SYSTEMIC LUPUS ERYTHEMATOSUS PRESENTING
AS INTERMITTENT INTESTINAL OBSTRUCTION

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Summary

Two patients are reported who presented to surgeons with intermittent intestinal obstruction of
the small intestine. The intestine was inflamed and oedematous and was resected. Subsequently
both patients developed systemic lupus erythematosus. One patient continued to have intestinal
obstruction until treated with steroids, to which both patients subsequently responded.

Histology showed a diffuse infiltration with eosinophils.

The relevant literature on gastro-intestinal lupus erythematosus is reviewed.

Systemic lupus erythematosus (SLE) is a disease with numerous manifestations, many of
which may give rise to the presenting symptoms. The frequency of any group of symptoms in a series often reflects the interests
of the author. Thus gastro-intestinal manifestations are said to be 'quite common' by Dubois(1) and to occur in 10 per cent of the
patients in neighbouring Singapore(2). Estes and Christian(3) note peritoneal serositis in 16 per cent. Enteritis is an unusual manifestation
of SLE and very rarely the presenting feature.

Two patients are described here who pre-
sented to surgeons with symptoms suggestive of
intestinal obstruction, who were found to have enteritis of the jejunum, and who subsequently
manifested other more typical features of SLE.
Both patients fulfil the proposed criteria for the
the classification of SLE(4).

Case 1

A 43 year old Chinese man presented with an
eight month history of recurrent episodes of
central abdominal pain, relieved by vomiting
and Hyoscine butylbromide (Buscopan). Each
attack was associated with the passage of loose
stools twice a day. He had two or three attacks
per month.

A barium meal and follow through showed a
narrow segment of terminal ileum. HE was
treated conservatively but subsequent attacks
of pain became more severe and exploratory
laparotomy was performed.

At laparotomy about 100cm of jejunum
were found to be thickened and oedematous
and showed a red discoloration. This abnormal
segment, which was sharply demarcated from
the adjoining normal gut, was excised and
continuity restored by end to end anastomosis.

Following laparotomy, there was complete
relief from pain, diarrhoea and vomiting. Four
months later he developed pain and swelling of
the small joints of the hands and aches in the
shoulders, elbows and neck. He had a weight
loss of about 18.3 kg over the previous seven
months. There was no past or family history,
and no history of Raynaud's phenomenon,
facial rash or mouth ulceration. Examination
found a thin man, BP 120/70 with no other
physical signs.

INVESTIGATIONS

Haematology

Hb 10.0 g/dl, white cell count 5.3 x 10^9/l
(P 66%, E 3%, M 1%). The ESR was 107 mm in
one hour. LE cells were positive on two
occasions, although the antinuclear factor was
negative initially. Rheumatoid factor was
negative.

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Biochemistry
Blood Urea 12.5 mmol/l, electrolytes were normal, Serum alanine aminotransferase 110 IU/l, serum globulin 38 g/l and serum bilirubin was 3.4 mmol/l.

Radiology
Postoperative barium meal and enema were normal, but the small intestinal follow through films showed that a short segment of the terminal ileum was narrowed although the mucosal pattern appeared normal, and the iliocaecal valve was patent.

Pathology
On microscopic examination there was severe oedema of all layers of the gut wall, maximal in the submucosa. A diffuse infiltration by eosinophilic polymorphonuclear leucocytes, present in all layers, was most prominent in the muscle coat. There were no granulomata or giant cells. Blood vessels were distended with blood but there was no evidence of vasculitis, necrosis or thrombosis.

An initial dose of Prednisolone 30mg/day gave good relief of his symptoms, and this was reduced to 15mg/day, following which he was given Cyclophosphamide 400mg I.V. Subsequently he developed cough and fever associated with bilateral upper lobe opacities on chest x-ray and a positive culture for acid fast bacilli.

He became cachectic with a high fever, and monilial patches were seen on the tongue and buccal mucosa. He began talking to himself, associated with inappropriate mood changes, and had one episode resembling a fit.

