LETTERS TO THE EDITOR

Spontaneous splenic rupture in Wegener's vasculitis

Sir: Spontaneous splenic rupture is a rare but well-known complication of several infections, haemoraphhic diseases, portal hypertension, and connective tissue diseases, such as rheumatoid arthritis, systemic lupus erythematosus, and polyarteritis nodosa. As far as we know, spontaneous splenic rupture has never been described in Wegener's vasculitis, though splenic involvement has been well documented in this condition.1,2 A 61 year old white man was admitted in July 1991 because of acute abdominal pain. He had always been healthy and denied any trauma and symptoms, such as malaise, fever, or weight change. Results of routine laboratory tests at admission were normal except for the erythrocyte sedimentation rate (ESR) 42 mm/h, haemoglobin 10 g/l, leucocytes 25·2×10⁹/l (1% eosinophils and 9% band forms), platelets 686×10⁹/l, and creatinine 173 µmol/l. The urine was positive for protein and the sediment contained more than 40 red cells per high power field. A nasal biopsy showed necrotising vasculitis. Antineutrophil cytoplasmic antibodies were strongly positive (titre 1/512) with cytoplasmic staining pattern and specificity for proteinase 3. A diagnosis of Wegener's vasculitis was established and treatment with prednisone and cyclophosphamide was started.

Early in the disease course, the patient received a pneumococcal vaccine. Although splenic vasculitis was notably absent in the spleen of our patient, the spontaneous splenic rupture coincided with the presence of glomerulonephritis. In addition, we found ANCA with autoantibodies against proteinase 3 at that time. The latter are specific markers for systemic vasculitis.1,11 Therefore, a causal relation between spontaneous splenic rupture and the autoimmune disease is highly suggestive.

Our patient's disease did not fulfil the classic definition of Wegener's granulomatosis as the presence of granulomas was not proved by biopsy. The combination of glomerulonephritis, the upper airway and eye disease, and the autoantibodies against proteinase 3, however, warranted the diagnosis of systemic vasculitis. It was not until the second attack that active vasculitis could be established beyond any doubt by a nasal biopsy. According to the classification system recently proposed by Jennette and Falk,12 our patient meets the criteria for Wegener's vasculitis. 

Splenic disease in Wegener's vasculitis is not a rare event at all.1,9 It is therefore the more surprising that a search of published reports on spontaneous splenic rupture did not disclose one single case report with Wegener's syndrome in its running title. However, we detected seven related cases in this survey in which spontaneous splenic rupture occurred in the course of connective tissue disease (table). All of these cases except two had signs of systemic vasculitis.

It is an interesting observation that disease activity decreased after splenectomy as renal function normalised and the erythrocytosis disappeared. Even more remarkable was the recurrence of disease activity after the pneumococcal vaccination. This might have been coincidental. It has been suggested, however, that infection with environmental antigens could lead to an exaggerated leucocyte activation and resultant vascular injury in patients with circulating ANCA.13 It is tempting to speculate that the pneumococcal vaccine might have triggered a similar activation of the vasculitic process.

This case again shows the protein nature of the vasculitides associated with ANCA. Early detection of ANCA in a patient with spontaneous splenic rupture is warranted since this could influence the subsequent follow up and treatment.

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Characteristics of eight patients with arammatous rupture of a normal sized spleen in connective tissue disease.

<table>
<thead>
<tr>
<th>Patient No</th>
<th>Sex/age</th>
<th>Diagnosis*</th>
<th>Splenic weight (g)</th>
<th>Pathologic examination of spleen</th>
<th>Extra-articular manifestation</th>
<th>Reference</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>M/60</td>
<td>Seropositive RA</td>
<td>300</td>
<td>Capillary fibrous thickening with rheumatoid nodule</td>
<td>Pleural effusion, pulmonary opacity, weight loss</td>
<td>2</td>
</tr>
<tr>
<td>2</td>
<td>F/67</td>
<td>Seropositive RA</td>
<td>150</td>
<td>Capillary fibrous thickening</td>
<td>Digital gangrene, vasculitic skin ulcers, myalgia</td>
<td>3</td>
</tr>
<tr>
<td>3</td>
<td>M/65</td>
<td>Seropositive RA</td>
<td>NA†</td>
<td>No significant abnormality</td>
<td>NA</td>
<td>8</td>
</tr>
<tr>
<td>4</td>
<td>F/56</td>
<td>Seropositive RA</td>
<td>NA</td>
<td>No significant abnormality</td>
<td>NA</td>
<td>5</td>
</tr>
<tr>
<td>5</td>
<td>F/31</td>
<td>SLE</td>
<td>NA</td>
<td>No significant abnormality</td>
<td>NA</td>
<td>5</td>
</tr>
<tr>
<td>6</td>
<td>M/59</td>
<td>PAN</td>
<td>NA</td>
<td>Vasculitis with fibrinoid necrosis of medium sized arteries</td>
<td>Muscular pain, weight loss, glomerulonephritis</td>
<td>6</td>
</tr>
<tr>
<td>7</td>
<td>M/28</td>
<td>PAN</td>
<td>NA</td>
<td>Vasculitis with fibrinoid necrosis of medium sized arteries</td>
<td>Muscular pain</td>
<td>7</td>
</tr>
<tr>
<td>8</td>
<td>M/61</td>
<td>WV</td>
<td>100</td>
<td>Capillary neutrophil infiltration</td>
<td>Glomerulonephritis</td>
<td>This study</td>
</tr>
</tbody>
</table>

*R=systemic lupus erythematosus; PAN=polyarteritis nodosa; WV=Wegener's vasculitis.
†NA=not available.