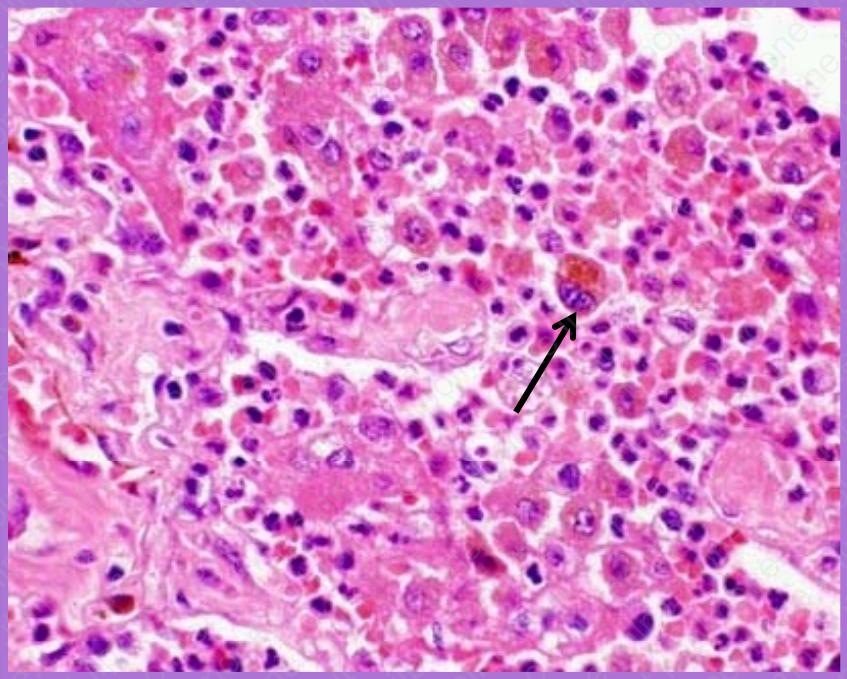
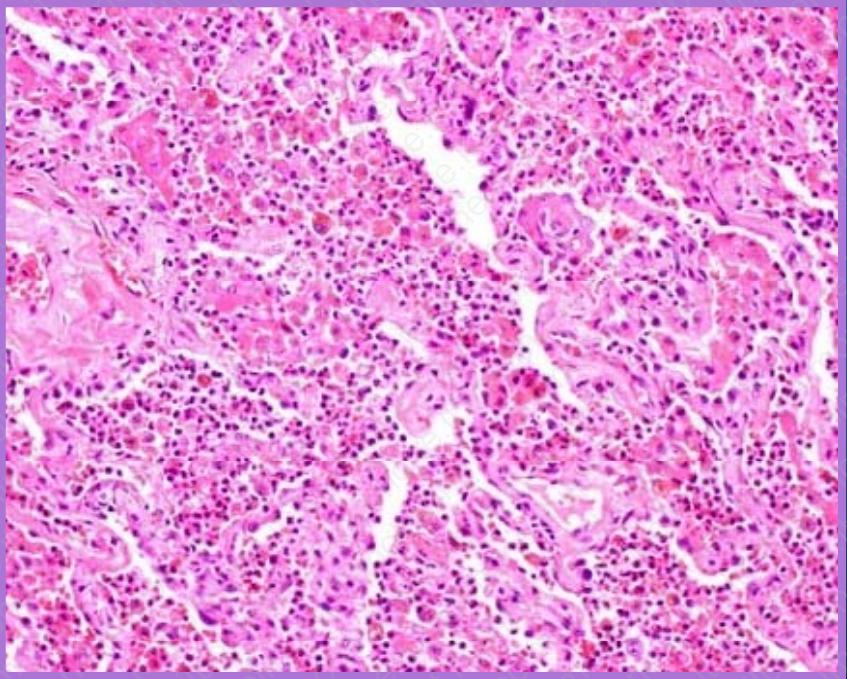
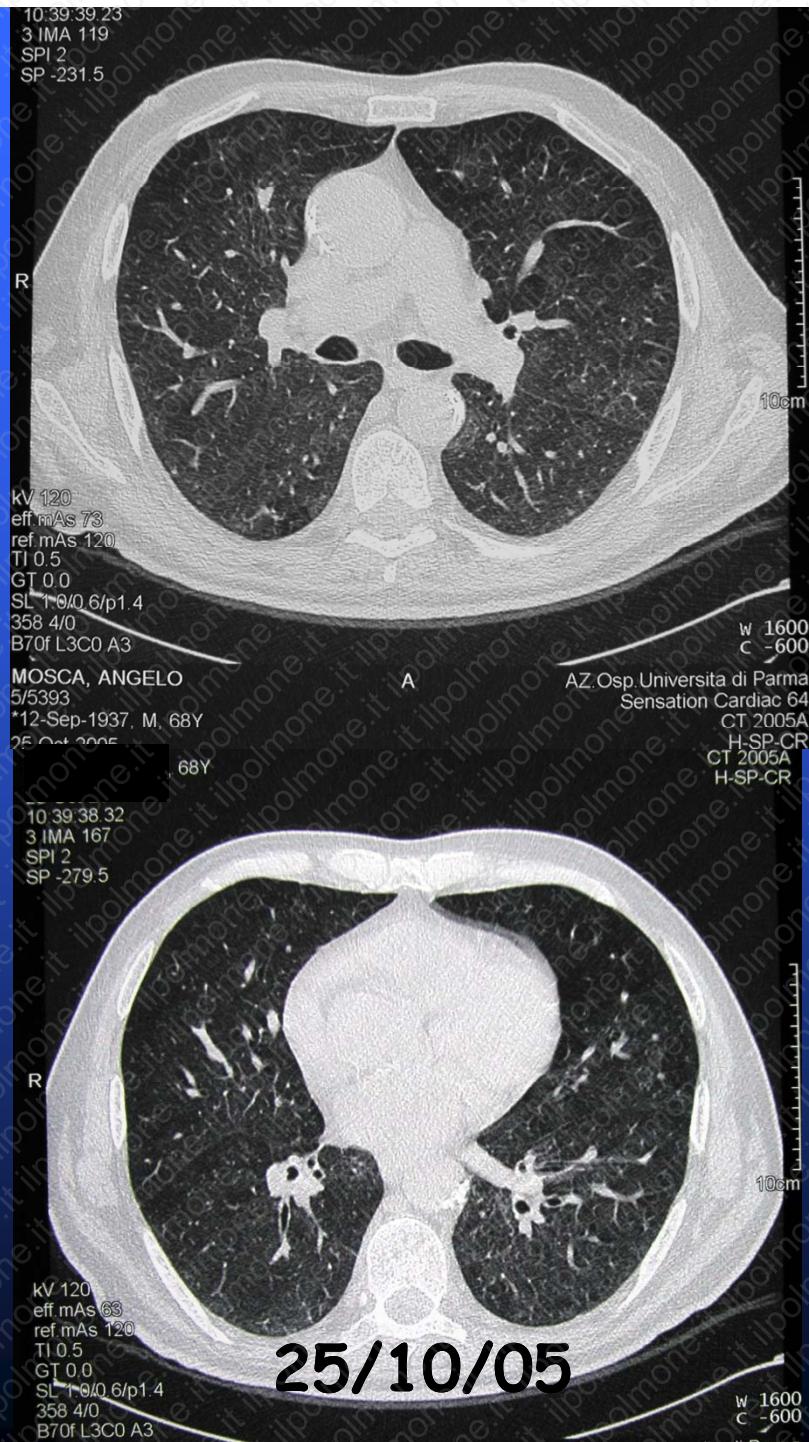
