Churg-Strauss Syndrome

Synonyms: allergic granulomatosis angitis, granulomatous small-vessel vasculitis, eosinophilic granulomatosis with polyangiitis (EGPA)

A rare diffuse vasculitic disease affecting coronary, pulmonary, cerebral, abdominal visceral and skin circulations. The vasculitis affects small- and medium-sized arteries and veins and is associated with asthma.

The aetiology is unknown although autoimmune and genetic factors have been implicated. Drug-induced Churg-Strauss syndrome (CSS) has been reported. Drugs implicated have included mesalazine, propylthiouracil, methimazole, freebase cocaine and leukotriene receptor antagonists.

The American College of Rheumatology has identified six criteria for the diagnosis of CSS:

- Asthma (wheezing, expiratory rhonchi).
- Eosinophilia of more than 10% in peripheral blood.
- Paranasal sinusitis.
- Pulmonary infiltrates (may be transient).
- Histological confirmation of vasculitis with extravascular eosinophils.
- Mononeuritis multiplex or polyneuropathy.

The presence of four out of six of these features has a high specificity and sensitivity for the diagnosis of CSS.

Epidemiology

- Rare with incidence of about 1-2 per million people and prevalence about 10-15 per million people.
- The age at onset usually varies from 15-70 years, although patients as young as 9 have been reported.

Presentation

The presentation will depend on which systems are involved. In any patient with asthma and/or nasal polyposis, any new or worsening general or constitutional symptoms, including fever, joint pain, diffuse muscle pain, major involuntary weight loss, chest pain, palpitations or abdominal pain, may be the first signs of a vasculitis, including CSS.

The most prominent symptoms and signs include:

- Pulmonary: asthma, pneumonitis and haemoptysis.
- Upper respiratory tract: allergic rhinitis, paranasal sinusitis, nasal polyposis.
- Cardiac involvement is common. This includes heart failure, myocarditis and myocardial infarction.
- Skin: purpura, skin nodules, leukocytoclastic angitis with palpable purpura, livedo reticularis, urticaria, necrotic bullae and digital ischaemia.
- Renal: glomerulonephritis, hypertension and advanced chronic kidney disease.
- Peripheral neuropathy: mononeuritis multiplex is the most frequent form. Less frequent symptoms include stroke and eye involvement.
- Gastrointestinal: vasculitis and bleeding, bowel ischaemia and perforation, appendicitis and pancreatitis.
- Cholestatic liver dysfunction has been reported.
- Malaise, fatigue, weight loss, fever, myalgia and arthralgia.
- Myositis following unaccustomed exercise has been reported.

Differential diagnosis

There are many possible differential diagnoses to consider but include:

- Other causes of systemic vasculitis:
  - Primary vasculitis: eg, polyarteritis nodosa, granulomatosis with polyangiitis (Wegener's granulomatosis).
  - Secondary vasculitis: a complication of a connective tissue disorder (eg, rheumatoid arthritis), infection, medication, or malignancy.
  - Infections: eg, helminth/nematode.
  - Primary eosinophilic syndrome.
  - Malignancy: eg, leukaemias, lymphomas, myeloproliferative neoplasms, solid cancers (especially gastrointestinal, breast or lung).
  - Myelodysplastic syndromes.
Further reading & references

Vasculitis UK

4. Information for doctors; Churg Strauss Syndrome Association