

## Autosplenectomy: rare syndrome in autoimmunopathy

J Leipe, A J Hueber, S Kallert, J Rech, H Schulze-Koops

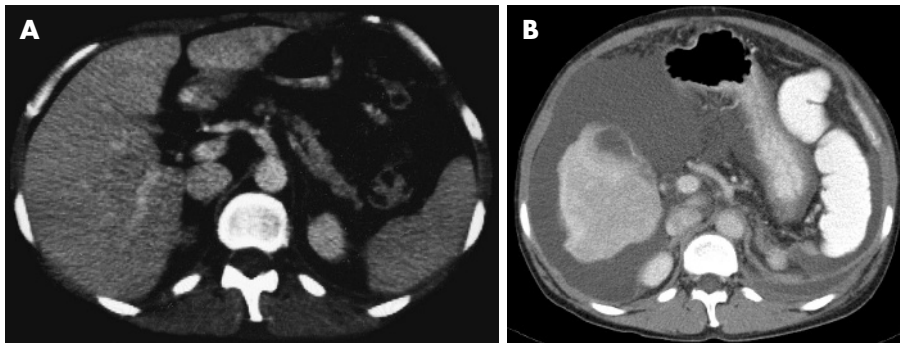
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**A**utosplenectomy has been described in association with systemic lupus erythematosus (SLE).<sup>1</sup> Although in patients with SLE small or atrophic spleens are usually seen, the complete absence of the spleen has been observed rarely. Here, we report a patient with an 18-year history of autoimmunopathy who developed anatomical asplenia during the disease.

A 51-year-old Caucasian woman was first diagnosed in 1988 as having autoimmunopathy of incompletely defined aetiology with antinuclear antibodies (ANA) positivity and immunocomplexes in conjunction with reduced complement levels and serositis. The patient presented with massive pleural and pericardial effusions.

Laboratory studies showed normal blood counts. Antibodies to extractable nuclear antigens or to phospholipids (lupus anticoagulant and anti-cardiolipin antibodies) could not be detected. Although suggestive of having SLE, the patient did not fulfill the classification criteria for the disease at any given time.<sup>2</sup> Notably, the spleen was normal in all respects as confirmed by ultrasound and CT scan, at the time of initial contact. Treatment was initiated with glucocorticoids and plasmapheresis, with good clinical response. Because of unresponsiveness to or unbearable toxicity

**Abbreviations:** AIH, autoimmune hepatitis; ANA, antinuclear antibody; SLE, systemic lupus erythematosus



**Figure 1** Demonstration of the presence and absence of the spleen. (A) Abdominal CT scan performed in 1992 showing a morphologically normal spleen. (B) Fourteen years later, the spleen is diminished; of note is the appearance of ascites and an atrophic nodular liver.

of subsequently started treatment with azathioprine, cyclophosphamide and ciclosporin, a regimen of repeated plasmapheresis during the following years was used to control pleural and pericardial effusions efficiently.

The patient was admitted to the hospital 18 years later because of the development of ascites during the preceding 2 weeks. Laboratory studies revealed an increased C reactive protein serum level (3,2 mg/dl), increased levels of immune complexes (IgG, IgM, IgA), decreased serum complement C3 and C4 and raised liver function tests. Peripheral blood counts were within normal limits. Howell–Jolly bodies were noticed in the blood smear. Indirect immunofluorescence showed a significant titre of circulating ANA (1:100) whereas antimitochondrial antibody, smooth muscle actin antibody and liver, kidney microsomal-1-antibodies were absent. As in the previous tests, extra nuclear antigen and anti-phospholipid antibodies were undetectable.

Ultrasound, CT scan and MRI demonstrated ascites, pleural effusions, pericardial effusion and an atrophic nodular-appearing liver with inhomogeneous contrast enhancement. Surprisingly, in contrast with previous investigations, the spleen was undetectable by ultrasound, CT scan and MRI (fig 1).

Liver biopsy revealed the presence of lymphocytic infiltrations, collapse fibrosis and central portal bridging necrosis, a picture potentially consistent with autoimmune hepatitis (AIH). After excluding other conditions predisposing to hepatitis, the patient fulfilled the criteria for AIH as defined by the International Autoimmune Hepatitis Group.<sup>3</sup>

Treatment for AIH with 1 mg/kg prednisolone with subsequent tapering was initiated, which yielded a partial clinical response.

Whereas functional asplenia occurs in about 5% of patients with SLE, anatomical asplenia has been seen in only a few cases.<sup>4</sup> The mechanism by which autosplenectomy develops in autoimmune diseases is rather unclear, but may relate to vasculitis with increased levels of immune complexes and subsequent silent splenic infarctions.<sup>4</sup> Our patient had increased levels of immune complexes since the first clinical appearance of the autoimmune disease. Although there were no obvious signs of

vasculitis, it is tempting to interpret the decrease in complement and presence of ANA and immune complexes as findings suggestive of unrecognised vasculitis that may have caused microinfarction and, hence, incremental atrophy of the spleen. Alternatively, AIH may have caused the anatomical asplenia. Notably, there is only one report demonstrating functional asplenia in a patient with non-autoimmune, aggressive hepatitis.<sup>5</sup>

In summary, we present the first case of a patient with anatomical autosplenectomy in the setting of an autoimmune-pathology different from SLE.

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