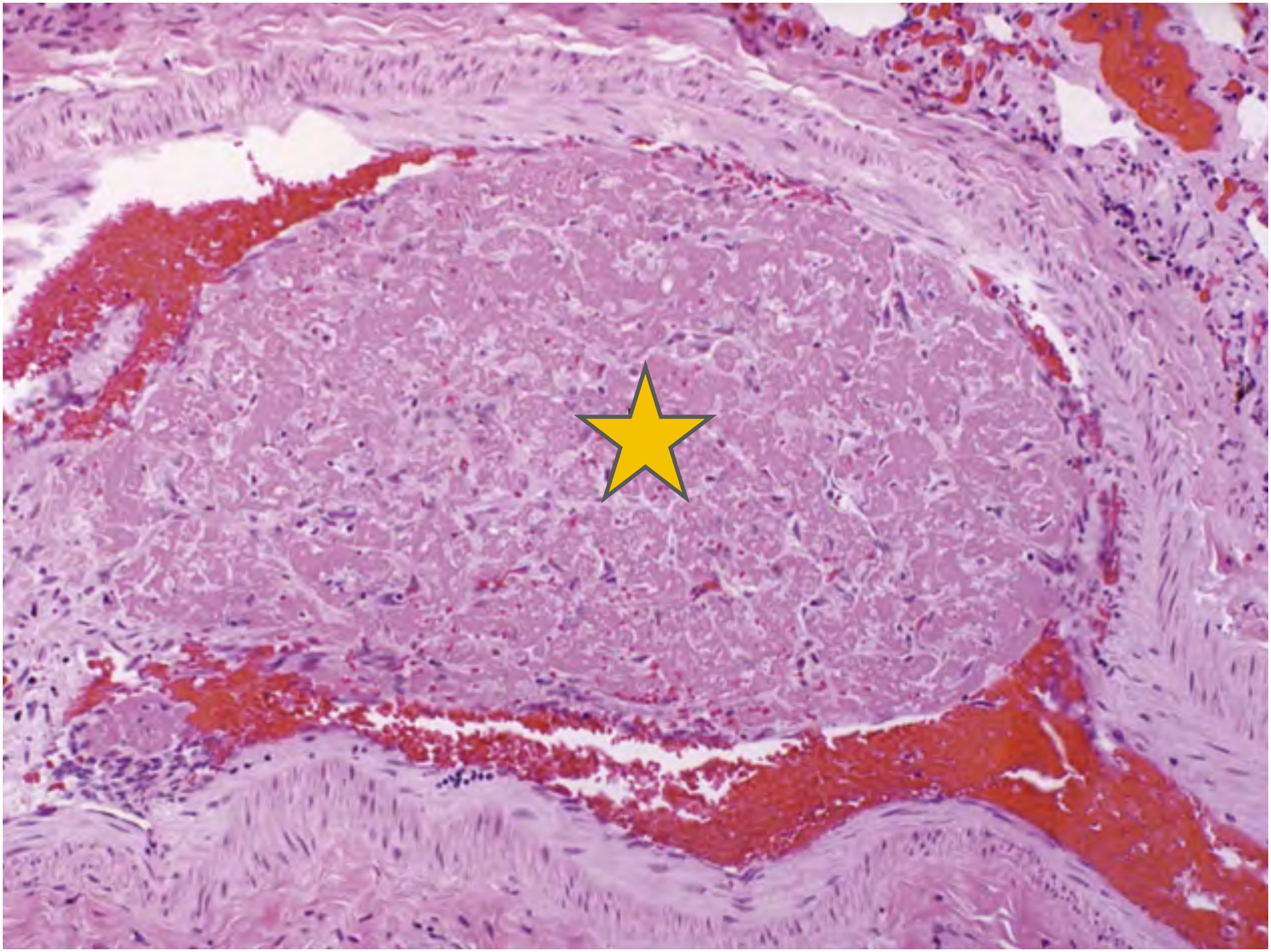


CLINICAL FEATURES

- 60% - 80% →clinically silent
- Small emboli
- embolic mass is rapidly removed by fibrinolytic activity.

- 5%→ death, acute right-sided heart failure, or cardiovascular collapse.
  - As in Massive pulmonary embolism: >60% of the total pulmonary vasculature is obstructed by a large embolus or multiple small emboli.

