Interessamento polmonare nelle vasculiti

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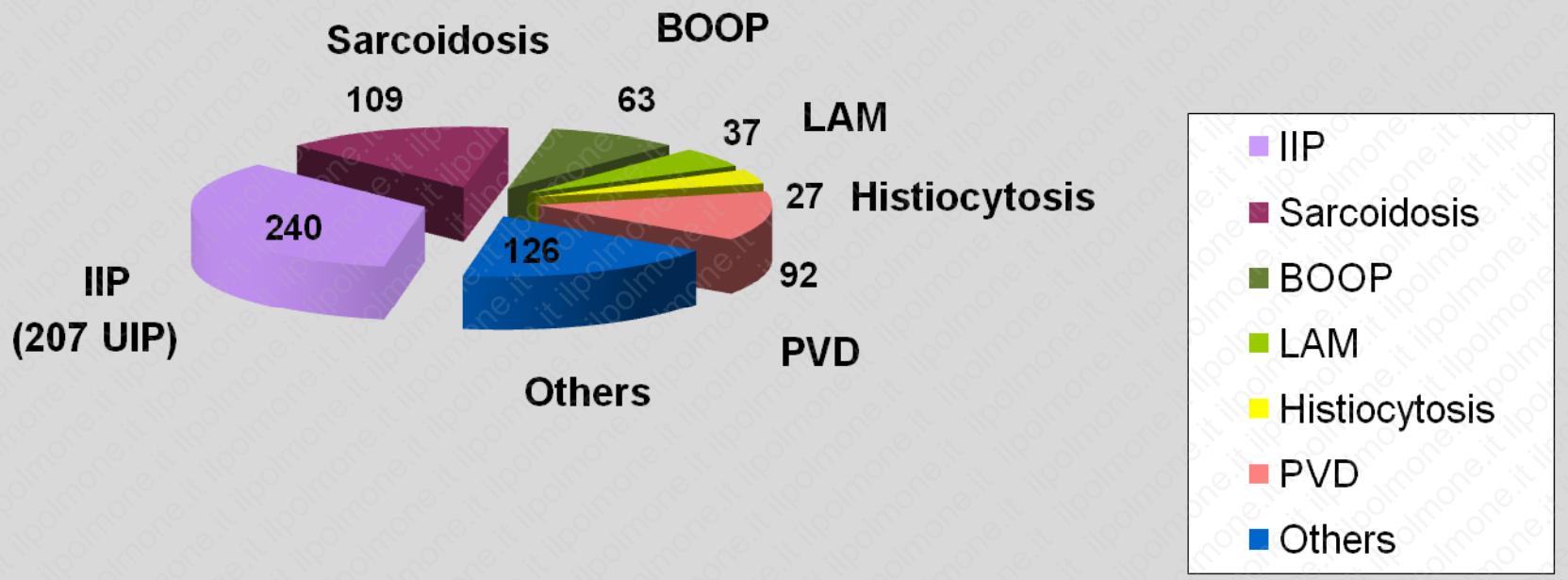
Rare Lung Diseases

Ospedale San Giuseppe Experience (2001- february 2009)

Tot. 694 pts



VASCULITI POLMONARI

Key message

The diagnosis and management of a systemic vasculitis is among the most demanding challenges in clinical medicine

DEFINITION

- can be pathologically defined by the presence of
  - cellular inflammation
  - vassel destruction and
  - tissue necrosis

The characteristics of the inflammation can be helfull in determining the underlying diagnosis, and granulomatous, eosinophilic, lymphoplasmacytic or neutrophilic patterns can be seen

DEFINITION

- The clinical features of each disease are determined by the site, size, and type of vessel involved and
- by the relative amounts of inflammation, vessel destruction and tissue necrosis