His ESR rose to 156mm/first hour, and the LE cell phenomenon was strongly positive with a positive antinuclear factor. Chest x-ray showed collapse of the left lower lobe. He was treated with high doses of Hydrocortisone, Streptomycin and Isoniazid.

Two months later when seen in this hospital the patient was weak, and walking with the help of a walking stick, but otherwise was asymptomatic. There was bilateral pitting oedema of the ankles with dullness to percussion at the left base, where a pleural rub was noted. The liver was palpable 3cm and the spleen was enlarged to percussion but not palpable. Urine examination showed SG 1015, proteinuria # (5.85g in 24 hours), red cells 58/ml, white cells 70/ml with occasional red cell, white cell and granular casts noted on several occasions.

He improved after increasing the dose of Prednisolone and adding oral Cyclophosphamide, and remains well two years after his original symptom developed.

Case 2
A 24 year old Chinese mother of two children presented with a nine month history of diarrhoea and weight loss. Due to weight loss she stopped taking the contraceptive pill (Ovulen) which she had been taking for the preceding two and a half years. Previously her bowel habits had been normal, but she began passing soft yellow stools, in the absence of blood and mucus, three or four times a day. Once in 2–3 days she had crampy lower abdominal pains lasting 2–3 hours. Subsequently she developed painful swelling of her proximal interphalangeal joints of both hands associated with morning stiffness. She lost 9.1kg weight over this period. On examination there was mild tenderness over the umbilicus. Barium meal with follow through suggested slow progress through the upper jejunum and exploratory laparotomy was performed. The second quarter of the small intestine was found to be inflamed and oedematous.

At laparotomy about 100cm of oedematous and hyperaemic jejunum were excised.

Following laparotomy the intermittent painful swelling of the proximal interphalangeal joints of both hands remitted but she continued to have diarrhoea and weight loss, and developed a flushed skin associated with a malar rash, subsequently found to be exacerbated by exposure to sunlight. Her general condition deteriorated with weight loss of a further 15kg and she was referred to the University Hospital, with persisting diarrhoea, occasional vomiting and weight loss, and of losing more hair than usual.

Examination found an emaciated woman with hyperaemic skin of the trunk and of the extensor surface of the arms. Palmar erythema was present. BP 120/70mm Hg. A midline lower laparotomy scar was noted, also hepatomegaly 2cm below the right subcostal margin, and bilateral axillary lymphadenopathy. She continued to pass frequent stools, and later developed vomiting of bile-stained fluid.
INVESTIGATIONS

Urine
RBC 2/ml, WBC 11/ml, and no casts were seen. The 24 hour urine protein was 0.7g, and urine culture was negative.

Haematology Hb 10.6g/dl, reticulocyte count 0.3%, white cell count 2.0 x 10^9/l, platelet count 64 x 10^9/l and the ESR was 55mm in one hour. Bone marrow showed hypoplasia with a suggestion of damage to the integrity of the cells. LE cells' were positive 1:500 WBC. Serum iron 1.3mmol/l, ifon-binding capacity 36.3mmol/l, serum folate 7.5mg/l and serum B was 558mg/l.

Immunology
Antinuclear antibody, rheumatoid factor and the Coombs test were negative. C3 45mg (lower limit of normal 80mg) and C4 5mg (lower limit of normal 20mg).

Biochemistry
Serum electrolytes: sodium 134mmol/l, potassium 2.9mmol/l, chloride 96mmol/l; serum albumin 28g/l, serum globulin 43g/l. Electrophoresis showed a diffusely raised gamma globulin with a decreased albumin. Aspartate aminotransferase 27 IU/l, alanine aminotransferase 7 IU/l and alkaline phosphatase was 105 IU/l; cholesterol 3.5 mmol/l.

