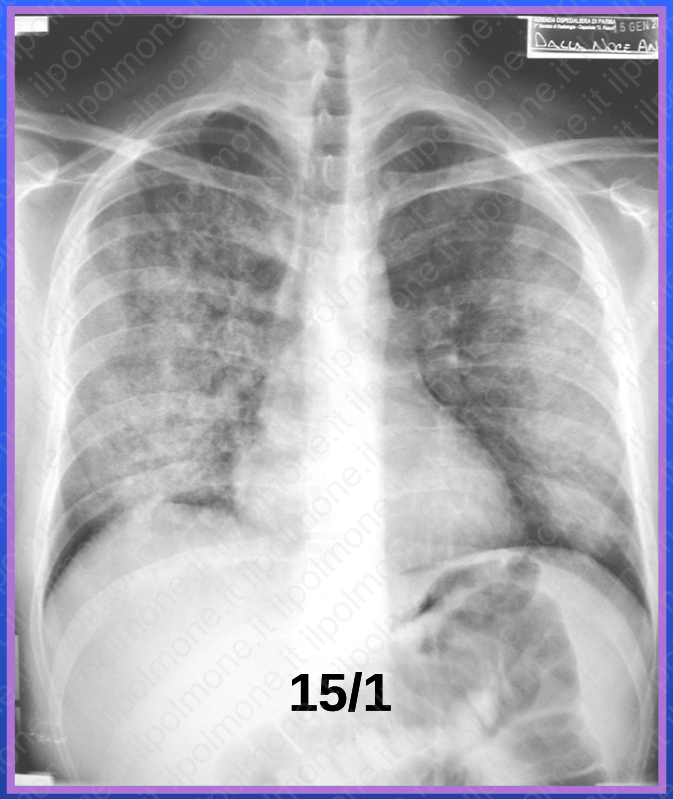

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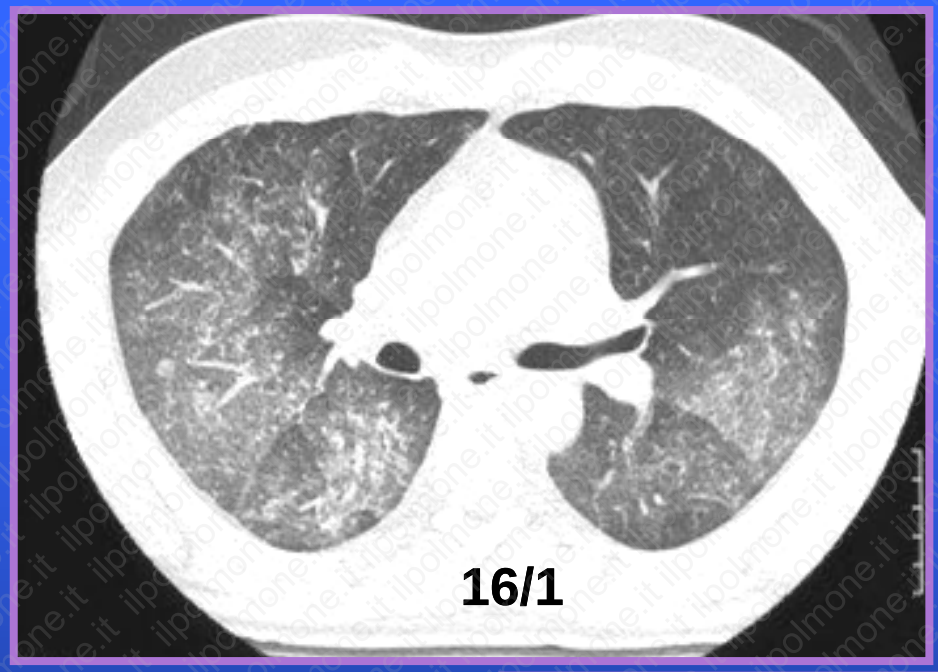

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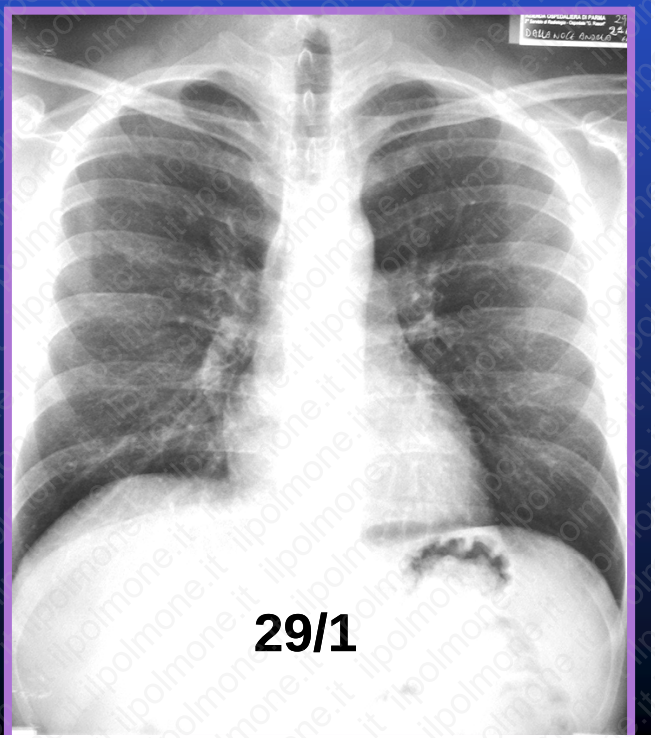
DAH in Microscopic Polyangiitis

