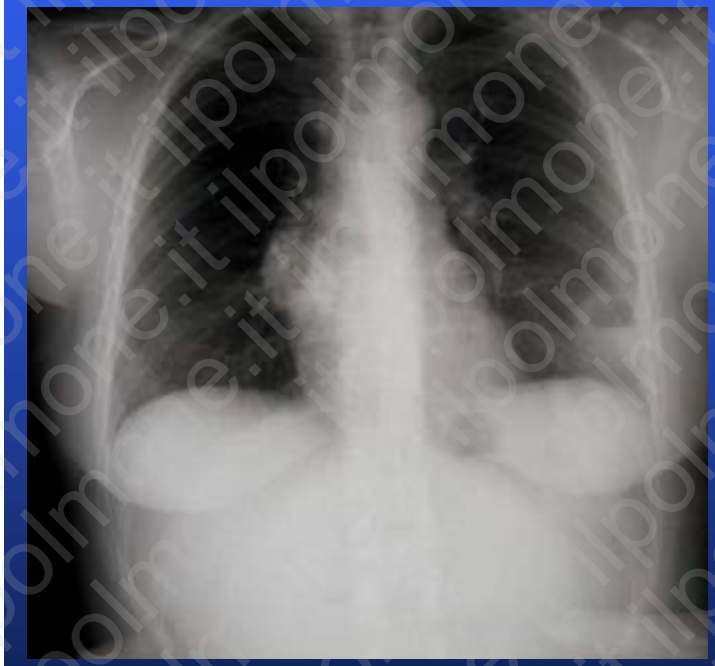
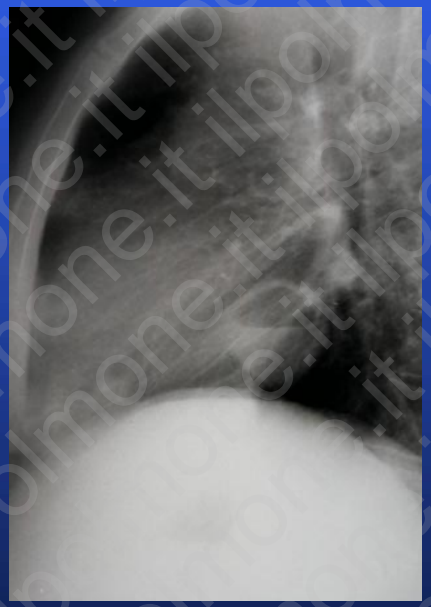
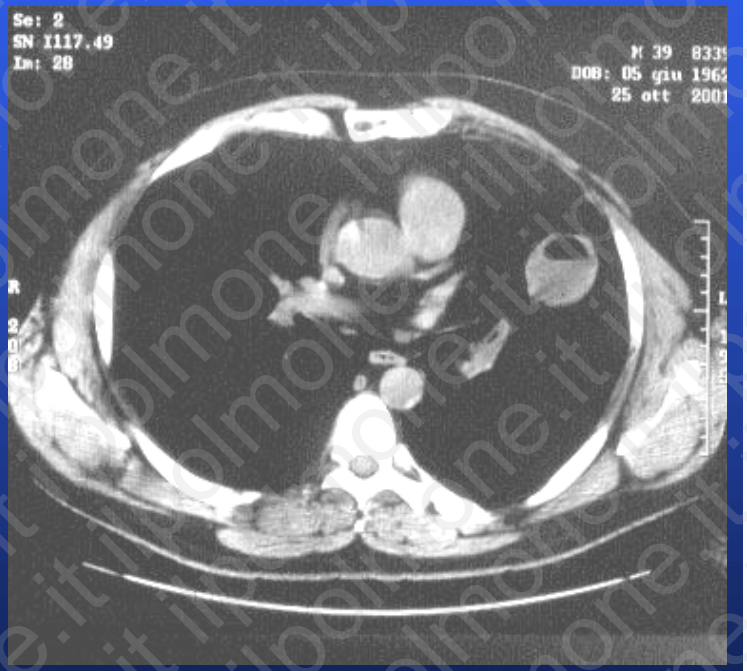
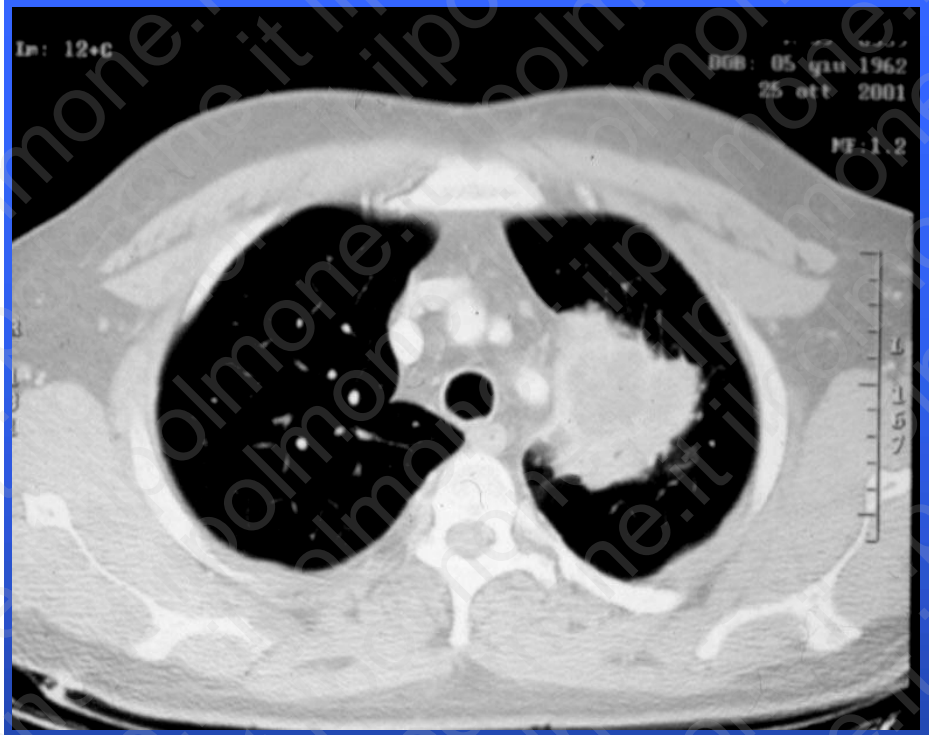
# Cavitating Nodules in Wegener's Granulomatosis



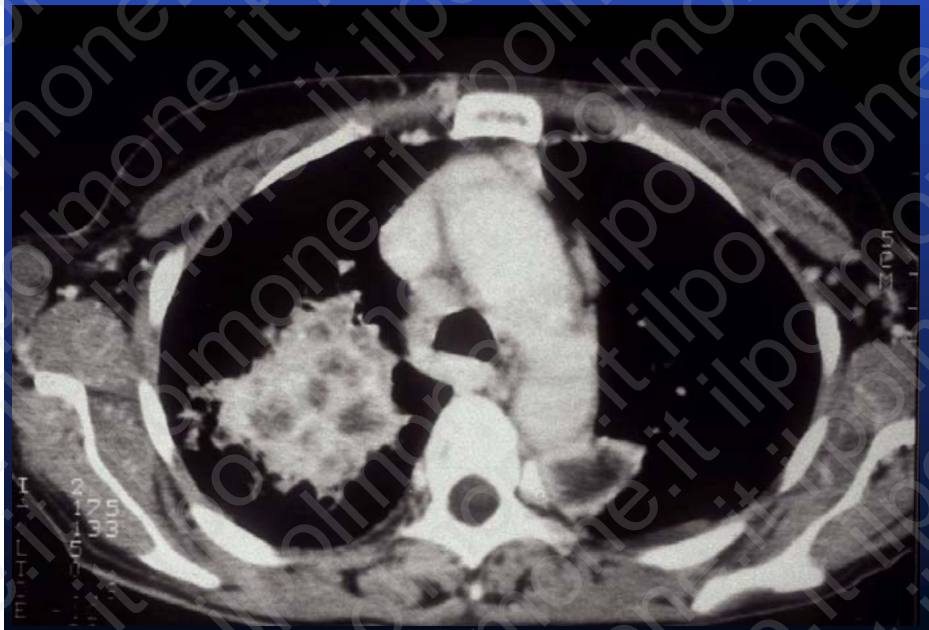


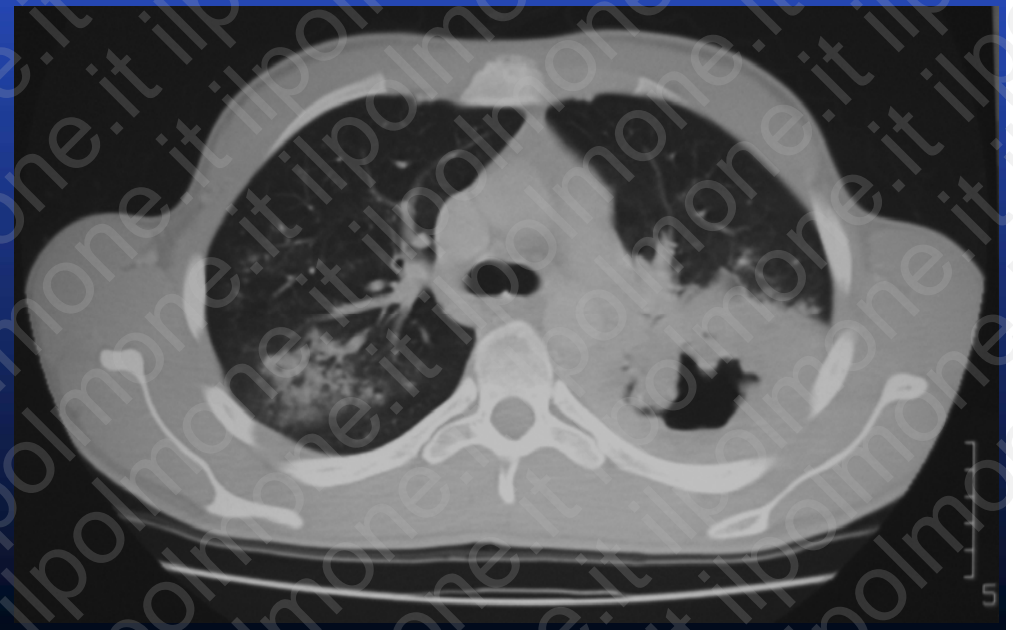



The cavity walls are often thick and irregular or shaggy, although thin-walled cavities may also be seen

