PULMONARY DISEASES OF VASCULAR ORIGIN

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PULMONARY DISEASES OF VASCULAR ORIGIN

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

THROMBOEMBOLISM:

- Almost all large pulmonary artery thrombi are embolic in Origin.

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

RISK FACTORS FOR VENOUS THROMBOSIS:

1. prolonged bed rest
2. Surgery
3. severe trauma
4. congestive heart failure
5. in women, the period around parturition or the use of OCPs
6. disseminated cancer
7. primary disorders of hypercoagulability

CONSEQUENCES

1. increase in pulmonary artery pressure and vasospasm

2. ischemia of the downstream pulmonary parenchyma.

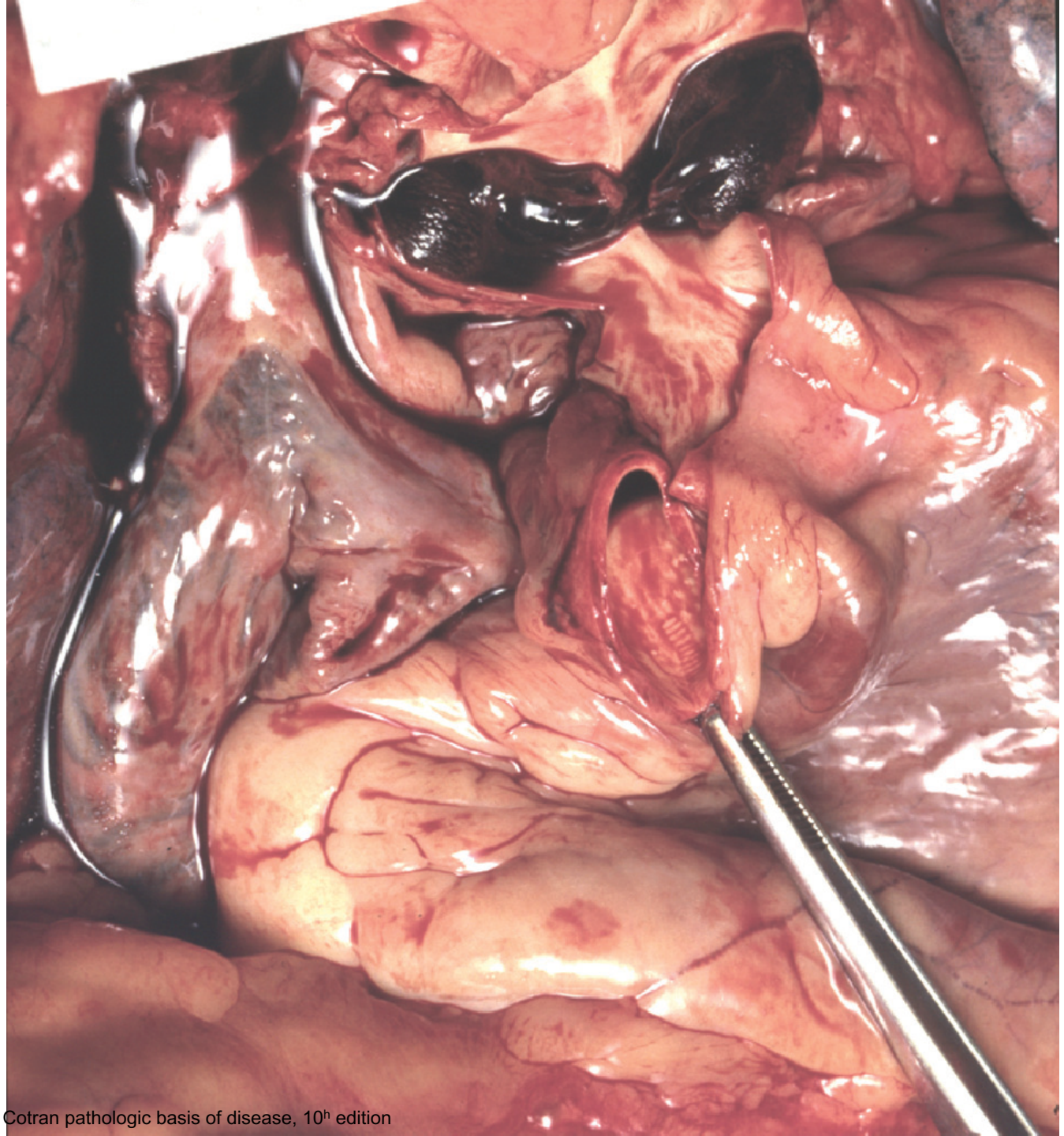
CONSEQUENCES:

