LETTERS TO THE EDITOR

Spontaneous splenic rupture in Wegener's vasculitis

Sir: Spontaneous splenic rupture is a rare but well-documented complication of several infections, haemorrhagic 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 red cell casts. Laparotomy disclosed a tear in the splenic capsule with active bleeding. A splenectomy was performed.

Pathological examination showed a small spleen (100 g). At the site of the rupture the capsule and the subcapsular zone were infiltrated with neutrophils. Some splenic follicles contained small eosinophilic depositions. There were no signs of vasculitis. The postoperative period was complicated by fever, a disseminated macular rash, and mild eosinophilia (maximum eosinophil count on the 10th postoperative day 7·5×10⁹/l). Repeated blood and urine cultures were negative. Chest radiography showed no abnormalities. Serological examination disclosed the presence of antineutrophil cytoplasmatic antibodies (ANCA) with a cytoplasmic staining pattern at a titre of 1/256 and specificity for proteinase 3. By the time this test result became known (the 10th postoperative day) the fever and rash had disappeared, serum creatinine and urine sediment were normal, and there were no signs of vasculitis. He received pneumococcal vaccine before discharge, three weeks after the spontaneous splenic rupture.

The patient was readmitted five weeks later owing to malaise, anorexia, and weight loss of 15 kg. Physical examination disclosed bilateral episceritis, rashes with subcapsular nodules, and bilateral otitis media. His ESR was 97 mm/h, haemoglobin 97 g/l, leucocytes 19·2×10⁹/l (eosinophils 0·39×10⁹/l), platelets 848×10⁹/l, and creatinine 2247 μmol/l. He was referred to our hospital for haemodialysis. Urine analysis showed proteinuria and the sediment contained more than 40 red cells per high power field. A nasal biopsy showed necrotising vasculitis. Antineutrophil cytoplasmatic antibodies were strongly positive (titre 1/512) with cytoplasmatic staining pattern and specificity for proteinase 3. A diagnosis of Wegener's vasculitis was established and treatment with prednisone and cyclophosphamide elicited remission.

Spontaneous rupture of a normal sized spleen is extremely rare. Adler found only 37 reported cases. 3 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. 4 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 5 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, 6 our patient meets the criteria for Wegener's vasculitis.

Splenecctomy in Wegener's vasculitis is not a rare event at all. 7, 8 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 except two had signs of systemic vasculitis.

It is an interesting observation that disease activity decreased after splenectomy as renal function normalised and the erythrocytura 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 one of these microbial antigens could lead to an exaggerated leukocyte activation and resultant vascular injury in patients with circulating ANCA. 9 It is tempting to speculate that the pneumococcal vaccine elicited a similar activation of the vasculitic process.

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

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Characteristics of eight patients with atraumatic 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-arteriolar manifestation</th>
<th>Reference</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>M/60</td>
<td>Seropositive RA</td>
<td>300</td>
<td>Capsular fibrous thickening with rheumatoid nodules</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>Capsular fibrous thickening</td>
<td>Digital gangrene, vasculitic skin ulcers, myelofibrosis</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>4</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>6</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>Capsular neutrophil infiltration</td>
<td>Glomerulonephritis This study</td>
<td>8</td>
</tr>
</tbody>
</table>

*RA=rheumatoid arthritis; SLE=systemic lupus erythematosus; PAN=polyarteritis nodosa; WV=Wegener's vasculitis.
†NA=not available.