Severe lupus enteritis complicated by intractable gastrointestinal hemorrhage

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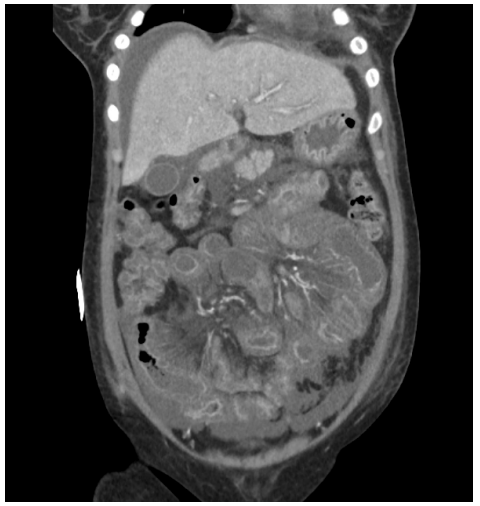
Background

- Lupus enteritis (LE) is a rare presentation of systemic lupus erythematosus (SLE).
- Symptoms can include abdominal pain, laboratory abnormalities, and abdominal imaging findings.
- The most severe complications of LE include gastrointestinal hemorrhage, bowel perforation, and death.
- Immunosuppression is the cornerstone for treatment of the underlying disease process, with endoscopic interventions being reserved for cases with identifiable sources of bleeding.

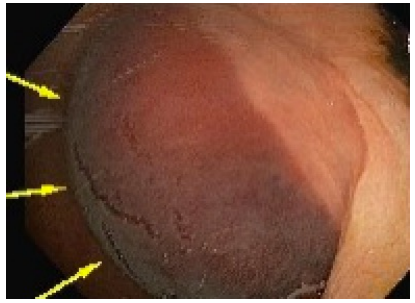
Case History

- A 22-year-old female with SLE, Sjögren syndrome, and bipolar disorder presented with 1 day of nausea, vomiting, non-bloody diarrhea, and severe abdominal pain.
- Patient was hospitalized and underwent abdominal imaging which was highly suggestive of LE (**Image A**). She later developed profound hematochezia and acute blood loss anemia
- **Treated with**: high dose systemic corticosteroids, IVIG, cyclophosphamide, and eventual plasmapheresis.
- **GI Procedures**: 4 colonoscopies, 2 anterograde enteroscopies, and 1 esophagogastroduodenoscopy each revealing diffuse ulcerations not amenable to interventions.
- **IR procedure**: mesenteric angiography with coil embolization of the ileal branch of superior mesenteric artery.
- Her GI hemorrhage was complicated by warm-autoimmune hemolytic anemia and disseminated intravascular coagulation resulting in a **nadir hemoglobin of 2.2 g/dL**, requiring a total of **43 units of packed red blood cell** transfusions.

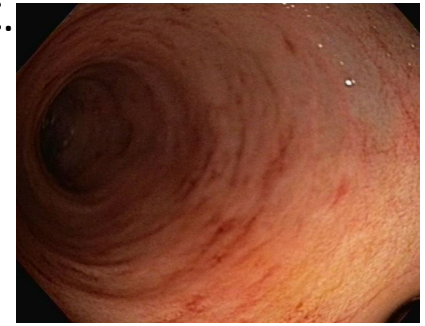
A.



B.



C.



Images: A. Computed tomography showing diffuse intestinal thickening and edema. **B.** Large cecal/ileocecal valve submucosal hematoma. **C.** Mid-distal ileal ulcers with punctate hemorrhagic lesions.

Case History

- **Other complications**: lupus nephritis, PRES syndrome, ascites, protein-losing enteropathy, cellulitis, pleural effusions, and critical illness myopathy
- After 8 weeks, she was discharged on prednisone, hydroxychloroquine, and cyclophosphamide
- She has remained in **clinical remission** at multiple clinic appointments

Discussion

- In the absence of hemodynamic instability, **repeat endoscopies after initial diagnostic endoscopy should be limited** due the low likelihood of achieving endoscopic hemostasis, while carrying a considerable risk of procedural and sedation-associated adverse events.
- In the setting of active extravasation, vascular embolization should be pursued cautiously.
- At all times, the focus of treatment should be on **early, aggressive immunosuppression** and initiation of advanced therapies such as plasmapheresis.

Conclusions

LE can be a life-threatening manifestation of SLE. Treatment should focus on immunosuppression and endoscopies should be limited if possible.