CLINICAL FEATURES

- 10-15%→ dyspnea
- Obstruction of small to medium pulmonary branches→ pulmonary infarction

- <3% →progressively worsening dyspnea
  - recurrent showers of emboli leading to pulmonary hypertension, chronic right-sided heart failure, and pulmonary vascular sclerosis.

MANAGEMENT:

- Prophylactic therapy: anticoagulation, early ambulation, elastic stockings, intermittent pneumatic calf compression, and isometric leg exercises for bedridden patients.

- anti-coagulation therapy for patients who develop pulmonary embolism

- thrombolytic therapy for hemodynamically unstable pts with massive pulmonary embolism

NONTHROMBOTIC PULMONARY EMBOLI:

- uncommon but potentially lethal
- such as:
  - air, fat, amniotic fluid embolism
  - foreign body embolism in intravenous drug abusers
  - Bone marrow embolism:
    - the presence of hematopoietic and fat elements within a pulmonary artery
    - after massive trauma and in patients with bone infarction secondary to sickle cell anemia

PULMONARY HYPERTENSION

- defined as pressures of **25 mm Hg or more at rest**

- may be caused by **increase in either** pulmonary vascular blood flow, pulmonary vascular resistance, or left heart resistance to blood flow.

CLASSIFIED AS FOLLOWING:

- **Pulmonary arterial hypertension (group 1):**
- affects small pulmonary muscular arterioles
  - heritable forms of pulmonary hypertension
  - Autoimmune diseases such as systemic sclerosis:
    - involve the pulmonary vasculature +/- interstitium→ increased vascular resistance and pulmonary hypertension.
  - connective tissue diseases, human immunodeficiency virus, and congenital heart disease with left to right shunts
  - When all known causes are excluded referred to as idiopathic pulmonary arterial hypertension
    - "idiopathic" is a misnomer, as up to 80% of "idiopathic" cases have a genetic basis

- **Pulmonary hypertension due to left-sided heart disease (group 2):**
    - including congenital or acquired heart disease
    - Eg: Mitral stenosis→ increase in left atrial pressure and pulmonary venous pressure→ eventually transmitted to the arterial side of the pulmonary vasculature→ hypertension.

- **Pulmonary hypertension due to lung diseases and/or hypoxia (group 3)**:
  - including COPD and interstitial lung disease and Obstructive sleep apnea
  - These diseases obliterate alveolar capillaries→ pulmonary resistance to blood flow → pulmonary blood pressure.
  - Obstructive sleep apnea is a common disorder associated with obesity and hypoxemia.