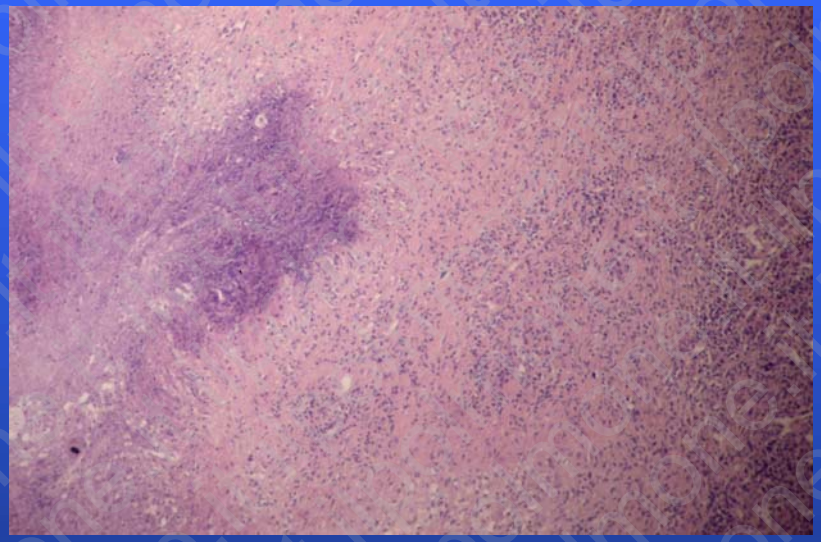


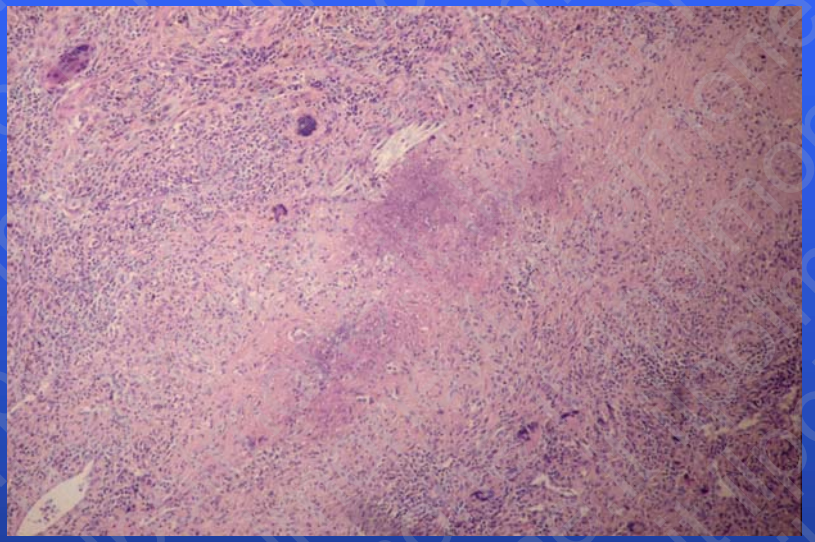
# Masses in Wegener's Granulomatosis

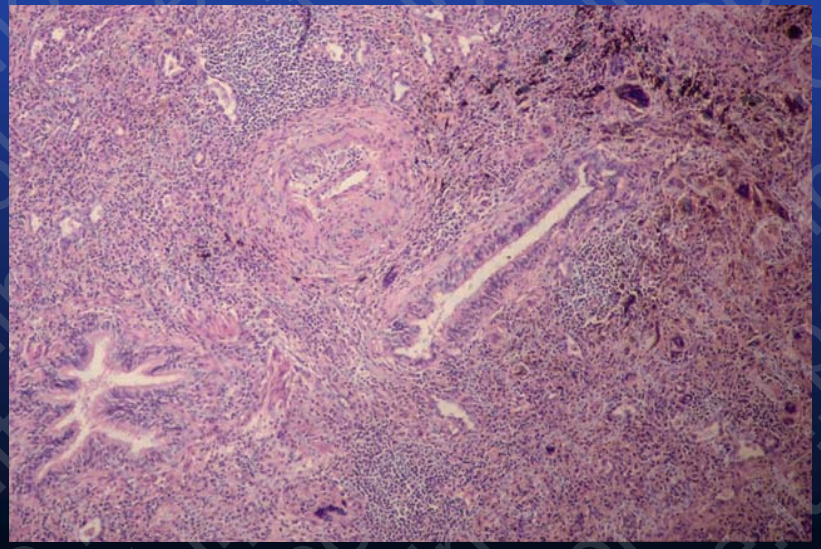


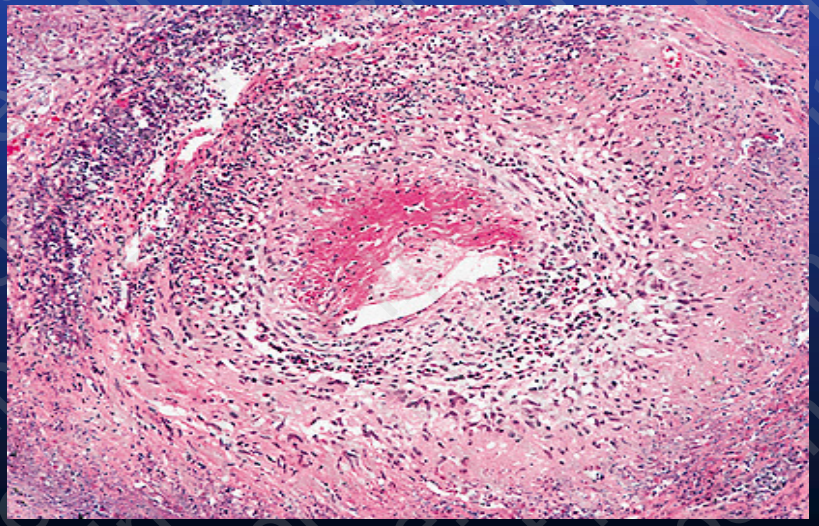


# Surgical Biopsy In WG nodules or masses Inflammatory background, necrosis and vasculitis

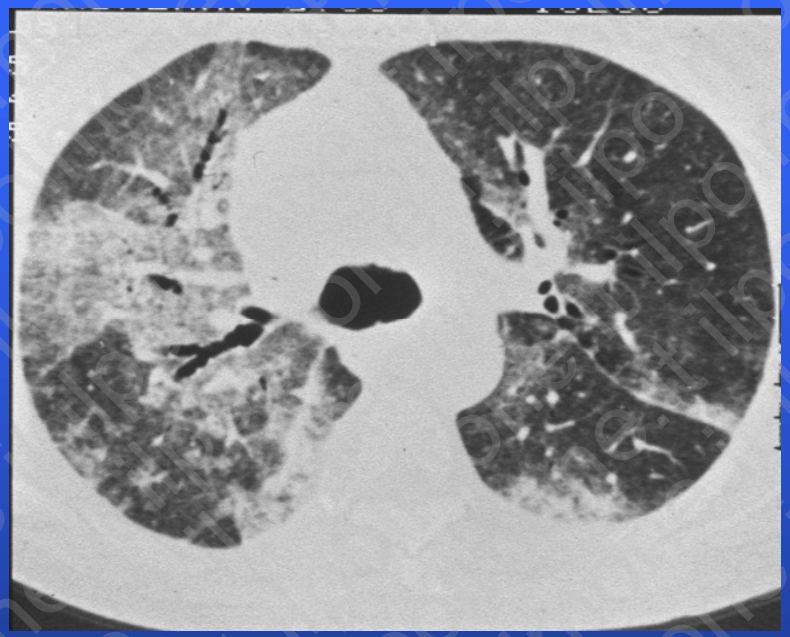


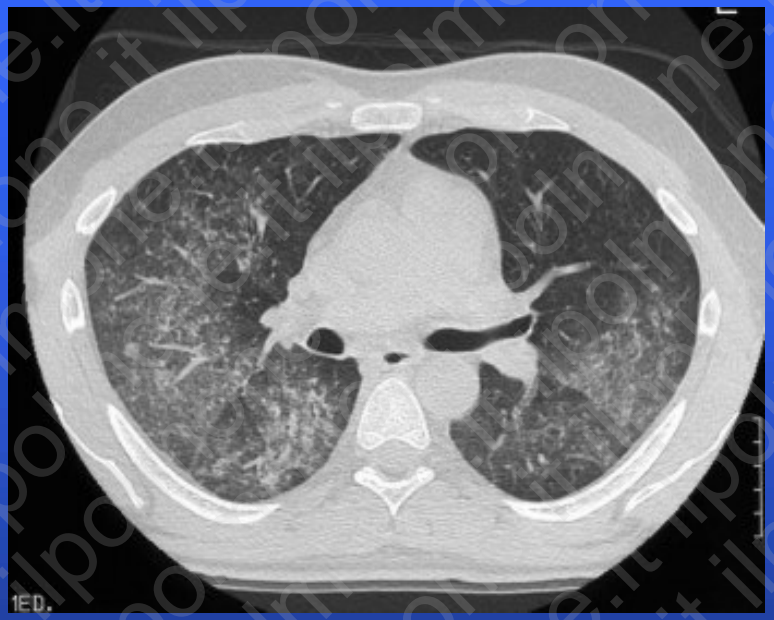


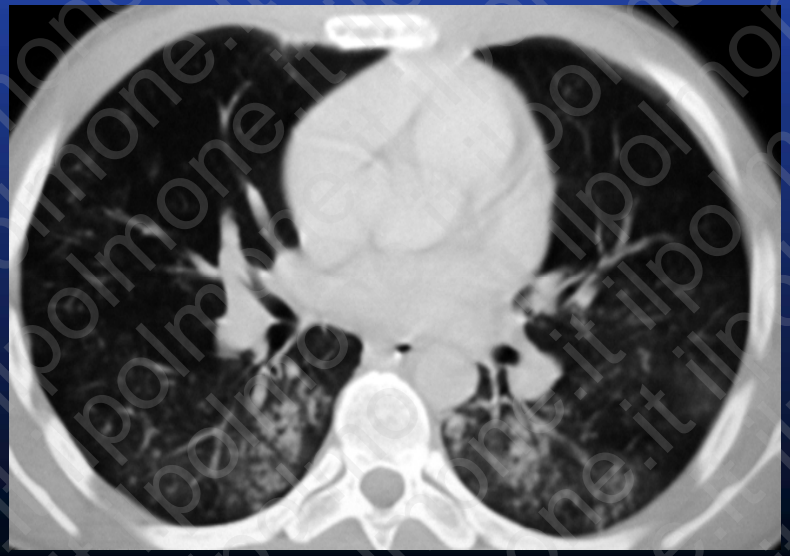




