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Single Case

Lupus Gastrointestinal Tract Vasculopathy: Lupus “Enteritis” Involving the Entire Gastrointestinal Tract from Esophagus to Rectum

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Keywords

Lupus enteritis · Lupus gastrointestinal tract vasculopathy · Upper and lower gastrointestinal tract

Abstract

Gastrointestinal symptoms are very common in systemic lupus erythematosus (SLE). Lupus “enteritis” is very responsive to treatment but can have devastating consequences if not detected. Most descriptions of enteritis involve the small and large bowel. This is the first report of lupus “enteritis” involving the entire gastrointestinal tract from the esophagus and stomach to the rectum. Lupus “enteritis” is another cause of upper gastrointestinal involvement in SLE (involving even the esophagus and stomach) in addition to involvement of the lower intestinal tract.

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Introduction

Gastrointestinal manifestations in systemic lupus erythematosus (SLE) were first described by Sir William Osler in 1895 [1]. SLE can affect every part of the gastrointestinal tract from mouth to rectum and in different ways [2]. In SLE-related abdominal pain, various diagnoses may refer to the same or very similar condition – for example lupus mesenteric

vasculitis, mesenteric arteritis, lupus arteritis, lupus vasculitis, gastrointestinal vasculitis, and acute gastrointestinal syndrome [3, 4]. The British Isles Lupus Assessment Group (BILAG) 2004 definition of lupus enteritis describes either vasculitis or inflammation of the small bowel with supportive imaging and/or biopsy findings [4, 5].

While lupus “enteritis” is relatively uncommon, most case reports have focused on involvement of the small and large bowel. We document a patient with SLE presenting with acute abdominal pain that had “enteritis” with inflammation along the entire gastrointestinal tract from the esophagus to the rectum. To our knowledge, this is the first description of such pan-gastrointestinal involvement including the esophagus.

Case Report

A 44-year-old female presented to the emergency department with a 24-hour history of abdominal pain. She had acute onset of severe right upper quadrant abdominal pain followed by nausea and non-bloody emesis. She had loose, non-bloody stools and felt dizzy and fatigued.

She had a 17-year history of SLE with synovitis, alopecia, malar rash, anemia, thrombocytopenia, elevated anti-dsDNA antibodies, and anti-Ro and anti-La antibodies. Initially she received only prednisone for flares. She was on hydroxychloroquine but stopped it 4 months before presentation due to cost. Four days prior to her admission, she saw her rheumatologist for increasing fatigue, achy joints, and a malar rash. Labs revealed an ANA titer of 1:640, markedly elevated anti-dsDNA antibodies, and undetectable C3 and C4 levels. She had positive anti-Sm, anti-RNP, anti-Ro, and anti-La antibodies. Lupus anticoagulant and anticardiolipin antibodies were negative. β 2 Glycoprotein I IgM/IgG antibodies were negative. β 2 Glycoprotein I IgA antibody was positive at >150 SAU (normal \leq 20 SAU). She was started on prednisone orally and hydroxychloroquine was reintroduced.

On examination she was afebrile but in distress. The abdomen was soft and non-distended. She had tenderness to direct palpation throughout with most pain noted centrally but without rebound tenderness. She had hypoactive bowel sounds. There was a malar rash, but no oral ulcerations or synovitis. Laboratory studies also included leucopenia, proteinuria, and hematuria. The total protein/creatinine ratio was 1.6 (normal <0.2).

A CT scan of the abdomen with IV contrast showed extensive, diffuse submucosal edema and bowel wall thickening involving the esophagus, stomach, duodenum, jejunum, ileum, colon, sigmoid colon, and rectum. The target sign was present as well as extensive mesenteric venous engorgement with the comb sign (Fig. 1). There was excellent flow demonstrated in the SMV, IMV, splenic, and portal venous systems without evidence for thrombus. Ascites was present. She was diagnosed with lupus “enteritis”. She received methylprednisolone 1 g IV daily for 3 days, then 250 mg IV daily for 2 days. She was on bowel rest which was advanced slowly as tolerated. She began to improve within 24 h of receiving IV steroids. A repeat CT scan 6 days after the initial scan showed marked improvement in the small bowel with resolution of the diffuse submucosal bowel wall edema, no mural thickening, or enhancement of the bowel wall. There was no stranding in the adjacent mesenteric fat or engorgement of vessels. There was resolution of the submucosal edema of the colon, esophagus, and stomach as well (Fig. 2). She was discharged on prednisone 60 mg daily, hydroxychloroquine, and mycophenolate mofetil. Approximately 3 weeks later she underwent a kidney biopsy which showed mesangial proliferative lupus nephritis (ISN/RPS class II, with index of activity 0/24 and index of chronicity 1/12).