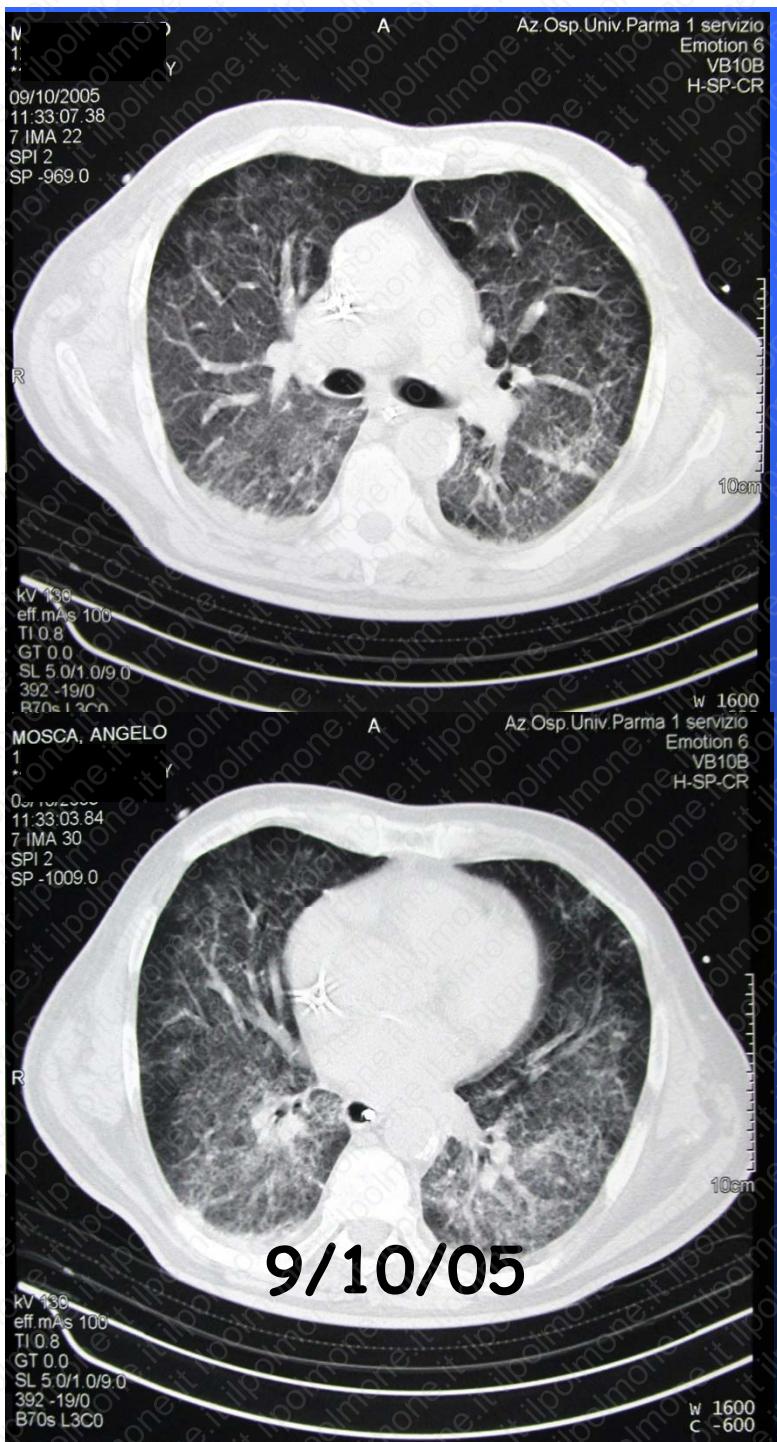
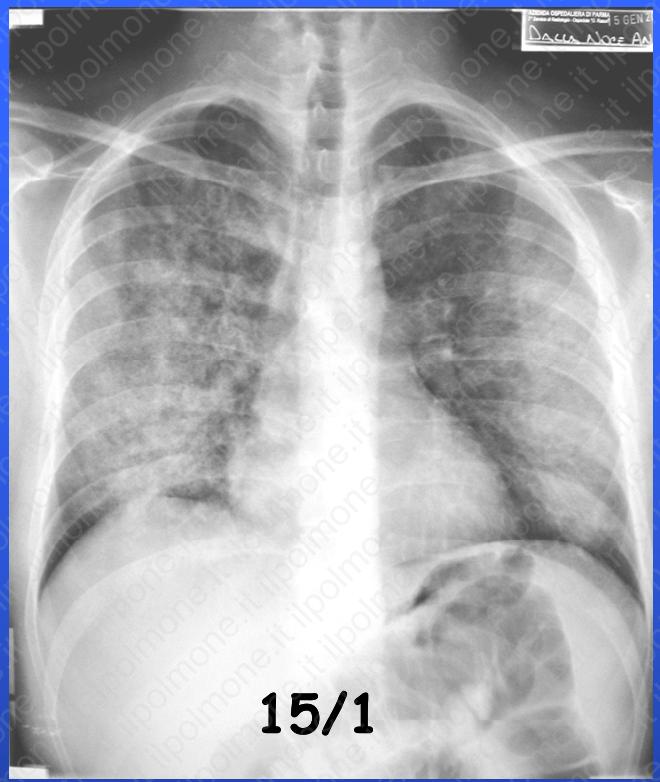