# Pulmonary Infiltrates in WG and MPA

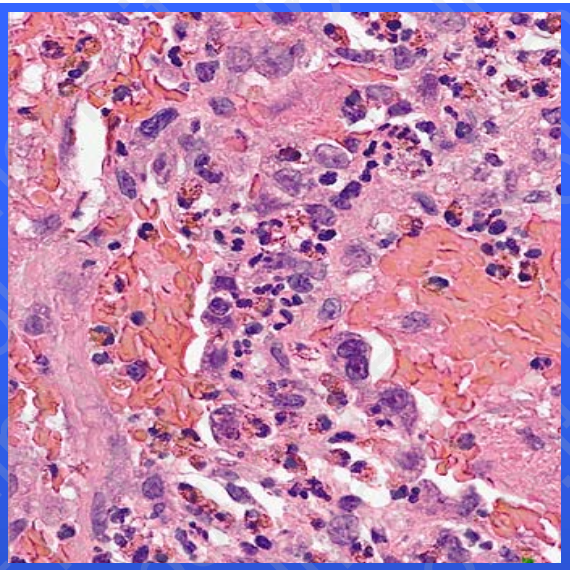






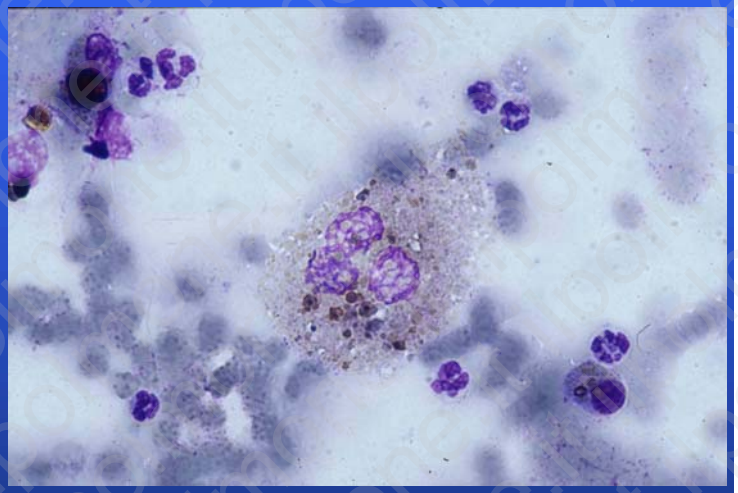
Consolidation or ground-glass opacity are usually related to pulmonary hemorrhage

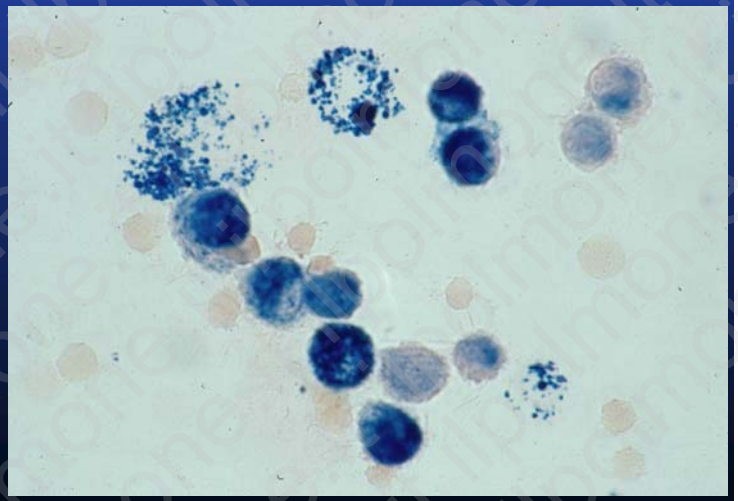
# BAL in Diffuse Alveolar Haemorrhage

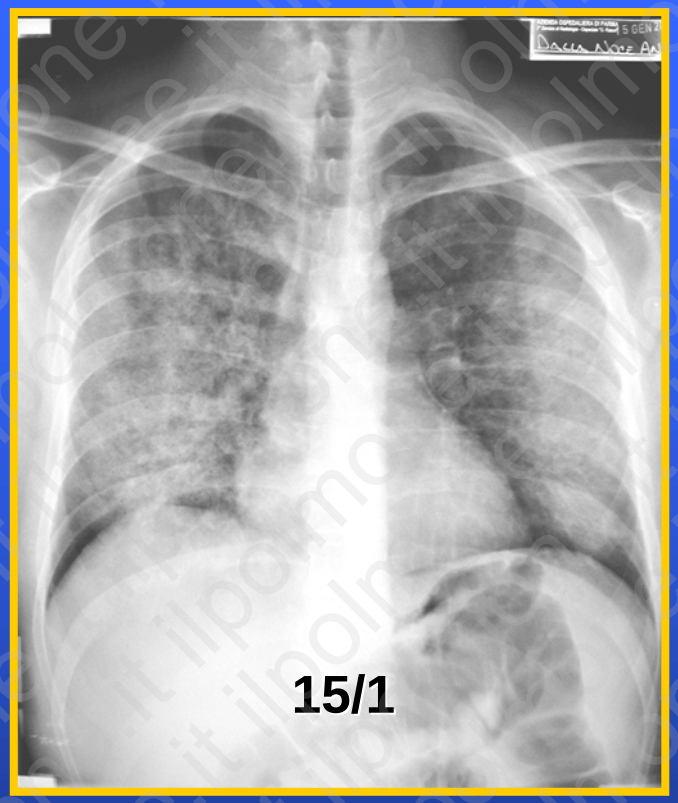


Biopsy

BAL







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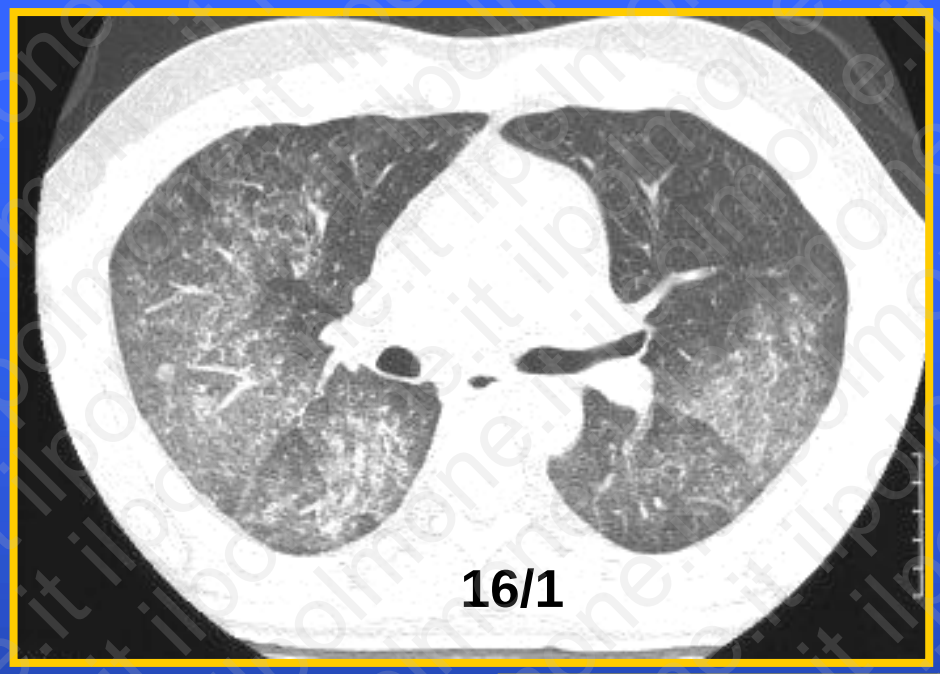