- **depend mainly on:**

1- size of the embolus:

- 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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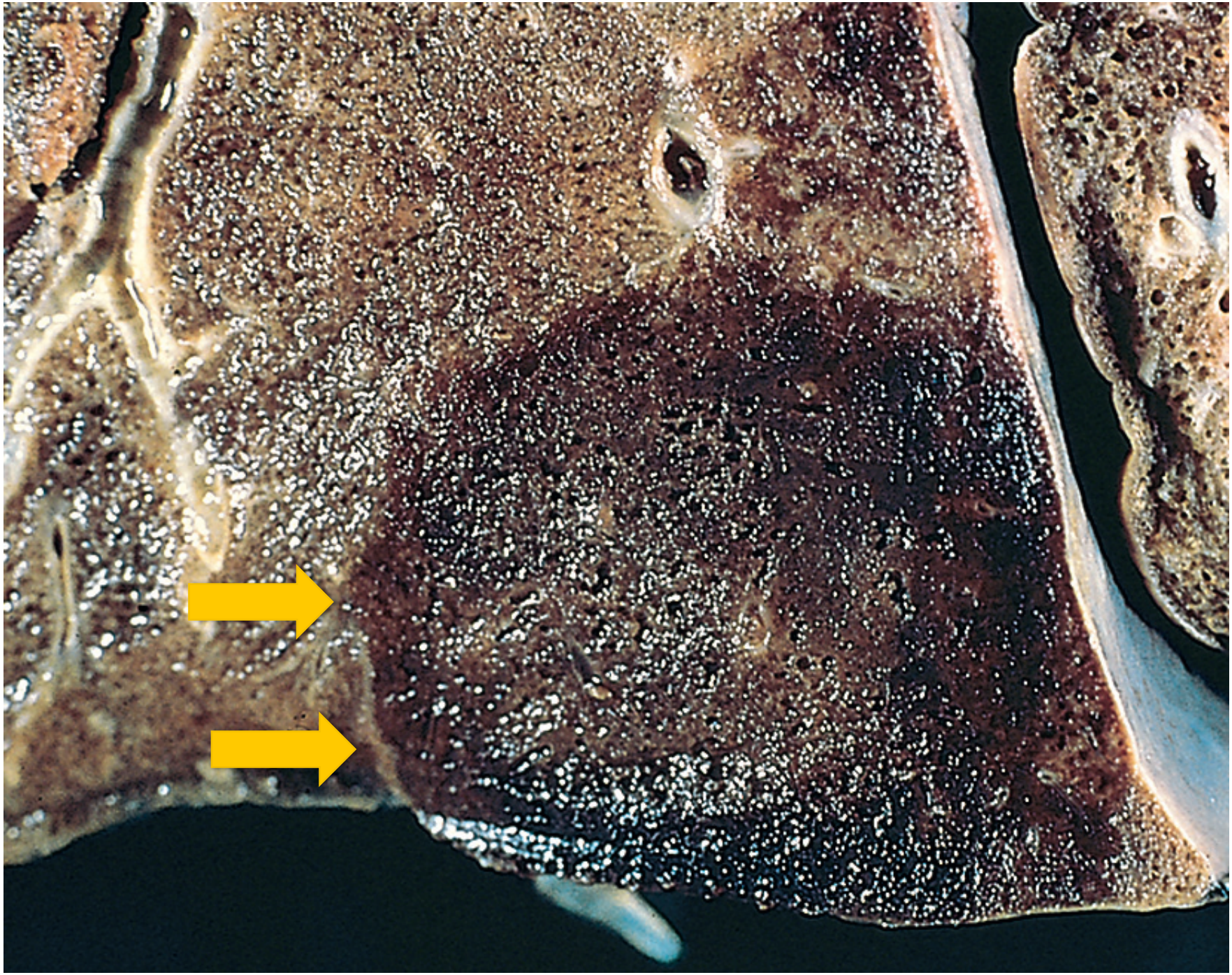
MORPHOLOGY:

- **No morphologic alternations**: large emboli
- **alveolar hemorrhage**: Smaller emboli
- **infarction :**
  - compromised cardiovascular status.
  - 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.