Reversibility after treatment



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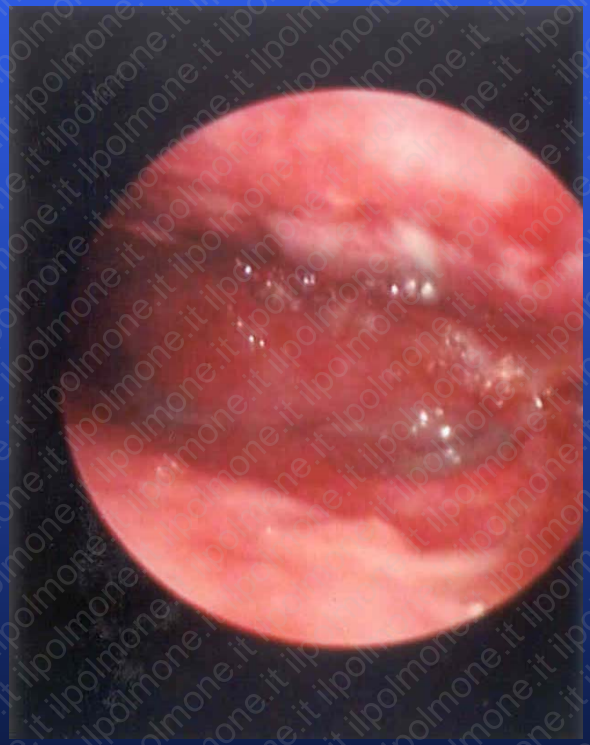
Churg Strauss Syndrome

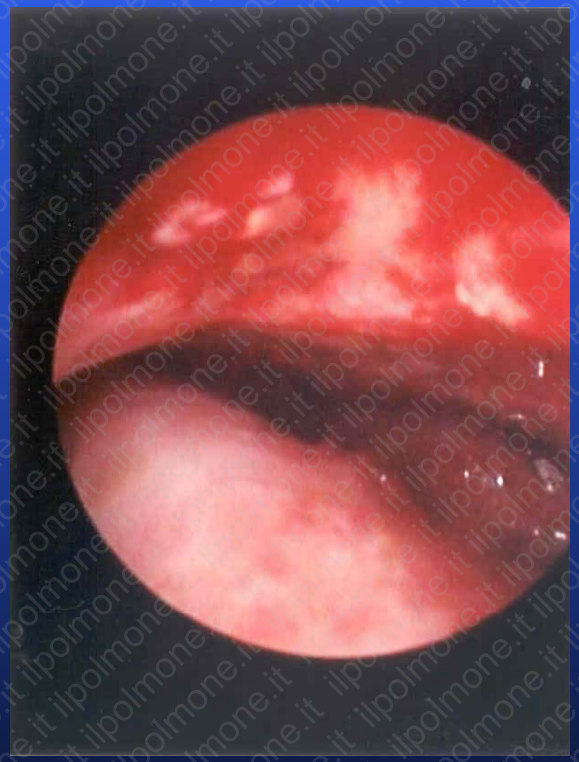
The syndrome is characterized by the triad of:

- asthma
- hypereosinophilia
- necrotizing vasculitis

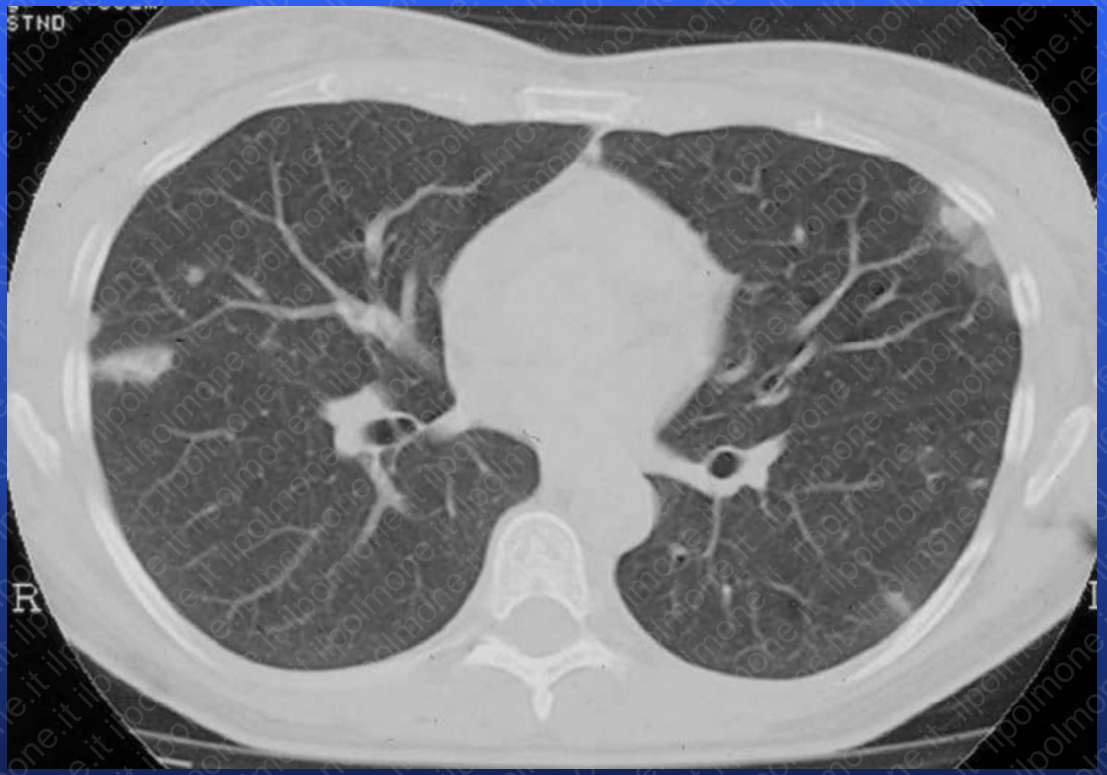
Clinical manifestations	Frequency (%)
Asthma	98-100
Constitutional (fever, chills, Weight loss, arthralgias/myalgias)	70-80
Nervous system (mononeuritis Multiplex, CNS, cerebral hemorrhage)	50-80
Cutaneous (purpora, urticaria, Subcutaneous nodules, exanthem)	50-80
Sinusitis	20-70
Cardiac (pericarditis, 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