Diffuse Alveolar Haemorrhage

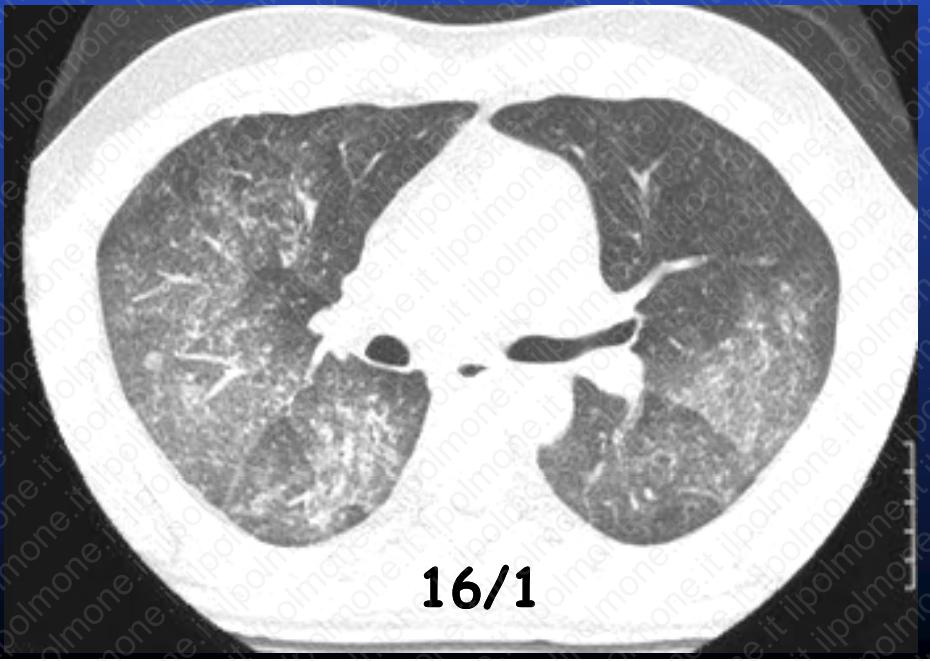


Diffuse alveolar hemorrhage



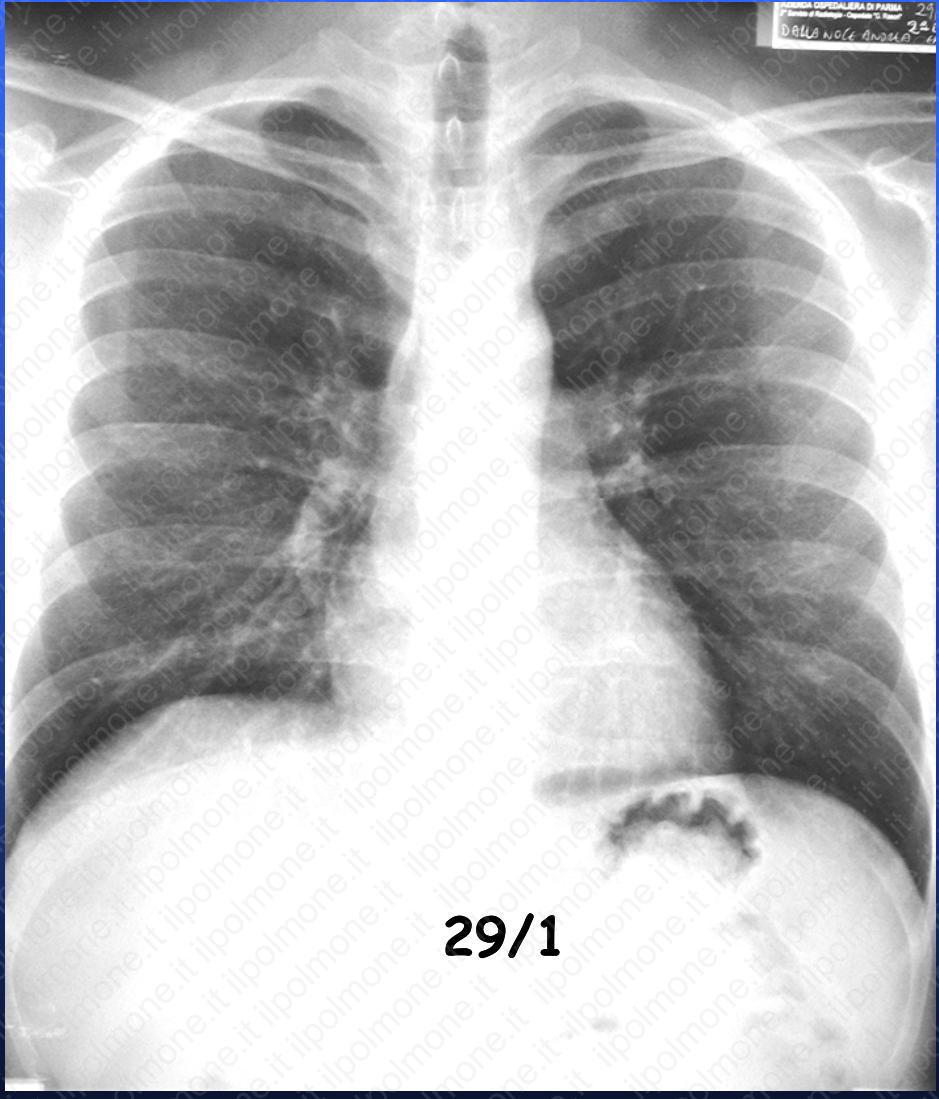


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Diffuse alveolar hemorrhage



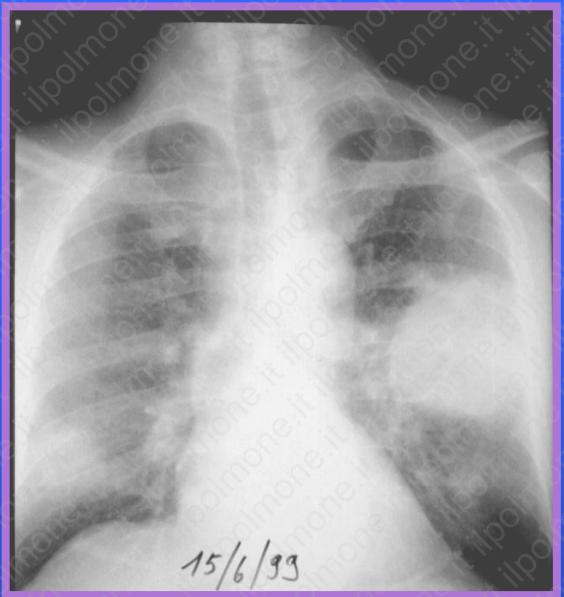
29/1

Comments

- DAH can be the *presenting* manifestation of WG
 - DAH is the result of *alveolar capillaritis*
 - DAH can cause *acute respiratory failure* requiring *mechanical ventilation*
 - *A rapid Hb drop and alveolar-interstitial pulmonary infiltrates on a chest radiograph should heighten suspicion of DAH, even in absence of hemoptysis or dyspnoea*
-



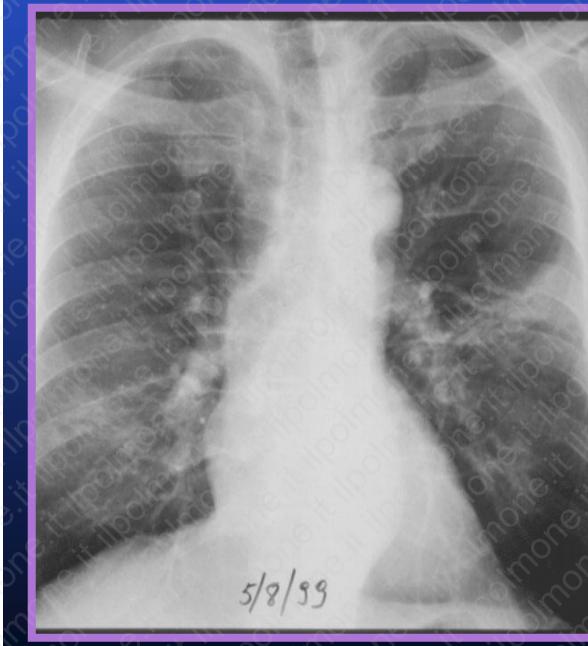
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3/7/99



5/8/99



Wegener's Granulomatosis

Reversibility
after treatment

Wegener's Granulomatosis

Renal involvement

Renal involvement

- ◆ *Rapidly progressive glomerulonephritis*