MORPHOLOGY:

- hemorrhagic raised
- red-blue areas of coagulative necrosis in the early stages
- fibrinous exudate covering adjacent pleura
- The occluded vessel is located near the apex of the infarcted area.
- The red cells begin to lyse within 48 hrs→ red-brown as hemosiderin is produced → fibrous replacement begins at the margins as a gray-white peripheral zone →scar.



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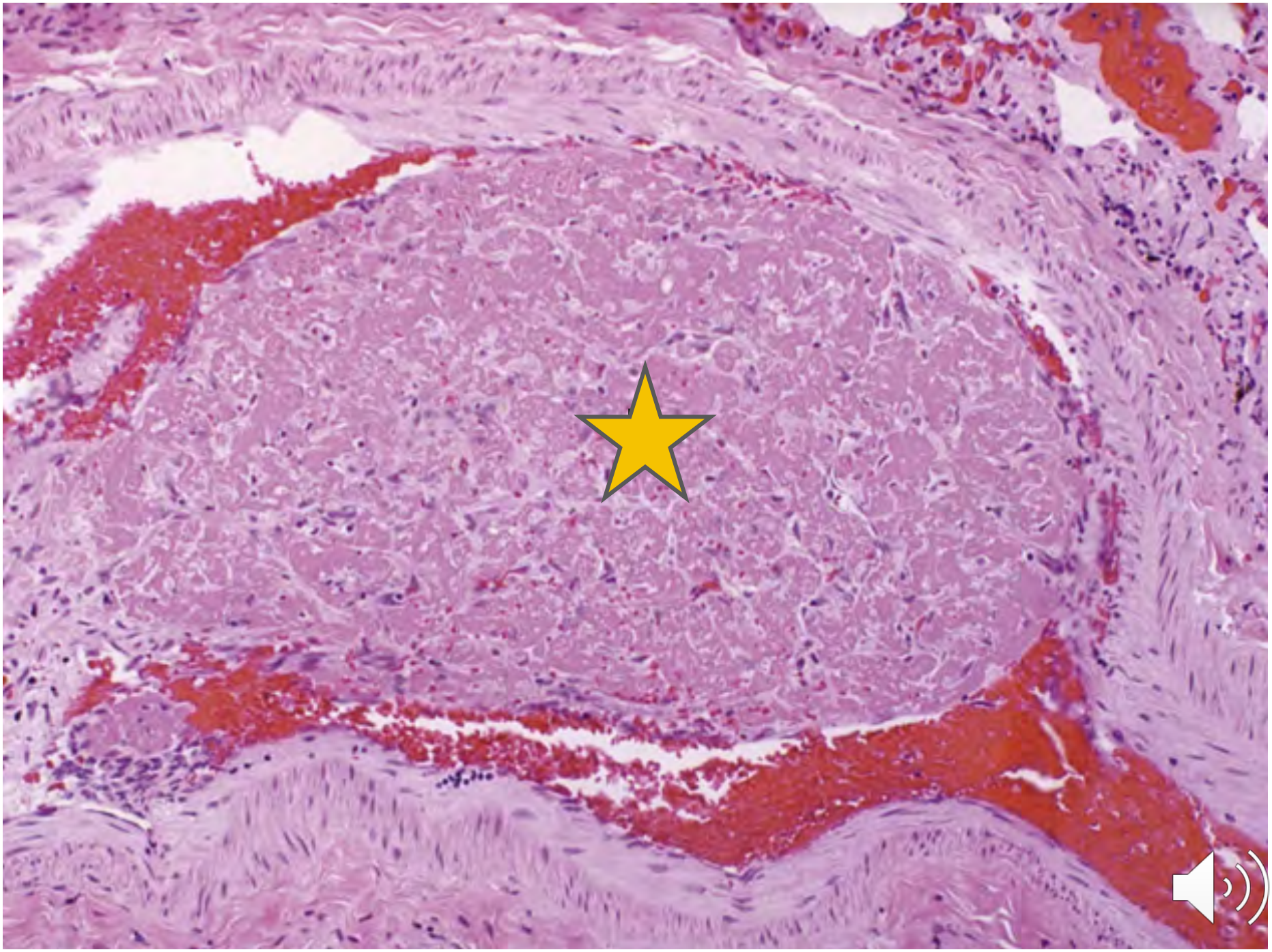


CLINICAL FEATURES

- 60% - 80% →clinically silent
- 5%→ death, acute right-sided heart failure, or cardiovascular collapse.
- 10-15%→ dyspnea
- <3% → pulmonary hypertension, chronic right-sided heart failure, and, pulmonary vascular sclerosis with progressively worsening of dyspnea.

