

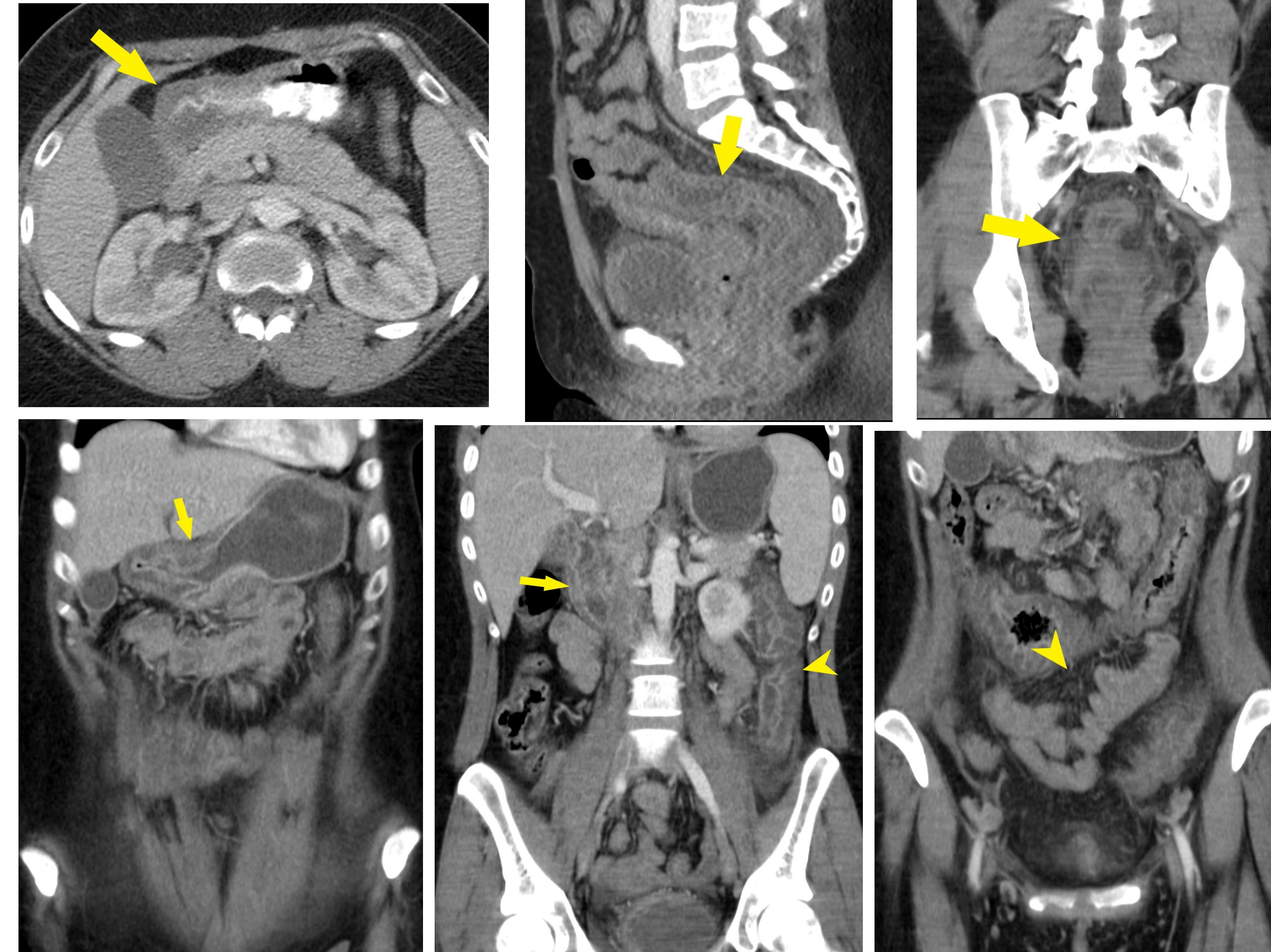
### INTRODUCTION

- The differential diagnosis of diarrhea in the setting of systemic lupus erythematosus (SLE) is broad and includes lupus-related as well as non-lupus-related etiologies.
- Lupus-related etiologies include but are not limited to mesenteric vasculitis, pseudo-obstruction, protein-losing enteropathy, and pancreatitis.
- Autoimmune lupus enteropathy (AIE) is rare in adults, with very few cases described in the medical literature.
- We report a case of pan-gastrointestinal lupus enteropathy that responded effectively to Belimumab.

