Systemic Lupus Erythematosus Presenting with a Fatal Intestinal Vasculitis: a Case Report

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ABSTRACT

This case report demonstrates fatal gastrointestinal vasculitis as a rare presentation of systemic lupus erythematosus. A 34-year-old woman presented with abdominal pain and diarrhea. Anti nuclear antibody was positive and high titre of anti-ds DNA antibody was also reported. Treatment with corticosteroid and supportive cares were started; however, her condition worsened. Eventually, she was considered as a candidate for diagnostic laparoscopy. Immediately after laparoscopy, she developed respiratory distress along with upper gastrointestinal bleeding. Soon after, the patient died because of disseminated intravascular coagulation.

KEYWORDS
Systemic lupus erythematosus;Vasculitis;Gastrointestinal manifestations

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INTRODUCTION

Systemic lupus erythematosus (SLE) is an autoimmune disease that may involve multiple organs.[1] Gastrointestinal (GI) manifestations of SLE are protean. Abdominal pain is the most common symptom but with different etiologies in SLE.[2] GI vasculitis is very important to be recognized and treated early. Clinicians can prevent morbidity and mortality related to this potentially grave entity. Herein, we report a case of fatal gastrointestinal vasculitis as a rare presentation of systemic lupus erythematosus in a 34-year-old woman.

CASE REPORT

A 34-year-old woman was admitted to our hospital because of progressive lower abdominal pain for duration of two weeks. She developed a few episodes of non-bloody, loose stool along with constant lower abdominal pain. She did not experience any fever, anorexia, or weight loss. On physical examination she was conscious, mildly pale, and afebrile with pulse rate: 100 beats/min, blood pressure: 120/80 mmHg, respiratory rate: 14/min, mild diffuse abdominal tenderness, and mild abdominal distension along with shifting dullness. Other exams were unremarkable.

Laboratory data showed white blood count (WBC):4700/mm^3 (neutrophil: 80%, lymphocyte: 9%, monocyte: 1%, eosinophil: 10%), hemoglobin: 12.8 g/dL, platelet count: 84,000/mm^3, ESR: 11 mm, CRP: 24 mg/dL, LDH: 610 IU/L (NL<460), AST: 20 IU/L, ALT: 13 IU/L, ALP: 165 IU/L, total bilirubin:

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0.3 g/dL, direct bilirubin: 0.1 g/dL, PT: 12 Sec, INR: 1, PTT: 25 Sec, total protein: 6.4 g/dL, albumin: 3.2 g/dL, BUN: 23 mg/dL, creatinin: 0.7 ng/mL.

Anti HCV antibody, HBS antigen, and HBS antibody were negative. Peripheral blood smear showed low platelet, as well as hypochromia, anisocytosis, and microcytosis in red blood cells. Stool smear and culture were normal. Abdominal ultrasonography showed moderate ascites. Abdominal Doppler ultrasonography was normal.

A diagnostic paracentesis was done, which showed: total protein: 5.1 g/dL, albumin: 2.6 g/dL, WBC: 160/mm^3 with 55% lymphocytes, RBC: 3700/mm^3. The serum-ascites albumin gradient (SAAG) was 0.5. Bacterial evaluation, acid fast staining, and cultures of the fluid were all negative.

Abdominopelvic computed tomography showed diffused wall thickening in the colon along with thickening of a loop in the small intestine at the left upper quadrant. Marked ascites and small left pleural effusion were also seen but no evidence of vascular thrombosis was detected.

Platelet count rapidly dropped to 20,000 during the next few days but reticulocyte count, d-dimer and coomb's test were normal. Nevertheless fibrinogen level was 14 mg/dL (NL<4). The patient remained in good condition and her diarrhea and abdominal pain improved with supportive treatment. Colonoscopy showed patchy mucosal red areas in mucosa without overt ulcers or erosions. Biopsy sample of the colon did not show any abnormality. Five days after taking biopsy samples, the patient's condition deteriorated as she complained of vomiting and diarrhea, yet again. At the same day she underwent another abdominopelvic computed tomography that showed wall thickening of the colon and target sign in the small intestine. Upper pole of the left kidney showed heterogeneous enhancement of suspected ischemic changes but no evidence of vascular thrombosis was detected again (figure 1).