 - ◆ *Isolated urinary abnormalities*
 - ◆ *Chronic renal failure*
-

Renal biopsy

- ◆ *Pauci-immune crescentic GN*
-

Wegener's Granulomatosis

Renal involvement

Rapidly progressive glomerulonephritis

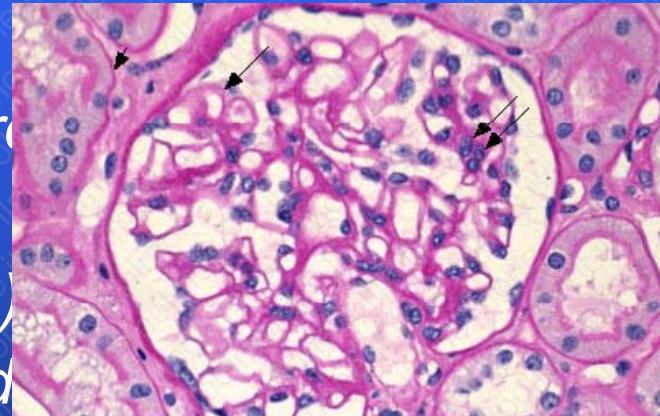
- ◆ *active urinary sediment*
- ◆ *serum creatinine ($\uparrow 0.7\text{-}1 \text{ mg\%}/\text{die}$)*
- ◆ *microscopic or macroscopic hematuria*
- ◆ *Proteinuria*
- ◆ *oligo-anuria*
- ◆ *Renal ultrasoud (enlarged and hyperechoic kidneys)*

Wegener's Granulomatosis

Renal biopsy

Glomerulonephritis

- ❖ *Crescentic (crescents in different types of glomerular lesions: fibrin, cellular and sclerotic)*
- ❖ *Necrotising (fibrinoid necrosis)*
- ❖ *pauci-immune (rare immune deposits)*



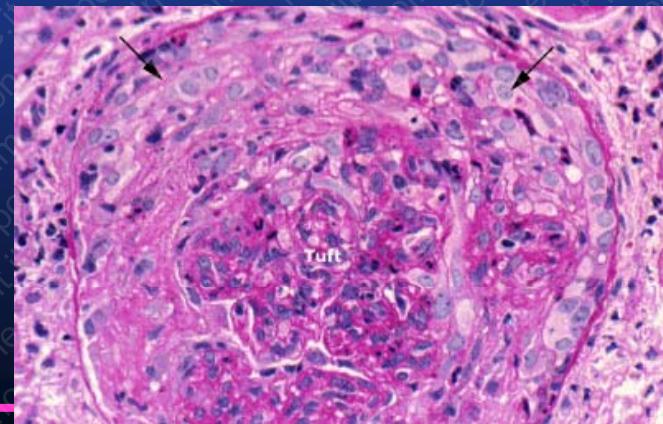
Normal glomerulus

Vasculitis (less frequent)

- ❖ *Interlobular arteries*
- ❖ *Arterioles*

Granuloma (rare)