### CASE PRESENTATION

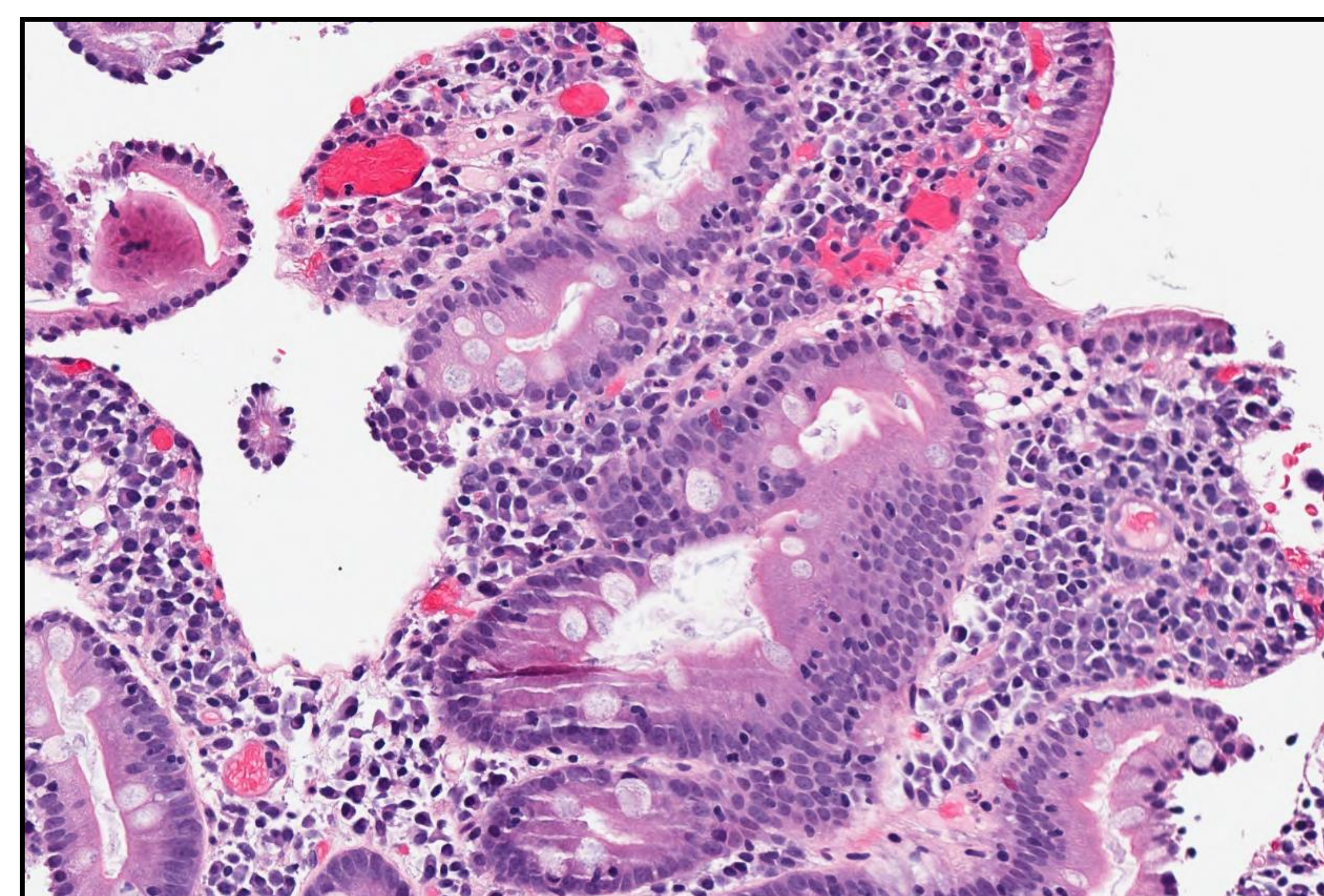
- A 31-year-old woman presented with intermittent ongoing diarrhea of six months duration.
- Workup for infectious causes : stool for ova and parasites, stool cultures, and C. difficile toxin, all of which were negative.
- Celiac disease antibody testing was negative.
- C3 and C4 complement protein levels were low.
- Positive results were seen for serum ANA at dilutions >1:1280 (normal 1:40) with a speckled pattern.
- Anti- dsDNA, Anti-SSA, anti-U1-RNP, anti-cardiolipin (IgG and IgM), and anti-B2-glycoprotein- I (IgA) antibody titers were elevated.
- Anti-MPO, and anti-PR 3 antibodies were within the normal range.
- CT abdomen with contrast : Wall thickening in the distal esophagus, gastric body and antrum, proximal bowel loops, sigmoid colon, and rectum. Minimal mesenteric vascular engorgement and inflammatory vascular stranding around the proximal bowel loops (figure).
- CT angiography of the abdomen : normal caliber aorta with patent celiac artery, superior mesenteric artery (SMA), and inferior mesenteric artery (IMA).
- EGD and colonoscopy : mild inflammation in the entire stomach, and the first and second portions of the duodenum, sigmoid colon and the rectum on colonoscopy. This was confirmed on histology (see figure)
- The imaging findings and the absence of infectious or other inflammatory etiology during the work-up pointed to the diagnosis of autoimmune lupus enteritis.
- High-dose intravenous steroids and Belimumab injections were initiated.
- The patient responded favorably and continued to remain in sustained remission for more than six years now.

