Systemic vasculitis:

Vasculitis is characterised by inflammation and necrosis of blood-vessel walls, with associated damage to skin, kidney, lung, heart, brain and gastrointestinal tract.

Large-Vessel Vasculitis

Takayasu's arteritis

Giant cell arteritis

Medium-Vessel Vasculitis

Polyarteritis nodosa

Kawasaki's disease

Small-Vessel Vasculitis

Anti-neutrophil Cytoplasmic Antibody–Associated Vasculitis

Cryoglobulinemic vasculitis

IgA vasculitis (Henoch-Schönlein purpura)

Clinical features of systemic vasculitis

Systemic

• Malaise

• Fever

• Night sweats

• Weight loss with arthralgia and myalgia

Rashes

• Palpable purpura

• Pulp infarcts

• Ulceration

• Livedo reticularis

Ear, nose and throat

• Epistaxis

• Recurrent sinusitis

• Deafness

Respiratory

• Haemoptysis

• Cough

• Poorly controlled asthma

Gastrointestinal

• Abdominal pain (due to mucosal inflammation or enteric ischaemia)

• Mouth ulcers

• Diarrhoea

Neurological

• Sensory or motor neuropathy

Large vessels vasculitis:

Takayasu arteritis

- Takayasu arteritis affects the aorta, its major branches and occasionally the pulmonary arteries.
- The typical age at onset is 25–30 years, with an 8 : 1 female-to-male ratio.
- It presents with claudication, fever, arthralgia and weight loss.
- Clinical examination may reveal loss of pulses, bruits, hypertension.
- Investigation will identify an acute phase response and normocytic, normochromic anaemia but the diagnosis is based on angiography.
- Treatment is with high-dose glucocorticoids and immunosuppressants.

Giant cell arteritis and polymyalgia rheumatic:

- Giant cell arteritis (GCA) is a granulomatous arteritis that affects any large (including aorta) and medium-sized arteries.

- It is commonly associated with polymyalgia rheumatica (PMR), which presents with symmetrical, immobility-associated neck and shoulder girdle pain and stiffness.
- Both diseases are rare under the age of 60 years. The average age at onset is 70, with a female-to-male ratio of about 3 : 1.

Clinical features

- The cardinal symptom of GCA is headache, which is often localized to the temporal or occipital region and may be accompanied by scalp tenderness.
- Jaw pain develops in some patients, brought on by chewing or talking.
- Visual disturbance can occur (most specifically amaurosis) and a catastrophic presentation is with blindness in one eye due to occlusion of the posterior ciliary artery.
- On fundoscopy, the optic disc may appear pale and swollen with haemorrhages, but these changes may take 24–36 hours to develop and the fundi may initially appear normal.
- In GCA, constitutional symptoms, such as weight loss, fatigue, malaise and night sweats, are common.
- With PMR, there may be stiffness and painful restriction of active shoulder movements on waking. Muscles are not otherwise tender, and weakness and muscle-wasting are absent

Investigations

- The typical laboratory abnormality is an elevated ESR, often with a normochromic, normocytic anaemia. CRP may also be elevated.
- There are three investigations to consider: temporal artery biopsy, ultrasound of the temporal arteries and 19fluorodeoxyglucose positron emission tomography (19FDG PET scan).
- A negative biopsy does not exclude the diagnosis.
- On ultrasound examination, affected temporal arteries show a ‘halo’ sign. A strongly positive 19FDG PET scan is highly specific but sensitivity is low.

Management

Prednisolone should be commenced urgently in suspected GCA because of the risk of visual loss. It is dramatic, such that symptoms will completely resolve within 48–72 hours of starting therapy in virtually all patients. It is customary to use

higher doses in GCA (60–80 mg prednisolone) than in PMR (15–20 mg). Most patients need glucocorticoids for an average of 12–24 months

Medium vessels vasculitis:

Kawasaki disease

- Kawasaki disease is a vasculitis that mostly involves the coronary vessels.
- It usually affecting children under 5 years.
- Presentation is with fever, generalised rash, including palms and soles, inflamed oral mucosa and conjunctival injection resembling a viral exanthema.
- Cardiovascular complications include coronary arteritis, leading to myocardial infarction.
- Treatment is with aspirin and IVIg .

