brittle or difficult-to-control asthma. When such patients are using appropriate pharmacotherapy and are clinically stable, there is understandably a disinclination to titrate therapy downwards, in case one offsets the control of asthma, which has been difficult to attain in the first instance. In conclusion, secondary care physicians need to have a heightened awareness in terms of stepping-down ICS therapy in stable asthmatics. Failing to do so may expose patients to unnecessary and prolonged treatment with high doses of ICS.

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Aortic regurgitation and Churg-Strauss syndrome

Sir,

A 56-year-old lady who was a lifelong non-smoker was admitted with acute pulmonary oedema. Echocardiography revealed severe aortic valve regurgitation and a left ventricular ejection fraction of 35–40% associated with moderate left ventricular dysfunction. Coronary angiography confirmed aortic regurgitation, as well as 60% stenosis in the left anterior descending artery with diffuse disease of the distal vessel. Three years earlier, she had been diagnosed with systemic hypertension and was prescribed an angiotensin-converting-enzyme inhibitor. Two months prior to this admission, she was admitted acutely with left ventricular failure and complete heart block, successfully managed with diuretics and insertion of a dual chamber permanent pacemaker.

A clinical diagnosis of Churg-Strauss syndrome had been made 20 years earlier, based on severe asthma, sinusitis, marked peripheral eosinophilia, and mononeuritis multiplex involving her legs and hands. She improved following a 2-year course of tapered steroids and azathioprine, although

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Table 1  Patient demographic details

<table>
<thead>
<tr>
<th>Patient</th>
<th>Sex</th>
<th>Age (years)</th>
<th>FEV$_1$ (l)</th>
<th>FEV$_1$ (%)</th>
<th>ICS</th>
<th>Dose (µg)</th>
<th>Second-line</th>
<th>Severity</th>
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<td>56</td>
<td>FP</td>
<td>500</td>
<td>SM, ML</td>
<td>S</td>
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<td>F</td>
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<td>73</td>
<td>FP</td>
<td>1000</td>
<td>SM</td>
<td>M</td>
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<td>76</td>
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<td>SM, ML</td>
<td>S</td>
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</tbody>
</table>

FEV$_1$, forced expiratory volume in 1 s; ICS, inhaled corticosteroid; FP, fluticasone propionate; BDP, beclomethasone dipropionate; SM, salmeterol; ML, montelukast; ZL, zafirlukast; S, severe; M, moderate.

References

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she still suffered from residual numbness in both hands. Since then, she had required intermittent steroid therapy for asthma exacerbations, although peripheral blood eosinophilia was not a feature. She denied ever taking a leukotriene receptor antagonist.

On this admission, peripheral blood eosinophil count rose to 18.7% of the total white cell count but serum anti-neutrophil cytoplasmic antibodies (ANCA) remained undetected and a high-resolution CT scan of her chest showed no parenchymal lung involvement. Surgical replacement of the aortic valve (Carbomedics ‘Tophat’ mechanical valve) was deemed necessary, along with coronary artery bypass grafting of her left internal mammary artery to her left anterior descending artery lesion. The histology of the removed aortic valve revealed necrotizing granulomatous inflammation with eosinophils infiltrating the aortic valve leaflets, with marked thickening, in part due to fibrosis, but to a larger extent to the presence of the florid inflammatory reaction (Figure 1). She was commenced on prednisolone 1 mg/kg/day in conjunction with oral cyclophosphamide 2 mg/kg (100 mg) daily, as well as osteoporosis prophylaxis with alendronate, and *Pneumocystis carinii* prophylaxis with cotrimoxazole 480 mg once daily, and clinically improved. Unfortunately, she subsequently developed bilateral Achilles tendon rupture, probably secondary to steroid therapy, requiring conservative management with orthopaedic splints.

Churg-Strauss syndrome (CSS) is a multi-system disorder characterized by four or more of the following: asthma, eosinophilia, mono/polyneuropathy, transient pulmonary opacities, paranasal sinusitis, and extravascular accumulation of eosinophils on biopsy. Three distinct phases occur: (i) an atopic prodromal phase, (ii) an eosinophilic phase, and (iii) a vasculitic phase. No laboratory tests are specific for CSS. Eosinophilia can be missed because of rapid, spontaneous fluctuations in eosinophil counts. Raised ANCA directed against myeloperoxidase with a perinuclear staining pattern (p-ANCA) can be seen in 44–66% of patients with CSS, but ANCA positivity is shared by other systemic vasculitides and is therefore non-specific.

Cardiac involvement is noted in approximately 50% of patients with CSS, ranging from asymptomatic electrocardiographic abnormalities to sudden death, and may be the first manifestation of CSS. Granulomatous eosinophilic infiltration of the myocardium and coronary artery vasculitis are the most common lesions noted, leading to severe congestive heart failure and coronary artery disease. Pericardial effusions may also occur leading to haemodynamic compromise or pericardial fibrosis. Complete heart block, as described in this case report, has rarely been described, and may be attributable to infiltration of the myocardium and the His-Purkinje system. Valvular heart disease (usually mitral regurgitation) secondary to CSS has previously been suggested to relate to endomyocardial fibrosis involving the papillary muscles, rather than direct valvular infiltration.

![Figure 1. Aortic valve biopsy containing an eosinophil-rich (upper right) lymphoplasmacytic inflammatory infiltrate.](image)

References


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