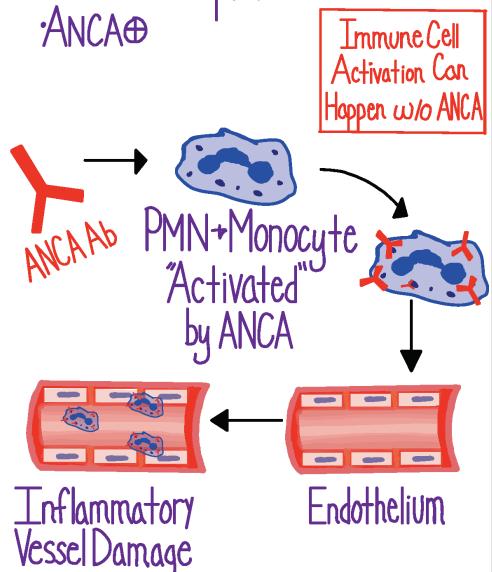


Granulomatosis with Polyangiitis

Overview

Small Vessel Vasculitis

- Normal complement
- ANCA \ominus



Epidemiology

- Rare
- Middle Age & Up
- M \approx F
- More Common in Caucasians

Clinical Manifestations

Inflammation

- Fevers
- Malaise
- Arthralgias
- Weight Loss



Ear, Nose, Throat 85%

- Recurrent Sinusitis
- Recurrent Otitis
- Saddle-Nose Deformity

Lower Resp. Tract 60%

- Hemoptysis
- Subglottic Stenosis
- Pulmonary Nodules

Renal 75%

- AKI
- Glomerulonephritis

Skin 30%

- Leukocytoclastic Vasculitis
- Necrosis, Ulceration

Neurologic 30%

- Mononeuritis Multiplex
- Cranial Neuropathy

Ophtho 40%

- Scleritis/Episcleritis
- Corneal Ulcers

Diagnosis

With Biopsy

- Preferred
- \oplus ANCA+Biopsy

Common

- Renal → Pauci-Immune GN
- Skin → Leukocytoclastic Vasculitis

Rare

- Lung → Granulomatous Inflammation

Without Biopsy

- NOT Preferred
- \oplus ANCA+Clinical Manifestations

Treatment

Glucocorticoids + Steroid-Sparing
ie. Methotrexate

\oplus ANCA in ~85%
of cases
 \hookrightarrow c-ANCA/PR3
Most Common