Initial investigations for infectious agents including strongyloides stercoralis and neoplasms such as lymphoma were negative. However, antinuclear antibody was elevated to 100 (Nl<10 EU). Intravenous hydrocortisone 300mg/day was started immediately and anti ds-DNA and complements (C3, C4, CH50) were requested to be tested.

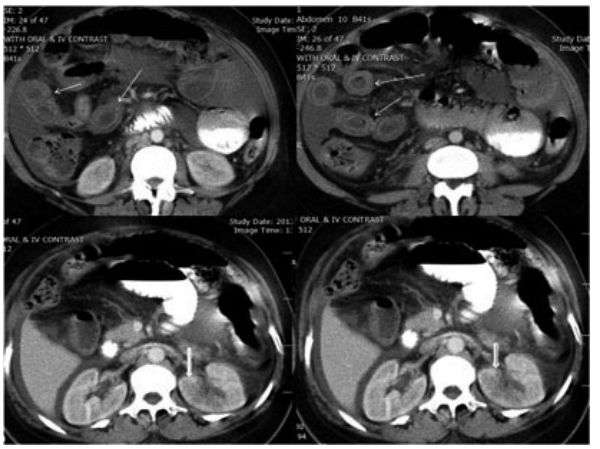

Fig. 1: Abdominal and pelvic tomogram with intravenous and oral contrast showed target sign (narrow arrows) and ischemic changes in left kidney (thick arrows).

Despite the use of high dose intravenous steroid therapy, the patient's symptoms did not improve. Her condition deteriorated precipitously. The patient underwent diagnostic laparoscopy after 3 days of corticosteroid therapy because of the new changes in her condition. Laparoscopy did not show any complications of GI vasculitis. During laparoscopy biopsy samples were taken from small and large intestine and peritonea. A few minutes after the procedure, abundant pinkish discharge was noted via the endotracheal tube and the patient also developed hypoxemia and tachycardia. On examination she had bilateral diffuse rales and ronchi along with a distended abdomen. Active upper GI bleeding was also seen through the nasogastric tube. The patient passed away despite immediate supportive treatments. Her death was due to disseminated intravascular coagulation (DIC) and subsequent respiratory failure. Laboratory data later showed high titer of anti ds-DNA [150 (NL<35 IU/ml)] and low complement levels. GI sample biopsies through laparoscopy and after death (open laparotomy) from intestine did not show any histological abnormality.

DISCUSSION

SLE enteritis is one of the most common causes of abdominal pain in SLE.[2] SLE can present with acute or chronic diarrhea. There are cases of SLE that presented with eosinophilic enteritis,[3] and ischemic colitis.[4] SLE vasculitis is rarely confirmed with histological examination.[4] In our case GI histology could not show any pathological problem

and was not helpful for accurate diagnosis.

Some laboratory data used to detect SLE activity such as anti phospholipid antibody (aPL), do not correlate with lupus enteritis except leucopenia.[5] Computed tomography can show and monitor pathological abnormalities in the abdomen.[6,7] In one study target sign was reported in 90% of patients with SLE who complained of acute abdominal pain. In all cases, bowel wall thickening was circumferential and symmetric and the most common sites of involvement were the jejunum and ileum.[6] Rectal involvement is rare.[5] Double halo or target sign,[8,9] can be seen due to submucosaledema of the small bowel.[8] In our case diffuse wall thickening was seen in the colon and rectum in early phase, which extended to the small intestine with target sign view.

Corticosteroid is the first line of treatment.[1,2,5,6,8] Most patients respond to high dose corticosteroid1,[2,6,8] but our patient did not. We performed diagnostic laparoscopy due to the rapid deterioration of her condition to detect any complications; however no macroscopic or microscopic abnormalities were found in the bowel and finally she died as a result of DIC. Although complications of diagnostic laparoscopy are rare, we cannot rule out the positive pressure effects on small vessels through gas insufflations. Small vessels are involved during GI vasculitis[8,10] and even minimal positive pressure might lead to reduce visceral perfusions in this setting.[11,12] We could not find the reason for the patient's rapid deterioration and death after diagnostic laparoscopy.

Clinicians can prevent morbidity and mortality of SLE related GI vasculitis if they can recognize and treat it as early as possible. Abdominal computed tomography is a useful tool for detecting some abnormalities in abdominal pain of patients with SLE.

SLE vasculitis is rarely confirmed with histology.

CONFLICT OF INTEREST

The authors declare no conflict of interest related to this work.

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