Classification of the vasculitides

Primary idiopathic vasculitis

Small vessel

Wegener's granulomatosis

Churg-Strauss syndrome

Microscopic polyangiitis

Idiopathic pauci-immune rapidly progressive glomerulonephritis

Isolated pauci-immune pulmonary capillaritis

Medium vessel

Polyarteritis nodosa

Kawasaki disease

Large vessel

Giant cell arteritis

Takayasu's arteritis

Primary immune complex-mediated vasculitis

Goodpasture's syndrome

Henoch-Schonlein purpura

Behcet's disease

IgA nephropaty

Secondary vasculitis

Classic autoimmune disease

Systemic lupus erithematous

Rheumatoid arthritis

Polymiositis/dermatomyositis

Scleroderma

Antiphospholipid antibody syndrome

Essential cryoglobulinemia

Inflammatory bowel disease

Hypocomplementemic urticarial vasculitis

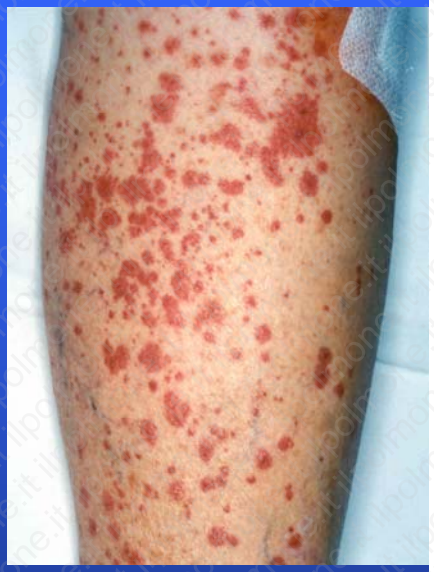
Drug-induced (e.g. propylthiouracil)