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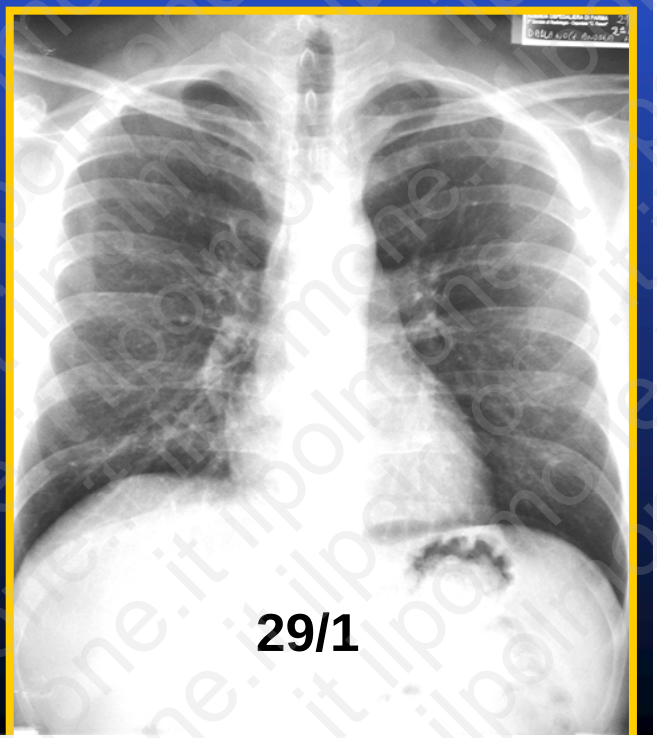


29/1

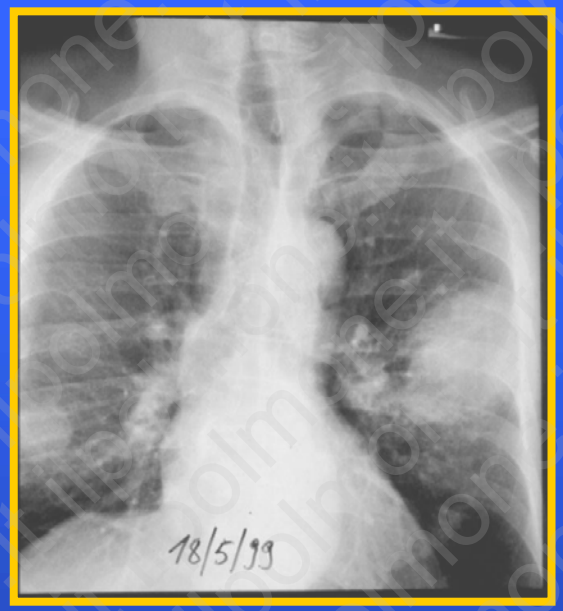
# DAH in Microscopic Polyangiitis

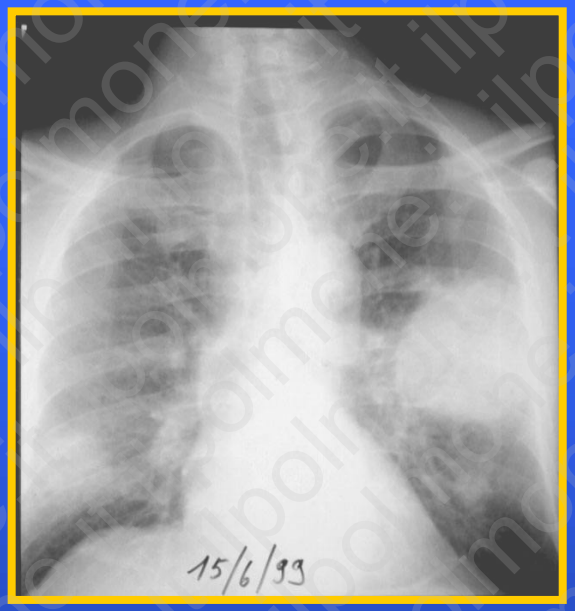
## Reversibility after treatment

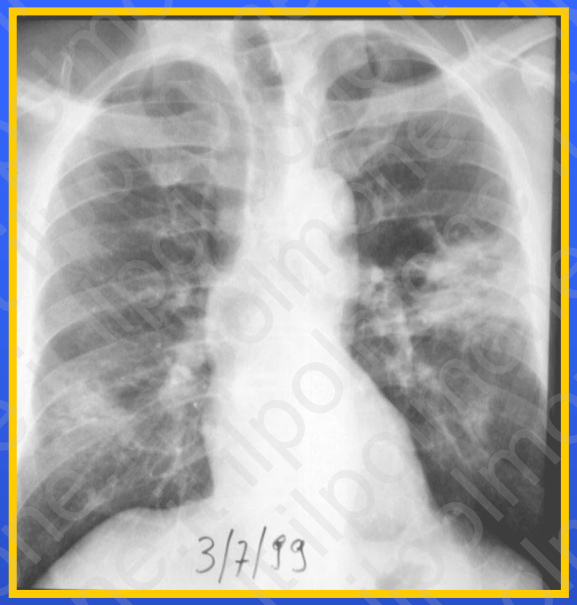
# MICROSCOPIC POLYANGIITIS

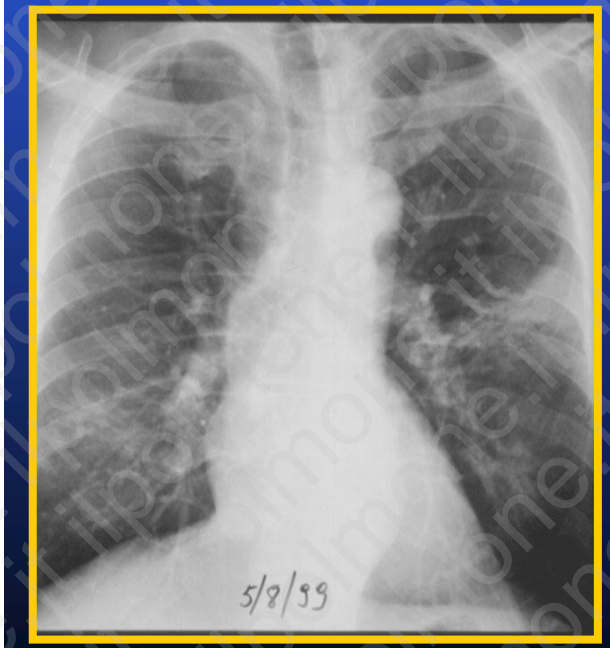
## clinical manifestations

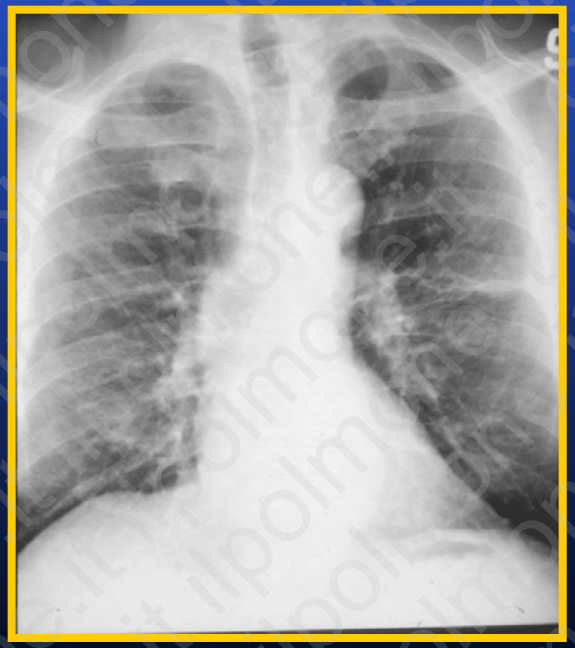
| Clinical manifestations | Frequency (%) |
|---|---|
| Rapidly progressive glomerulonephritis | 100 |
| Pulmonary (hemorrage, hemoptysis) | 10-30 |
| Constitutional symptoms (fever, chills, weight loss, arthralgias/myalgias) | 70-80 |
| Cutaneous (purpura, urticaria, subcutaneous nodules, exanthem)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