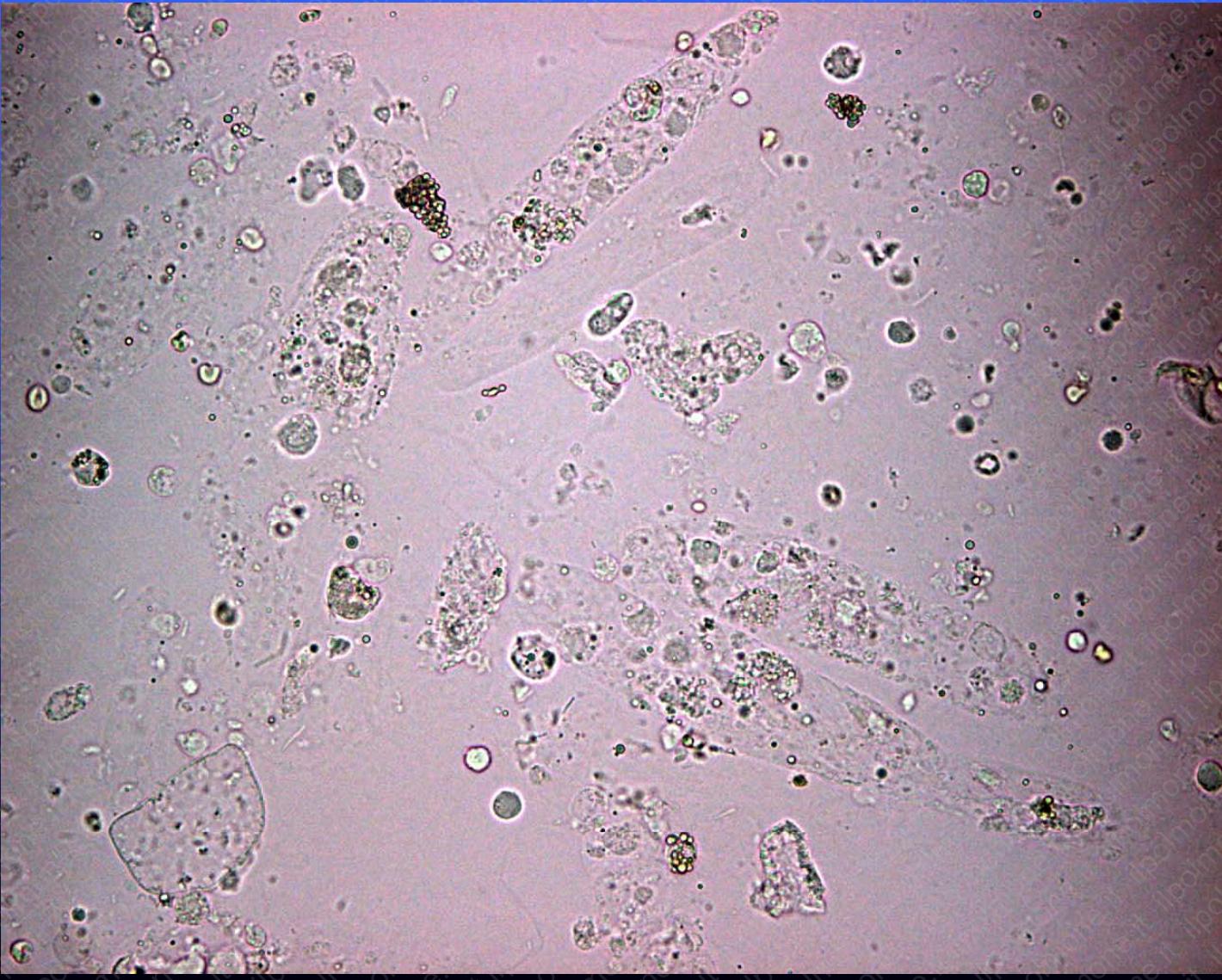
RPGN/Crescentic GN