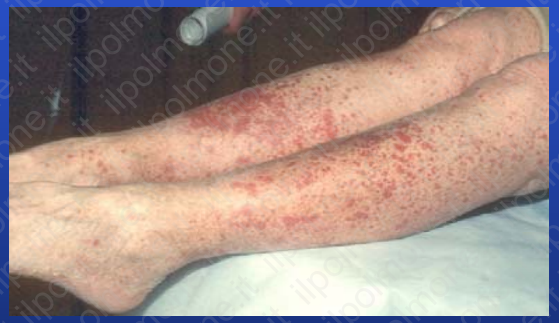
Paraneoplastic

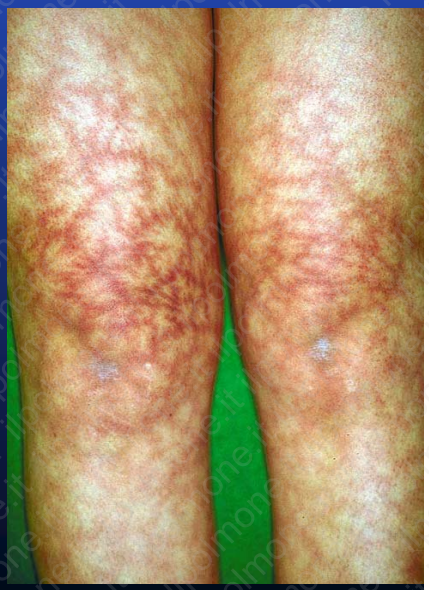
Infection

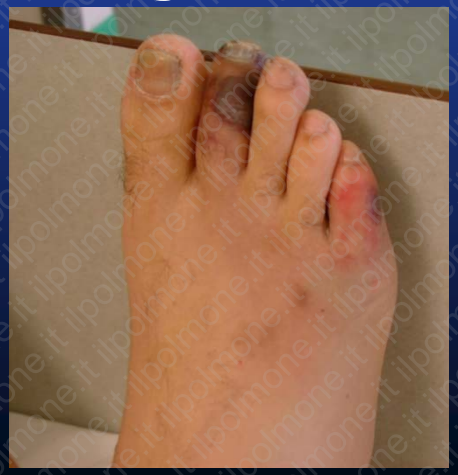
Proteiform Systemic Vasculitides

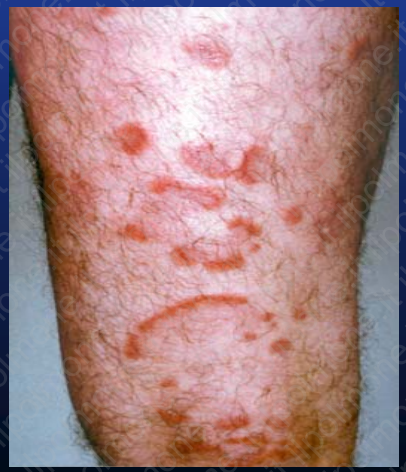
Systemic vasculitides may change into many shapes. Systemic vasculitides can affect virtually one or more organ and/or system resulting in a wide variety of signs and symptoms, and are challenging to diagnose and to treat