# Wegener's Granulomatosis

Reversibility after treatment

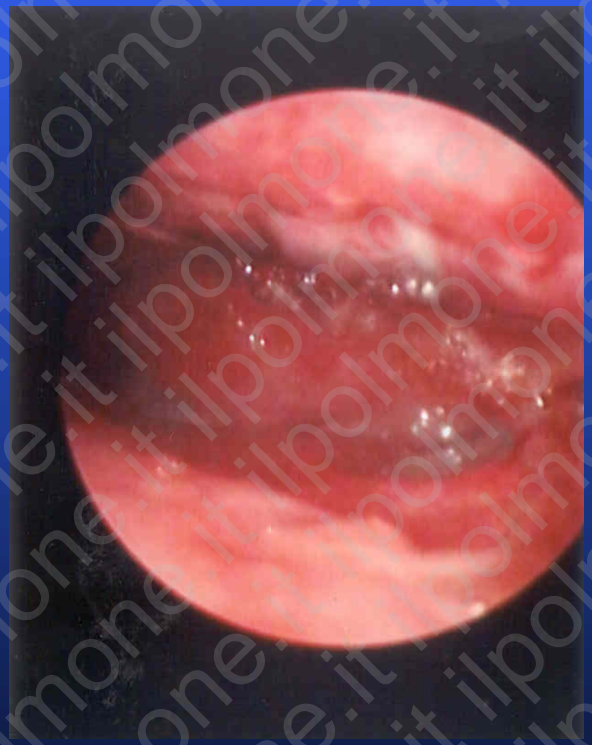
# Churg Strauss Syndrome

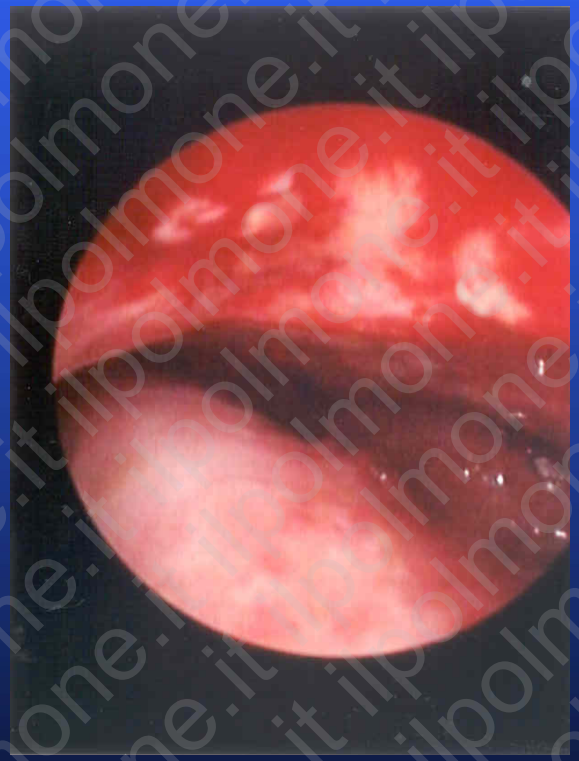
The syndrome is characterized by the triad of:

- asthma
- hypereosinophilia
- necrotizing vasculitis

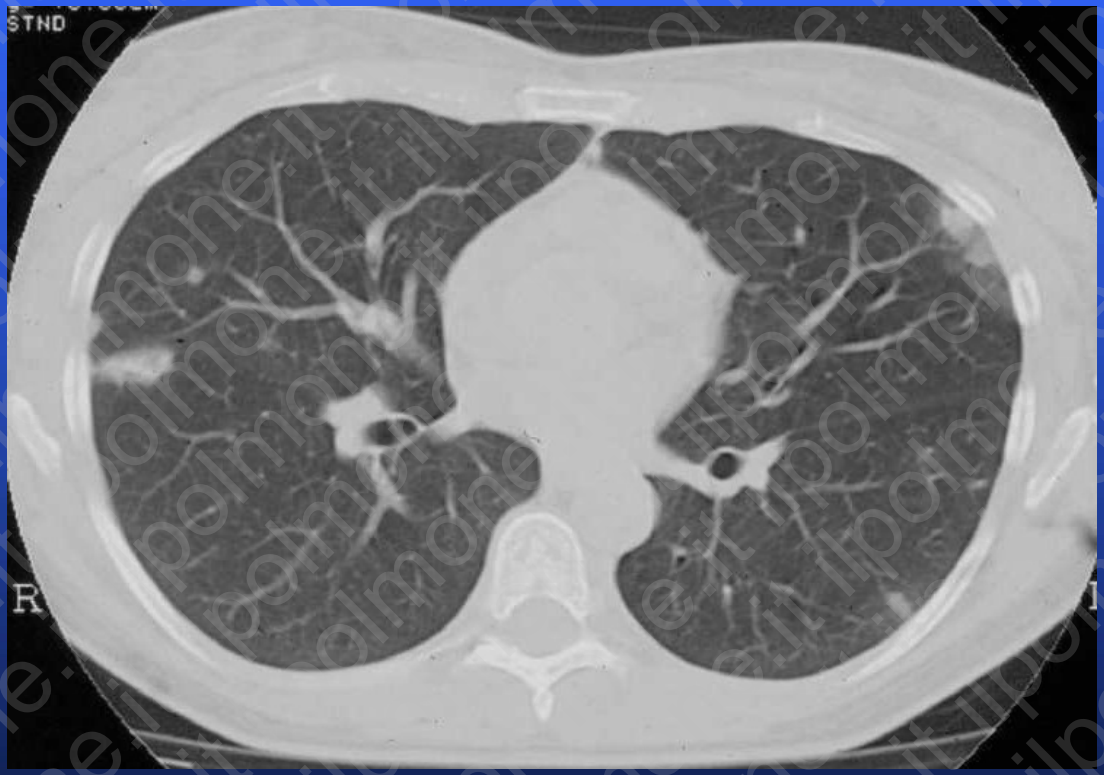
| Clinical manifestations | Frequency (%) |
|---|---|
| Asthma | 98-100 |
| Constitutional (fever, chills, Weight loss, arthralgias/myalgias) | 70-80 |
| Nervous system (mononeuritis Multiplex, CNS, cerebral hemorrhage) | 50-80 |
| Cutaneous (purpora, urticaria, Subcutaneous nodules, exanthem) | 50-80 |
| Sinusitis | 20-70 |
| Cardiac (pericarditis, heart failure, coronary vasculitis) | 35-50 |
| GI (diarrhea, GI bleeding, colitis, pain) | 30-60 |
| Renal (proteinuria, hematuria) | 10-50 |

