SIR, A 27-yr-old woman was referred for evaluation of mild anaemia, leucocytosis and thrombocytosis. History revealed a diagnosis of Rothmann–Makai syndrome (lipogranulomatosis subcutanea), a rare variant of Weber–Christian disease, at the age of 13 yr. At that time the patient had presented with a minor fracture of the left ankle, and when the plaster was removed she was found to have irregular red discoloration on the outer part of the ankle. This subsequently spread to involve both shins. A biopsy specimen was obtained from one of the erythematous nodules. In the histopathological sections there was a moderate lymphocytic infiltration in the lobules of the panniculus and fibrocytes invading the lobules from the fibrous septae. Work-up at that time also included antinuclear antibodies (ANA), rheumatoid factor (RF), C-reactive protein (CRP), anti-double-stranded DNA, antiautoantibodies, anti-Sel-70 and cryoglobulins, which were all negative. A complete blood count was normal. With these findings and lack of any systemic manifestations, a diagnosis of Rothmann–Makai syndrome was established. The patient received an 8-month course of steroids. The erythema resolved and the patient was left with two slightly atrophic areas in both calves. No relapse has occurred. History also revealed Hashimoto’s thyroiditis diagnosed at the age of 14 yr and the onset of alopecia areata at the age of 20 yr.

The patient has developed progressive increase in her leucocyte count [(8–12) × 10⁹/l] with a normal distribution and platelet count [(450–600) × 10⁹/l] over the past 8 yr. When seen in our clinic she was feeling well. Except for obesity, the physical examination was unremarkable. Haemoglobin was 14 g/dl with normal red cell indices, the platelet count was 668 × 10⁹/l and the leucocyte count was 16.2 × 10⁹/l with a normal distribution. A peripheral blood smear revealed anisopoikilocytosis and numerous Howell–Jolly bodies, indicating functional hyposplenism. Computed tomography revealed a small spleen and no flow signals could be derived by colour Doppler measurements from the spleen.

Rothmann–Makai syndrome is a rare form of Weber–Christian disease, usually seen in adolescent and middle-aged women [1]. Subcutaneous nodules develop during the course of the disease whereas systemic manifestations are absent. Multiple painless nodules appear throughout the muscle and subcutaneous tissues. The primary pathological process is lobular panniculitis, which passes through the same three stages as Weber–Christian disease. In the first stage, there is acute inflammation of the fat lobules with fat cell degeneration accompanied by an infiltrate of neutrophils, lymphocytes and macrophages. The second stage is characterized by many foamy histiocytes, and the infiltrate is discretely localized to the fat lobules. Finally, the foam cells are replaced by fibroblasts. The first two histological stages correspond to clinically apparent induration, while in the third stage atrophy of the skin may develop. There are no diagnostic laboratory findings. The diagnosis is based upon clinical and pathological findings. Corticosteroid treatment does not prevent new lesions or seem to alter the course of the self-limited disease.

Infiltrative, inflammatory or thromboembolic processes in the parenchyma of the spleen can cause a functional loss of the organ. This phenomenon is called functional asplenia and occurs as a complication, especially in sickle cell disease, lupus erythematosus (SLE) and after bone marrow transplantation [2–4]. Functional asplenia has complicated the course of autoimmune diseases other than SLE, such as candidiasis endocrinopathy syndrome and alopecia areata [5].

This is the first report of functional asplenia occurring in the setting of Rothmann–Makai syndrome. The aetiology and pathogenesis of Rothmann–Makai syndrome is still unknown. Christian–Weber disease often occurs in patients with autoimmune disorders [6–8], as is the case with our patient, reinforcing the view that Rothmann–Makai syndrome, a variant of Christian–Weber, may also have an autoimmune basis.

The authors have declared no conflict of interest.

A. PSYRRI and T. ECONOMOPOULOS

2nd Department of Internal Medicine, Attikon University Hospital, Athens, Greece

Accepted 2 September 2005

Correspondence to: A. Psyrrir, Attikon Hospital, Rimini 1, Haidari, Athens, Greece. E-mail: Diamando.Psyrrir@yale.edu

Advance Access publication 18 October 2005

Infliximab infusions for persistent back pain in two patients with Schmorl’s nodes

SIR, Schmorl’s nodes (SNs) are herniations of the nucleus pulposus (NP) material through the vertebral endplates into the trabecular bone. SNs are the most common non-vertebral disc herniations, occurring in about 10% of adults [1]. SNs may cause back pain either by producing local symptoms or by triggering chronic low back pain and radiculopathy [2]. A recent study of 129 patients undergoing lumbar spine surgery reported 15% of patients had uncomplicated SNs [3]. However, local symptoms due to SNs are infrequently reported. We describe two cases of persistent back pain due to SNs and the role of infliximab in their management.

The first patient was a 36-yr-old woman who had undergone a total hip replacement for a right hip fracture at the age of 32 yr. Three months after the operation she presented with acute back pain radiating to the right lower leg. A systemic examination was normal. Magnetic resonance imaging (MRI) of the lumbar spine revealed SNs at L2-L3, L3-L4, and L4-L5. Conservative management including analgesics, physiotherapy, and non-steroidal anti-inflammatory drugs was ineffective. Infliximab was administered intravenously at 3 mg/kg at weeks 0, 2, 6, 10, and 14, and at 6 mg/kg at 20 weeks. MRI of the lumbar spine performed at week 20 showed resolution of the SNs at L2-L3, L3-L4, and L4-L5, and the patient was pain-free at week 26.

The second patient was a 38-yr-old woman with a 10-yr history of idiopathic low back pain. MRI of the lumbar spine revealed SNs at L1-L2 and L2-L3. Over the ensuing 5 yr, her symptoms remained stable with radicular symptoms predominantly in the right leg. At 42 yr she presented with severe back pain radiating to the right lower leg. A systemic examination was normal. MRI of the lumbar spine revealed SNs at L1-L2 and L2-L3, with evidence of adjacent disc herniation at L2-L3. Conservative management including analgesics, physiotherapy, and non-steroidal anti-inflammatory drugs was ineffective. Infliximab was administered intravenously at 3 mg/kg at weeks 0, 2, 6, 10, and 14, and at 6 mg/kg at 20 weeks. MRI of the lumbar spine performed at week 20 showed resolution of the SNs at L1-L2 and L2-L3, and the patient was pain-free at week 26.

Infliximab is a chimeric monoclonal antibody against tumour necrosis factor alpha (TNF-α), a pro-inflammatory cytokine. TNF-α is involved in the pathogenesis of chronic inflammatory diseases including ankylosing spondylitis (AS), rheumatoid arthritis (RA), and Crohn’s disease [4]. Infliximab has been shown to be effective in the treatment of AS [5], RA [6], and Crohn’s disease [7]. It has also been shown to improve outcomes in patients with lumbar spinal stenosis [8], and decreases the number of lumbar disc herniations in patients with rheumatoid arthritis [9]. In a recent study of 30 patients with AS, infliximab reduced the number of SNs and was associated with an improvement in clinical symptoms [10]. Further studies are needed to investigate the role of infliximab in the management of SNs.

The authors have declared no conflict of interest.

Haidari, Athens, Greece. E-mail: Diamando.Psyrri@yale.edu

Rheumatology 2005;44:1588 Letters to the Editor
doi:10.1093/rheumatology/kei144
Advance Access publication 18 October 2005

Functional asplenia in a patient with Rothmann–Makai syndrome: a pathogenetic relationship?

Letters to the Editor