Discussion

The clinical presentation of lupus enteritis is nonspecific but is usually associated with abdominal pain and vomiting. Missing the diagnosis of lupus enteritis can lead to devastating consequences including perforation [6]. This case is the first to show lupus “enteritis” involving the entire gastrointestinal tract from the esophagus to the rectum.

Janssens et al. [4] reported 7 new patients who met criteria for lupus enteritis as well as an additional 143 patients from the literature. Jejunum was involved in 83%, ileum in 84%, colon in 19%, duodenum in 17%, and rectum in 4%. Kwok et al. [7] described 41 SLE patients with lupus enteritis. The jejunum was involved in 80% and ileum in 73.3%. Lee et al. [8] reported on 17 patients with a total of 21 episodes of lupus enteritis. The ileum was involved in 80% of cases, jejunum in 85% of cases, and rectum in 14%. Nineteen of the 21 cases had bowel involvement in multiple vascular territories as lupus enteritis may affect several vessels at once [9]. In these retrospective reviews, patients responded well to steroids [4, 7, 9]. Therefore, making the correct diagnosis is of the utmost importance. With availability of CT scans, the mortality rate is believed to be lower and the disease is now thought to have a more benign clinical course [10].

This case adds to the literature in providing evidence that the same underlying pathophysiological process may involve the entire gastrointestinal tract from the esophagus to the rectum. Lupus enteritis is thought to be due to immune-complex deposition and complement activation in association with primed endothelial cells which can induce neutrophil-endothelial cell adhesion and predispose to a leuko-occlusive vasculopathy [11, 12]. Systemic complement activation can lead to diffuse microvascular injury and increase vascular permeability which may be present in the mesenteric circulation leading to intestinal capillary leak resulting in submucosal edema [13]. Similarly, Musaev et al. [14] reported on 12 children with SLE and chronic gastroduodenitis. Biopsies of the stomach and duodenum during disease exacerbation revealed immune complex deposition in the arteriolar walls [14]. The same pathophysiology may be present in lupus enteritis. Occasionally, microvascular thrombosis is noted [4]. This patient had positive $\beta 2$ glycoprotein I antibodies but did not fit criteria for the antiphospholipid syndrome.

In summary, lupus “enteritis” is a serious gastrointestinal manifestation of SLE which is very responsive to steroids. This report confirms that the entire length of the gastrointestinal tract may be involved. As lupus “enteritis” suggests involvement of just the small bowel, perhaps lupus gastrointestinal tract vasculopathy would be a more fitting, descriptive term.

Statement of Ethics

The work done for this paper has the approval of the Institutional Review Board and all rules were followed.

Disclosure Statement

The authors declare that there is no conflict of interest.

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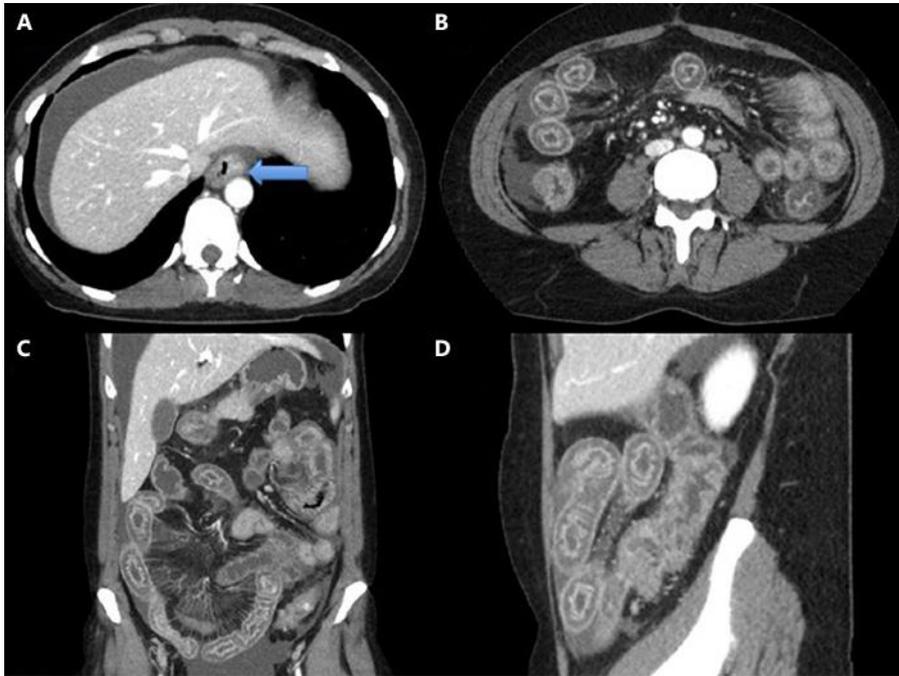