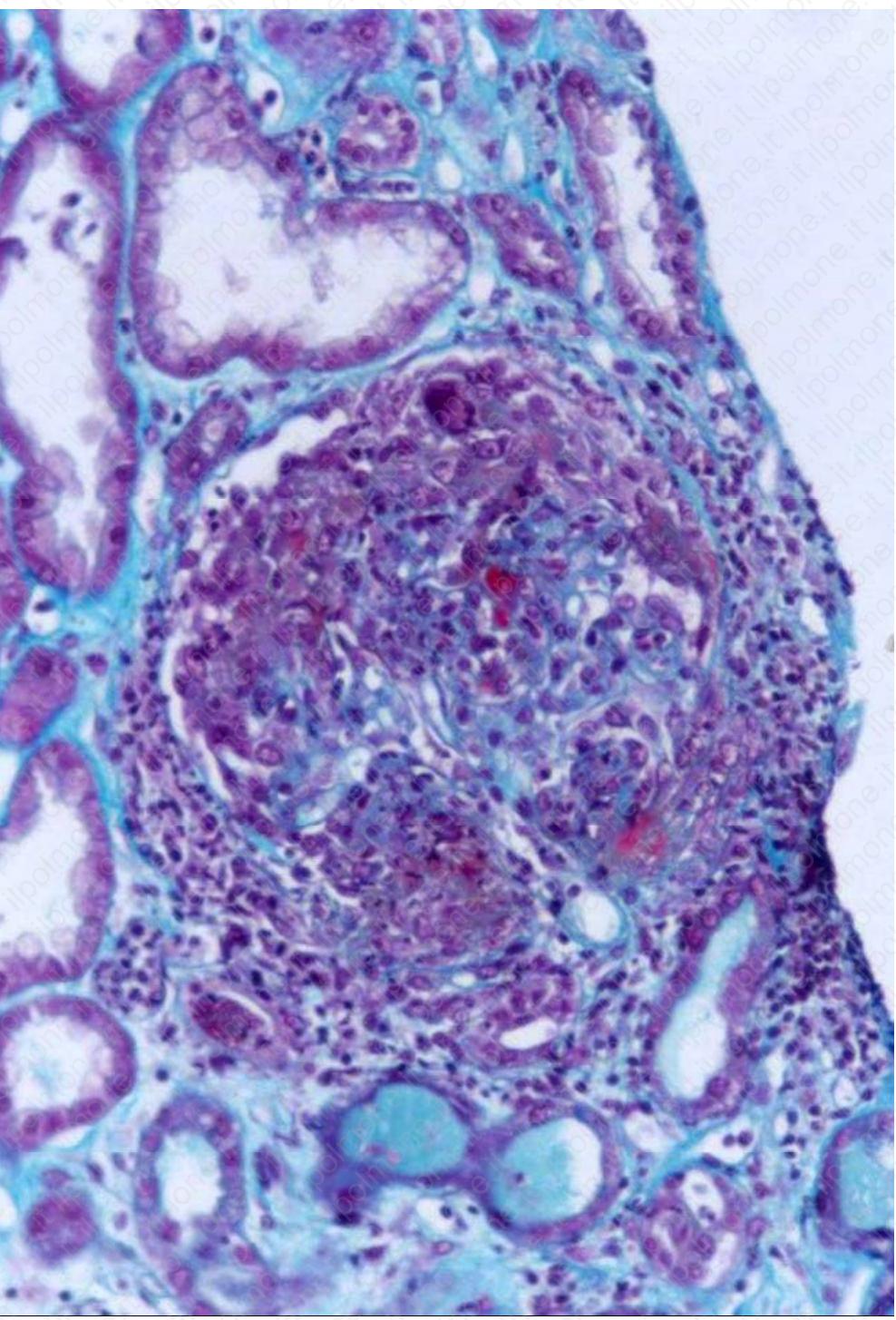
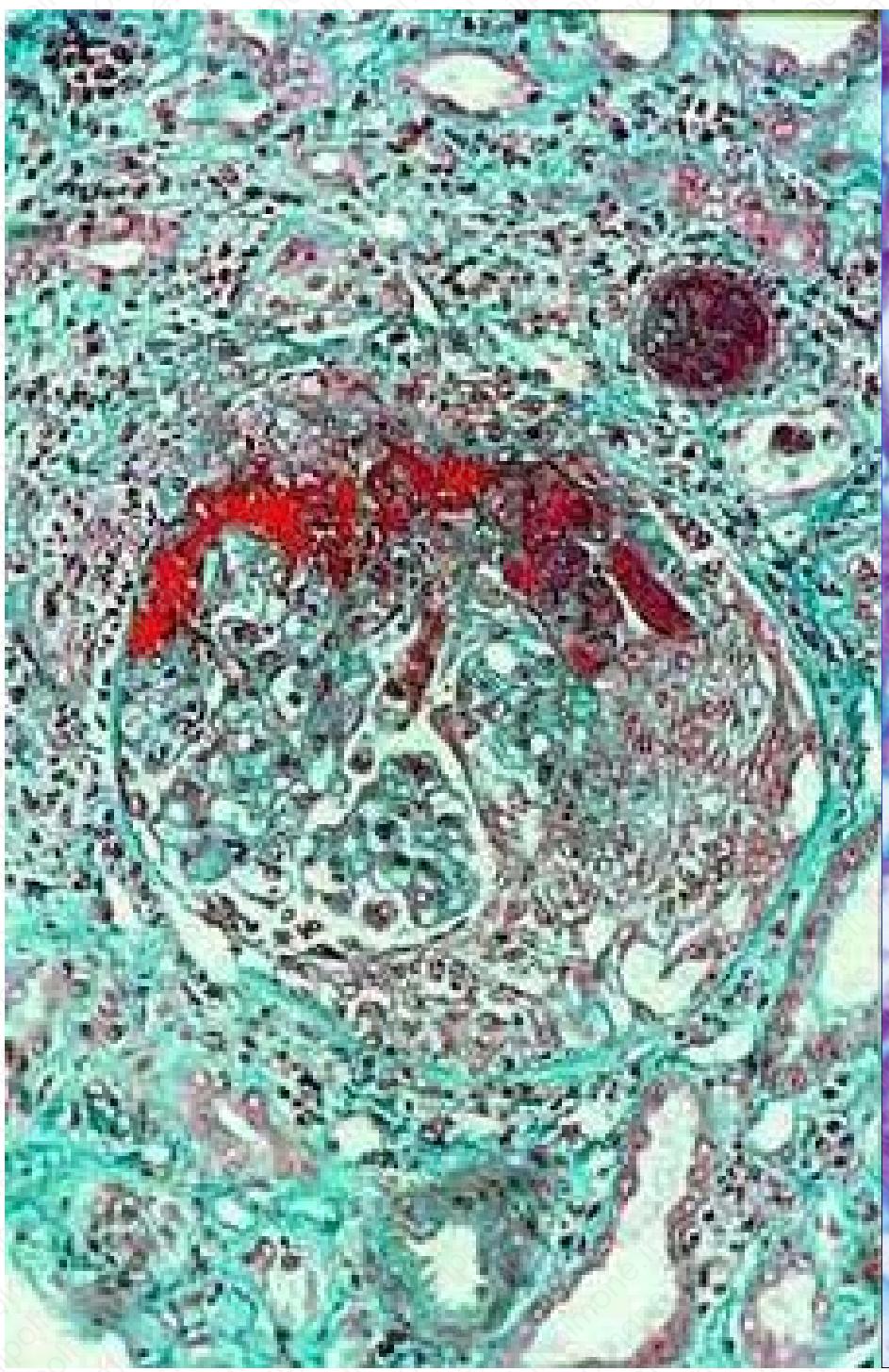