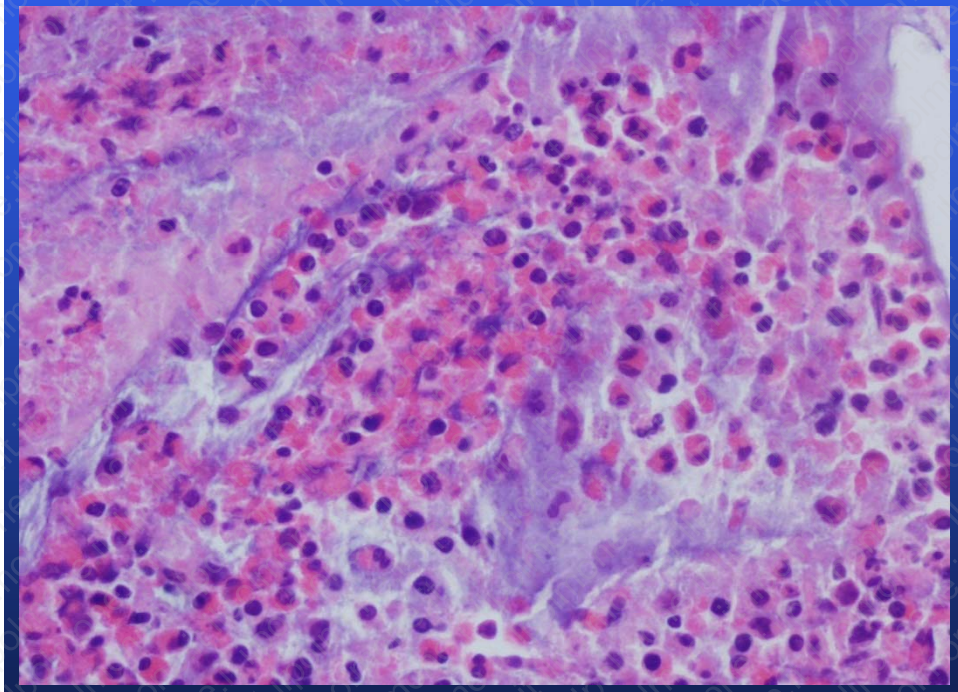


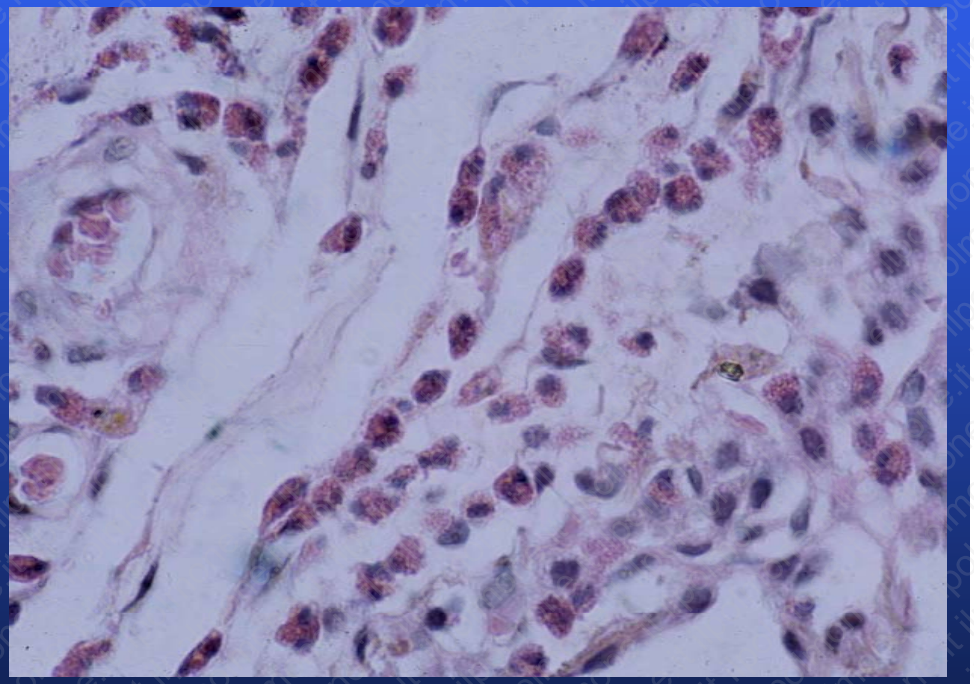
Nodules in Churg Strauss Syndrome



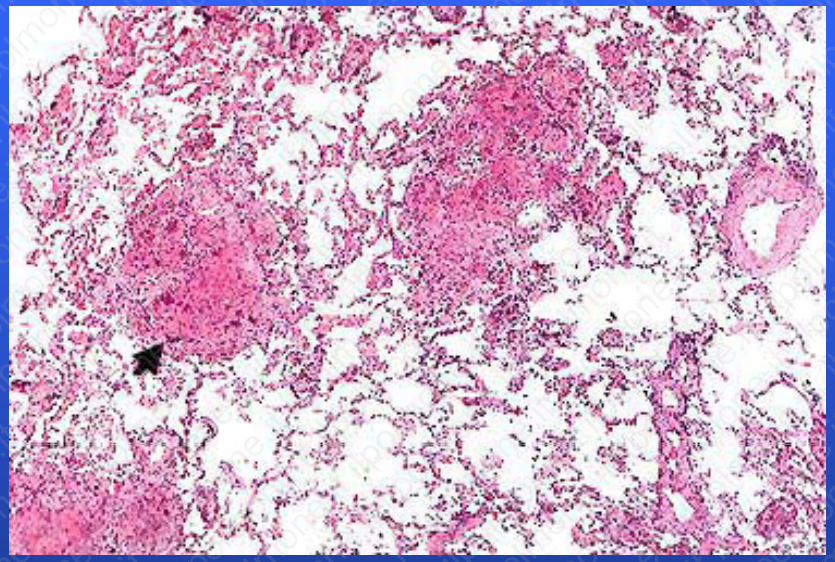
Multiple nodules ranging from 0.5 to 3.5 cm, which may contain air bronchograms or cavitate

