A Perplexing Case Of Episodic Abdominal Pain And The Role Of Mast Cells Gastroenteritis

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### Objective

Outline the clinical course of a patient with mast cell gastroenteritis and describe typical presentation, interpretation of results, and suggested treatments for this evolving diagnosis.

### History of Presenting Illness

38 y/o woman with hx of RA and MCTD, has several recent hospitalizations for colitis/enteritis of unknown etiology.

### Current constellation of symptoms:

- 4 days of nausea and intractable vomiting
- new-onset rectal fullness with constipation for 4 days
- mucoid rectal discharge for 2 days

Symptoms that started 3 months ago:

- Intermittent intractable vomiting, diarrhea, and abdominal pain
- Complete resolution of symptoms in between episodes
- 3 different admissions at 3 different hospitals

### Social History

Negative for drug use including marijuana. Travel history includes frequent travel for long periods of time to Mexico to visit family.

### Physical Exam

Patient had mild tachycardia but had an otherwise benign exam including a normal rectal. Clear mucoid discharge was seen on her bed sheets.

### Diagnostic Workup

#### Workup prior to admission

- Positive lactoferrin
- QuantiFERON Gold TB screen positive
- CT scan showing inflammatory small and large bowel patterns consistent with inflammatory bowel disease
- Upper and lower endoscopy with biopsies— inconclusive studies without signs of inflammation
- Exploratory laparoscopy within normal limits
- Serum inflammatory markers inconclusive

#### Workup during admission

- Normal CBC with diff
- 2+ protein and 3+ blood on UA
- ESR 10 and CRP 2.4
- Complement C3/C4 low, with normal C1 esterase
- Repeat abdominal imaging showed diffuse small bowel and colon inflammatory wall thickening with severe rectal wall thickening.
- Flexible sigmoidoscopy showed significantly congested rectal mucosa and a patch of mildly erythematous mucosa
- Rheumatologic workup negative for acute flare

### Results

In hopes of discovering a cause for the patient’s symptoms, empiric treatments were held off for almost a week.

- Collitis ruled out on endoscopy and biopsies
- Vasculitis was ruled out due to symptoms being localized to GI system only and no signs of vasculitis seen on rectal biopsy
- Proteinuria and hematuria found to be secondary to menstruation and not part of a vasculitis
- No signs of serosis or rheumatologic flare found
- Underlying mixed connective tissue disease confirmed on rheumatologic workup but not likely contributing to GI symptoms
- Hereditary angioedema unlikely with normal C1 esterase

### Differential Diagnosis

<table>
<thead>
<tr>
<th>Inflammatory bowel disease</th>
<th>Acute intermittent porphyria</th>
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<tbody>
<tr>
<td>Irritable bowel disease</td>
<td>Eosinophilic gastroenteritis</td>
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<tr>
<td>Small vessel vasculitis</td>
<td>Intra-abdominal TB</td>
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<tr>
<td>Hereditary angioedema</td>
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### Radiographic Imaging

**Image 1-CT Abdomen/Pelvis**

Severe rectal wall thickening (see arrow) seen in the setting of constipation on admission. Similar changes observed in more proximal segments of the colon in previous images

**Image 3-CD117 Stain**

Example of a positive CD117 stain for mast cells. Testing is done on a biopsy of the gastrointestinal tissues and cells are seen within the lamina propria.

### Direct Imaging

**Image 2-Flexible Sigmoidoscopy**

Direct visualization of the rectum shows no signs of inflammation. Other than nonspecific swelling no other findings were found on sigmoidoscopy.

### Treatment Pathway

**Figure 1-Mast Cell Inhibitors**

I. Uninhibited mast cell degranulation
II. Decreased mast cell recruitment due to dampened immunologic response from steroids
III. Mast cell stabilizers prevent release of granules
IV. Inhibition of mast cell mediators after degranulation

#### Steroids

- Prednisone
- Budesonide

#### H1 blockers

- Cetirizine
- Loratadine

#### H2 blockers

- Lansoprazole

#### Leukotriene antagonists

- Montelukast

#### Cromolyn

- Quercetin

#### Mast cell stabilizers

- Ketotifen
- Cromolyn
- Steroids

### Discussion

- Mast cells in the GI tract can cause intractable vomiting and diffuse small and large bowel inflammation, as demonstrated in this case.
- Activation of mast cells may be the primary cause of GI pathology, or secondary to another process.
- Within the lamina propria mast cells are stained with CD117 and found with an average of 13 cells/hpf.
- No specific criteria exists at this time for mast cell gastroenteritis, but one study suggests a cutoff of 20 cells/hpf within the lamina propria.
- Mast commonly proposed cause of MCG is food allergen.
- Treatments suggested for GI mast cell activation include H2 antihistamines, cromolyn, antileukotriene drugs, and budesonide.

### References