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: hemodynamically unstable patients with massive pulmonary embolism

NONTHROMBOTIC PULMONARY EMBOLI:

- uncommon but potentially lethal
- such as air, fat, amniotic fluid embolism, and foreign body embolism in intravenous drug abusers
- Bone marrow embolism (the presence of hematopoietic and fat elements within a pulmonary artery)
- 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 a **decrease** in the cross-sectional area of the pulmonary vascular bed or by **increased** pulmonary vascular blood flow.

CLASSIFIED AS FOLLOWING:

- **Pulmonary arterial hypertension:**
  - heritable forms of pulmonary hypertension and diseases that cause pulmonary hypertension by affecting small pulmonary muscular arterioles;
    - Examples: connective tissue diseases, human immunodeficiency virus, and congenital heart disease with left to right shunts

- **Pulmonary hypertension due to left-sided heart disease:**
  - including systolic and diastolic dysfunction and valvular disease

- **Pulmonary hypertension due to lung diseases and/or hypoxia**:
    - including COPD and interstitial lung disease
- **Chronic thromboembolic pulmonary hypertension**
- **Pulmonary hypertension with unclear or multifactorial mechanisms**

PATHOGENESIS

- *Chronic obstructive or interstitial lung diseases*
- *congenital or acquired heart disease*
- *Recurrent thromboemboli*
- *Autoimmune diseases*

- **Obstructive sleep apnea**

- **idiopathic pulmonary arterial hypertension**
  - Also called primary pulmonary hypertension
  - Uncommon
  - all known causes are excluded
  - Up to 80% has a genetic basis:
    - inherited in families as an autosomal dominant trait. incomplete penetrance

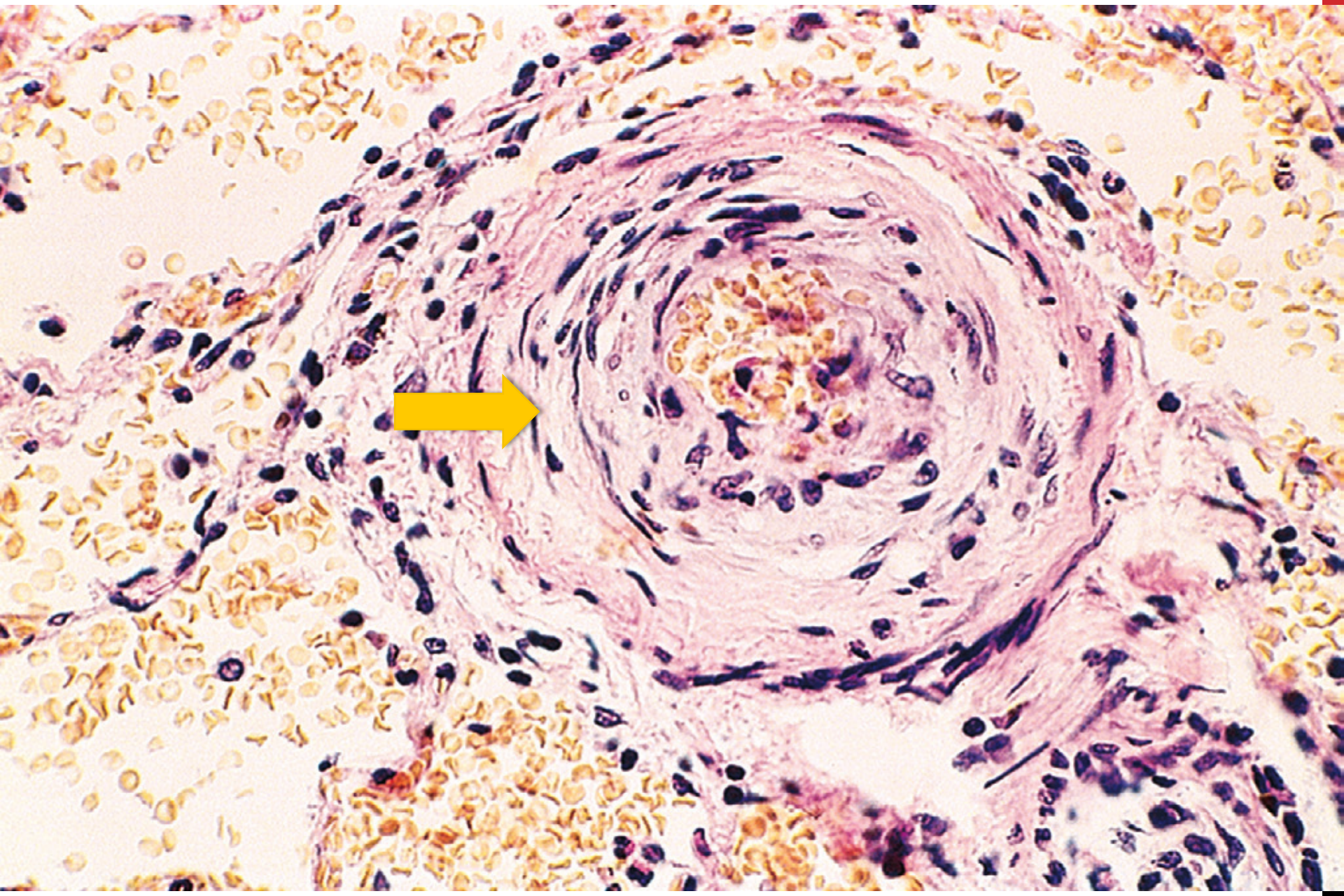
MORPHOLOGY:

