

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, haemopietic diseases, portal hypertension,¹ and connective tissue diseases, such as rheumatoid arthritis,²⁻⁴ systemic lupus erythematosus,⁵ and polyarteritis nodosa.^{6,7} 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.^{8,9}

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 101 g/l, leucocytes $25.2 \times 10^9/l$ (1% eosinophils and 9% band forms), platelets $686 \times 10^9/l$, and creatinine 173 $\mu\text{mol/l}$. 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 $0.75 \times 10^9/l$). Repeated blood and urine cultures were

negative. Chest radiography showed no abnormalities. Serological examination disclosed the presence of antineutrophil cytoplasmic 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 episcleritis, rhinitis with mucosal crusts, and bilateral otitis media. His ESR was 97 mm/h, haemoglobin 97 g/l, leucocytes $19.2 \times 10^9/l$ (eosinophils $0.39 \times 10^9/l$), platelets $848 \times 10^9/l$, and creatinine 2247 $\mu\text{mol/l}$. 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 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.

Atraumatic rupture of a normal sized spleen is extremely rare. Adler found only 37 reported cases.¹⁰ 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.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 granulomatosis⁸ 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,¹¹ our patient meets the criteria for Wegener's vasculitis.

Splenic disease in Wegener's vasculitis is not a rare event at all.^{8,9} 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 erythrocyturia 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 infections or other environmental antigens could lead to an exaggerated leucocyte activation and resultant vascular injury in patients with circulating ANCA.¹² 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 since this could influence the subsequent follow up and treatment.

C F M FRANSSEN
J C TER MAATEN
S J HOORNTJE
Department of General Internal Medicine
Catharina Hospital
Michelangelolaan 2
5623EZ Eindhoven
The Netherlands

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

Patient No	Sex/age	Diagnosis*	Splenic weight (g)	Pathologic examination of spleen	Extra-articular manifestation	Reference
1	M/60	Seropositive RA	300	Capsular fibrous thickening with rheumatoid nodule	Pleural effusion, pulmonary opacity, weight loss	2
2	F/67	Seropositive RA	150	Capsular fibrous thickening	Digital gangrene, vasculitic skin ulcers, myopathy	2
3	M/63	Seropositive RA	NA†	No significant abnormality	—	3
4	F/56	Seropositive RA	NA	No significant abnormality	—	4
5	F/31	SLE	NA	No significant abnormality	NA	5
6	M/59	PAN	NA	Vasculitis with fibrinoid necrosis of medium sized arteries	Muscular pain, weight loss, glomerulonephritis	6
7	M/28	PAN	NA	Vasculitis with fibrinoid necrosis of medium sized arteries	Muscular pain	7
8	M/61	WV	100	Capsular neutrophilic infiltration	Glomerulonephritis	This study

*RA=rheumatoid arthritis; SLE=systemic lupus erythematosus; PAN=polyarteritis nodosa; WV=Wegener's vasculitis.

†NA=not available.

- Cochrane J P S. Ruptured spleen. *Br J Hosp Med* 1980; 24: 398-404.
- Haskard D O, Higgins C S, Temple L N, Currey H L. Spontaneous rupture of the spleen in rheumatoid arthritis. *Ann Rheum Dis* 1983; 42: 411-4.
- Pena J M, Garcia-Alegria J, Crespo M, Gijon J, Vazquez J J. Spontaneous rupture of the spleen in RA. *Ann Rheum Dis* 1984; 43: 539.
- Marcus R W. Rupture of the spleen in rheumatoid arthritis. *Ann Rheum Dis* 1985; 44: 142.
- Krauser R E. Spontaneous rupture of the spleen in systemic lupus erythematosus. *JAMA* 1976; 236: 1149.
- Fallingborg J, Laustsen J, Jakobsen J, Winther P, Svanholm H. Atraumatic rupture of the spleen in periarteritis nodosa. *Acta Chir Scand* 1985; 151: 85-7.
- Ford G A, Bradley J R, Appleton D S, Thiru S, Calne R Y. Spontaneous splenic rupture in polyarteritis nodosa. *Postgrad Med J* 1986; 62: 965-6.
- Godman G C, Churg J. Wegener's granulomatosis. Pathology and review of the literature. *Arch Pathol* 1954; 58: 533-53.
- Pinching A J, Lockwood C M, Pussell B A, et al. Wegener's granulomatosis: observations on 18 patients with severe renal disease. *Q J Med* 1983; 52: 435-60.
- Adler G G. Zur problematik der spontanen ruptur einer normalen milz. *Brunns Beitrage fuer Klinische Chirurgie* 1969; 217: 413-20.
- Jennette J C, Falk R J. Diagnostic classification of antineutrophil cytoplasmic autoantibody-associated vasculitides. *Am J Kidney Dis* 1991; 18: 184-7.
- Jennette J C, Falk R J. Antineutrophil cytoplasmic autoantibodies and associated diseases: A review. *Am J Kidney Dis* 1990; 15: 517-29.