# Upper Airway Involvement in Churg Strauss Syndrome



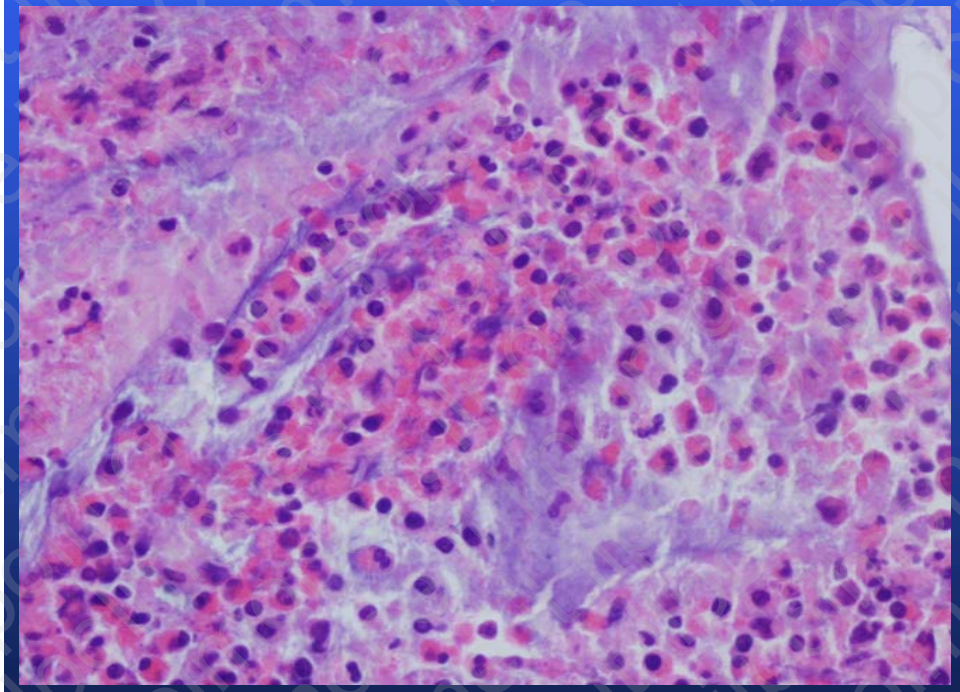


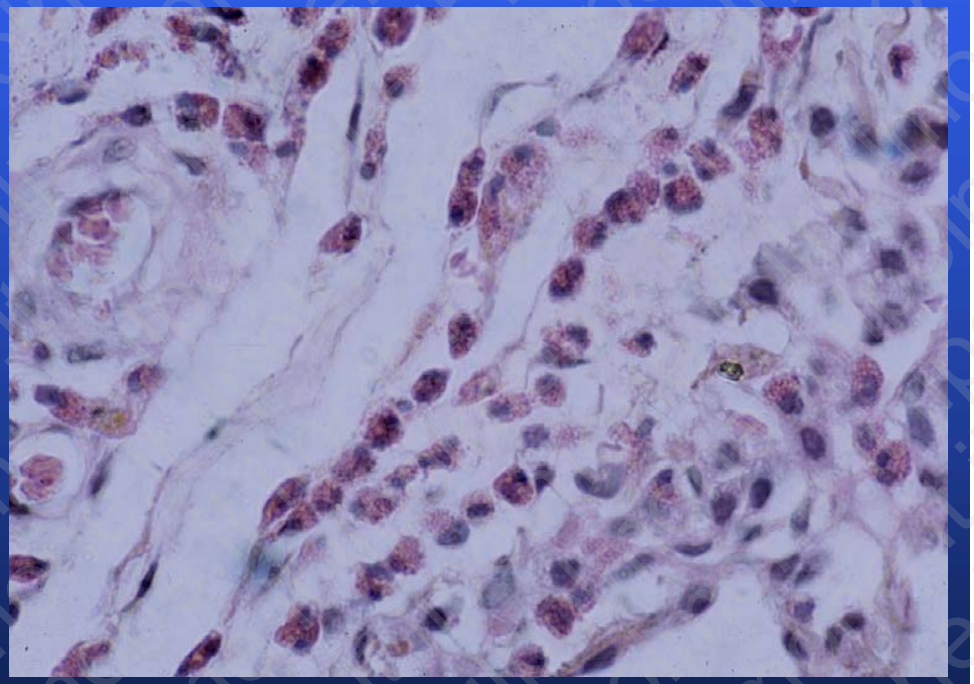
# Nodules in Churg Strauss Syndrome



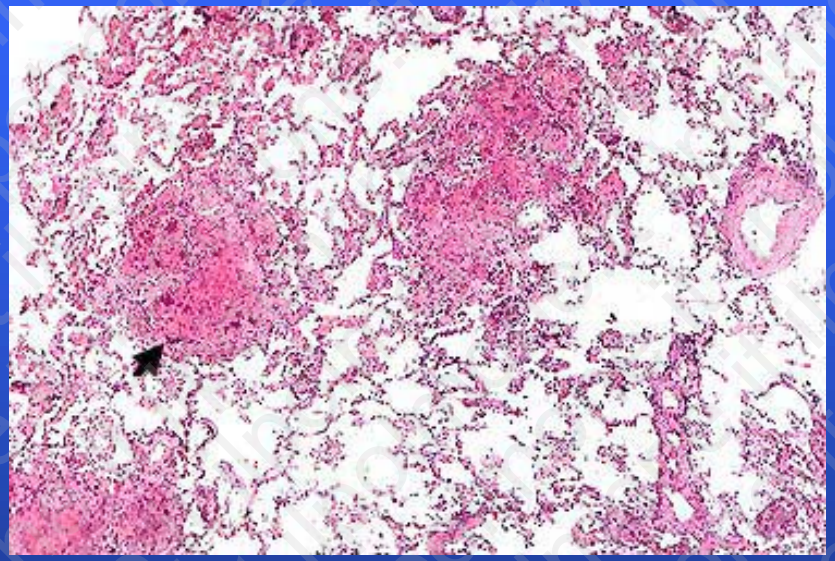

Multiple nodules ranging from 0.5 to 3.5 cm, which may contain air bronchograms or cavitate

