since no official scoring system such as the Behçet's Disease Current Activity Form (BDCAF) was available at the start of anti-TNF α treatment in our centre.

It is unknown how long anti-TNF α treatment must be given, but anti-TNF α treatment in patients with rheumatic arthritis is continued for >2 years and continued until there is a settled response.¹⁰ In our patients, infliximab was discontinued after complete response of >3 months or acceptable improvement of (eye) symptoms. In five of the six patients, relapses after infliximab did not necessitate immediate restart of anti-TNF α treatment. In this period (mean duration 562, range 136–1093 days), immunosuppressive therapy could be adjusted until the symptoms required a restart of anti-TNF α treatment. Adalimumab was considered to be equal potential, but more convenient, and was added in cases of severe relapse with patients' informed consent. In addition, formation of auto-antibodies to infliximab when restarted was considered. All patients responded and most of them showed dramatic and quick improvement. Subsequently, immunosuppressive therapy could again be tapered (table 1).

Patient 6 had a severe BD-associated colitis and was periodically treated with infliximab and other immunosuppressive agents for nearly 3 years. Despite intensified immunosuppressive therapy, the colitis worsened and became refractory and life threatening. Subsequently, a high dose of adalimumab 40 mg/week was started subcutaneously, yielding a complete response of >1 year. Adalimumab was briefly combined with 30 mg of prednisone, which was tapered rapidly to prevent central retinal serosa ablation that developed in a previous period in which steroids had been used. Later, mesalazine and rectal budesonide were also given. Apart from some minor flares, the patient remained stable for nearly 2 years. Until now, all patients are receiving adalimumab, except patient 5 who discontinued 4 months after complete remission was achieved (table 1). In general, few side effects were observed. Three patients (1, 3 and 6) developed lichenoid-like lesions that were treated with local steroids by a dermatologist.

This report on patients with treatment refractory BD indicates that adalimumab treatment is promising and can be prescribed safely for a prolonged period. To our knowledge, this is the first case series in which patients with BD with systemic

disease treated with adalimumab are presented. More studies on this subject are warranted.

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Autosplenectomy: rare syndrome in autoimmuneopathy

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

Autosplenectomy has been described in association with systemic lupus erythematosus (SLE).¹ 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 autoimmuneopathy who developed anatomical asplenia during the disease.

A 51-year-old Caucasian woman was first diagnosed in 1988 as having autoimmuneopathy 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.² 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.³

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.⁴ 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.⁴ 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.⁵

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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