# Letters to the Editors

## Relapsing isolated lupus peritonitis

Sirs,

A 42-year-old Caucasian woman with no personal or family history of gastrointestinal disorders, allergy or autoimmunity presented with acute peritonitis.

Work-up disclosed massive ascites and oedematous thickening of the small intestine, a high titer of antinuclear, anti-SSA and anti-double-stranded DNA antibodies, and low C3 and C4. A paracentesis of 3500 ml of yellow and clear fluid showed an exudate with a serum ascitic albumin gradient <1.1, high protein level, mesothelial cells, lymphocytes and macrophages; search for malignant cells and culture and stains for bacteria, mycobacteria, and fungi were negative. We diagnosed lupus peritonitis after malignancies, eosinophilic gastroenteritis, protein-losing enteropathy and other causes of peritonitis and ascites were appropriately ruled out.

The patient achieved full recovery after two weeks of prednisone (1 mg/kg daily). Azathioprine (100 mg daily) was added and prednisone gradually tapered and stopped. Peritonitis and ascites relapsed three months later with recovery again achieved after prednisone therapy. Remission was maintained with an increased dose of azathioprine (150 mg) until 28 months after the first presentation when a new flare-up of peritonitis and ascites was abated with steroids. The patient displayed no other SLE features over more than two years of close observation. She was thereafter lost to follow-up. This patient fulfilled the Systemic Lupus Collaborating Clinics criteria for systemic lupus erythematosus (SLE) (1). Up to 70% of SLE cases have peritoneal involvement. However, only a a minority suffer full-blown peritonitis and ascites (2, 3). No SLE features were recognised in this case other than relapsing peritonitis and massive ascites over more than two years. This supports the view that SLE could remain confined to the peritoneum for several years after the first onset. The mechanisms underlying such an unusual natural history are unknown. Lupus peritonitis could represent the initial and predominant feature of SLE with an acute or chronic course (4, 5). Acute peritonitis is a sudden development of painful serositis with small amounts of intraperitoneal fluid that is commonly associated with other SLE features such as fever, arthritis and rashes. Chronic peritonitis is notably painless with massive ascites that develops over several months, but has no other SLE features, and frequent steroid-resistant relapses. Lupus peritonitis has a broad differential including both SLE-related and non-SLE-related entities (Table I). Patients with milder disease are especially challenging because of their subtle and confounding clinical features. The laparoscopic finding of a hyperaemic, thickened, nodular or adhesive peritoneum, histology showing peritoneal infiltration with mononuclear cells, vasculitis and LE cells, a serum ascitic albumin gradient <1.1 and increased leukocyte count and protein content in the peritoneal fluid are helpful diagnostic tools (2, 3, 6). We did not find LE cells in the peritoneal fluid from our patient. Most patients quickly respond to systemic or intraperitoneal steroids; severe and refractory or relapsing cases need long-term steroid courses and azathioprine or other immunosuppressants typically with clinically significant efficacy (1, 2, 7). The role of hydroxychloroquine and non-steroid anti-inflammatory drugs in the management of lupus peritonitis is unclear. Ischaemic and infarcted bowel caused by the associated mesenteric SLE vasculitis may progress to bowel-wall haemorrhage, perforation and death. However, their detection can be masked or delayed in steroid-treated patients (2, 3). Pneumatosis cystoides intestinalis is a late complication (8). Many problems should be elucidated in lupus peritonitis. It is unknown whether isolated lupus peritonitis should be categorised into a pathophysiologically distinct SLE subset with a better outcome than peritonitis associated with other features of the disease. The question whether unidentified SLE is an underestimated cause of ascites is also unanswered.

**Table I.** Causes of ascites in SLE patients.

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| SLE-related | Lupus peritonitis (acute and chronic) |
| | Nephrotic syndrome (Lupus glomerulonephritis, renal vein thrombosis) |
| | Mesenteric vasculitis with bowel infarction |
| | Lupus enteritis with protein-losing enteropathy |
| | Constrictive pericarditis |
| | Libmann-Sachs endocarditis causing congestive heart failure |
| | Budd-Chiari syndrome |
| Non SLE-related | Liver cirrhosis |
| | Liver cancer |
| | Pancreatitis |
| | Peritoneal carcinomatosis |
| | Peritoneal mesothelioma |
| | Peritoneal tuberculosis |
| | Atherosclerotic and cardioembolic bowel infarction |
| | Non SLE-related constrictive pericarditis |
| | Non SLE-related congestive heart failure |
| | Non SLE-related Budd-Chiari syndrome |

G. FAMULARO[1], *MD, PhD*
G. MINISOLA[2], *MD*
L. GASBARRONE[1], *MD*

*[1]Department of Internal Medicine and [2]Department of Rheumatology, San Camillo Hospital, Rome, Italy.*

*Address correspondence to:*
*Dr Giuseppe Famularo, Department of Internal Medicine, San Camillo Hospital, Circonvallazione Gianicolense, 00152 Rome, Italy.*
*E-mail: gfamularo@scamilloforlanini.rm.it*

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