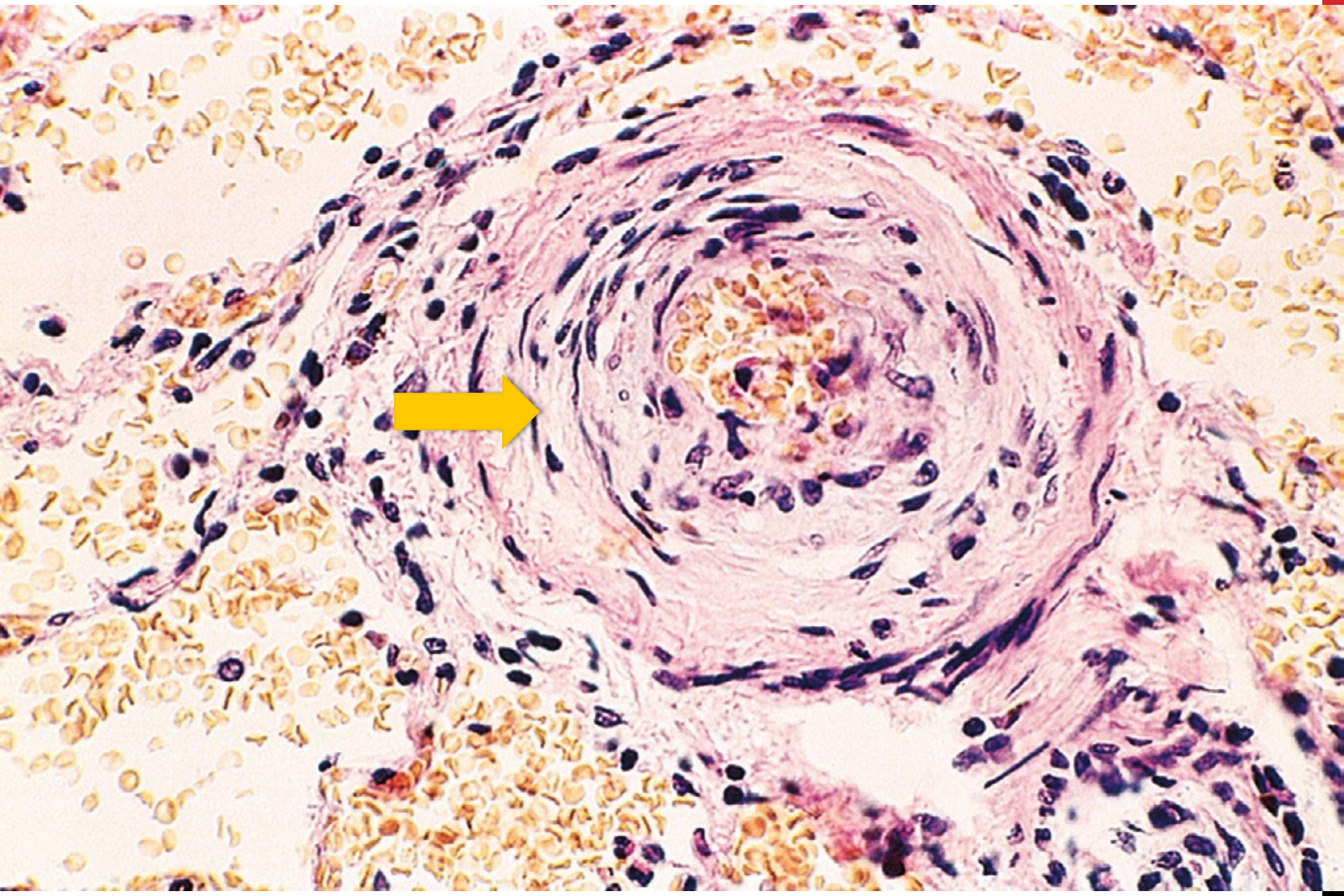
- **Chronic thromboembolic pulmonary hypertension (group 4):**

Recurrent pulmonary emboli cause pulmonary hypertension by reducing the functional cross-sectional area of the pulmonary vascular bed→ increase in pulmonary vascular resistance.

- **Pulmonary hypertension with unclear or multifactorial mechanisms (group 5)**

MORPHOLOGY:

- **Medial hypertrophy of the pulmonary muscular and elastic arteries**
- **Medial hypertrophy and intimal fibrosis of the arterioles and small arteries**
- **Right ventricular hypertrophy**
- **plexiform lesion:**
  - Uncommon
  - tuft of capillary formations producing a network within the lumens of dilated thin-walled, small arteries and may extend outside the vessel.
  - Plexiform lesions are most prominent in group 1& 2.