- **Medial hypertrophy of the pulmonary muscular and elastic arteries**
    - arterioles and small arteries
- **Pulmonary arterial atherosclerosis**
    - pulmonary artery and its major branches
- **Right ventricular hypertrophy**

- **Plexiform lesion:**

  - uncommon

  - a tuft of capillary formations producing a network, or web, that spans the lumens of dilated thin-walled, small arteries and may extend outside the vessel.



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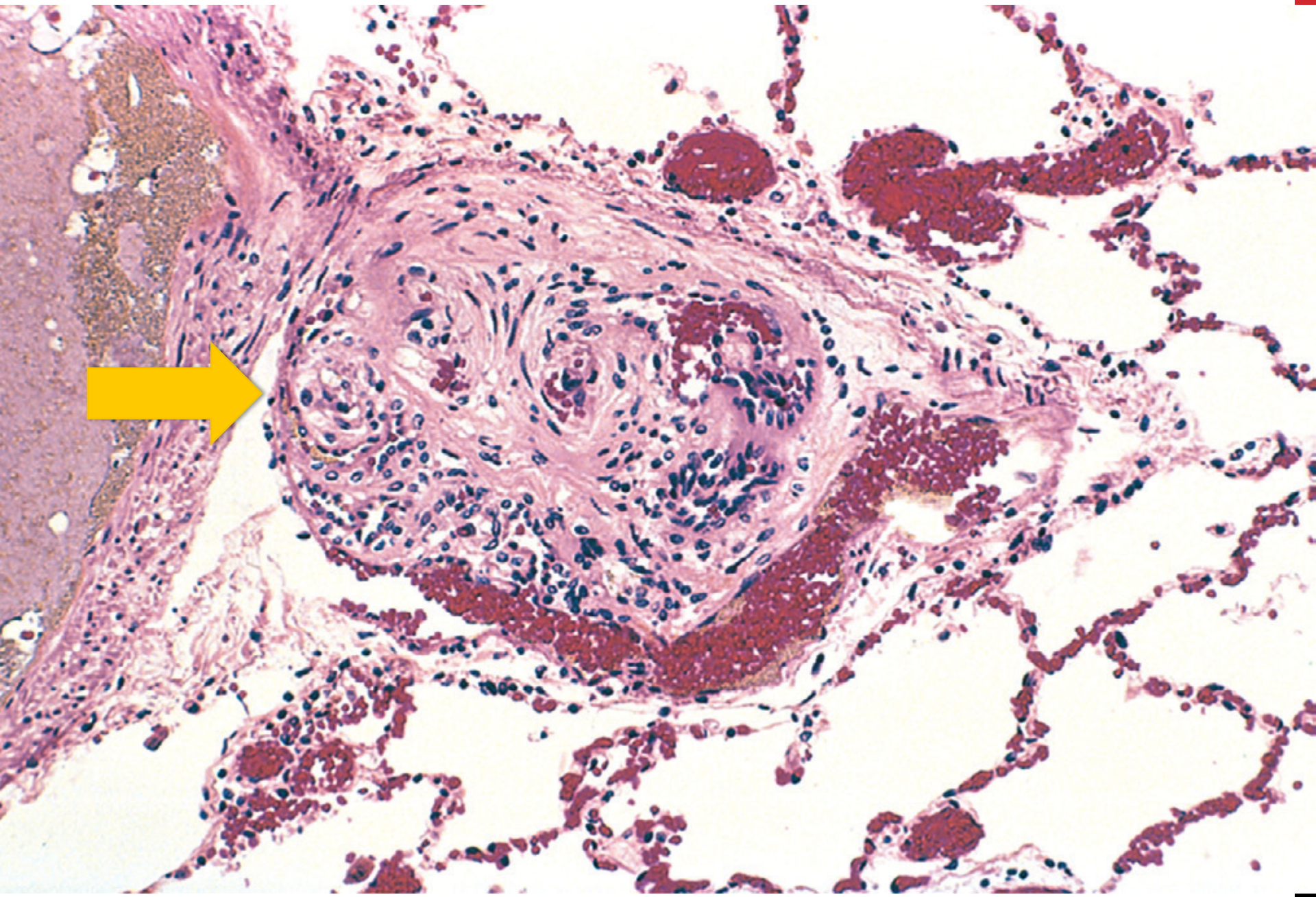