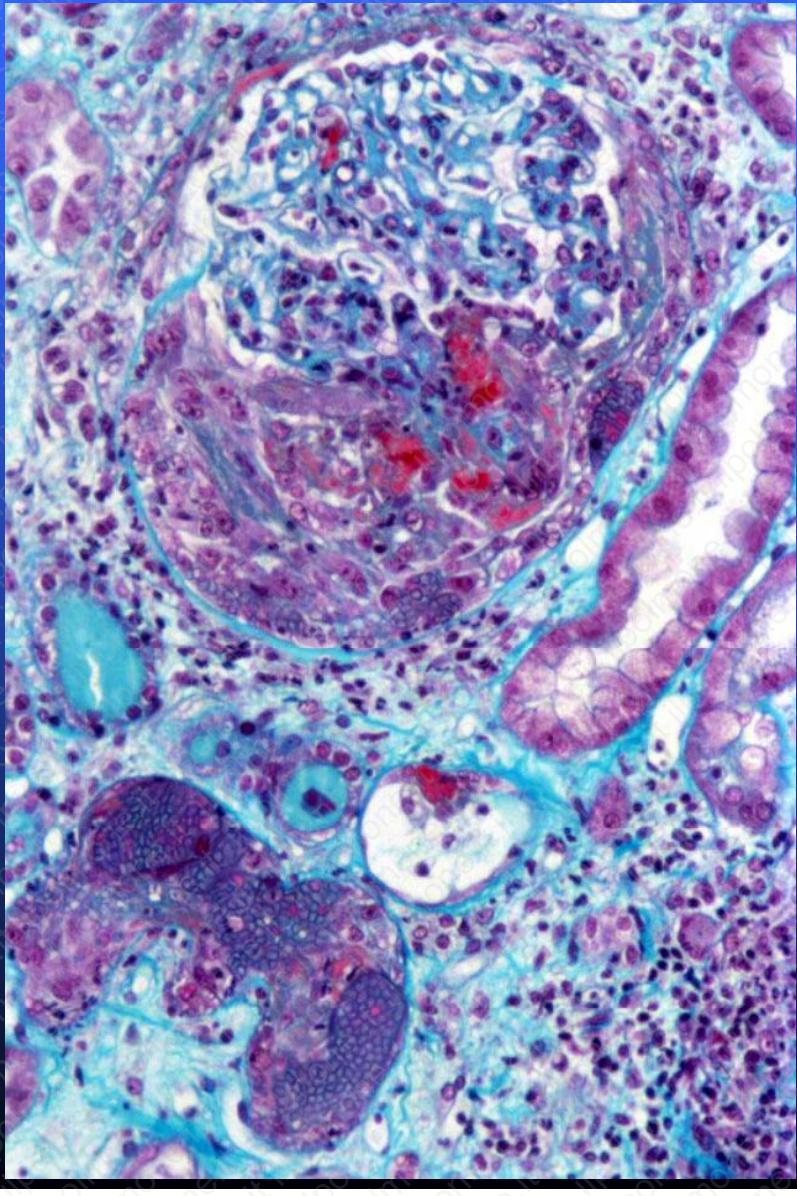
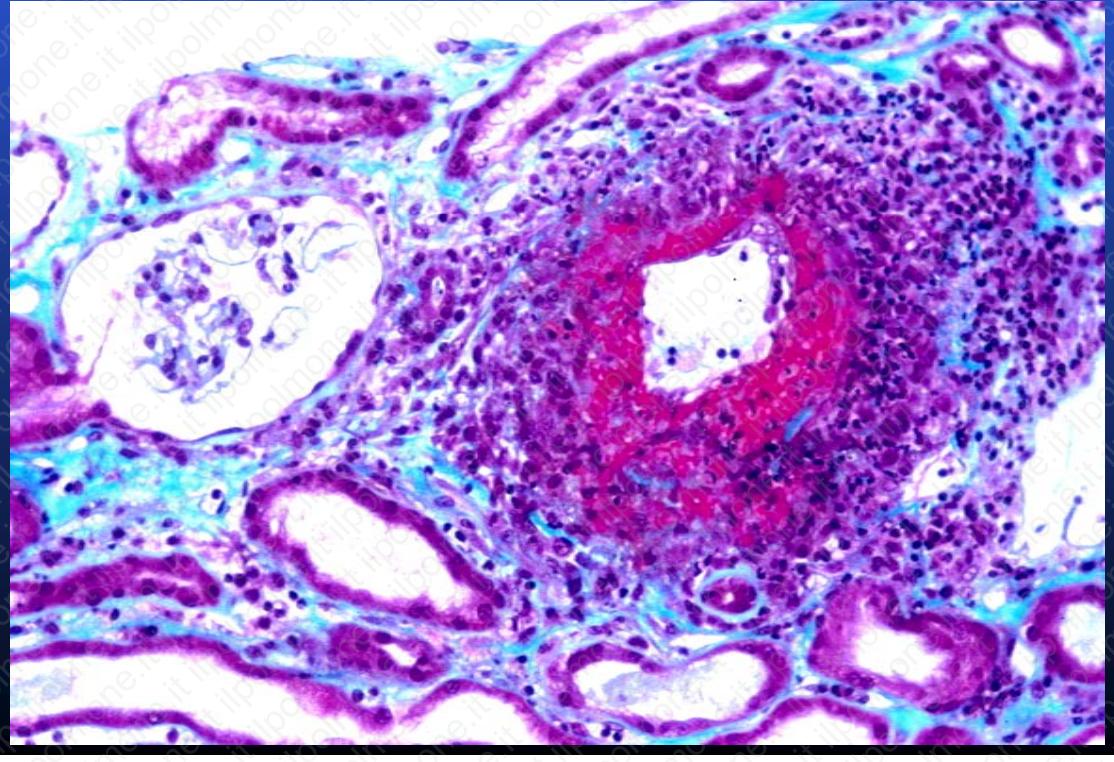
Rapidly progressive glomerulonephritis