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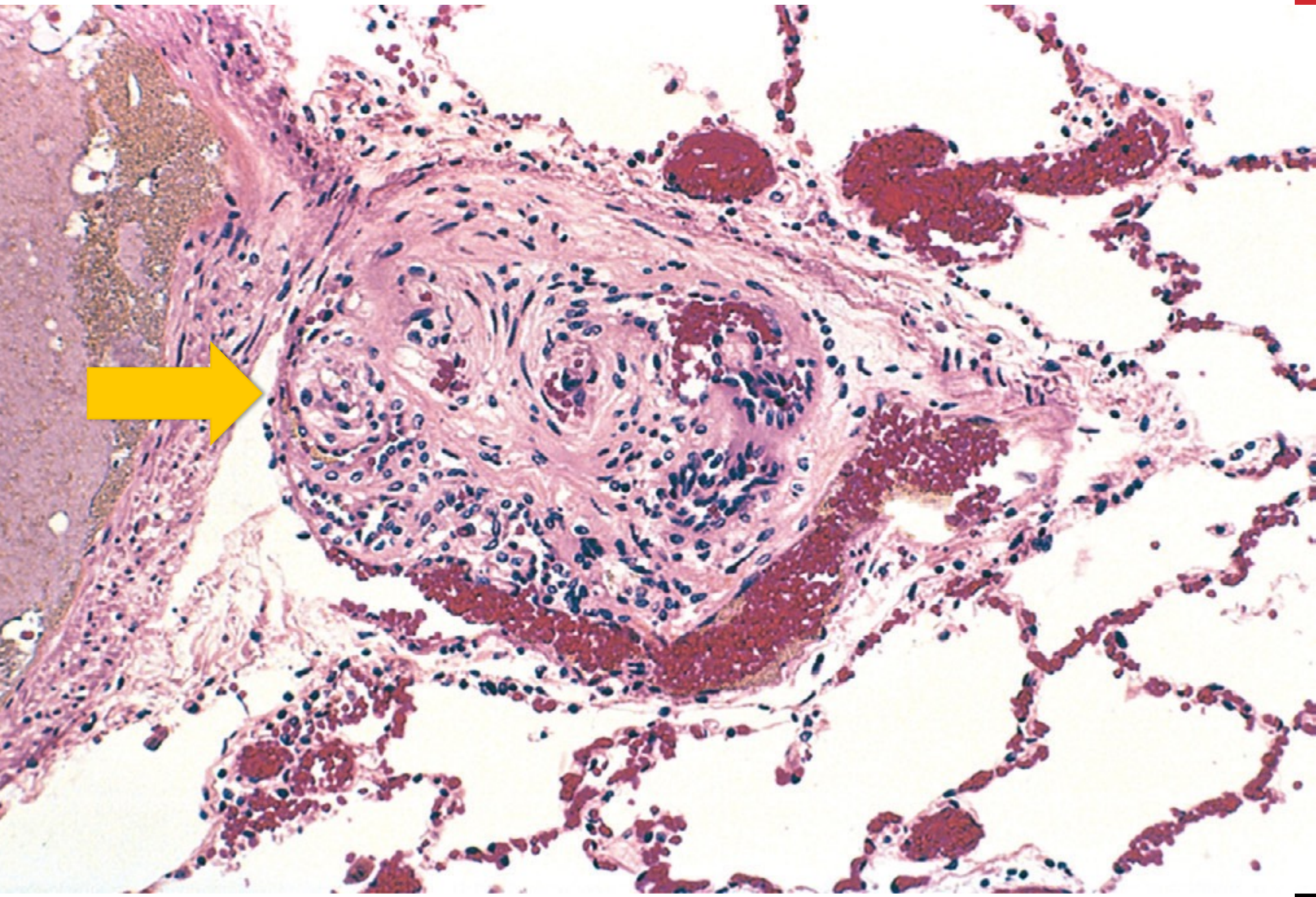

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DIFFUSE ALVEOLAR HEMORRHAGE SYNDROMES

Includes:

1. **Goodpasture syndrome**
2. **Idiopathic pulmonary hemosiderosis**
3. **vasculitis-associated hemorrhage:**
   - hypersensitivity angiitis
   - granulomatosis with polyangiitis
   - systemic lupus erythematosus

GOODPASTURE SYNDROME:

- **Called Anti–Glomerular Basement Membrane Antibody Disease With Pulmonary Involvement**

- **Is an uncommon autoimmune disease in which lung and kidney injury are caused by circulating autoantibodies against certain domains of type IV collagen.**
    - type IV collagen is intrinsic to the basement membranes of renal glomeruli and pulmonary alveoli

GOODPASTURE SYNDROME:

- The antibodies trigger destruction and inflammation of the basement membranes in pulmonary alveoli and renal glomeruli → Results in **necrotizing hemorrhagic interstitial pneumonitis and rapidly progressive glomerulonephritis**.