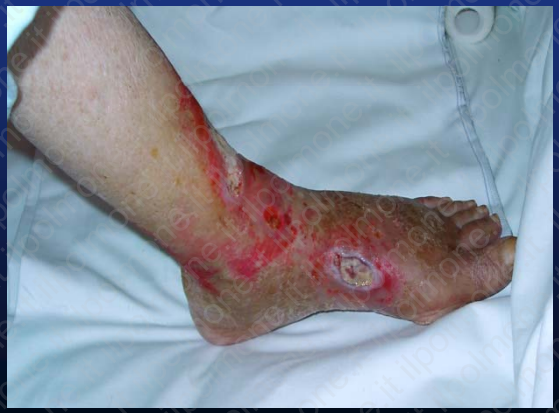




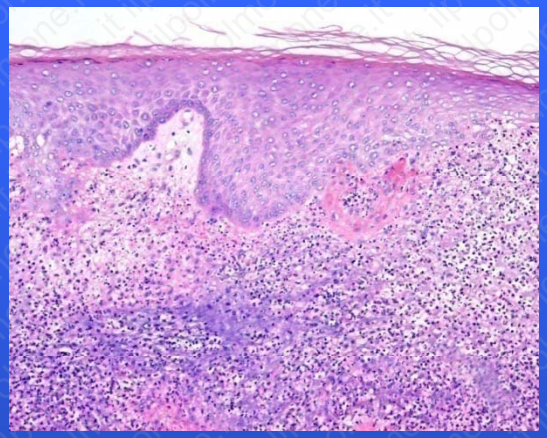


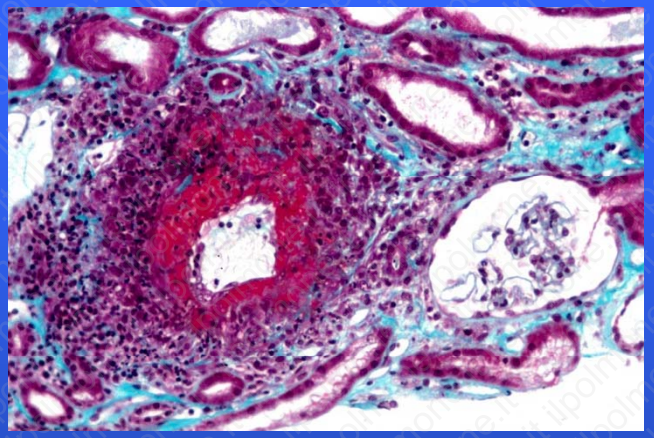


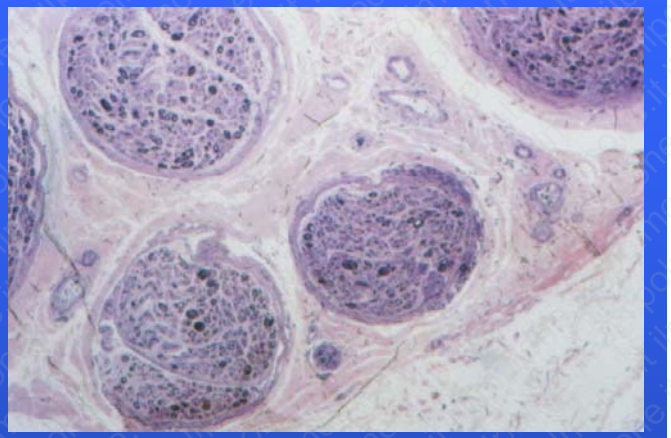