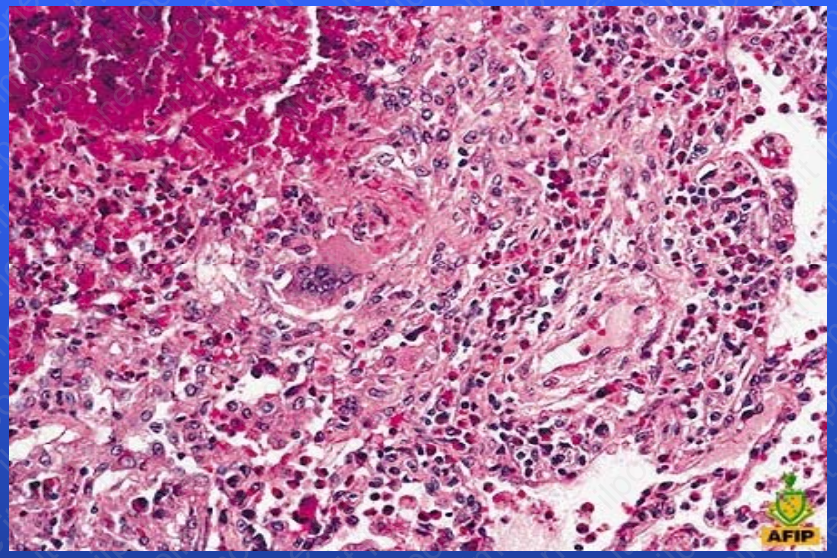
Transbronchial Biopsy in CSS

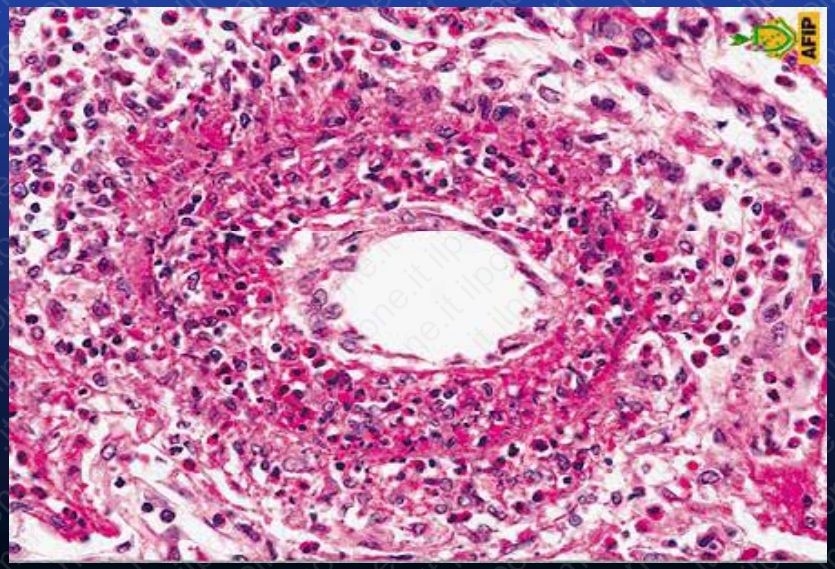


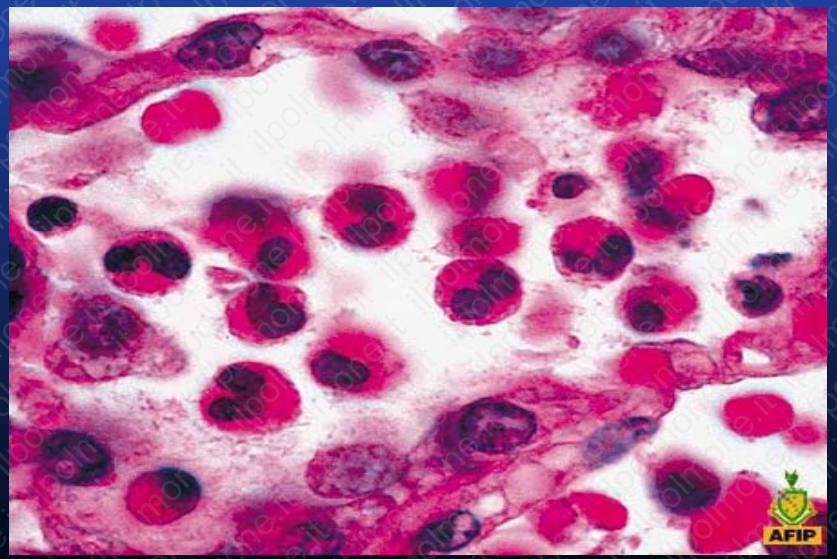


Surgical Biopsy in CSS by Travis et al

