A challenging case of Lupus Mesenteric Vasculitis

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Case presentation

- 41 year old male with PMH of antiphospholipid syndrome, Sjogren's syndrome, Systemic lupus erythematosus, stable on hydroxychloroquine and azathioprine since 2011, presented with:

1. Acute onset of diffuse abdominal pain
2. Watery diarrhea
3. Nausea and vomiting
PHYSICAL EXAM

Vitals:  T:97.8, RR: 16, Pulse: 110, BP 102/80

HEENT:  Dry mucus membranes

CVS:  S1,S2 normal , no murmur , rubs or gallops

Lungs:  clear to auscultation
Physical Exam

Abdomen: soft, non-distended, **diffuse tenderness to palpation**, +rebound, no guarding/rigidity, BS active

Extermities: trace pedal edema.

Skin: Malar rash, diffuse erythematous blanching rash on upper neck
WBC 8,000, hemoglobin 13, Platelets 240,
- Alb 1.7, TP 3.7
- AST 54, ALT 33, T. bilirubin 0.6, Cr 0.5,
- Lipase 80, lactic acid 1.2
- CRP: 16.7
- Stool for ova, parasites, c. diff, bacterial pathogen negative
- Blood culture: no growth
- UA clean
- ANA: 1:164
- Complement level: C3 normal, C4 low
- ANCA titer: negative
CTA

- Generalized edema involving the entire small bowel wall. Mesenteric fat attenuation.

- No CTA evidence of large/medium sized vessel abnormalities or large vessel thrombosis
Fig 1. Microphotograph showing the duodenal mucosa virtually replaced by a band of granulation tissue (H&E stain, magnification x200).
Fig 2. Microphotograph of the duodenal mucosa excessive plasma cells in the lamina propria (rectangle) and vasculitis in the mucosal capillaries (arrow) (H&E stain, magnification x400).
Lupus mesenteric vasculitis
Lupus Mesenteric vasculitis

- Vasculitis of small vessels
- One of the most serious GI complication in SLE
- If untreated can lead to ischemia, bowel perforation and severe morbidity and mortality = 50%
- Global prevalence ranges from 0.2-9.7%
- Male : female ratio 1:14

Lupus Mesenteric vasculitis

- Symptoms - diffuse abdominal pain, nausea, vomiting, diarrhea, melena and postprandial fullness.

- Symptoms varies from mild to severe GI bleeding.

- Signs - abdominal distension, rebound, guarding, hypoactive bowel sounds - only 30%
Diagnosis

- No single lab data is considered to be pathognomonic for LMV.
Sagittal section of abdominal CT shows engorgement of mesenteric vessels (Comb appearance)
Arrow
Cross sectional CT images shows bowel wall edema, thickening (Target sign) small arrow, fat attenuation and ascites (asterix)
Endoscopy

- Abnormal in only 40%
  - Typically findings are multiple small ulcers and scars.
  - Jejunum and ileum is most commonly involved
  - Involvement of duodenum is nearly always indicative of vasculitis
Pathology

- Cellular infiltration of submucosal and muscular layers with or without edema or vasculitis.

- Vasculitis has been characterized as necrotizing vasculitis with fibrinoid necrosis.
Evidence

- Janssens et al.

- 150 patients

- All received steroids as first line treatment either IV methylprednisolone (87%) or oral prednisone (13%)

- 141 patients received corticosteroids alone

- Cyclophosphamide was used as additional immunosuppression in 9 patients with severe organ involvement and unresponsiveness to steroids (1)
Evolution

- Recurrence was reported in 34% patients.
- Retreated with corticosteroids.
- Four patients died after median follow up 18 months, yielding mortality of 2.7%.
Our patient
Treatment

- Initially received 70mg IV Methylprednisolone with minimal symptom improvement

- Plan was to start pulse dose steroids due to severity of illness

- Developed fever, tachycardia and became hypotensive

- Concern for sepsis
Our patient

- Treated with IV cyclophosphamide
- Symptoms improved
- Following in rheumatology clinic
- Getting monthly Cyclophosphamide- 3rd dose
- Prednisone 30mg
Take home points

- Lupus mesenteric vasculitis is one of the most serious GI complication of SLE.

- Early diagnosis is crucial

- Based on case series steroids is the first line treatment

- LMV carries high risk of gut bacterial translocation and sepsis and use of pulse dose steroids can be challenging.

- IV cyclophosphamide may be safer in these situation with lower risk of worsening infection