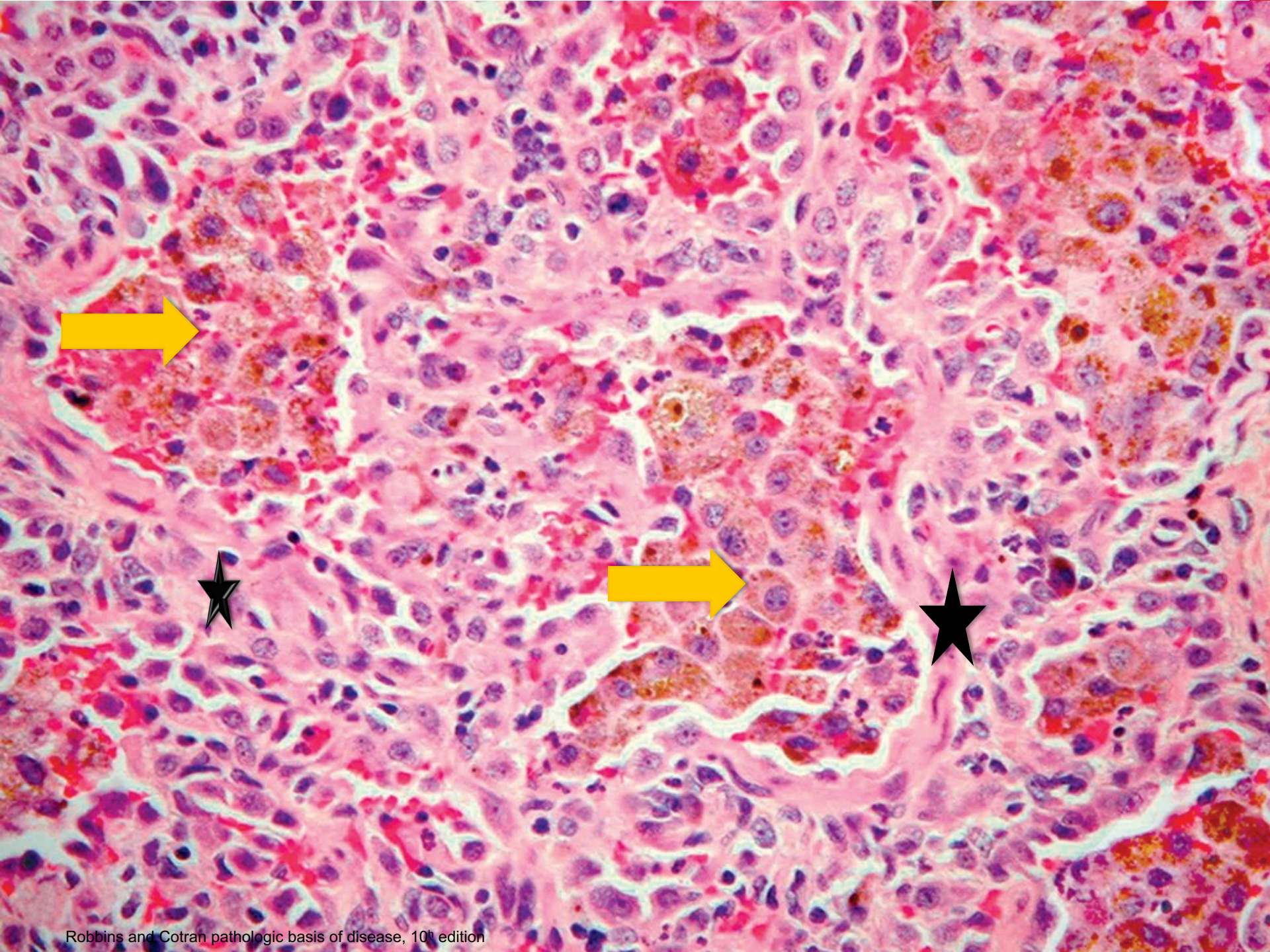
MICROSCOPICALLY:

- Focal necrosis of alveolar wall with intraalveolar hemorrhage, Abundant hemosiderin laden macrophages

Later:

- Fibrous thickening of septa , Hypertrophic type II pneumocytes and organization of blood in alveolar spaces**.**

- **DIF: Linear pattern of immunoglobulin deposition (IgG, sometimes IgA or IgM) seen along the basement membranes of alveolar septa.**