Active urine sediment

Pauci-immune crescentic glomerulonephritis

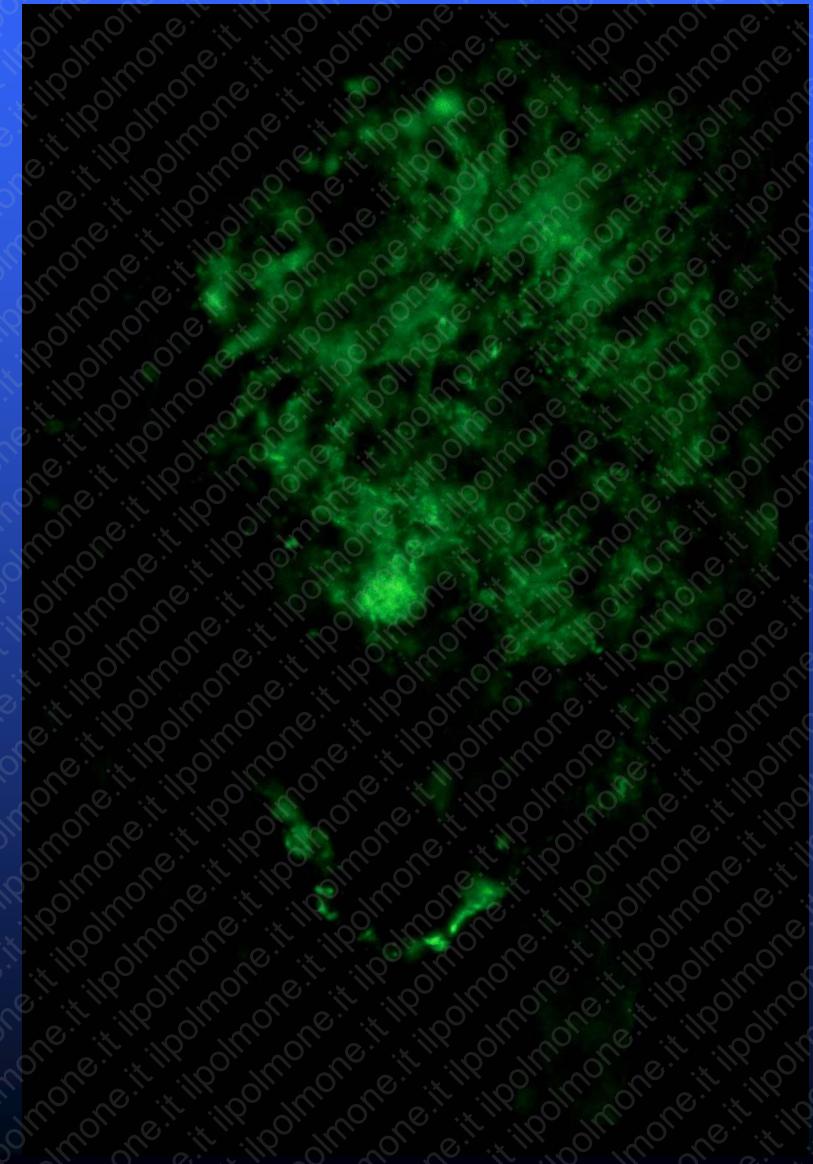




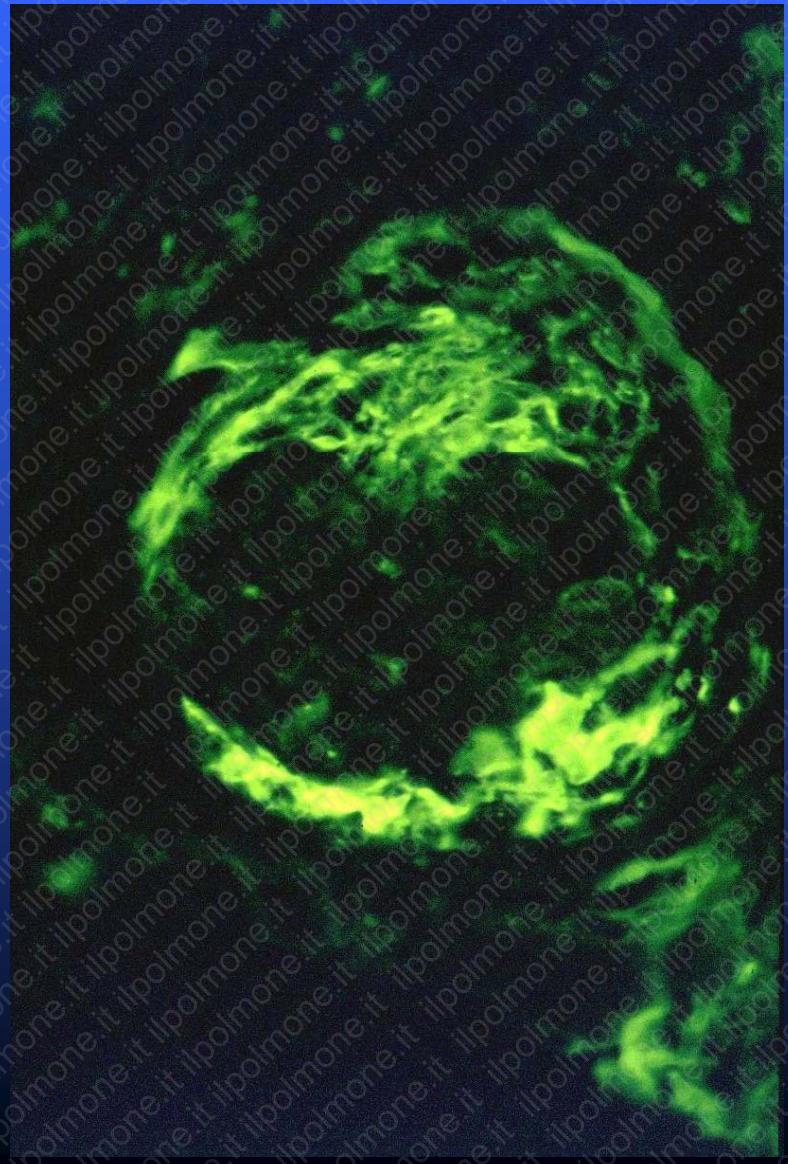


"Pauci-immune" crescentic glomerulonephritis

*Mild IgG deposits
mesangial*



*Fibrinogen deposits predominantly
in the urinary space*



Wegener's Granulomatosis

Prognosis

- ◆ Poorer outcomes with advanced age, severe renal impairment, DAH
- ◆ Mortality >75% if untreated with median survival of 5 months. Drastic improvement since 1970s in mortality
- ◆ Permanent morbidity:
 - CKD 42%
 - Hearing Loss 35%
 - Nasal Deformity 28%
 - Tracheal Stenosis 13%
 - Severe Infection 50% (Treatment)

Microscopic Polyangiitis

clinical manifestations

Clinical manifestations	Frequency (%)
Rapidly progressive glomerulonephritis	100
Pulmonary (hemorrhage, hemoptysis)	10-30
Constitutional symptoms (fever, chills, weight loss, arthralgias/myalgias)	70-80
Cutaneous (purpura, urticaria, subcutaneous nodules, exanthem)	50-65
Nervous system (mononeuritis multiplex)	15-50
GI (pain, GI bleeding, infarction, perforation)	30-45
Ocular (conjunctivitis, uveitis)	0-30
Cardiac	10-20
Upper airway	0-15

Microscopic Polyangiitis

■ Clinical Presentation:

- ◊ Systemic, multi-organ complaints along with constitutional symptoms
- ◊ Pulmonary involvement in approximately 30-50%
- ◊ Milder upper respiratory disease than pts with WG
- ◊ Necrotizing glomerulonephritis is common (79%)

■ Laboratory Findings:

- ◊ ANCA: + p-ANCA in 50-75% and + c-ANCA in 10-15%

■ Diagnosis:

- ◊ Biopsy reveals necrotizing vasculitis and non-granulomatous inflammation