

## Vasculitis

Vasculitis	Pathology	Epidemiology	Clinical picture
<b>Takayasu's arteritis</b> ("pulseless disease")	Granulomatous <b>large vessel vasculitis</b> involving <b>aortic arch vessels</b>	Young women and children	Absent upper extremity pulse Visual defects, stroke
<b>Giant cell (temporal) arteritis</b>	Granulomatous <b>large vessel vasculitis</b> involving <b>superficial temporal and ophthalmic arteries</b> ; thrombi contain microabscesses	Adults > 50 years of age	Temporal headache, jaw claudication (pain when chewing) Blindness on ipsilateral side Polymyalgia rheumatica (muscle and joint pain; normal serum creatine kinase) Increased ESR
<b>Polyarteritis nodosa</b>	Necrotizing <b>medium-sized vessel vasculitis</b> involving <b>renal, coronary, mesenteric</b> arteries ( <u>sparer pulmonary arteries</u> )	Middle-aged men Association with <b>HBsAg</b> (30%)	Vessels at all stages of acute and chronic inflammation Focal vasculitis produces aneurysms (detected with angiography) Organ infarction in kidneys (renal failure), heart (acute MI), bowels (bloody diarrhea), skin (ischemic ulcer)
<b>Kawasaki disease</b>	Necrotizing <b>medium-sized vessel vasculitis</b> involving <b>coronary arteries</b>	Children < 4 years of age	Desquamating rash, swelling of hands and feet, cervical adenopathy, oral erythema Abnormal ECG (e.g., acute MI) <u>Corticosteroids contraindicated</u> (danger of vessel rupture)
<b>Thromboangiitis obliterans (Buerger's disease)</b>	<b>Medium-sized vessel vasculitis</b> with <b>digital vessel</b> thrombosis	Men 25-50 years of age who smoke cigarettes	Foot claudication, Raynaud's phenomenon, ulceration, gangrene
<b>Raynaud's disease</b>	<b>Medium-sized vessel vasculitis</b> involving <b>digital vessels</b> in fingers and toes	Young women Exaggerated vasomotor response to cold or stress	Paroxysmal digital color changes (white-blue-red sequence) Ulceration and gangrene in chronic cases
<b>Raynaud's phenomenon</b>	<b>Medium-sized vessel vasculitis</b> involving <b>digital vessels</b> in fingers and toes	Adult men and women Secondary to other diseases (e.g., systemic sclerosis, CREST syndrome)	Systemic sclerosis and CREST syndrome: digital vasculitis with vessel fibrosis, dystrophic calcification, ulceration, gangrene
<b>Wegener's granulomatosis</b>	Necrotizing <b>medium-sized and small vessel vasculitis</b> involving <b>upper respiratory tract, lung, renal vessels</b>	Childhood to middle age	Necrotizing vasculitis in upper respiratory tract (nasopharynx, sinuses, trachea), lower respiratory tract (pulmonary vessels; infarction, pneumonia), kidneys (crescentic glomerulonephritis) c-ANCA antibodies (>90% of cases) Treatment: corticosteroids, cyclophosphamide

<b>Microscopic polyangiitis</b>	<b>Small vessel vasculitis</b> involving skin, lung, brain, GI tract, postcapillary venules and glomerular capillaries	Children and adults Precipitated by drugs (e.g., penicillin), infections (e.g., streptococci), immune disorders (e.g., SLE)	Vessels at same stage of inflammation Palpable purpura, glomerulonephritis <i>p</i> -ANCA antibodies (>80% of cases)
<b>Churg-Strauss syndrome</b>	<b>Small vessel vasculitis</b> involving skin, lung, heart vessels	Children and adults	Allergic rhinitis, asthma <i>p</i> -ANCA antibodies (70% of cases), eosinophilia
<b>Henoch-Schönlein purpura</b>	<b>Small vessel vasculitis</b> involving skin, GI tract, renal, joint vessels	Children and young adults Most common vasculitis in children IgA immunocomplexes	Palpable purpura of buttocks and lower extremities Polyarthritits, glomerulonephritis, GI bleeding
<b>Cryoglobulinemia</b>	<b>Small vessel vasculitis</b> involving skin, GI tract, renal vessels	Adults Association with <i>HCV</i> , type I MPGN	Cryoglobulins: immunoglobulins that gel at cold temperatures Palpable purpura, acral cyanosis of nose and ears and Raynaud's phenomenon (reverses when in warm room)
<b>Serum sickness</b>	<b>Small vessel vasculitis</b> involving immunocomplex deposition in skin vessels	Children and adults Complication of treatment with horse- or sheep-based antivenin	Fever, urticaria with vasculitis, arthralgia, GI pain with melena
<b>Infectious vasculitis</b>	<b>Small vessel vasculitis</b> involving skin vessels	Children and adults Involves all microbial pathogens	Rocky Mountain spotted fever: tick transmission of <i>Rickettsia rickettsiae</i> ; organisms invade endothelial cells; petechiae on palms spread to trunk Disseminated meningococcemia due to <i>Neisseria meningitidis</i> Capillary thrombosis produces hemorrhage into skin and confluent ecchymoses