

Hirschprung's Disease: A Rare Cause of Intestinal Obstruction in Adults

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Abstract

Hirschprung's disease (aganglionosis) in the adult is a rare condition but needs to be considered as a diagnosis in any case of intractable constipation. We report a case of twenty-year-old woman with adult hirschprung's disease, presenting as intestinal obstruction. Diagnosis is discussed and operative management evaluated.

Key Words

Adult Hirschprung's Disease, Intestinal Obstruction, Constipation

Introduction

Presentation and recognition of Hirschprung's Disease in adulthood is unusual (1). We report here an adult patient with this disease. Clinical, radiological and operative findings of this case are presented.

Case Report

A twenty-year-old female presented with features of large bowel obstruction. She had a history of constipation since three years, which was partially relieved by enemas and laxatives. On examination, abdomen was distended and bowel sounds absent. Plain x-ray of the abdomen showed a dilated left colon. Barium enema demonstrated a dilated sigmoid colon and rectum proximal to a narrowed segment (Fig. 1). Laparotomy revealed a dilated sigmoid colon and upper rectum. A right transverse loop colostomy was done. A full thickness rectal biopsy was taken that showed absence of ganglia on histopathology (Fig. 2).

Six weeks later, a Duhamel's operation was done. Colostomy was subsequently closed. The patient is on a regular follow up for one year now and has normal bowel habits.

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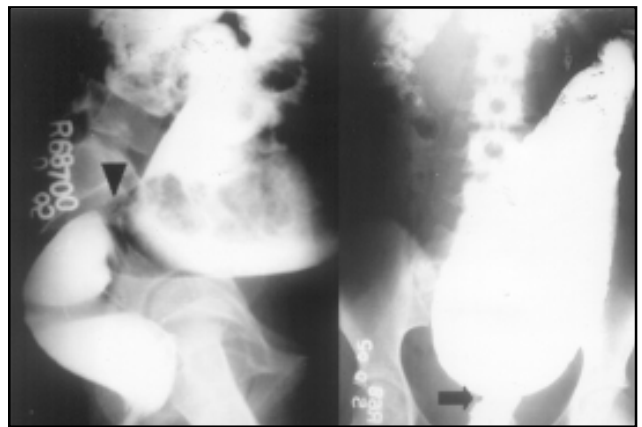


Fig. 1 Barium enema showing a dilated sigmoid colon and rectum proximal to a narrowed segment (arrow). left: lateral view; right: AP view.

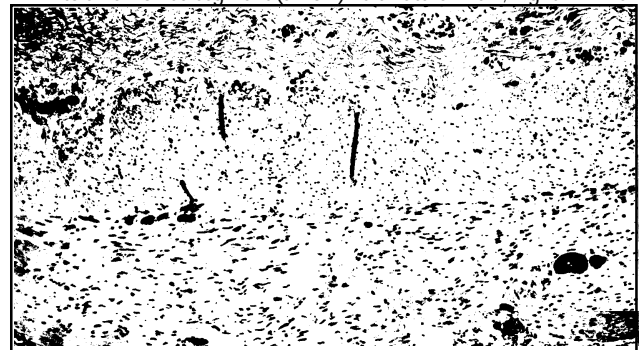


Fig. 2 Photomicrograph of rectal biopsy showing absence of ganglia in the myenteric plexus (H & E x 40).

Discussion

Hirschprung's Disease is a congenital aganglionosis of the submucosal and myenteric neural plexuses principally affecting the rectosigmoid or rectal segments of varying lengths (2). It occurs in approximately 1:5000 live births with a preponderance in males (2). Adult Hirschprung's Disease, usually defined as a diagnosis after 10 years of age is rare and very few cases are reported in literature (4). It should be considered in the differential diagnosis of chronic constipation, especially when the history goes back to infancy (4).

Untreated Hirschprung's Disease leads to complications that include intestinal obstruction, enterocolitis, perforation, dehydration and severe malnutrition (3).

Diagnostic studies include barium enema, rectal biopsy and anorectal manometry (1). Barium enema findings include proximal megacolon and anorectal narrowing (5). Biopsies from distal bowel reveal absence of ganglion cells and increased acetylcholinesterase activity in mucosal nerve fibers. Anorectal manometry would show inability of internal sphincter to relax on distension of the rectum (1).

Aim of the surgical treatment is to remove the aganglionic segment and non-functioning megacolon. Two treatment approaches for Hirschprung's Disease have evolved. The first involves anastomosis of ganglion-containing bowel to the anus. The Swenson, Duhamel and Soave procedures are based on this principle. The other anorectal myectomy, involves division of the internal sphincter and muscle of a distal aganglionic segment of rectum (1). A modification of Duhamel operation in which gastrointestinal anastomosis (GIA) staplers are used to appose the posterior wall of rectum and anterior wall of

the pulled through colon is the most commonly used operation (4). In infants staged procedures using a protective colostomy is the standard procedure. In adults, staged procedures are preferred. Limited pelvic dissection required in Duhamel operation and retention of most of the rectum, albeit aganglionic, results in a low morbidity operation with good functional results (3).

Soave's endorectal pull through and Swenson's rectogonoidectomy are the other most commonly used procedures (2). Only a small number of Soave and Swenson procedures have been reported in adults. Although functional results in long term are reported to be satisfactory, the incidence of septic complications is high and neither of these procedures leave a compliant reservoir (3). Anorectal myectomy has been used in treatment of short segment disease, but has not been found to be uniformly successful (1).

To conclude, adult Hirschprung's disease must always be suspected in the context of chronic constipation. Surgery is recommended as soon as the diagnosis is made.

References

1. Wu JS, Schoetz DJ Jr, Collier JA, Veidenheimer MC. Treatment of Hirschprung's disease in the adult. Report of five cases. *Dis Colon rectum* 1995; 38(6): 655-59.
2. Luukkonen P, Heikkinen M, Huikuri K, Jarvinen H. Adult Hirschprung's disease: Clinical features and functional outcome after surgery. *Dis Colon Rectum* 1990; 33(1): 65-69.
3. Keating JP, Frizelle FA, Brasch HD. Low anterior resection and colonic J-pouch in the treatment of adult Hirschprung's disease. *ANZ J Surg* 2001; 71: 124-26.
4. Kim CY, Park JG, Park KW, Park KJ, Cho MH, Kim WK. Adult Hirschprung's disease: Results of the Duhamel's procedure. *Int J Colorectal Dis* 1995; 10: 156-60.
5. Mindelzun RE, Hicks SM. Adult Hirschprung's disease: radiographic findings. *Radiology* 1986; 160: 623-25.