

Hirschsprung's Disease in Adult Treated by Coloproctectomy and Colo-Anal Anastomosis

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Abstract

Hirschsprung's disease is a rare pathology. Most cases become manifest during the neonatal period, but in rare instances, the disease is initially diagnosed in adult patients.

We report a case of the 26 years old man who presented a Hirschsprung disease. Developed severe intestinal obstruction and presented to the emergency department with significant abdominal distension. A computed tomographic scan confirmed significant fecal loading of the entire colon and rectum. The diagnosis was established through peroperatively biopsy. A rectal biopsy showing aganglionic anorectum. The treatment consist in a coloproctectomy with coloanal anastomosis.

The postoperative course was uneventful with complete resolution of the symptoms. Hirschsprung's disease should be considered in adults who have long-standing and refractory constipation.

Keywords: Hirschsprung's disease; Adult; Surgery

Introduction

Hirschsprung's disease or congenital aganglionic megacolon, occurs in approximately 1 in every 5000 live births and is about 4 times more frequent in males than in females. Hirschsprung disease is the most frequent aetiology of intestinal occlusion in child. This disease is characterized by lack of ganglionic cells in the submucous membrane and in the myenteric plexuses of the digestive tract in its distal part.

Observation

A 26-year-old man suffering from recurrent abdominal pain and constipation distention over a period of 6 years was admitted to our institution for increasing abdominal pain, distension, and symptoms of colonic obstruction by for a 4-day period for colonic obstruction. He is complaining of many years of constipation. He underwent emergency laparotomy. The left colon and the rectum are markedly dilated (Figure 1); a diverting loop colostomy was performed associated with a biopsy of the rectum and about 7 kg of feces was evacuated through the colostomy. Full-thickness rectal biopsies demonstrated a paucity of ganglion cells, suggestive of HD. colonoscopy, and laboratory studies were normal. After few weeks of examination, we found a significant abdominal distension and tympany to palpation. A plain x-ray film of the abdomen revealed extensive fecal retention throughout the colon. Computed tomographic scan in the axial plane through the abdomen confirmed the severe colonic dilatation along with extensive fecal loading and generalized colonic wall thickening (Figure 2). The rectum was markedly dilated and there was no obstructive lesion identified. Another rectal manometry that revealed a lack of normal rectoanal inhibitory reflex. Sigmoidoscopy with biopsy of rectum and sigmoid colon showed no ganglion cells. Colonoscopy through the colostomy and biopsy was normal.

He underwent a total proctectomy with sigmoidectomy and colo-anal anastomosis between healthy colon and anal mucosa with diverting loop ileostomy. Histological staining revealed absence of ganglion cells in the myenteric plexus in the narrow segment consistent with the diagnosis of adult HD. The post operative course was uneventful. Her diverting ileostomy was taken down three months later. Actually two years after the intervention he has been well with complete resolution of symptoms and absolutely normal continence and sexual potency and good quality of life.

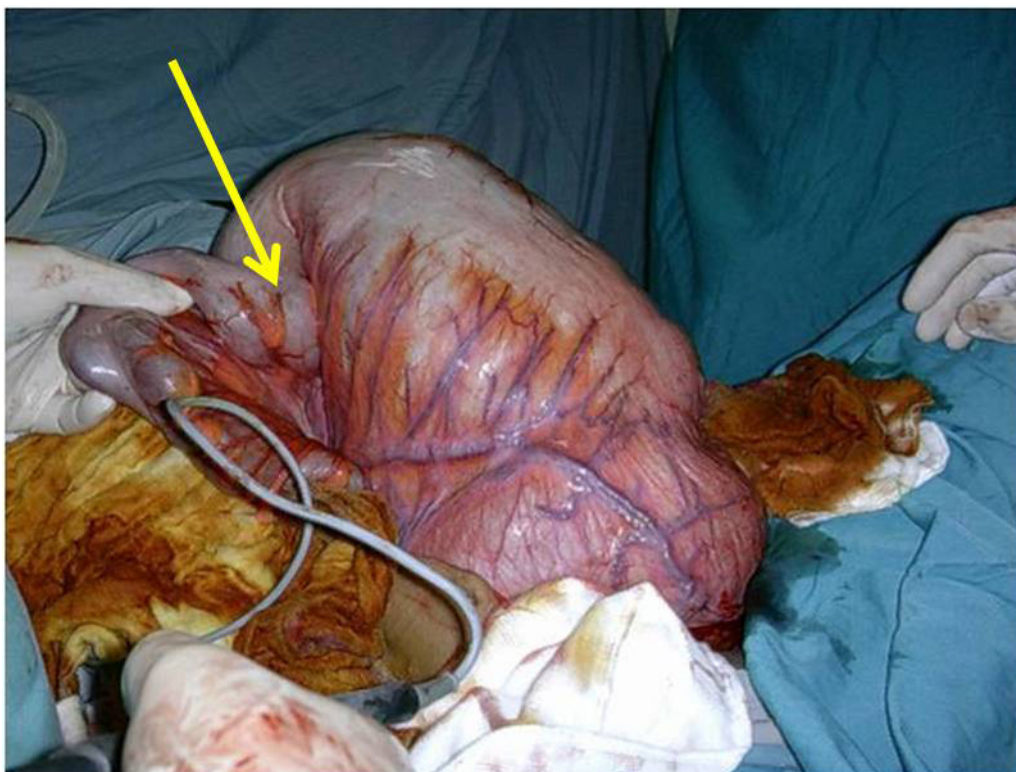


Figure 1: Per operatively view: The descending colon transverse and right colon is markedly dilated and sigmoid rectum were narrow

