Diaphragm Disease: A Crohn's Mimicker?

Abigail Schubach, MD, MS, Natalie C. Penn, MD, Rabih Salloum, MD, Yanseng Hao, MD, PhD, Maisa Abdalla, MD, MPH

University of Rochester Medical Center, Rochester, NY

Introduction:

- Diaphragm disease of the small bowel is a rare disease typically associated with NSAID use[1,2].
- The disease is characterized by multiple strictures appearing as 'pinhole' sized lumens caused by mucosal thickening.
- Often the condition goes misdiagnosed until the lumen is visualized, with clinical presentations initially consisting of recurrent bowel obstructions, anemia and/or protein loosing enteropathy[3,4].

Case Description:

- A 50-year-old male with a history of hypertension, incarceration, and prior recurrent small bowel obstructions and prior exploratory laparotomy with lysis of adhesions, presented with abdominal pain, nausea, vomiting and constipation.
- A year prior to presentation, the patient was hospitalized for similar symptoms, with colonoscopy demonstrating a fibrotic stricture at the ileocecal valve, which was only able to be traversed after balloon dilation. The mucosa appeared endoscopically normal, however, biopsies of the terminal ileum demonstrated chronic active focal ileitis, suggestive of Crohn's disease.
- Patient then experienced two more hospitalizations for small bowel obstruction, which resolved with supportive care and steroid administration.
- During this hospitalization, CT scan demonstrated small bowel obstruction with several transition points in the mid small bowel (Figure 1).

Disease Course Before Hospital Presentation

2 years prior to presentation
Reported exploratory laparotomy with lysis of adhesions for small bowel obstruction.

7 months prior to presentation-
Admitted for medical management of small bowel obstruction, discharged after four days

6 months prior to presentation-
Colonoscopy demonstrated focal fibrotic stricture at the IC valve, which was traversed after balloon dilation. The mucosa of the colon and terminal ileum appeared normal but biopsies showed chronic active ileitis.

5 months prior to presentation-
CT enterography with thickening and enhancement of terminal ileum, Crohn's disease suspected.

3 months prior to presentation-
Seen in emergency department with small bowel obstruction, discharged home same day as patient responded well to PO trial

1 month prior to presentation-
Hospitalized for recurrent small bowel obstruction, which responded to IV steroids, discharged with PO steroid taper. Plan to start infliximab in outpatient setting.

Case Continued:

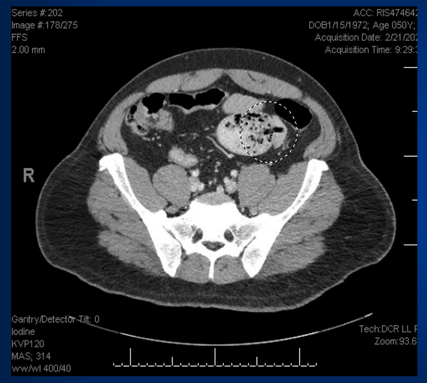

Figure 1: CT abdomen and pelvis with contrast demonstrated multifocal strictures throughout the small bowel.

- IV corticosteroids were started, and nasogastric tube was placed with improvement. He adamantly denied any current or past prolonged NSAID use.
- It was decided that patient would benefit from a surgical small bowel exploration.
- Several strictures were seen in the proximal small bowel, and therefore proximal and distal resection of jejunum with a primary end-to-end enteroenterostomy anastomosis was performed.

Case Resolution:

Histopathological evaluation of the resected specimen showed small bowel with circumferential membranes with submucosal neuromuscular and vascular hyperplasia, fibrosis, displaced mucosal glands, and associated mucosal active and chronic ileitis with ulceration, compatible with a diagnosis of diaphragm disease.

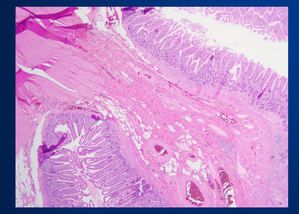

Figure 2: H&E stain, submucosal fibrosis and neuromuscular and vascular hyperplasia.

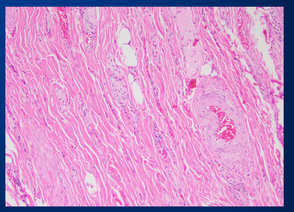

Figure 3: H&E 10 x submucosal fibrosis and neuromuscular and vascular hyperplasia.

Conclusions:

- To our knowledge, this is only the second reported case of diaphragm disease without a reported history of NSAID use[5].
- Diaphragm disease can often clinically mimic Crohn's disease, however, they are histologically distinct in that diaphragm disease is devoid of typical Crohn's markers such as full-thickness bowel wall involvement, scarring of the muscularis propria, transmural lymphoid aggregates, transmural inflammation, or granuloma.

References:

1. Lang JPrice ABLevi AJBurke MGumpel JMBjarnason I Diaphragm disease: pathology of disease of the small intestine induced by non-steroidal anti-inflammatory drugs. *J Clin Pathol* 1988;41516- 526
2. Bjarnason IPrice ABZanelli G et al. Clinicopathological features of nonsteroidal antiinflammatory drug-induced small intestinal strictures. *Gastroenterology* 1988;941070- 1074
3. Smith JAPineau BC Endoscopic therapy of NSAID-induced colonic diaphragm disease: two cases and a review of published reports. *Gastrointest Endosc* 2000;52120- 125
4. Onwudike MSundaresan MMelville DWood JJ Diaphragm disease of the small bowel: a case report and literature review. *Dig Surg*
5. Santolaria S, Cabezali R, Ortego J, Castiella T, Salinas JC, Lanas A. Diaphragm disease of the small bowel: a case without apparent nonsteroidal antiinflammatory drug use. J Clin Gastroenterol. 2001;**32**:344–346.