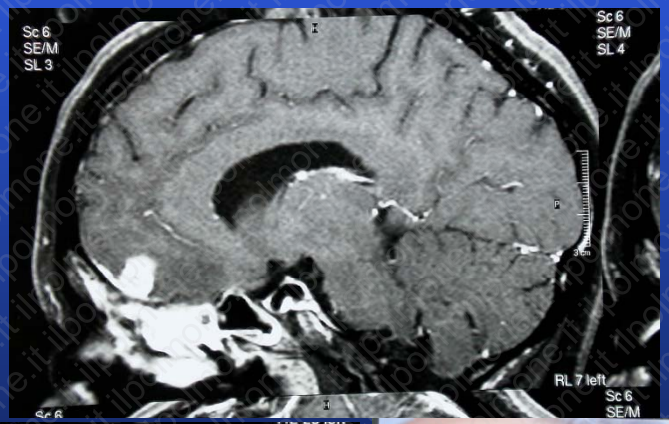


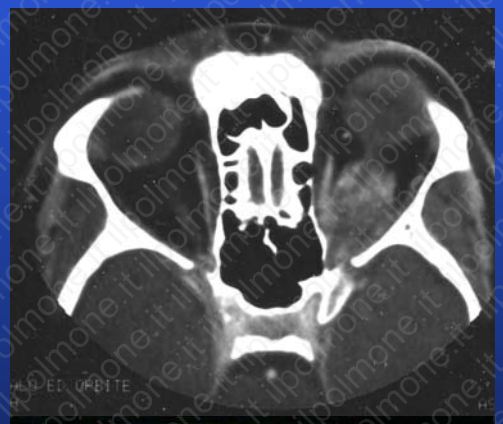
Proteiform Systemic Vasculitides

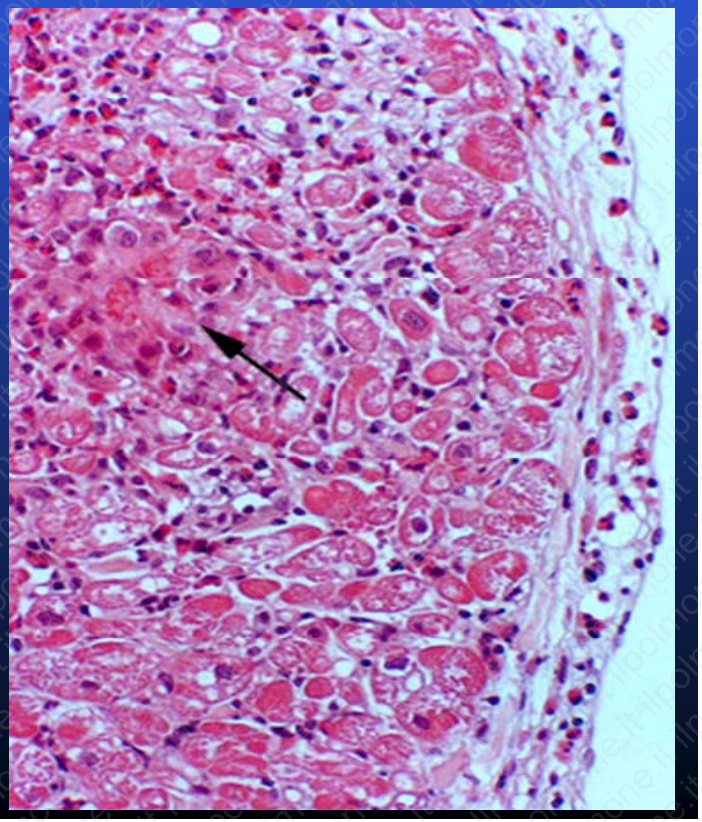


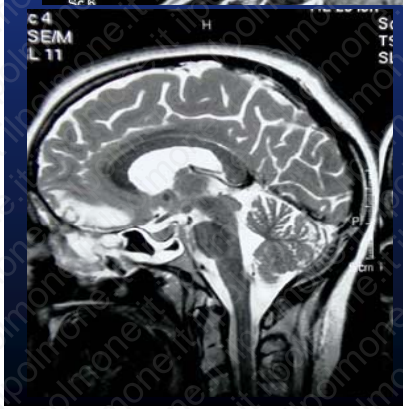


