

Granulomatosis With Polyangiitis (Wegeners) Characteristics

Wegener's granulomatosis, more recently termed granulomatosis with polyangiitis, is a small and medium vessel necrotizing vasculitis that typically affects the nose, lungs, and kidneys. Wegener's is characterized by granulomatous inflammation against a nonspecific inflammatory background thought to be caused by the presence of C-ANCA or cytoplasmic anti-neutrophil cytoplasmic antibodies. These antibodies react with proteinase 3, an enzyme within neutrophil granulocytes. The lungs and upper airways are classically involved and can cause chronic sinusitis, otitis media, perforation of the nasal septum, and saddle nose deformity. Individuals also commonly complain of cough and hemoptysis. The kidneys are also classically involved and can cause glomerulonephritis with hematuria. Cyclophosphamide and corticosteroids can be used as immune suppressants in the treatment of granulomatosis with polyangiitis.



PLAY PICMONIC

Pathophysiology

Granulomatosis With Polyangiitis

Granny-llama and Poly-angel

Wegener's granulomatosis, more recently termed granulomatosis with polyangiitis, is a small vessel vasculitis that typically affects the nose, lungs, and kidneys. Wegener's granulomatosis was named after the German pathologist Friedrich Wegener who was part of the Nazi Party in 1932. After the discovery of his Nazi past, a campaign was begun to rename Wegener's disease to granulomatosis with polyangiitis.

Vasculitis

Blood-vessels

Vasculitis is a general term of inflammation of the blood vessel walls. Clinical features of various vasculitides largely depend on the vascular beds affected. This is a small and medium vessel vasculitis that typically affects the nose, lungs, and kidneys.

Necrotizing

Necrosis-crow

Granulomatosis with polyangiitis is commonly described as a necrotizing vasculitis due to necrosis of the vessels it affects. It is also characterized by necrotizing granulomas and focal necrotizing renal disease.

C-ANCA

C Anchor

Wegener's is characterized by granulomatous inflammation against a nonspecific inflammatory background thought to be caused by the presence of C ANCA or cytoplasmic anti-neutrophil cytoplasmic antibodies. These antibodies react with proteinase 3, an enzyme within neutrophil granulocytes.

Lungs And Upper Airway

Lungs

The lungs and upper airways are classically involved with granulomatosis with polyangiitis and can present with chronic sinusitis, ulcerative lesions of the nose and palate, and nodules and cavities in the lungs.

Crescentic Glomerulonephritis

Crescent on Glowing-mare

The kidneys are classically involved in granulomatosis with polyangiitis and can exhibit a spectrum of disease including focal necrosis of glomeruli to diffuse necrosis and parietal cell proliferation to form crescents seen in crescentic glomerulonephritis.

Treatment

Cyclophosphamide

[Cyclops-phosphate-P](#)

This is a nitrogen mustard alkylating agent that can be used to treat various types of cancer and some autoimmune disorders including granulomatosis with polyangiitis. Hemorrhagic cystitis is a common complication of drug use that can be prevented with mesna and adequate fluid intake.

Corticosteroids

[Quarter-on-steroids](#)

Corticosteroids are commonly used in the treatment of granulomatosis with polyangiitis due to immunosuppressive effects.