Radiology
X-rays of the chest, sacro-iliac joints and knees were normal. Barium meal and follow through showed that the whole of the small bowel was abnormal with thickened folds of mucosa. The second and third parts of the duodenum were dilated with retention of the barium. The caecum appeared contracted with possible ulceration of the mucosa. Barium enema was normal. Intravenous Urogram showed normal kidneys, but the ureters were dilated and full throughout their length, including the post-micturition film.

Pathology
As in Case 1, histological examination showed widespread oedema of the gut wall, most prominent in the submucosa. A moderate, diffuse infiltration by eosinophils was also present, especially in the muscle layer, and this was associated with a few lymphocytes and other mononuclear cells. There were no granulomata. Blood vessels were numerous, particularly in the submucosa. They were patent, engorged with blood, and did not show vasculitis, thrombosis or necrosis. However, intimal thickening and endothelial swelling were present in occasional small blood vessels.

She was treated with Prednisolone 60mg daily with complete remission of her symptoms. She remained well until her facial rash recurred, exacerbated by sunlight, which she attributed to her Prednisolone and so she stopped therapy. Within one week she had recurrent abdominal pains and diarrhoea and developed signs of a paralytic ileus which regressed with Prednisolone.

Two years following the development of her initial symptoms she developed a nephrotic syndrome and renal biopsy showed a severe epi-membranous nephropathy.

DISCUSSION
Gastro-intestinal manifestations are quite common in SLE, but they are very rarely the presenting features. Brown et al.(5) noted minor gastro-intestinal symptoms in 29% of 87 patients, and major symptoms in 8%. One of Brown's patients (case 2) presented with weight loss, vomiting and belching, and diarrhoea. This 'Duodenal ileus' was attributed to compression by the superior mesenteric artery. The diagnosis of SLE in this case rested on positive LE cell preparations and a leucopenia only. None of the patients reported by Kurlander and Kirsner(6) appear to resemble our two patients. Our two patients most nearly resemble the patient described by Pachas et al.(7) although his patient had no crampy abdominal pains. The other features of diarrhoea, vomiting and weight loss however are very similar to those of our two patients, as was the general clinical appearance with rash, alopecia and leucopenia. Another similar patient was described by Trentham and Masi(8) again with the absence of crampy abdominal pains. However in our second case, relapse of the SLE by self-withdrawal of treatment produced a recurrence of the presenting symptoms of diarrhoea and abdominal pain. On this occasion there was existence of an active facial rash, whilst on her initial admission palmar erythema had been noted.
The case reported by Bazinet and Marin(9) had similar abdominal symptoms, with an acute onset, but here the presenting feature was a Coomb's positive haemolytic anaemia.

The patient reported by Shafer and Gregory(10) presented with nephritis and pleurisy and later an acute abdomen, with features at operation somewhat similar to the above two patients. Unfortunately, no biopsy material was available as regional enteritis was diagnosed, and the subsequent post-mortem examination would undoubtedly have been influenced by the steroid therapy given.

In neither of our patients was peritoneal irritation prominent, although this has been noted(11–13).

More specific symptomatology has been described in patients with mesenteric arteritis(14,15) and arteriolonecrosis(16). Dubois(1) reports a similar patient with 'lupus vasculitis' with an acute abdomen. She had had SLE for seven years. Dubois called this a 'lupus vasculitis' of the ileum, but the histology of our two patients as with that of Dubnow et al.(11) showed no evidence of vasculitis, thrombosis or necrosis. Since neither of the two patients had had previous steroid therapy, the lack of histological evidence of vasculitis cannot be attributed to immunosuppression.

The significance of the eosinophilic infiltration is uncertain. Siurala et al.(17) found eosinophils in 16 out of 36 patients with 'collagen disease' who had been untreated with steroids and in only 2 of 15 patients treated with steroids. No eosinophils were noted by Dubnow et al.(11) and they were not seen by Pollak et al.(13) in a patient on steroids. However, the presence of eosinophils in a section of oedematous bowel wall should alert the surgeon to the possibility of a connective tissue disorder.

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REFERENCES


