CASE REPORTS

Chronic lupus peritonitis with ascites

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Abstract

A 28 year old woman with systemic lupus erythematosus who developed chronic lupus peritonitis and ascites is described. Lupus peritonitis appeared with abdominal fullness, postprandial abdominal discomfort, and painless ascites. Four months later the patient developed vertigo, headaches, visual disturbances, serositis, and glomerulonephritis. Lupus peritonitis and the other disease manifestations responded to treatment with intravenous pulse methylprednisolone (four 1 g/m² injections at one week intervals), oral azathioprine (200 mg daily), and diuretics.

Serositis is a feature of systemic lupus erythematosus (SLE).¹ Peritoneal involvement has been found in two thirds of patients with SLE who undergo necropsy,² though it is detected clinically only in 4%³ to 16%⁴ of them. Ascites may present acutely or chronically, with or without pain, and it may be due to lupus peritonitis,⁵⁻¹⁵ visceral infarction,¹⁶ congestive heart failure,¹⁶ constrictive pericarditis,¹⁷ nephrotic syndrome,⁴ Budd-Chiari syndrome,¹⁸ protein losing enteropathy,¹⁹ and acute pancreatitis.²⁰ Chronic ascites in patients with SLE is extremely rare and as far as we know only 11 cases have been published.¹⁵ We report the case of a patient with inactive SLE who developed unexplained ascites several months before flaring of her disease with polyserositis and glomerulonephritis.

Case report

In 1986 a 25 year old white woman was diagnosed as having SLE when she presented with fever, pericarditis, anaemia, leucopenia, and positive serology. She was treated successfully with steroids and hydroxychloroquine. Prednisone was tapered to less than 10 mg daily and the patient had no clinical or laboratory findings of active disease until September 1988 when she noticed morning stiffness and transient Raynaud's phenomenon. One week later she felt abdominal fullness, heartburn, and postprandial abdominal discomfort. Upper gastrointestinal series showed gastritis and treatment with antacids was started. In October 1988 she had pain in the left upper part of the abdomen, and physical examination showed, for the first time, shifting dullness. Prednisone was increased to 35 mg daily. Between October 1988 and January 1989 she continued to have the same abdominal symptoms and new ones, which included tachycardia, recurrent trouble-

some vertigo, headaches, and decreased visual acuity. Physical examination showed no pericardial rub, eye, ear, nose, or throat findings. An echocardiogram was normal. Computed tomography of the brain was normal. Despite treatment with higher doses of prednisone her symptoms worsened, the amount of ascitic fluid increased, and in December 1988 a small amount of pericardial effusion was detected in a second echocardiogram. On admission the following month she had fatigue, vertigo, tachycardia, headaches, and decreased visual acuity. A complete physical examination showed normal vital signs, pericardial and pleural rub, tense ascites, and woollen exudates in both fundi. There was no lymphadenopathy, arthritis, Raynaud's phenomenon, rash, oedema, hepatosplenomegaly, or neurological findings; pelvic examination was normal.

Laboratory investigation showed: erythrocyte sedimentation rate (Westergren) 50 mm/h, packed cell volume 0.38, haemoglobin 125 g/l, white blood cell count $9.4 \times 10^9/l$ (polymorphs 82%, lymphocytes 10%, monocytes 7%, and basophils 1%). Platelet count and prothrombin time were normal. Blood urea was 9.8 mmol/l, serum creatinine 88.4 $\mu\text{mol/l}$. Antinuclear antibodies were positive at a titre of 1/320 (homogeneous pattern) and anti-dsDNA antibodies were 85 U/ml (normal <30). Antibodies to extractable nuclear antigen, ribonucleoprotein, and Ro (SS-A) antigen were positive. C reactive protein was 2+, latex test was positive, lactate dehydrogenase was 469 U/l (normal <225), haemolytic complement CH₅₀, and serum C3, C4 concentrations were below normal, total protein was 59.4 g/l, (albumin 30 g/l and γ -globulin 15.6 g/l). Cryoglobulins were positive (IgG and IgM κ). Urine analysis showed trace protein, five to eight red blood cells per high power field. Serum cholesterol was 7.4 mmol/l. Urine cultures were negative. Electrolytes, fibrinogen, liver and thyroid function tests, serum uric acid, and antibodies to cardiolipin were all normal or negative. The Mantoux test was negative.

Computed tomography of the liver showed no abnormalities. Echocardiogram showed a small amount of pericardial fluid and the chest radiograph a small amount of pleural effusion at the right base. Computed tomography and ultrasonic examination showed ascitic fluid but no lymphadenopathy or other abnormal findings. Electrocardiogram was normal. Computed tomography of the brain was normal. Central venous pressure was 8 cmH₂O.

Ascitic fluid was aspirated and contained 0.15×10^9 cells/l (90% lymphocytes) and 35 g/l

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protein but no malignant cells. Cultures for bacteria and mycobacteria gave no growth.

The patient was treated with 100 mg of prednisone and diuretics. Ten days later she developed pericardial rub, S3 gallop, while the amount of ascitic fluid increased. Urine analysis showed cellular casts and 1 g protein was excreted daily. Bolus cyclophosphamide treatment was offered, which the patient rejected, and treatment was started with azathioprine 200 mg daily. A week later she developed pitting oedema of the feet, while the ascites enlarged; packed cell volume dropped to 0.3 and 4.5 g protein was excreted daily. At that point she was given four intravenous pulse methylprednisolone (1 g/m²) injections at one week intervals. Two months after her admission the ascitic fluid was remarkably reduced and there were no pericardial or pleural effusions. A renal biopsy showed segmental hyperplastic glomerulonephritis (WHO class IIIa) with activity index of 7 and chronicity index of 3.

One month later (April 1989) both visual disturbances and vertigo subsided, and there was no detectable ascitic fluid. A 24 hour urine specimen contained 1.08 g protein. Eight months later she remains free from symptoms while continuing to take prednisone (25 mg daily) and azathioprine (150 mg/daily).

Discussion

Our patient had chronic lupus ascites which appeared several months before flaring of her disease and subsided months later with intravenous pulse methylprednisolone and oral azathioprine treatment. Only 11 cases of chronic lupus ascites have been reported so far, indicating that it is a rare manifestation of the disease.⁶⁻¹⁵ Other causes of ascites^{15 18 19 21} were excluded, and it was thus attributed to her disease. Nephrotic syndrome, which occurs in 10% of patients with SLE,⁴ was ruled out because ascites developed months before the kidney disease.

The patient had no signs of congestive heart failure or constrictive pericarditis. The favourable response to pulse methylprednisolone and azathioprine treatment suggests a diagnosis of peritonitis or vasculitis. Vertigo, headaches, and visual disturbances, which were dominant presenting symptoms in our patient, might be explained by a generalised vasculitis. These symptoms subsided several weeks after the activity of the disease was controlled. Similar findings have been associated with cerebral vasculopathy.²² Computed tomography of the brain in our case was normal. In such cases a

cerebral angiogram, which was not performed in our patient, might have been indicated, particularly if these symptoms had persisted and neurological findings had been present.^{22 23}

The pathogenesis of chronic lupus peritonitis with ascites is not entirely clear.¹⁵ Possibly, deposition of immune complexes on the peritoneum may explain the chronic lupus peritonitis.¹⁵ Treatment requires diuretics, high dose of steroids, and in non-responding cases immunosuppressive drugs.⁶⁻¹⁰

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