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CLINICAL FEATURES:

- **Asymptomatic**
- women 20-40 & young children
- dyspnea and fatigue, anginal chest pain
- 80% of patients within 2-5 years→ respiratory distress, cyanosis, and right ventricular hypertrophy and death from decompensated cor pulmonale

DIFFUSE ALVEOLAR HEMORRHAGE SYNDROMES

- Complication of some interstitial lung disorders.
- Includes:

1. Goodpasture syndrome
2. Idiopathic pulmonary hemosiderosis
3. Granulomatosis with polyangiitis

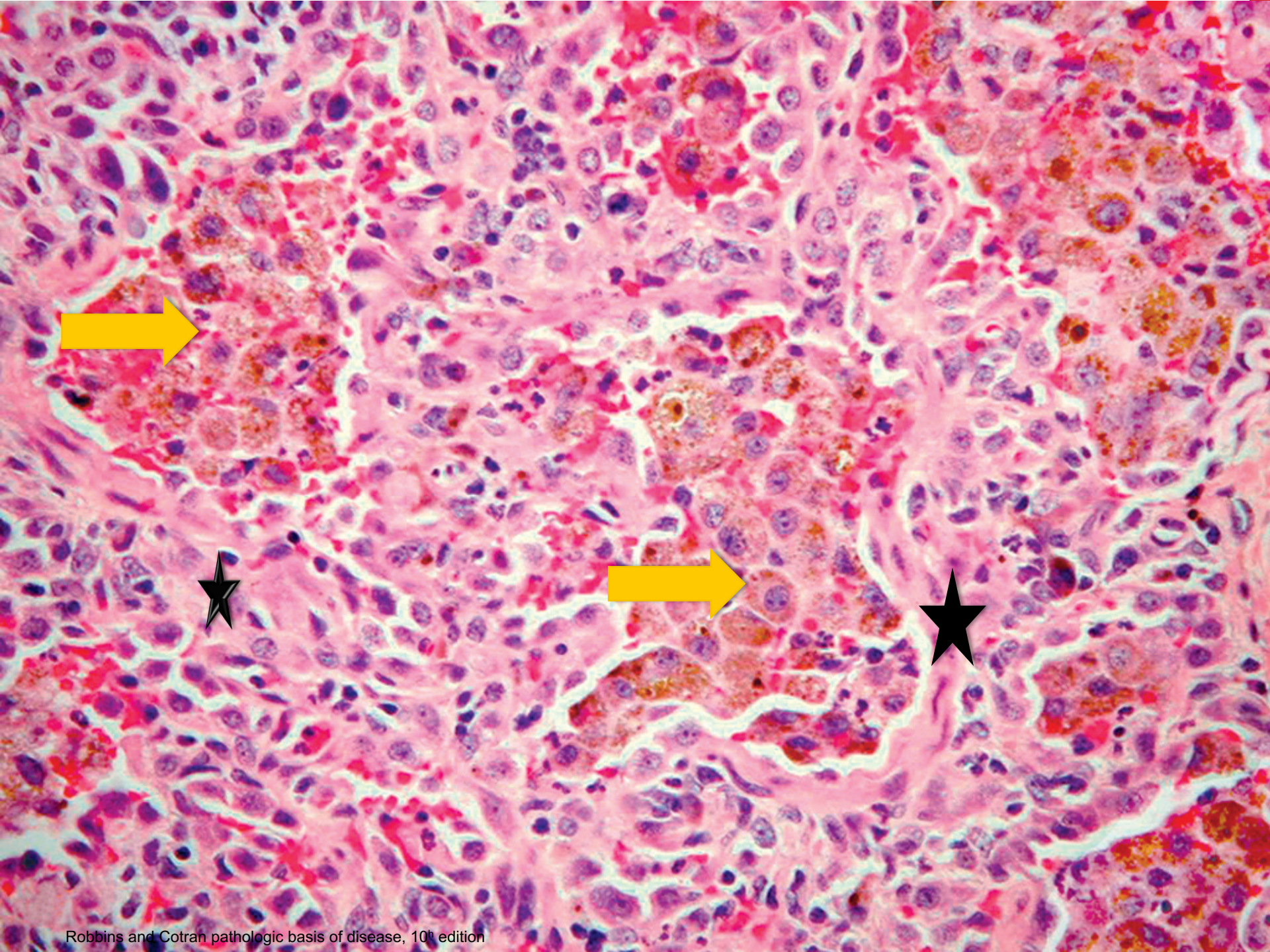
GOODPASTURE SYNDROME:

- **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
- **Results in necrotizing hemorrhagic interstitial pneumonitis and rapidly progressive glomerulonephritis.**

MORPHOLOGY:

- **Grossly,** red-brown consolidation due to **diffuse alveolar hemorrhage**

- **Microscopically:**
  - Focal necrosis of alveolar wall with intraalveolar hemorrhage,
  - Fibrous thickening of septa,
  - Hypertrophic type II pneumocytes.
  - Abundant hemosiderin
  - Linear pattern of immunoglobulin deposition (IgG, sometimes IgA or IgM) seen along the alveolar septa.



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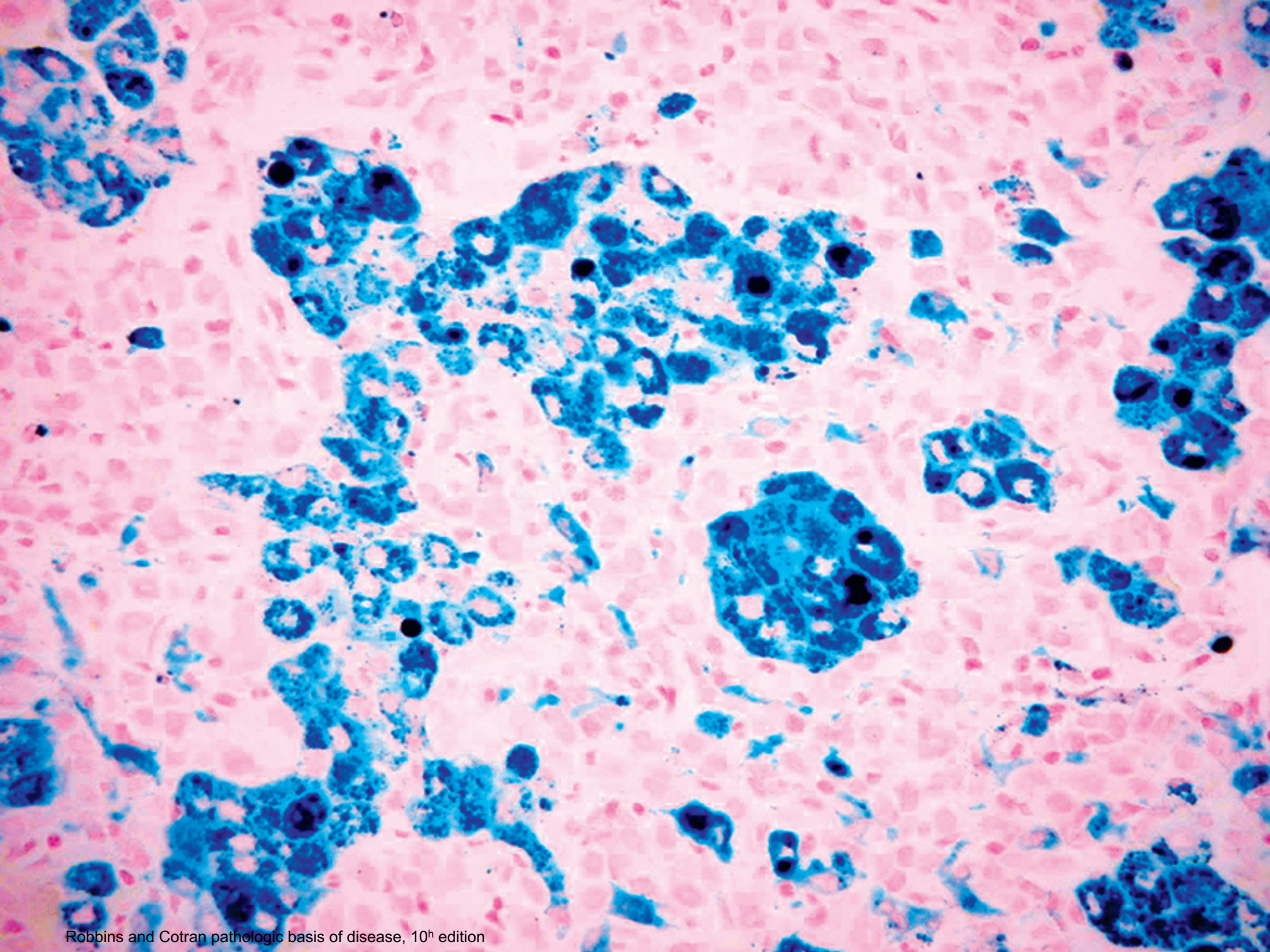

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CLINICAL FEATURES:

- Teens and twenties
- Males > females
- Active smokers
- Plasmapheresis and immunosuppressive therapy , renal transplantation

GRANULOMATOSIS AND POLYANGIITIS

- Formerly called **Wegener granulomatosis**
- >80% of patients develop upper-respiratory or pulmonary manifestations.
- The lung lesions are characterized by a combination of necrotizing vasculitis (“angiitis”) and parenchymal necrotizing granulomatous inflammation.

- The signs and symptoms stem from involvement of the upper-respiratory tract (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

