Lupus Enteritis as a Lone Manifestation of SLE: Rare but Possible.

Eduardo Quintero 1, Grace Salacup 1, Jerald Pelayo 1, Kevin Bryan Lo1.

1. Department of Medicine, Einstein Medical Center Philadelphia, Pennsylvania, USA.

Introduction

Lupus enteritis (LE) is a rare manifestation of systemic lupus erythematosus (SLE) and only affects 0.2% - 5.8% of patients. There are only 11 cases reports in which lupus enteritis was the initial presentation of SLE which made our case a diagnostic challenge.

Case Presentation

A 28-year-old female with asthma and cholecystectomy 3 months ago presented with abdominal pain, diarrhea, and vomiting. Her abdominal pain started 3 days prior, was diffuse, intermittent, sharp, non-radiating, associated with 20-30 non-bloody bowel movements, and bilious emesis. She denied fever, rash, arthritis, oral ulcers, sick contacts, travel, changes in diet, or antibiotic use. No pertinent family or social history.

She was hemodynamically stable, afebrile. The physical exam was unremarkable except for hyperactive bowel sounds. Laboratory results showed anemia and leukocytosis.

The initial differentials were inflammatory bowel disease, infectious enteritis, biliary leak, malabsorption, and irritable bowel syndrome. CT scan of the abdomen showed ascites with wall thickening of the third, and fourth portion of the duodenum as well as thickening and mucosal hyperenhancement of the jejunum and ileum (figure 1). Paracentesis was non-diagnostic.

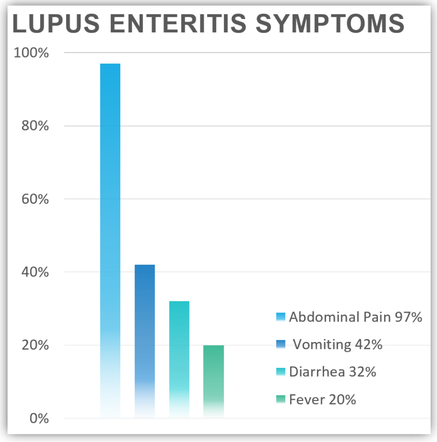
The infectious workup was negative. MRI enterography showed both small and large bowel wall thickening. Since non-invasive studies were inconclusive, upper endoscopy and colonoscopy with biopsies were performed which were unremarkable.

At this point, other less common conditions such as autoimmunity were considered. Her ANA was elevated with hypocomplementemia. Anti-dsDNA and Anti-Smith were positive suggestive of SLE. High dose steroids were given and symptoms rapidly improved. She was discharged on oral steroids. On outpatient follow up, her condition was improved with hydroxychloroquine and prednisone.

Discussion

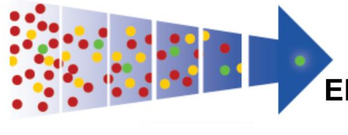
SLE presents with diffuse abdominal pain, vomiting, diarrhea and fever. Laboratory testing is a useful tool to aid in the diagnosis. Our patient had anemia, positive ANA, elevated anti-Smith antibodies, and hypocomplementemia, which correlate with prior case reports. SLE manifesting as enteritis with no other prominent associated signs and symptoms can present as a diagnostic dilemma.

The initial imaging study of choice is an abdominal CT scan. Cardinal features of LE on CT include bowel wall thickening or 'target sign'(Figure 2), intestinal dilatation, engorgement of mesenteric vessels/ "comb sign" and mesenteric fat attenuation. Endoscopic procedures have a low yield.



Differential Diagnosis

- -Infection
- -Inflammatory Bowel Disease
- -VIPoma
- -Serositis
- -Pancreatitis
- -Intestinal vasculitis.
- -Protein-losing enteropathy.
- -Celiac disease.
- -Intestinal pseudo-obstruction



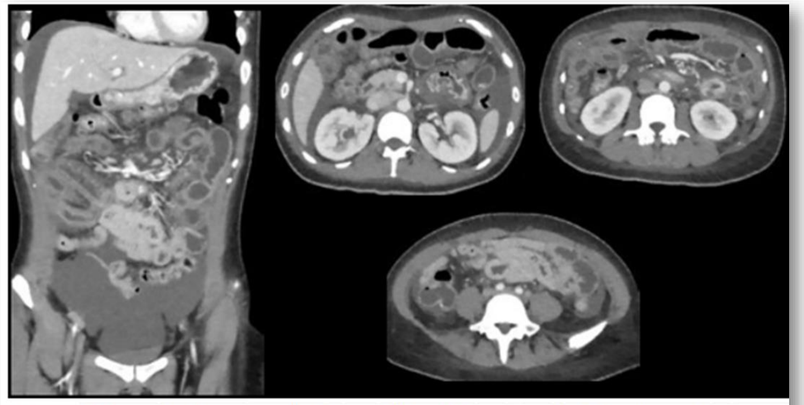


Figure 1. Computed tomography Scan of the abdomen and pelvis with contrast, coronal view and transversal view

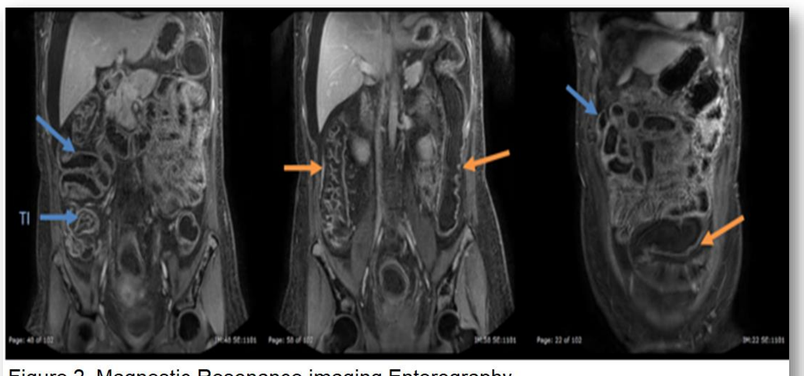


Figure 2. Magnestic Resonance imaging Enterography

The evidence is poor on LE treatment, but steroids have been used as first-line agents. Prognosis is generally excellent for patients with LE has given a good response to steroids.

Conclusion

Lupus enteritis as the sole presenting manifestation of SLE is rare and requires a high index of suspicion together with a multidisciplinary approach. Evidence for treatment is scarce but steroids may be utilized