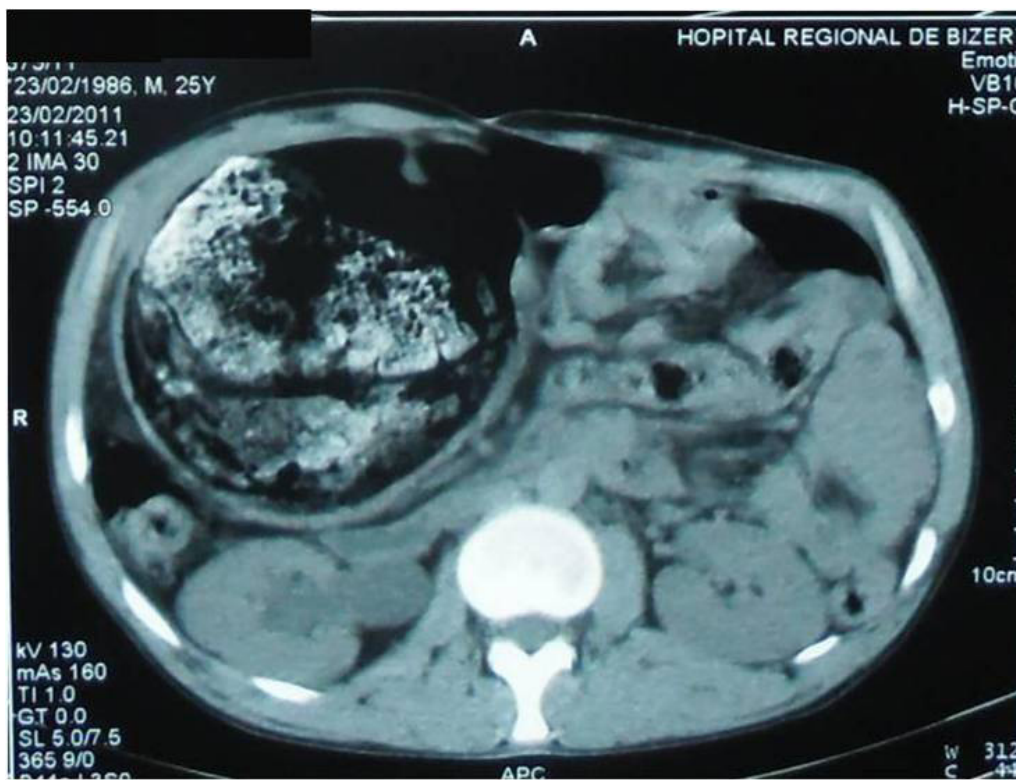


Figure 2: CT abdominal scan: severe colonic dilatation along with extensive fecal loading and generalized colonic wall thickening

Discussion

Congenital aganglionic megacolon was first described more than 100 years ago by Harald Hirschsprung, who published his classic description of the disease in 1888 [1]. The term adult HDQ has been arbitrarily applied by some investigators to cases in which the patient is older than 10 years when the diagnosis is established [2,3], whereas others have defined adult HD as cases in which

the diagnosis was made after age 18 or 19 years [4]. Ninety-four percent of these cases are diagnosed before the patient is aged 5 years. In rare cases of these disorders, the patient may not receive a diagnosis until he or she reaches adulthood [5,6]. In our case the symptoms appear in adulthood. Nearly 300 cases with at least some features of adult or adolescent HD have been documented, some of which were diagnosed by rectal biopsy or resection. Fairgrieve [2,7] documented HD in 7 men whose ages varied from 17 to 34 years.

The primary defect in adult HD is identical to that seen in infancy or childhood and is characterized by the total absence of intramural ganglion cells of the submucosal (Meissner) and myenteric (Auerbach) plexuses in the affected segment of the bowel.

The typical adult patient with HD has a history of longstanding constipation since infancy or early childhood; the male to female ratio is approximately 4:1. Patient age ranges from 10 to 73 years, and the average age is 24.1 years. Half of the patients are younger than 30 years [2-8,9]. Other symptoms include abdominal discomfort, distension, and abdominal pain. Physical examination frequently reveals palpable fecal masses [10,11].

The treatment of this affection is exclusively surgical. The surgical procedures developed to treat the disease in children have been applied to adults. Many procedures were reported in the literature: Swenson abdominoperineal pull-through, Duhamel retrorectal pull-through, Soave endorectal pull-through, Lynn myectomy, LAR combined with myectomy, LAR with colectomy (State procedure). In our case we have used a coloproctectomy with a coloanal anastomosis [2,12-15].

The Duhamel procedure appeared to be associated with a lower rate of major postoperative complications than the other procedures, and the rate of good long-term results was higher [12]. Swenson [16] reported their collective results in 282 patients treated with the Swenson procedure, including 71 adults, and noted a 5% incidence of anastomotic leak and 90% rate of excellent long-term results. Lynn procedure for posterior anorectal myectomy was advocated its usefulness as an initial approach or complementary procedure approach to short segment Hirschsprung's disease because of the low morbidity and absence of technical difficulty. The long-term results of a combination of anorectal myectomy and anterior resection showed excellent results with no major complications [13,14,17,18].

All the procedures can now be performed laparoscopically and the results were excellent [13].

Conclusion

Hirschsprung's disease is an uncommon cause of chronic constipation in adults, it should be considered in the differential diagnosis of this entity in similar clinical cases.

Surgical treatment is still the only curative treatment of this disease. Results are excellent in most cases. The quality of life of the patients after surgery was excellent.

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