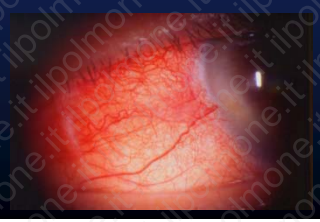


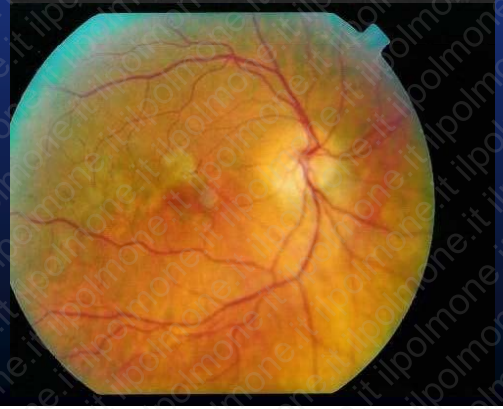




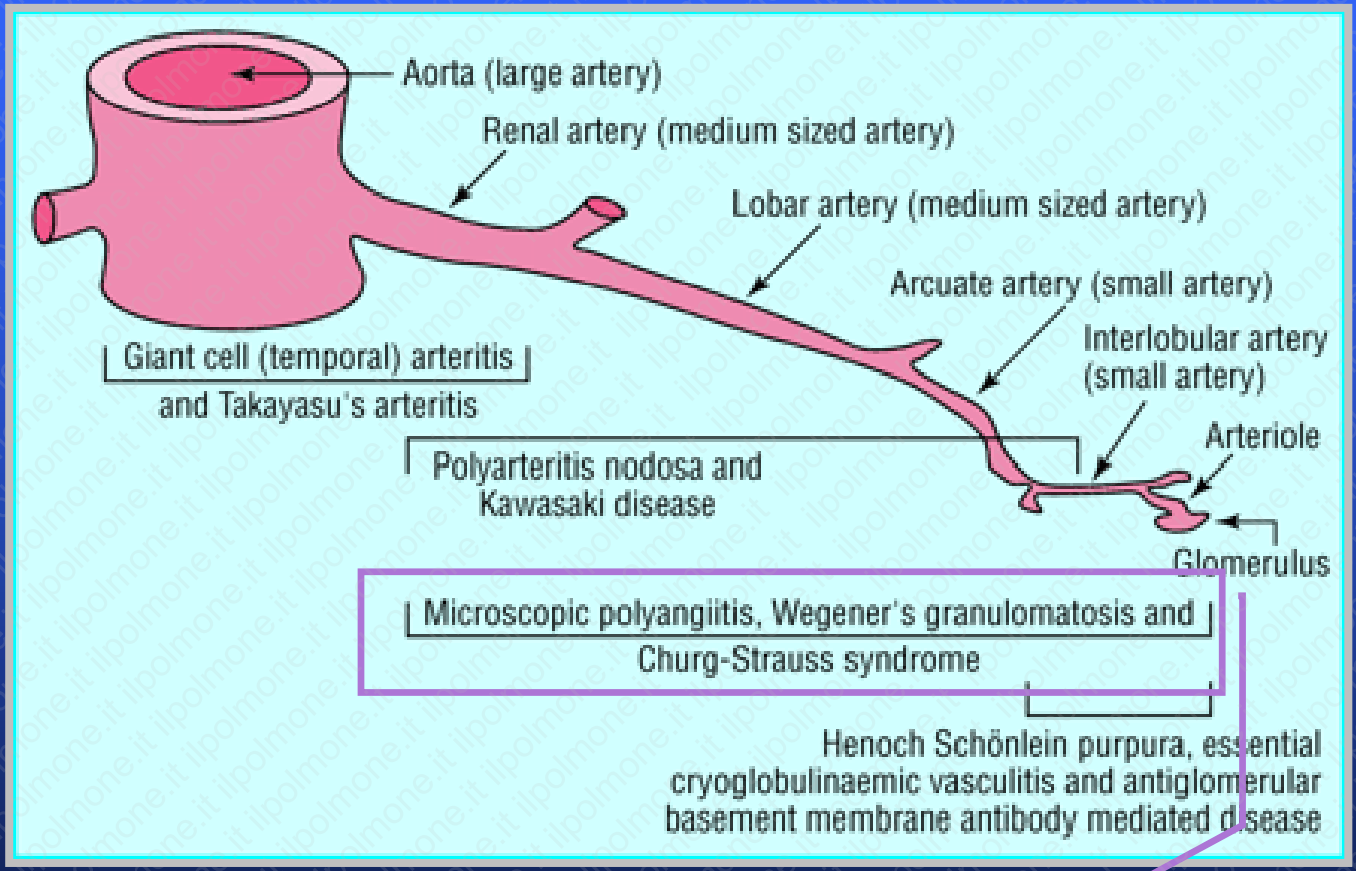






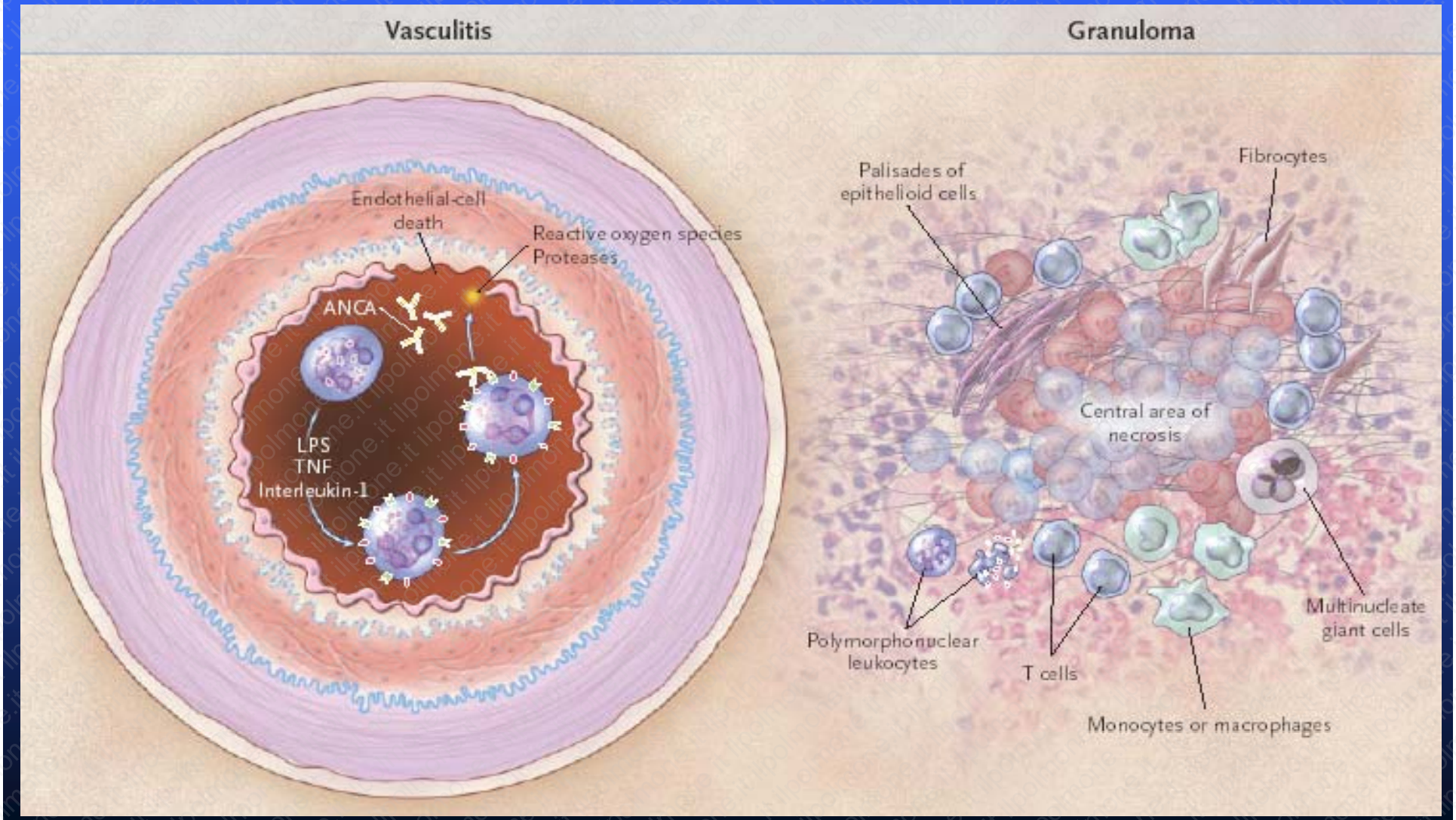


Systemic Vasculitides and Respiratory System



The respiratory system may be involved in all systemic vasculitides, although more frequently in the small vessel ANCA-associated vasculitis