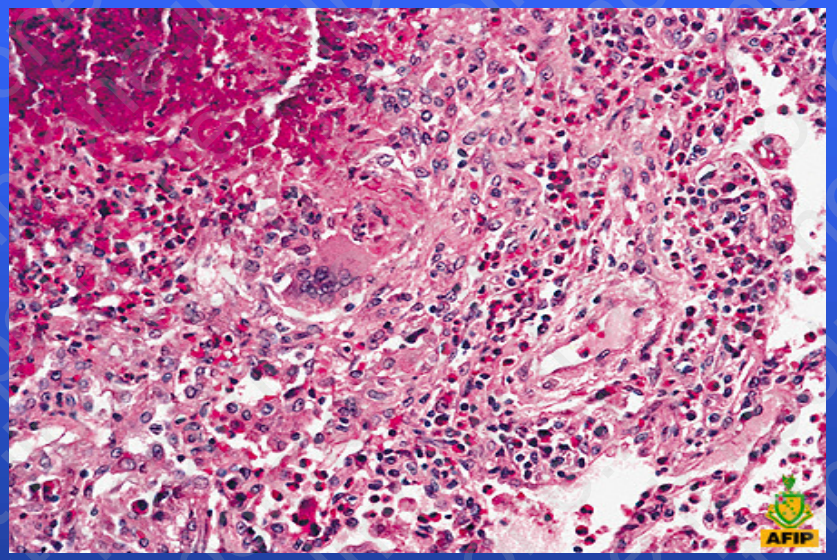
# Transbronchial Biopsy in CSS

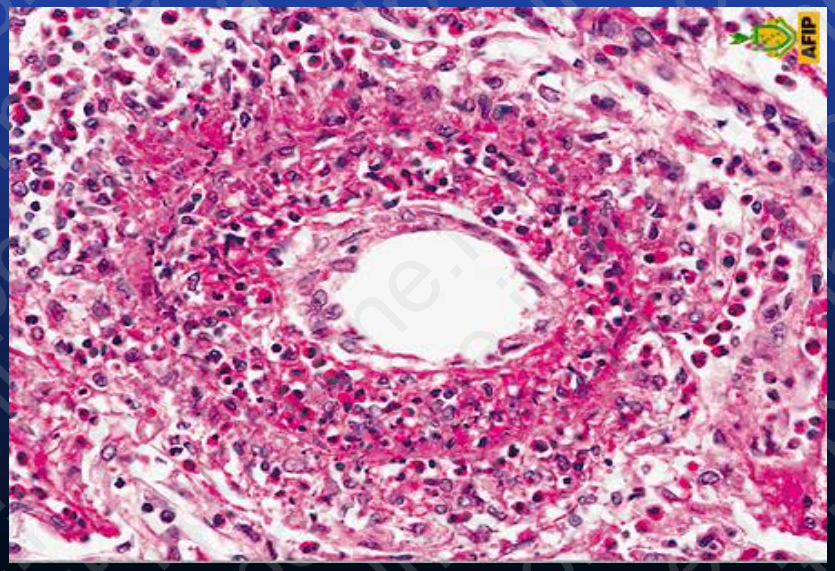


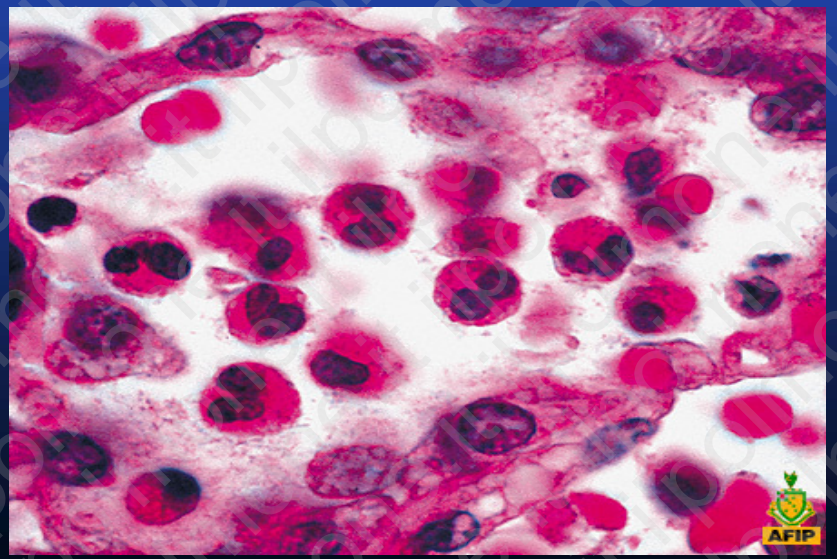


# Surgical Biopsy in CSS by Travis et al

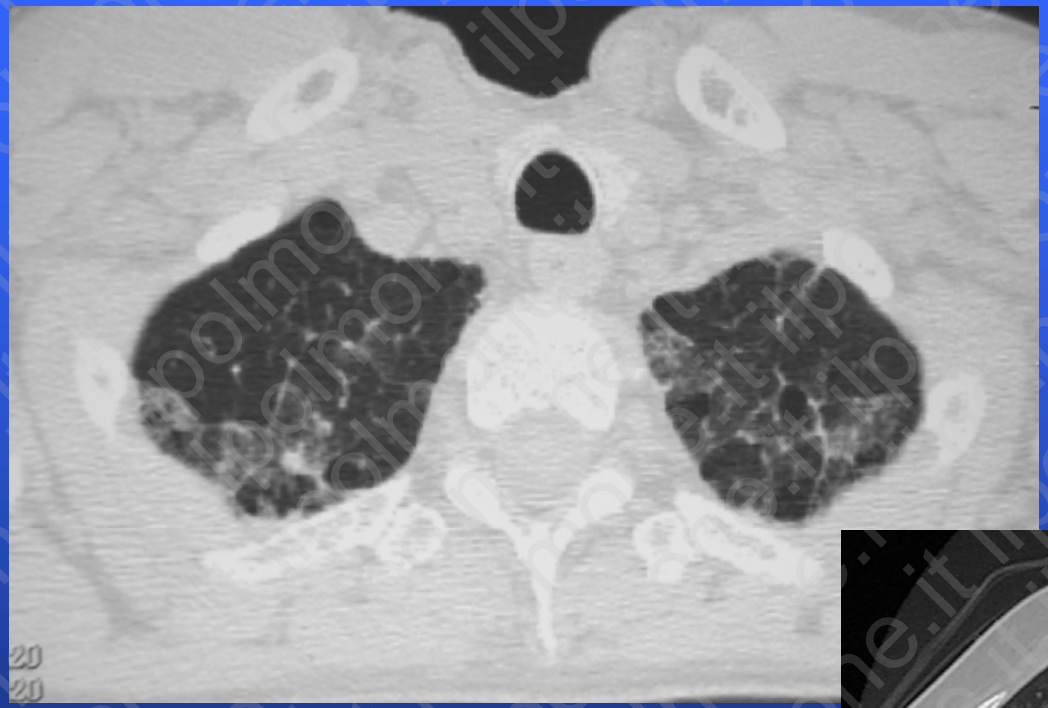


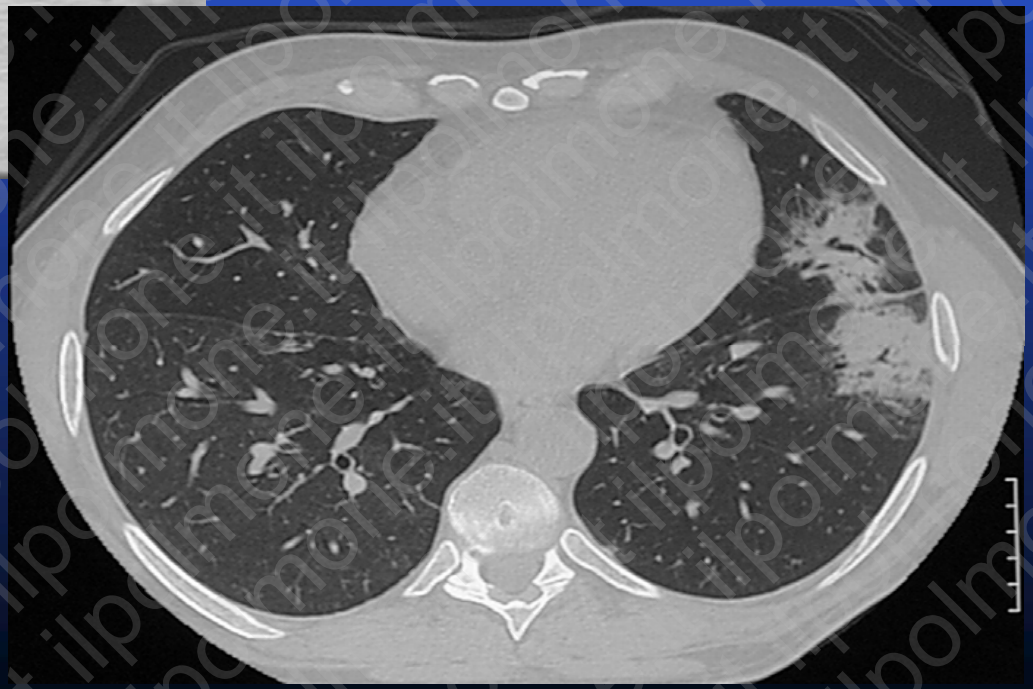


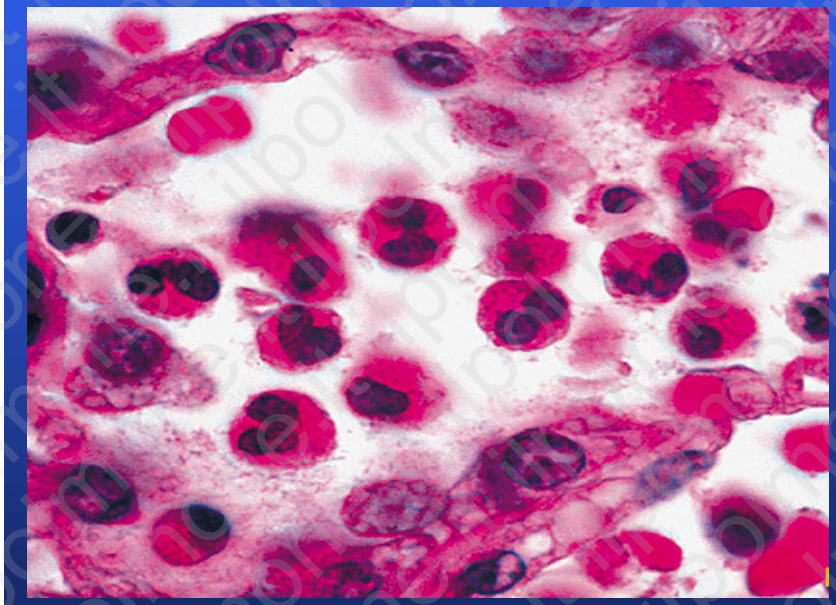



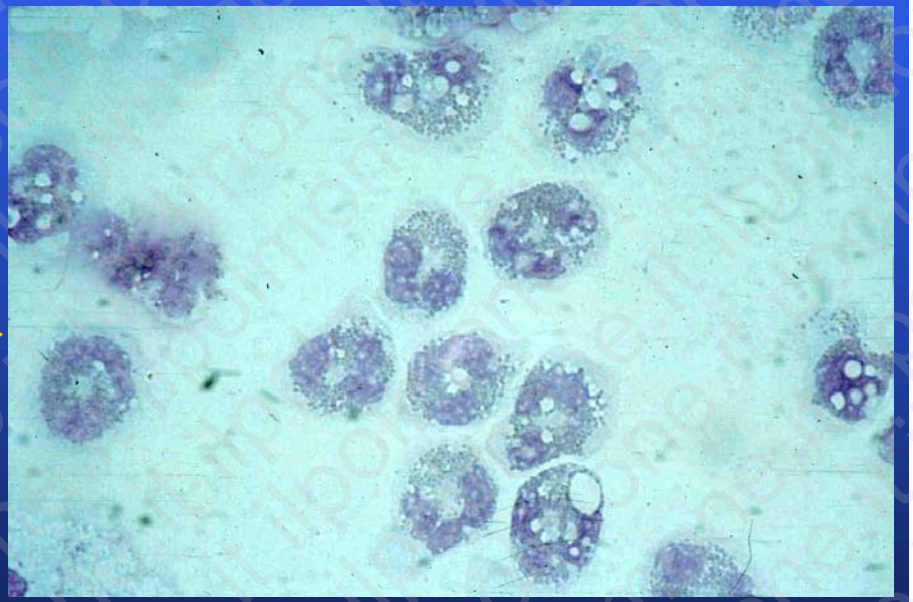
# Pulmonary Infiltrates in CSS





# Eosinophilic alveolitis in BAL fluid of CSS patients





Biopsy

BAL (Eosinophils> 33%)

# Relapse rate of 67 patients with ANCA-associated vasculitides

The CSS had the highest relapse rate at the first and second year (27 and 35%), followed by WG (16 and 26%), and MPA (10 and 19%)

# Prognostic factors for death of 67 patients with ANCA-associated vasculitides

The global risk of death was 22% and 35% after 5 and 10 years of follow up

at 5 years follow up, MPA patients have a risk of death (35%) higher than those affected by both WG (27%) and CSS (0%)

# Complications

In ANCA-associated systemic vasculitides after patients have begun treatment with corticosteroids and cytotoxic drugs, the recurrence of pulmonary symptoms suggesting a flare indicates a careful search for an opportunistic lung infection or iatrogenic pulmonary complication

# Iatrogenic Pulmonary Complications

**Chronic Interstitial Pneumonitis (NSIP)**
cyclophosphamide, methotrexate

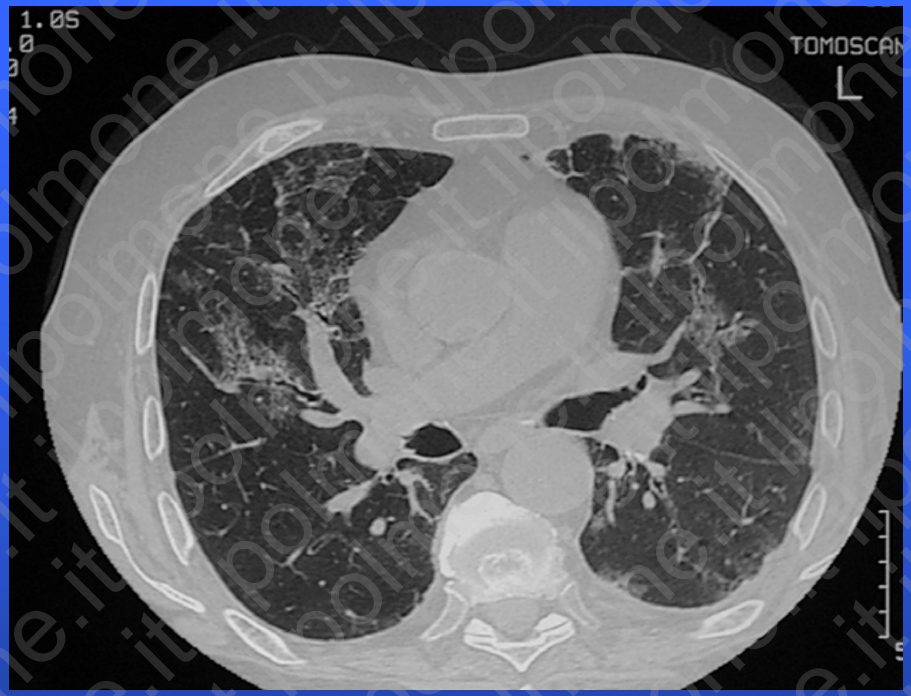
**Diffuse Alveolar Damage**
azathioprine, cyclophosphamide, methotrexate