A 45-year-old gentleman has chronic cough for the last 11 months. Physical examination, shows nasopharyngeal ulcers. on auscultation, the lungs have diffuse crackles bilaterally. Laboratory studies include a serum urea nitrogen level of 75 mg/dL and a creatinine concentration of 6.7 mg/dL. Urinalysis shows 50 RBCs per high-power field and RBC casts. His serologic titer for C-ANCA (proteinase 3) is elevated. A chest radiograph shows multiple, small, bilateral pulmonary nodules. A transbronchial lung biopsy specimen shows a necrotizing inflammatory process involving the small peripheral pulmonary arteries and arterioles. Which of the following is the most likely diagnosis?

A. **Granulomatosis with polyangiitis**

B. **Pulmonary hypertension**

C. **Goodpasture syndrome**

D. **Idiopathic pulmonary hemosiderosis**

E. **Polyarteritis nodosa**

ANOTHER CASE?!

A 33-year-old gentleman, medically free, presented with acute onset of hemoptysis. he is afebrile, with normal heart rate, increased respiratory rate and blood pressure. A transbronchial lung biopsy, shows focal necrosis of alveolar walls associated with prominent intraalveolar hemorrhage. Two days later, he has decreased urine output with abnormal serum creatinine and urea nitrogen. Which of the following antibodies is most likely involved in the pathogenesis of his condition?

A **Anti–DNA topoisomerase I antibody**

B **Anti–glomerular basement membrane antibody**

C **Antimitochondrial antibody**

D **Anti–neutrophil cytoplasmic antibody**

E **Antinuclear antibody**

FOR YOUR QUESTIONS:

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Or E-learning

THANK YOU!