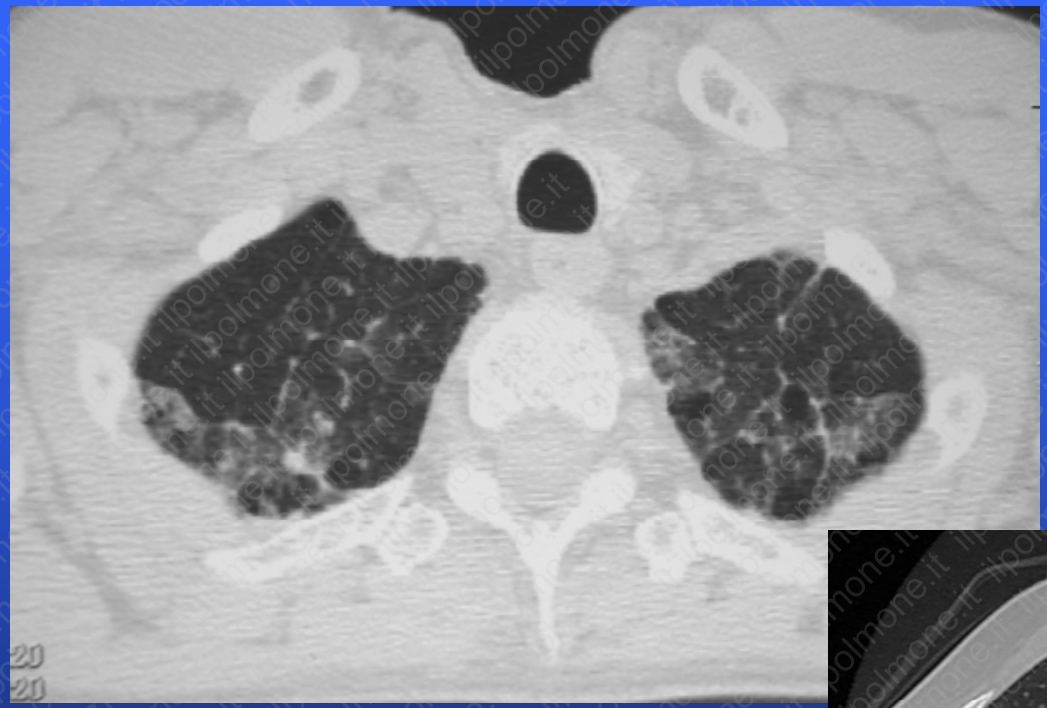


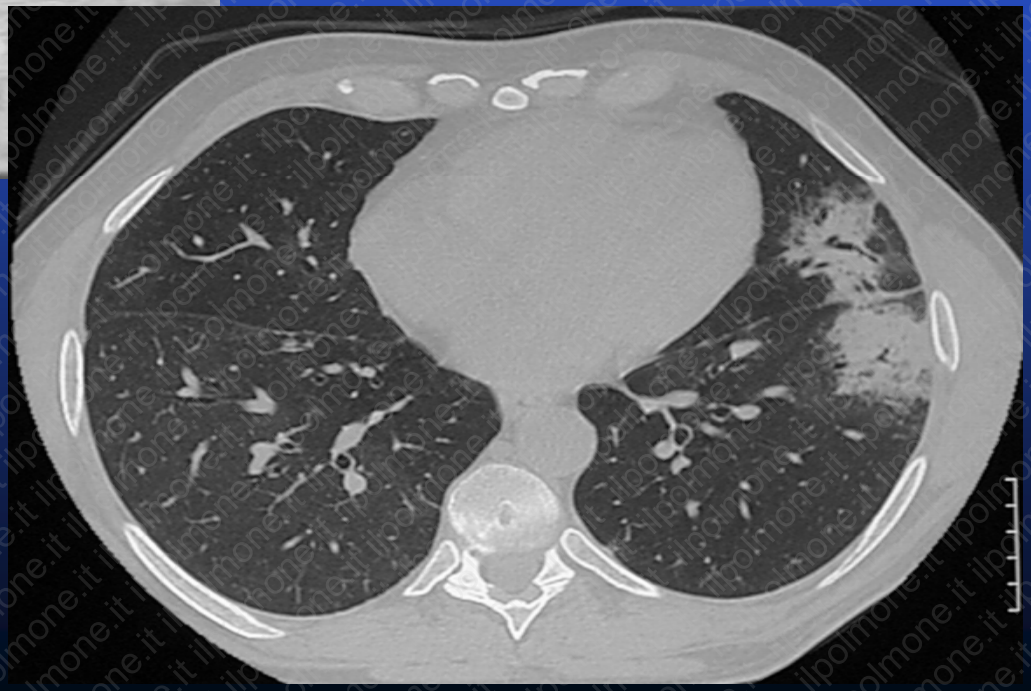




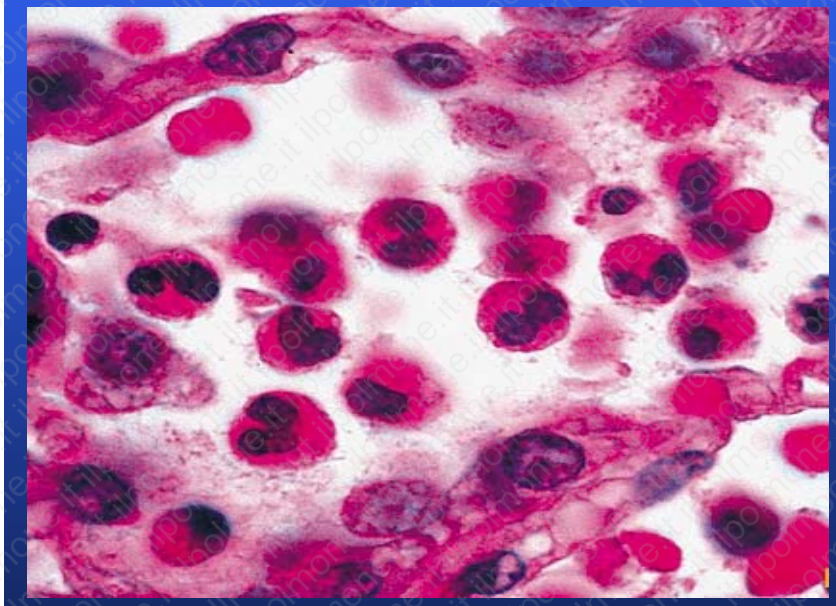


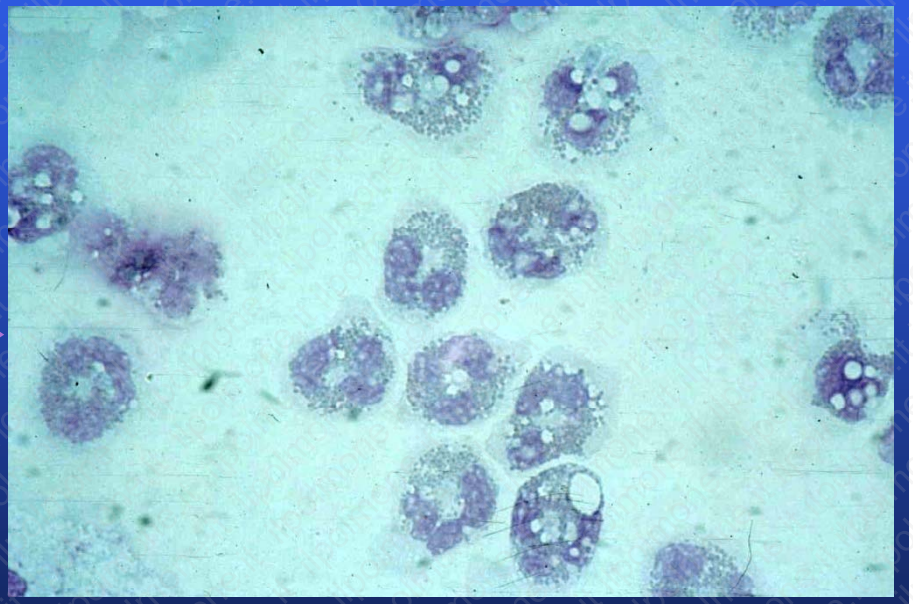
Pulmonary Infiltrates in CSS





