Pulmonary, cardiac and renal syndrome

A 30-year-old man presented with symptoms of intermittent wheeze and breathlessness. A diagnosis of asthma was made and he received a 5-day course of oral prednisolone and inhaled beta-2 agonists. Five months later he became unwell with fatigue, anorexia, weight loss and was again wheezy. He received a course of amoxycillin without any symptomatic improvement. He was admitted to hospital and was found to have multiple splinter haemorrhages and a vasculitic rash particularly affecting the palms. Investigations included a normal chest radiograph but a substantially elevated blood eosinophil count of $35 \times 10^9/l$. There was evidence of mild renal impairment with a measured creatinine clearance of 69 ml/min and protein excretion of 0.15 g/24 h. An ECG showed widespread T-wave inversion.

Questions

What is your diagnosis?
What further investigations would be helpful?
What treatment would you give?
Churg-Strauss syndrome is a rare systemic vasculitis with an incidence of 2–3/million/year and which can cause renal disease [1]. The typical clinical pattern includes a prodrome of late onset asthma, peripheral blood eosinophilia, eosinophilic tissue infiltrates and vasculitis affecting extrapulmonary sites. The renal histological findings include focal necrotizing glomerulonephritis, a vasculitis often accompanied by a marked eosinophilic infiltrate and an eosinophilic interstitial nephritis has also been described. In other systemic vasculitides, antibodies to neutrophil cytoplasmic antibody (ANCA) may be present, may correlate with disease activity and might be of pathogenetic importance. In Churg-Strauss syndrome such antibodies appear to be predominantly against myeloperoxidase and in one series were detected in 60% of cases [2].

As occurred in this patient, cardiac involvement is well recognized in Churg-Strauss syndrome and may be associated with an adverse prognosis. Other features associated with poor prognosis are renal impairment, proteinuria, gastrointestinal tract involvement and CNS signs [3]. Cardiac involvement may manifest as cardiomyopathy, coronary vasculitis, myocardial fibrosis and mitral regurgitation [4].

Churg-Strauss syndrome is usually treated with corticosteroids which often produce significant clinical improvement. As in treatment of other systemic vasculitides, the administration of cytotoxic agents such as cyclophosphamide is recommended in severe disease, whilst the use of plasma exchange is unproven [1].

Suggested reading

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Fig. 1. Histology of the renal biopsy appearances in this patient with Churg-Strauss Syndrome showing (a) tubules with a prominent interstitial infiltrate including large numbers of eosinophils and (b) an artery showing vasculitis with fibrinoid change in part of its wall and further interstitial infiltrate.