Polyarteritis nodosa

- Polyarteritis nodosa has a peak incidence between the ages of 40 and 50, with a male-to-female ratio of 2 : 1.
- Hepatitis B is an important risk factor, in combination with manifestations of multisystem disease.
- The most common skin lesions are palpable purpura, ulceration, infarction and livedoreticularis.
- neuropathy, which is typically symmetrical and affects both sensory and motor function.
- Severe hypertension and/or renal impairment may occur due to multiple renal infarctions but glomerulonephritis is rare.
- The diagnosis is confirmed by conventional or magnetic resonance angiography, which shows multiple aneurysms and smooth narrowing of mesenteric, hepatic or renal systems, or by muscle or sural nerve biopsy.
- Treatment is with high-dose glucocorticoids and immunosuppressants

small vessels vasculitis:

Antineutrophil cytoplasmic antibody-associated vasculitis:

Antineutrophil cytoplasmic antibody-associated vasculitis (AAV) is a life-threatening disorder characterised by inflammatory infiltration of small blood vessels, fibrinoid necrosis.

1- Microscopic polyangiitis is a necrotising small-vessel vasculitis found with rapidly progressive glomerulonephritis, often in association with alveolar haemorrhage, Patients are usually myeloperoxidase (MPO) antibody-positive
2- granulomatosis with polyangiitis (formerly known as Wegener's granulomatosis) is characterised by granuloma formation, mainly affecting the nasal passages, airways and kidney. A minority of patients present with glomerulonephritis. The most common presentation of granulomatosis with polyangiitis is with epistaxis, nasal crusting and sinusitis, but haemoptysis and mucosal ulceration may also occur. Deafness may be a feature due to inner ear involvement, and proptosis may occur because of inflammation of the retro-orbital tissue. Untreated nasal disease ultimately leads to destruction of bone and cartilage. Migratory pulmonary infiltrates and nodules occur in 50% of patients (as seen on high-resolution CT of lungs). Patients with granulomatosis with polyangiitis are usually proteinase-3 (PR3).
3- Eosinophilic granulomatosis with polyangiitis (formerly known as Churg–Strauss syndrome: It is associated with eosinophilia. Some patients have a prodromal period for many years, characterised by allergic rhinitis, nasal polyposis and late-onset asthma that is often difficult to control. The typical acute presentation is with a triad of skin lesions (purpura or nodules), asymmetric mononeuritis multiplex and eosinophilia. antibodies to MPO or PR3 can be detected in up to 60% of cases

The diagnosis should be confirmed by biopsy of the kidney or lesions in the sinuses and upper airways.

Management for organ-threatening or acute–severe disease is with high-dose glucocorticoids (e.g. daily pulse intravenous methylprednisolone 0.5–1 g for 3 days, then oral prednisolone 0.5 mg/kg) and intravenous cyclophosphamide (e.g. 0.5–1 g every 2 weeks for 3 months), followed by maintenance therapy with lower-dose glucocorticoids and azathioprine, methotrexate or MMF. Plasmapheresis

should be considered for fulminant lung disease. Rituximab in combination with high-dose glucocorticoids is equally effective as oral cyclophosphamide at inducing remission in AAV.

Henoch–Schönlein purpura

- Henoch–Schönlein purpura is a small-vessel vasculitis caused by immune complex deposition following an infectious trigger.
- It is predominantly a disease of children and young adults.
- The usual presentation is with purpura over the buttocks and lower legs, accompanied by abdominal pain, gastrointestinal bleeding and arthralgia. Nephritis can also occur and may present up to 4 weeks after the onset of other symptoms.
- Biopsy of affected tissue shows a vasculitis with IgA deposits in the vessel wall.
- Henoch–Schönlein purpura is usually a self-limiting disorder that settles spontaneously without specific treatment.
- Glucocorticoids and immunosuppressive therapy may be required in patients with more severe disease,

Cryoglobulinaemic vasculitis

- This is a small-vessel vasculitis that occurs when immunoglobulins precipitate out in the cold.
- The typical presentation is with a vasculitic rash over the lower limbs, arthralgia, Raynaud's phenomenon and neuropathy.
- Some cases are secondary to hepatitis C infection and others are associated with other autoimmune diseases.
- Glucocorticoids and immunosuppressive therapy are often used

Read more in:

- Davidson's Principles and Practice of Medicine, 23rd edition
- Kelley & Firestein's Textbook of Rheumatology, 10th edition