# Vasculitis

| Disease | Site | Special | Cause | Clinical Features | Diagnosis & Treatment |
|---|---|---|---|---|---|
| **Giant-Cell (Temporal) Arteritis** | large to small-sized arteries temporal arteries<br>Rarely the aorta | Chronic, granulomatous inflammation | Unknown cause<br>Likely immune, T cell-mediated | > 50 years<br>Vague symptoms<br>facial pain or headache<br>painful to palpation | Biopsy<br>Treatment: corticosteroids |
| **Polyarteritis Nodosa ( PAN )** | Systemic<br>Small or medium-sized muscular arteries ONLY involving renal and visceral vessels but **sparing** the pulmonary circulation | all stages of activity coexist in different vessels or in the same vessel<br>30% of patients have hepatitis B antigenemia | Complications :<br>Vessel rupture<br>Ulcerations<br>Infarcts<br>Ischemic atrophy<br>Haemorrhages in the distribution of affected vessels may be the first sign of disease | Largely young adults<br>Fever and weight loss<br>Renal (arterial) cause death<br>Hypertension,<br>Abdominal pain and melena<br>Diffuse muscular aches and pains<br>Peripheral neuritis | Biopsy<br>corticosteroids<br>results in remissions or cures in 90% |
| **Microscopic Polyangiitis** | capillaries as well as arterioles and venules<br>*pulmonary capillaritis are particularly common* | *All lesions tend to be of the same age*<br>*Necrotizing glomerulonephritis (90% of patients)*<br>p-ANCAs are present in more than 70% of patients<br>autoantibodies are specific for myeloperoxidase (MPO) | antibody response to antigens, drugs (penicillin), microorganisms (streptococci), heterologous proteins, or tumor proteins is the presumed cause<br>This can result in immune complex deposition, or it may trigger secondary immune responses | Depending on the organ involved, major clinical features include:<br>Hemoptysis<br>Hematuria and proteinuria<br>Bowel pain or bleeding<br>Muscle pain or weakness<br>Palpable cutaneous purpura | |
| **Wegener Granulomatosis** | small to medium-sized vessels<br>Associated with C-ANCAs target antigen is proteinase-3 (PR3) | *Focal necrotizing, often crescentic, glomerulitis Acute necrotizing granulomas* of the upper and lower respiratory tract | 40-50 years<br>Without Rx -> 80% die<br>With Rx -> 90% live (not cured)<br>The Rx -> immunosuppression | | |
| **Churg-Strauss syndrome** | small vessels<br>Associated with p-ANCAs. autoantibodies are specific for myeloperoxidase (MPO) | Eosinophil-rich and granulomatous inflammation involving the respiratory tract | strong association with<br>Allergic rhinitis<br>Bronchial asthma<br>Eosinophilia | Vessels in the lung, heart, spleen ... are frequently involved by:<br>intravascular and extravascular granulomas<br>infiltration of vessels and perivascular tissues by eosinophils is striking | |
| **Thromboangiitis obliterans (Buerger disease)** | of medium-sized and small arteries<br>Tibial and radial arteries, with occasional secondary extension into extremity veins and nerves | Segmental, thrombosing acute and chronic inflammation | vascular insufficiency caused by atherosclerosis, in Buerger disease the insufficiency tends to be accompanied by **severe pain,** even at rest, related undoubtedly to the neural involvement<br>Unknown cause | Chronic ulcerations of the toes, feet, or fingers may appear, perhaps followed in time by frank gangrene | Heavy smoker<br>Begins before the age of 35<br>Superficial nodular phlebitis<br>Cold sensitivity of the Raynaud type in the hands<br>Pain in the instep of the foot induced by exercise (so-called *instep claudication*) |