Chronic Intestinal Obstruction in Surgically Treated Hirschsprung's Disease: A Rare Presentation

Robin David, MD[1], Kajali Mishra, MD[2], Keith Bruninga, MD[3]
[1]Department of Internal Medicine, [2]Division of Nutrition and Gastroenterology
Loyola University Medical Center, Maywood, IL
[3]Department of Gastroenterology, Edward Hines, Jr. VA Hospital, Hines, IL

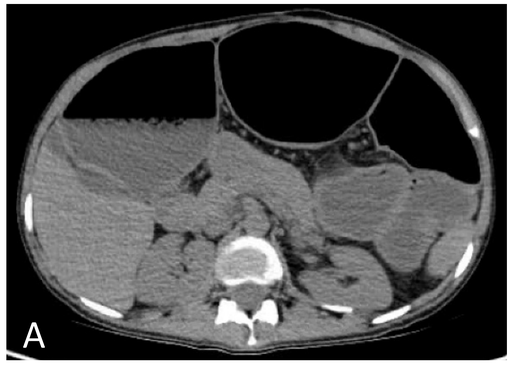
Introduction

- Hirschsprung's Disease (HD) is exceedingly rare and represents a failure of ganglion cell migration to the gut.
- Longitudinal studies examining long-term outcomes of surgically treated HD are extremely limited.
- We discuss a patient who underwent surgical treatment for HD as an infant and presented in adulthood with severe abdominal pain and weight loss.

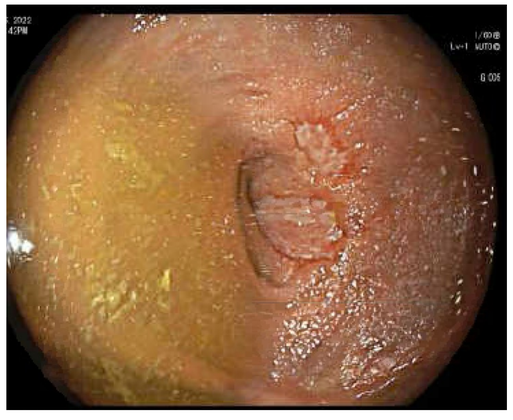
Case Presentation

- A 34-year-old female presented with severe abdominal pain and 15-pound weight loss over 1 month. She continued to pass gas; bowel movements were at baseline.
- Past medical history: HD that involved the entire colon diagnosed at 6 months which was treated with the Duhamel procedure at age 1.
- Her postoperative course was complicated by recurrent small bowel obstruction (SBO) requiring multiple surgeries for adhesiolysis.
- Physical exam revealed mild distention and tenderness in the periumbilical region.

Figures



CT demonstrating areas of narrowing and marked dilation proximal to the anastomosis



Endoscopic view of the ileum demonstrating ulceration and stenosis at the anastomosis site

Hospital Course

Imaging: CT showed marked small bowel dilation proximal to the anastomosis, multiple areas of focal narrowing, and significant stool burden (Figure A).

Studies: Manometry revealed impaired recto-anal inhibitory reflex possibly due to residual HD and Type 1 dyssynergic defecation.

Endoscopy: nonobstructive ileal stenosis proximal to the anastomosis and ileal ulcerations with biopsies showing acute and chronic inflammation with cryptitis (Figure B).

Interventions:
1) Empiric treatment for small intestinal bacterial overgrowth and Hirschsprung-associated enterocolitis were initiated without improvement
2) Trialed on mesalamine with no benefit.
3) Started on prucalopride with some improvement of her abdominal pain.

Conclusion

- The curative treatment for HD is surgical resection.
- Limited studies on long-term outcomes of these patients have demonstrated that constipation or diarrhea are common, and obstructive symptoms or dyssynergia are rare.
- Potential reasons for the complications presented in this case include mechanical obstruction from strictures and possible residual aganglionosis from insufficient resection.
- Additionally, prolonged bowel dilation may lead to impaired motility, evacuation difficulties, chronic dilation, and ischemic changes.
- There is a need for longitudinal studies at dedicated colorectal centers to provide effective transition of care from childhood to adulthood.
- A formalized approach to the management of postoperative complications of HD in adulthood is needed, especially as these issues closely relate to functional status and quality of life.