Fig. 1. **A** Edema of the esophagus (arrow). **B** Target signs of the small bowel with mesenteric vascular engorgement suggestive of the comb sign. **C** Coronal view showing target signs of the small bowel as well as engorgement of the mesenteric vasculature. **D** Sagittal image showing edema of the small and large bowel.

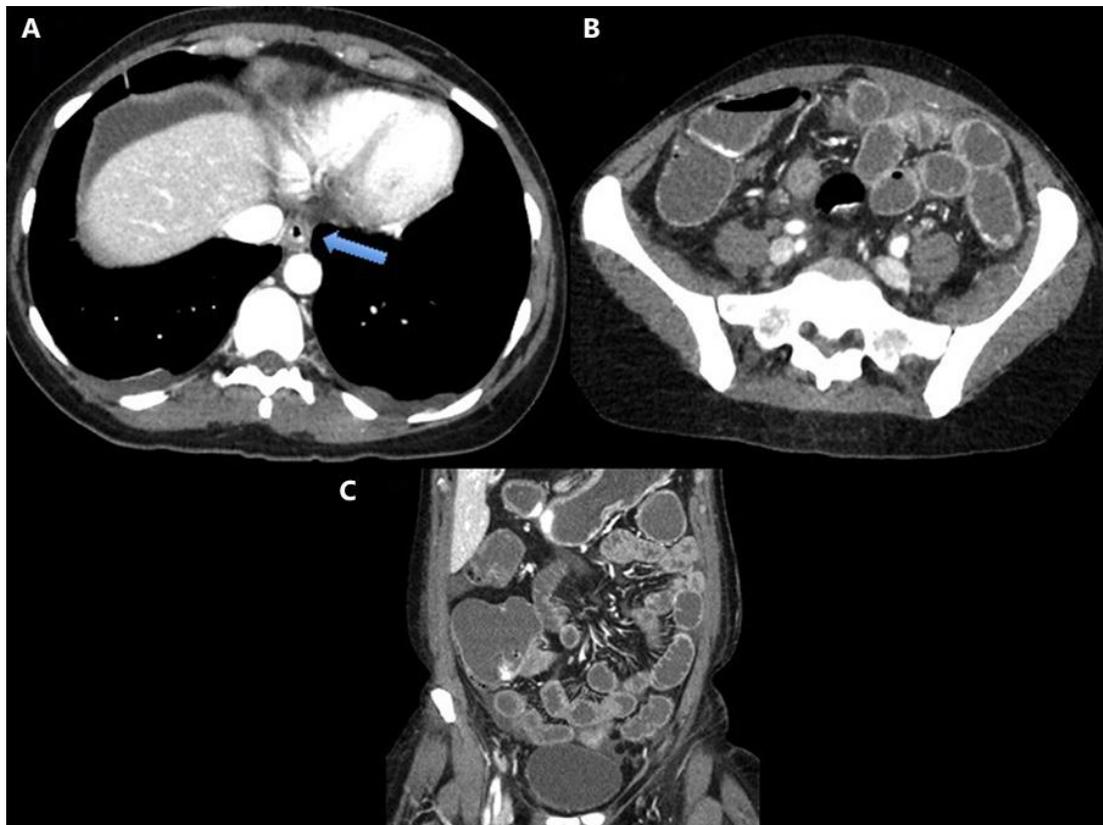


Fig. 2. **A** Resolution of edema of the esophagus (arrow). **B** Resolution of the small bowel edema and target signs. **C** Coronal image showing resolution of comb sign and target signs.