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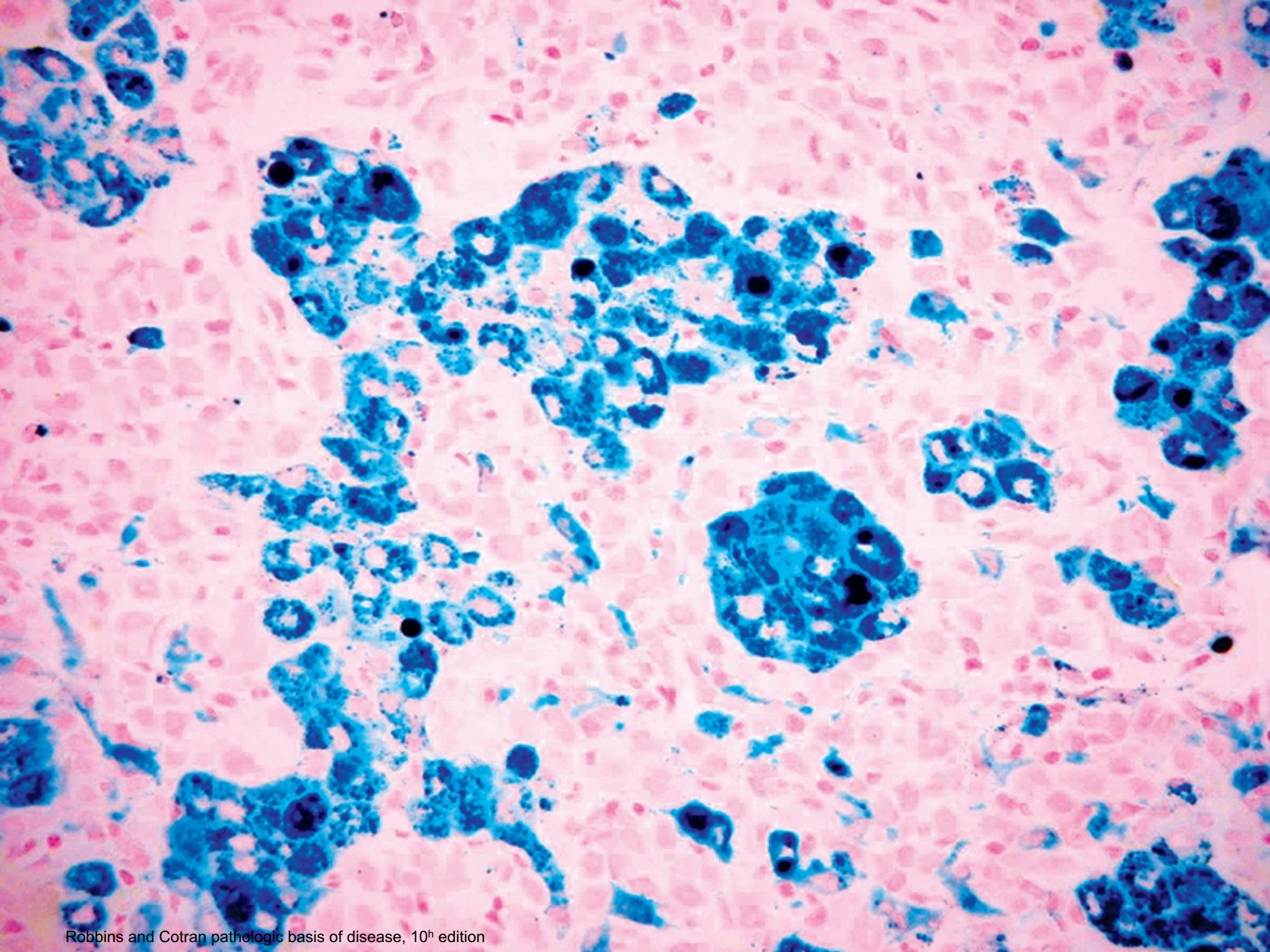

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CLINICAL FEATURES:

- Males> females, Teens and twenties, Active smokers
- Most cases begin with hemoptysis
- Soon, manifestations of glomerulonephritis appear, leading to rapidly progressive renal failure.
- Plasmapheresis and immunosuppressive therapy , renal transplantation

GRANULOMATOSIS WITH POLYANGIITIS (GPA):

- Formerly called **Wegener granulomatosis**

- **Triad of:**
  - necrotizing angiitis
  - aseptic necrosis of upper respiratory tract and lungs
  - focal glomerulonephritis (necrotizing, often crescentic, glomerulonephritis)

- **Lung histology:**
  - necrotizing angiitis of arteries and veins
  - parenchymal necrotizing poorly formed granulomatous inflammation.

- signs and symptoms of the upper-respiratory tract involvement (chronic sinusitis, epistaxis, nasal perforation) and the lungs (cough, hemoptysis, chest pain).

- Anti-neutrophil cytoplasmic antibodies (PR3- ANCAs) are present in close to 95% of cases

THANK YOU!