


Rheumatology 2005;44:1588
doi:10.1093/rheumatology/kei44
Advance Access publication 18 October 2005

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

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 septa. Work-up at that time also included antinuclear antibodies (ANA), rheumatoid factor (RF), C-reactive protein (CRP), anti-double-stranded DNA, anticardiolipin, 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] × 10^9/l) with a normal distribution and platelet count ([450–600] × 10^9/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 × 10^9/l and the leucocyte count was 16.2 × 10^9/l with a normal distribution. A peripheral blood smear revealed anisopoikilocytosis and numerous Howell–Jolly bodies, indicating functional hypoplasmen. 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. 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. Psyrri, Attikon Hospital, Rimini 1, Haidari, Athens, Greece. E-mail: Diamando.Psyrri@yale.edu


Rheumatology 2005;44:1588–1590
doi:10.1093/rheumatology/kei155
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
Two women aged 52 and 49 yr with SNs received infliximab because of severe, long-standing back pain (duration of pain 24 and 18 months, respectively) without radicular symptoms. There was a partial response of pain to treatment with NSAIDs and corticosteroids in both patients, but severe spinal pain recurred after discontinuation of the above agents. SNs of the L3 and L5 vertebrae in the first patient and of the T12, L1 and L2 vertebrae in the second patient were depicted by MRI. There were MRI findings of active SN (oedema of adjacent bone marrow and contrast enhancement of SN) in the L5 vertebra for the first patient and in the L2 vertebra for the second patient. After obtaining ethics committee approval and written consent from the patients, the patients scheduled to receive infliximab infusions at a dose of 3 mg/kg at weeks 0, 2, 6 and 14. Response was evaluated using a 0–100 mm visual analogue scale (VAS) for back pain. Furthermore, the first patient was evaluated for radiological progression of the lesions by sequential MRI examination.

Prior to infliximab treatment, the VAS score for back pain for the two patients was 90 and 85, respectively. The first patient responded promptly to infliximab with a decrease in VAS for back pain to 7, and back pain completely resolved after the second infusion. Due to a severe allergic reaction just after the second infusion, she discontinued the infliximab regimen. Six months later, MRI examination revealed disappearance of contrast enhancement of SN of the L5 vertebra and of bone marrow oedema (Fig. 1). She has remained asymptomatic for 30 months.

The second patient completed four infliximab infusions. She also responded promptly to treatment. VAS for back pain ranged from 15 to 20 at weeks 2, 6 and 14. She was missed 5 months after the last infliximab infusion, and had VAS for back pain ~20 over the follow-up period.

MRI is useful in differentiating between symptomatic and asymptomatic SNs. In an analysis of MRI findings of SNs [6], the vertebral body marrow surrounding the SN was seen as low signal intensity on T1-weighted images and as high signal intensity on T2-weighted images in all symptomatic cases. These MRI findings were not seen in asymptomatic individuals. Histological examination of two symptomatic cases demonstrated bone marrow oedema and inflammatory cell infiltration in the affected vertebral body. In a small number of cases of acute SNs [7], there had been improvement in back pain by the time the follow-up MRI (between 6 weeks and 7 months after the initial MRI) showed reduction in bone marrow oedema. Furthermore, there was resolution of oedema with fatty marrow change in two cases with longer follow-up at MRI (10 and 18 months after the initial MRI, respectively). In addition to surrounding bone marrow oedema, MRI can reveal vascularized tissue in SNs after the intravenous administration of contrast material [8]. Contrast-enhancing SNs are larger and more frequently associated with bone marrow oedema in patients with back pain than in asymptomatic patients.

Our patients had not only severe back pain, which was induced by active SNs (L5 vertebra in the first patient and L2 vertebra in the second patient), but also long-standing symptoms, which could be explained by previous active SNs that were in the healing stage at the time of MRI examination (L3 vertebra in the first patient and T12 and L1 vertebra in the second patient). A single infliximab infusion for HID-induced sciatica produced not only a sustained but also an immediate improvement (within hours) [4]. The first infliximab infusion led to prompt and significant improvement in back pain in our patients within the first 24 h. There was a complete response to the second infliximab infusion in the first patient, but the second patient did not have a complete improvement of back pain with the four-dose infliximab regimen. A pilot study is required to validate the results of our two case reports.