### CT ABDOMEN



**Diffuse circumferential wall thickening and edema of the gastric wall (arrow, a), most prominent at the pylorus. There is mild diffuse circumferential wall thickening of the colon, most prominent at the recto-sigmoid region (arrow, b) with mucosal hyper-enhancement, pericolonic fat stranding (arrow, c) and small ascites. CT eight months later shows similar findings including diffuse wall thickening and edema involving the stomach (arrow, d), duodenum (arrow, e), and colon (arrowhead, e) including rectosigmoid. Again, diffuse mesenteric edema and engorgement (arrowhead, f)**

### DUODENAL BIOPSY



**Neutrophils within the lamina propria and apoptotic bodies in the glands, indicating active inflammation and glandular injury. There is also an increase in the plasma cells within the lamina propria**

### DISCUSSION

- Lupus enteritis is defined as either vasculitis or small-bowel inflammation with accompanying radiological and/or histological evidence<sup>1</sup>
- While it is not uncommon for multiple vascular territories to be involved, the entire gastrointestinal tract is rarely affected<sup>2</sup>.
- It is organ-specific manifestation of systemic inflammatory response syndrome (SIRS) that accompanies a lupus flare<sup>3</sup>.
- It is best explained by the Schwartzman phenomenon in which complement activation in tandem with immune complexes and primed endothelial cells are presumed to induce neutrophil-endothelial cell adhesion and leuko-occlusive vasculopathy. The resultant effect is widespread damage to microvasculature and an increase in vascular permeability<sup>3</sup>.
- Early diagnosis with CT abdomen is pivotal to avoiding sequelae including bowel ischemia, perforation, and hemorrhage<sup>4</sup>.
- Imaging findings include dilated bowel, focal or diffuse bowel wall thickening, abnormal bowel wall enhancement (double halo or target sign), mesenteric edema, engorged mesenteric vessels (Comb sign), and increased attenuation of mesenteric fat, and ascites<sup>4</sup>.
- Definitive diagnosis requires endoscopy and biopsy demonstration of vasculitis in the mesenteric arterioles. As the submucosa is difficult to access, a negative biopsy does not rule out the diagnosis
- Immune modulatory treatment is often initiated with high-dose intravenous corticosteroids.
- Belimumab is a monoclonal antibody directed against B-cell survival factor (BAFF) authorized for the treatment of non-renal lupus<sup>5,6</sup>.
- It is infrequently used in the treatment of lupus enteropathy. This is one of the few cases successfully treated with Belimumab.

### REFERENCES



### CONTACT INFORMATION

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