Pathogenetic Role of ANCA



ANCA-associated vasculitides

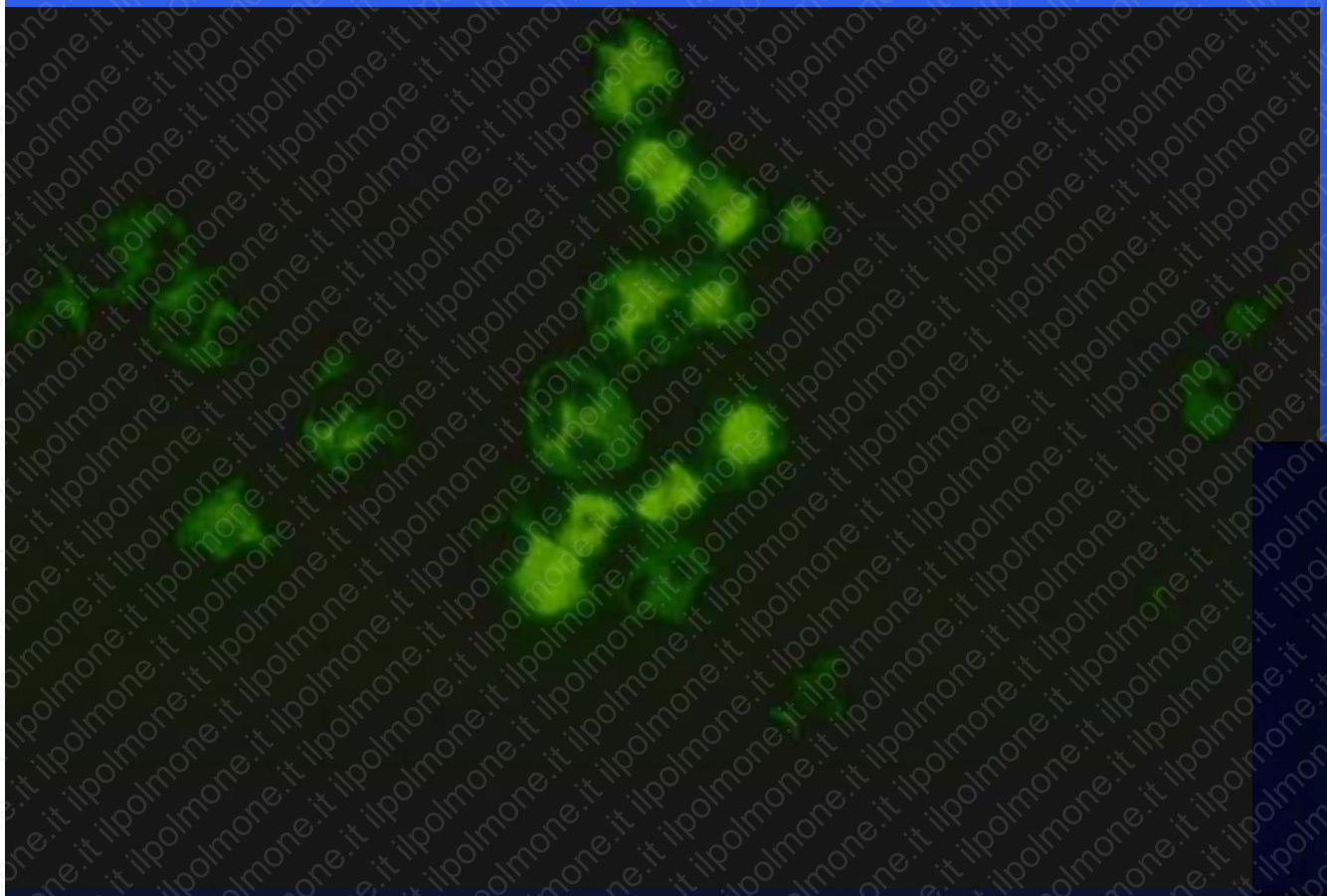
Key message

Are grouped together because of common clinical features, pathologic involvement of the small vessels, similar responses to immunosuppressive interventions and ANCA positivity (which is common but not universal)

Autoanticorpi Anti Citoplasma dei Neutrofili (ANCA)

	Antigene bersaglio	**Malattie Associate**
c-ANCA	Proteinosi 3 (CAP 57)	Granulomatosi di Wegener (90%) Poliangite Microscopica, S. di Churg-Strauss
p-ANCA	Mieloperossidasi, Elastasi, Catepsina G, Lisozima, Lattoferrina	Vasculite Renale, RPGN, S. di Churg-Strauss (50%), Poliangite microscopica (70%), connettiviti
Atipici (x) ANCA O p-ANCA	Lattoferrina, Lisozima, Beta-glucuronidasi, Catepsina G	Colite Ulcerosa, Epatite Autoimmune, Colangite Sclerosante Primitiva

ANCA
Indirect immunofluorescence staining



c-ANCA

p-ANCA

ANCA-associated vasculitides

- Each pattern is associated with antibodies against intracellular antigen(s) found in neutrophils and monocytes
- The sensitivity, specificity, and PPV of c-ANCA for WG and p-ANCA for MPA, CSS and idiopathic pauci-immune RPGN are critical in determining their diagnostic utility
- If these tests are not applied selectively to high-risk populations, then the PPV of the testing declines

ANCA-associated vasculitides

- In patients with higher risk for an ANCA-associated vasculitis, the PPV of the tests increased without reducing sensitivity
- The combination of ANCA indirect immunofluorescent testing plus ELISA testing maximizes their sensitivity
- c-ANCA is highly sensitive (90-95%) in active, systemic WG, with a specificity of approximately 90%