Eosinophilic alveolitis in BAL fluid of CSS patients





Biopsy

BAL (Eosinophils> 33%)

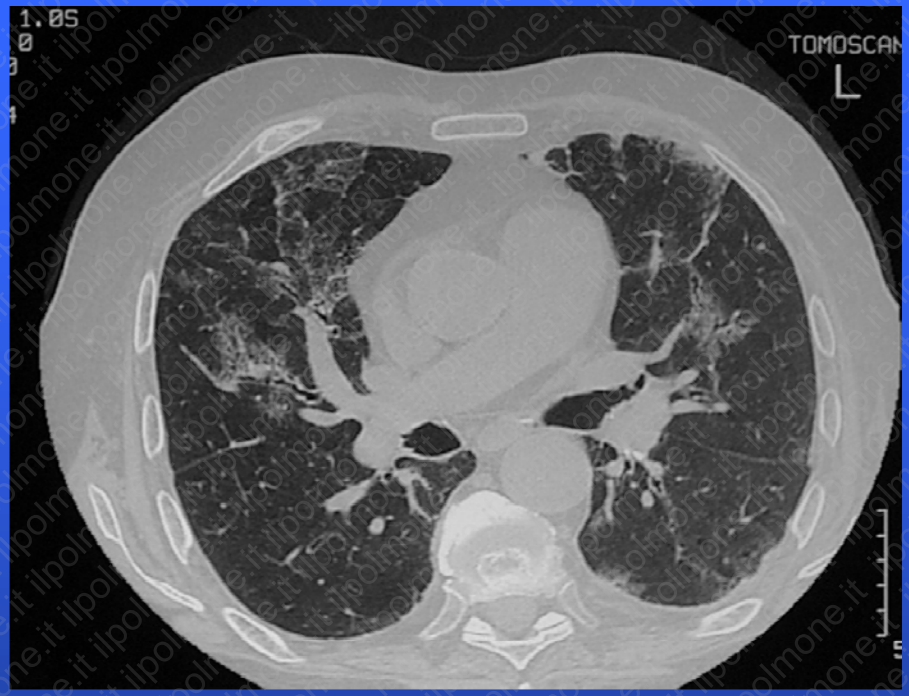
Relapse rate of 67 patients with ANCA-associated vasculitides

