Association of paralytic ileus, interstitial cystitis and hydronephrosis with systemic lupus erythematosus

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We report the case of a 25-year-old woman who presented with paralytic ileus, interstitial cystitis and bilateral hydronephrosis with systemic lupus erythematosus being the probable underlying aetiology. This uncommon association—the first case described in our locality—may represent a specific subgroup found in lupus patients.

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Case report

A 25-year-old woman with previously good health presented with repeated episodes of abdominal pain and vomiting of more than one month's duration. Physical examination revealed abdominal distension and generalised tenderness with decreased bowel sound. Abdominal X-ray showed a dilated small bowel with multiple fluid levels. An emergency laparotomy was performed. At operation, ascites and an inflamed small bowel were found, but there was no definite obstruction.

An operative ileal biopsy showed marked submucosal oedema and acute inflammatory cell infiltration but no evidence of tuberculosis, Crohn's disease or malignancy. Immunofluorescent staining of the biopsy material revealed no specific abnormalities; neither did colonoscopy. Rectal biopsy showed no evidence of vasculitis or amyloidosis. On echocardiography, a small pericardial effusion was detected. Laboratory investigations showed an increased erythrocyte sedimentation normal range, rate (168 mm/h, normal range, 0-30), positive anti-nuclear antibody (1:360 dilution, normal range, <1:40), positive anti-DNA (43%, normal range, <40%) as well as depressed C3 levels (0.42g/L, normal range, 0.6-1.2 g/L). Serial urine, blood, and stool cultures revealed no pathogens. Although serum albumin, urea, and creatinine were initially normal, hypoalbuminaemia occurred two weeks after admission and persisted for the period of hospitalisation. Urine for porphobilinogen was negative. She was given total parenteral nutrition because of vomiting and poor oral intake.

One month after admission, she developed increased frequency of micturition and urinary incontinence. An intravenous pyelogram showed bilateral hydronephrosis and hydroureters down to the level of the ureterovesical junction. An abdominal ultrasound demonstrated a contracted urinary bladder and bilateral hydronephrosis. Cystoscopy showed mucosal oedema and a shrunken bladder. A urinary bladder biopsy demonstrated chronic inflammatory cells and stromal oedema suggestive of chronic interstitial cystitis. Immunofluorescent studies of the specimen showed no significant findings. The 24-hour urine for protein was 0.31 g. There were no dysmorphic red blood cells or white cells on urinalysis. The clinical diagnosis was most probably systemic lupus erythematosus (SLE).

Two months following admission, the patient was given high-dose prednisone (80 mg daily). Her urinary symptoms improved but the paralytic ileus persisted. She was then also given a one-pulse intravenous injection of cyclophosphamide (1 g) after receiving high dose prednisone for two weeks. Four days after receiving the cyclophosphamide, she started to pass watery stools and the bowel sound reappeared. The abdominal distension was diminished.

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Three days following her initial improvement, she developed *Escherichia coli* septicaemia and disseminated intravascular coagulation despite being given multiple antibiotics. She was not neutropenic. Her clinical course was complicated by gastrointestinal bleeding and intracerebral haemorrhage. She died after a period of active resuscitation.

An autopsy was performed. The mucosa of the oesophagus and stomach was markedly congested and haemorrhagic. The small bowel was normal. Three warty excrescences were noted behind the posterior cusp of the mitral valve. Microscopic examination revealed sub-endothelial accumulation of fibrinoid material and subintimal fibrosis with mucoid interstitial changes suggestive of Libman-Sacks endocarditis. Ascites and pericardial effusion were present and the mediastinal and abdominal lymph nodes were enlarged. On section, the lymph nodes were congested, with a number of megakaryocytes in the sinusoids. Some of the sinusoids showed changes of early vascular transformation but there were no other abnormalities. Histological evidence of lupus nephritis was absent in light microscopy and immunofluorescence studies.

Discussion

The diagnosis of SLE in our patient was made clinically because this was a multisystemic illness, and positive anti-nuclear antibody, positive anti-double-stranded DNA, and low complement levels were present. Subsequent autopsy also revealed Libman-Sacks endocarditis.

Patients with small bowel manifestations of SLE may present with vomiting, diarrhoea, protein-losing enteropathy, malabsorption, or paralytic ileus. There are four reported cases in the literature of SLE patients who presented with both paralytic ileus and chronic interstitial cystitis.[1-4] Two of the cases also had cutaneous vasculitis; glomerulonephritis was present in three cases; and one of the cases had central nervous system symptoms. Our patient had paralytic ileus involving the small intestine for more than three months. Following immunosuppression with high-dose prednisone and cyclophosphamide, bowel transit was recovered. Her apparent response could either have been due to the cyclophosphamide or the prior use of high-dose prednisone, which later took effect. Lupus enteropathy is thought to be due to intestinal venulitis.[5] However, in this case, the small intestinal biopsy showed only non-specific inflammation without evidence of vasculitis or villous atrophy. A normal immunofluorescence result, as occurred with this patient, can occur even in proven cases of lupus involvement of the bowel.[4]

Chronic interstitial cystitis associated with SLE was first reported by Shipton in 1965.[6] Boye et al[7] suggested an immune complex-mediated pathogenesis by demonstrating immunoglobulin and complement deposition in the bladder vessels. Only 16 cases of lupus cystitis have been reported as a major manifestation of SLE. Thirteen of the 16 (81%) reported patients with lupus cystitis had a variety of gastrointestinal manifestations.[1-4,8-10] However, there is no known causal relationship between enteropathy and cystitis in SLE. Orth et al reported six cases of lupus cystitis; five of which had either vesicoureteric reflux or bilateral hydroureteronephrosis.[1] Corticosteroid may reverse the vesical wall inflammation. Recognition of bladder involvement is important as it may be a partially reversible cause of renal failure in a patient with SLE.

In summary, we have presented the first local report of a case of probable SLE whose main manifestations were paralytic ileus and chronic interstitial cystitis. This subgroup of patients usually respond to prednisone therapy. For lupus enteropathy not improved by prednisone, cyclophosphamide has been reported to be of use.[4] We hope that this report will draw our colleagues' attention to the protean manifestations of SLE, some of which may seem rare and unrelated, but which may actually be features of a well-defined syndrome requiring intensive therapy.

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