**Bronchiolitis Obliterans – Organizing Pneumonia**
cyclophosphamide, methotrexate

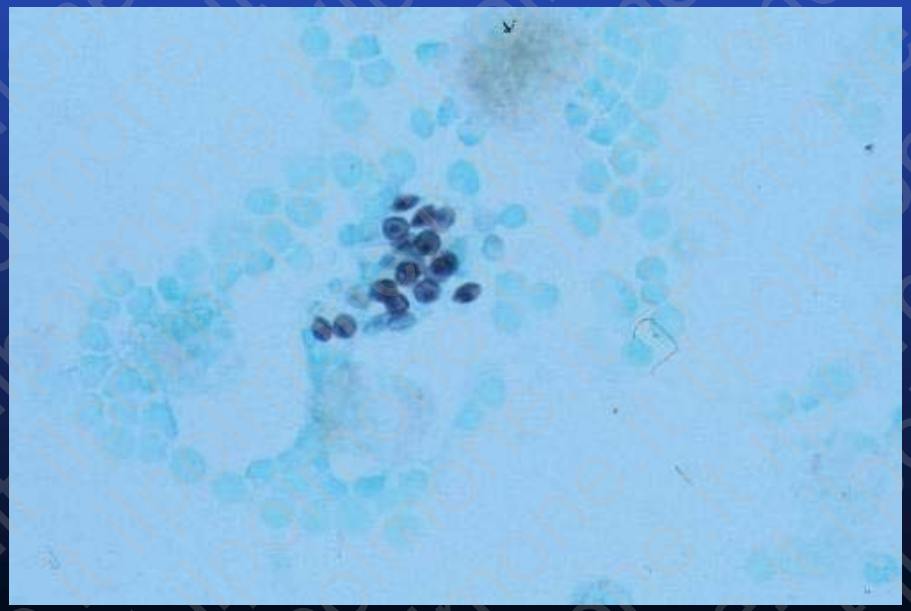
**Haemorrhagic Alveolitis**
cyclophosphamide

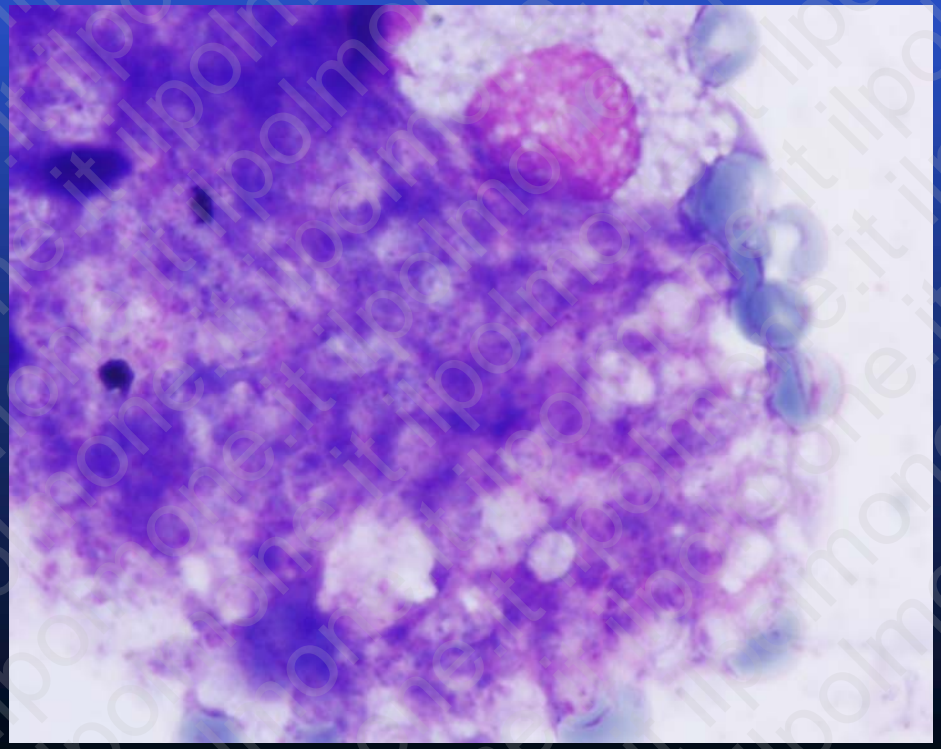
**Pulmonary Oedema**
methotrexate

**Granulomatous reaction**
methotrexate




# Pneumocystis Carinii Pneumonia





# Distinguishing Features of Pulmonary Involvement in ANCA-associated Systemic Vasculitides

| | Wegener's Granulomatosis | Churg-Strauss Sindrome | Microscopic Polyangiitis |
|---|---|---|---|
| **Lung Involvement** | Common | Common | Common |
| **Pulmonary Function Tests** | Obstructive/Restrictive pattern, DLCO ↓ or ↑ if DAH | Obstructive pattern | DLCO ↑ if DAH, obstructive pattern (rare) |
| **Most common HRCT findings** | Multiple nodules, cavitary lesions, infiltrates (DAH) | Fleeting ground-glass and/or consolidations | Ground-glass and/or consolidations (DAH) |
| **ANCA prevalence** | 80-90% | 30-50% | 70-90% |
| **ANCA pattern** | PR3 >> MPO | MPO > PR3 | MPO >> PR3 |
| **Main lung pathologic findings** | Necrotizing granulomatous vasculitis, capillaritis | Eosinophilic infiltrates and vasculitis, necrotizing granulomatous vasculitis | Capillaritis and DAH |
| **Prominent BAL findings** | Neutrophilia, blood red cells and siderophages (>30%) if DAH | Eosinophilia (>33%) | Blood red cells and siderophages (>30%) if DAH |

# THERAPY

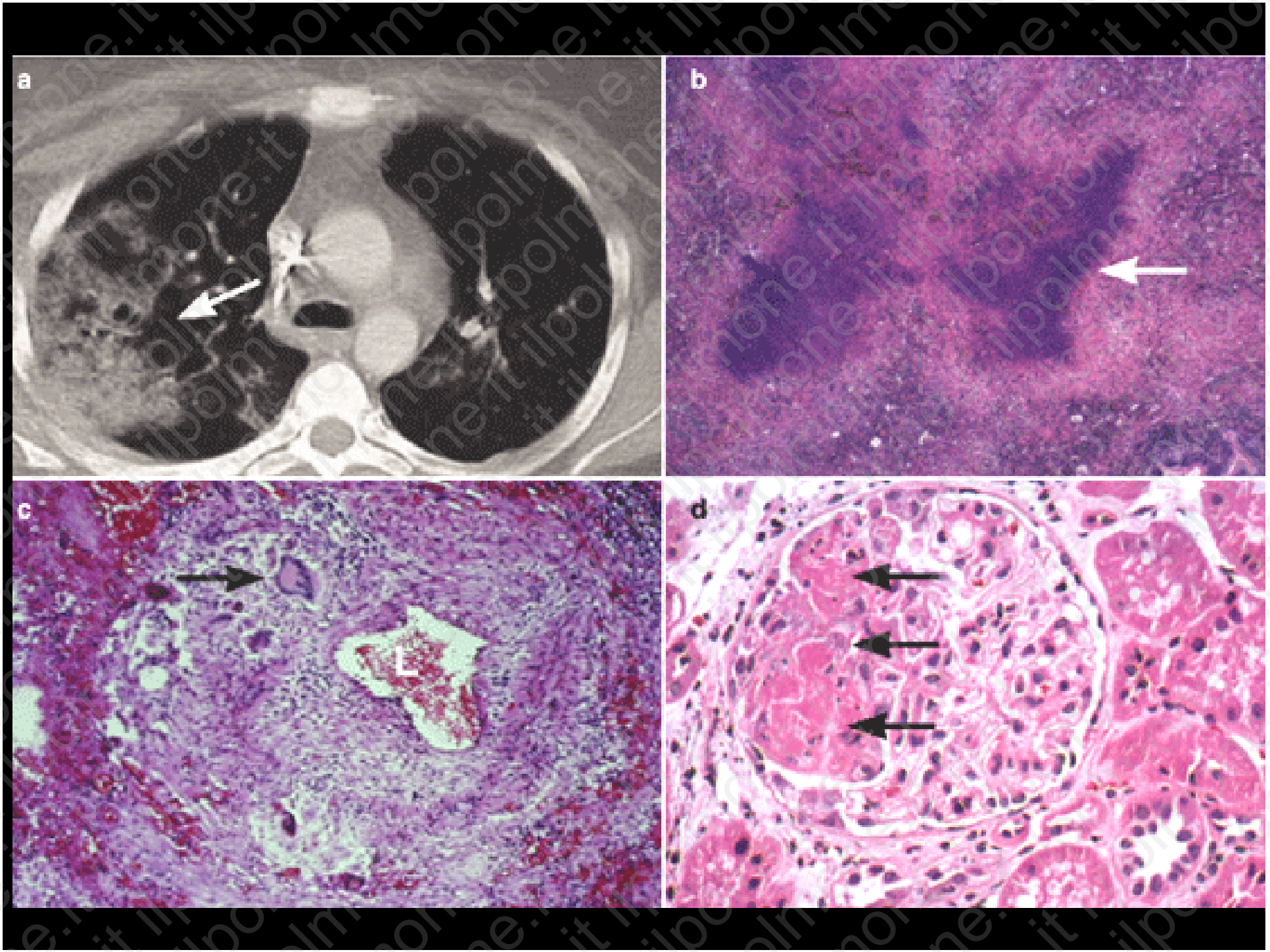
- Before immunosuppressive therapy, the mortality rate of pts with systemic vasculitis was 75%, with a median survival of 5 mo

## KAY MESSAGE

Despite this impressive progress, the survival of treated pts with a systemic vasculitis remains significantly lower than that of the general population

# GOALS OF THERAPY

- Prevention of disease mortality and morbidity
- Minimization of treatment-related complications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