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%)

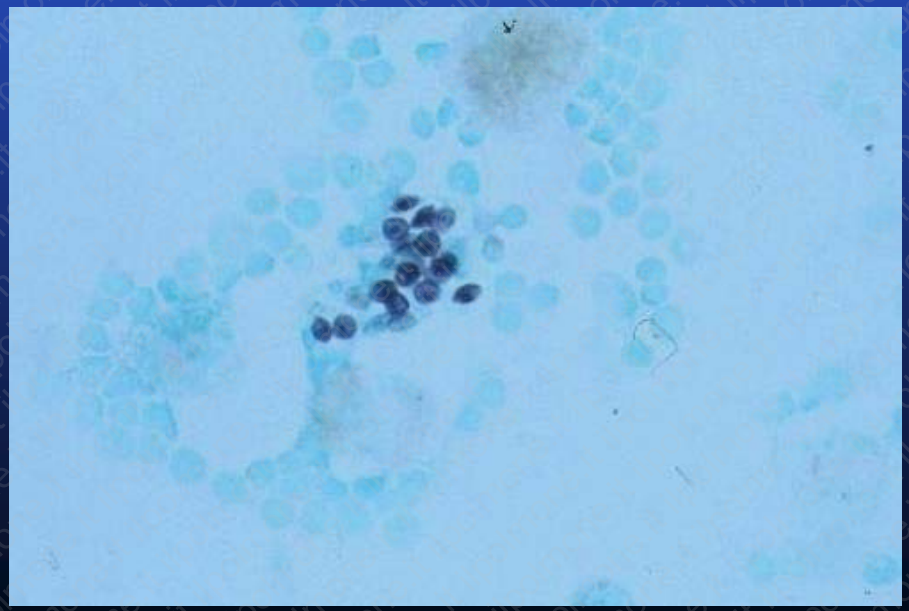
Prognostic factors for death of 67 patients with ANCA-associated vasculitides

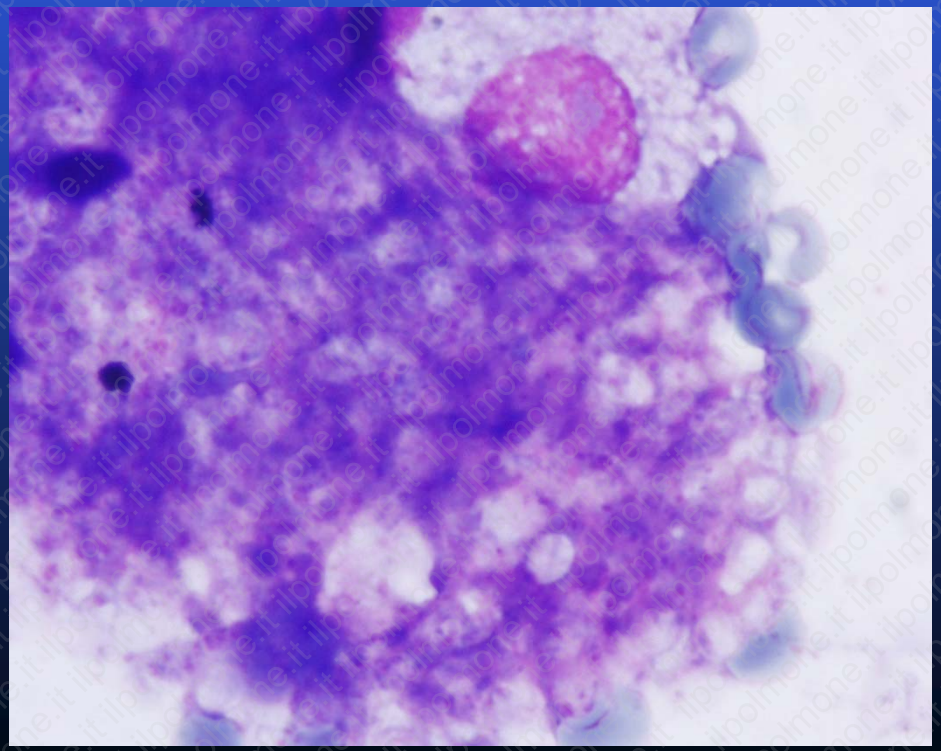
The global risk of death was 22% and 35% after 5 and 10 years of follow up

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



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

Key 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

Therapy

- Induction of remission: 12 months
- Maintenance: 12-18 months
  - Cyclophosphamide → azathioprine/methotrexate
  - Additional agents: mycophenolate mofetil (MMF), leflunomide, cyclosporine
  - Pneumocystis carinii prophylaxis with trimetroprim-sulfametoxazole