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Surgical management of adult Hirschsprung's disease by subtotal colectomy: a possible alternative

Suaugusiųjų Hirschprungo ligos chirurginis gydymas subtotaline kolektomija: galima alternatyva

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Adult Hirschsprung's disease (AHD) is a rare and often undiagnosed condition. At present, only a limited number of cases have been reported. We managed a 19-year-old woman with rich history of chronic constipation and abdominal discomfort. Abdominal radiography and barium enema examination demonstrated massively dilated colon (up to 15 cm in diameter). Full-thickness rectal biopsy showed absence of ganglionic cells. AHD was diagnosed and subtotal colectomy with ileorectal anastomosis was performed. At five-year follow up, the patient had significantly improved defecation and quality of life and was extremely satisfied with the operation. AHD should be considered in patients with long-standing and refractory constipation with megacolon. We advocate a subtotal colectomy with ileorectal anastomosis as one of the possible surgical alternatives in such young patients.

Key words: adult Hirschsprung's disease, constipation, megacolon, aganglionosis

Suaugusiųjų Hirschprungo liga yra reta ir sunkiai diagnozuojama vidurių užkietėjimo priežastis. Straipsnyje aprašoma 19 metų mūsų gydytos moters istorija. Pacientė kreipėsi dėl visą gyvenimą trunkančio vidurių užkietėjimo ir pilvo skausmų. Radiologinio ir endoskopinio tyrimo duomenimis, tiesioji žarna normalaus pločio, o gaubtinė žarna labai plati ir pilna turinio. Viso storio tiesiosios žarnos biopsija patvirtino aganglinę tiesiąją žarną. Pacientė buvo operuota ir jai atlikta subtotalinė kolektomija ir ileorektostomija. Penkerių metų stebėjimas po operacijos parodė, kad pacientė gyvena kokybiškai. Manome, kad subtotalinė kolektomija ir ileorektostomija yra viena iš jaunų pacientų chirurginio gydymo alternatyvų.

Reikšminiai žodžiai: suaugusiųjų Hirschprungo liga, vidurių užkietėjimas, didelė gaubtinė žarna, agangliozė

Introduction

Hirschsprung's disease (HD) is characterized by congenital absence of ganglion cells of the submucosal (Meissner's) and myenteric (Auerbach's) neural plexuses in the distal bowel, beginning at the internal anal sphincter and extending proximally to varying distance [1, 2]. Most often HD is diagnosed in the newborns and infants (usually before the age of five years), but occasionally, especially if the symptoms are "mild", HD is diagnosed later [1–3]. An affected colon produces mechanical obstruction because of failure to relax during peristalsis [1–3]. A dozen of operation types have been described in the literature to correct this condition, usually eliminating or avoiding an affected (aganglionic) segment. We report the representative case of AHD successful surgical management by subtotal colectomy.

Results

A 19 year-old woman was admitted to the Department of Surgery complaining of generalized abdominal pain, nausea, constipation and fever (39 °C).

She had a significant history of chronic constipation from childhood (bowel movements once in 3–4 days, abdominal discomfort, sometimes pain and distension after meals). Previously she undergone laparotomy because of suspected perforated viscus in the regional hospital – grossly dilated colon full of fecaliths was found and they were removed through colotomy. Patient's brother had undergone operation in the children's hospital for diagnosed HD.

On examination large masses were palpable on both sides of the abdomen. Plain abdominal radiography showed dilated colon (up to 15 cm in diameter on the right) with large amount of feces and gas with elevated both hemi-diaphragms up to the third intercostal space (Fig. 1). Single contrast enema and retrograde proctoscopy visualized dilated proximal colon with large amount of content and non-dilated, elastic, well contracted after defecation rectum and distal segments of sigmoid colon (Fig. 2). An attempt to clean out the colon with enemas was unsuccessful; antegrade colonic lavage was not attempted.

HD was suspected and full-thickness lower rectal biopsy was performed under anesthesia – no ganglionic cells were present in the specimen and the diagnosis was confirmed.

The patient underwent laparotomy. She was found to have grossly dilated stool-filled colon from the right side up to the mid-sigmoid, where the colon appeared normal. Small bowel appeared normal. Subtotal colectomy was performed, removing 6.5 kg weighing colon

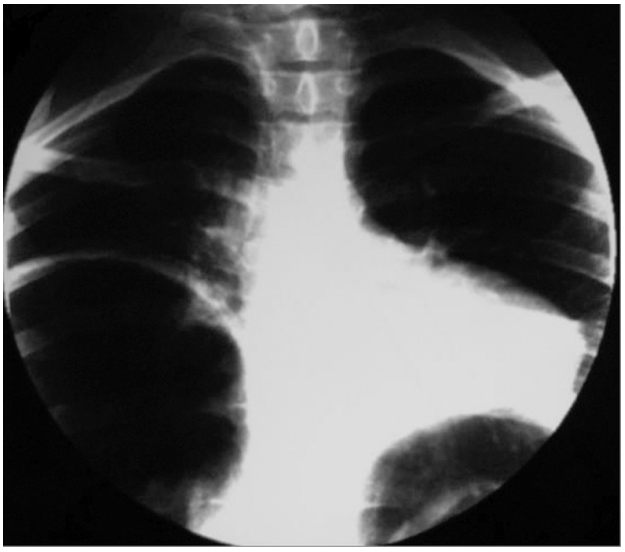

Figure 1. Chest X-ray of the patient demonstrating elevated both hemidiaphragms

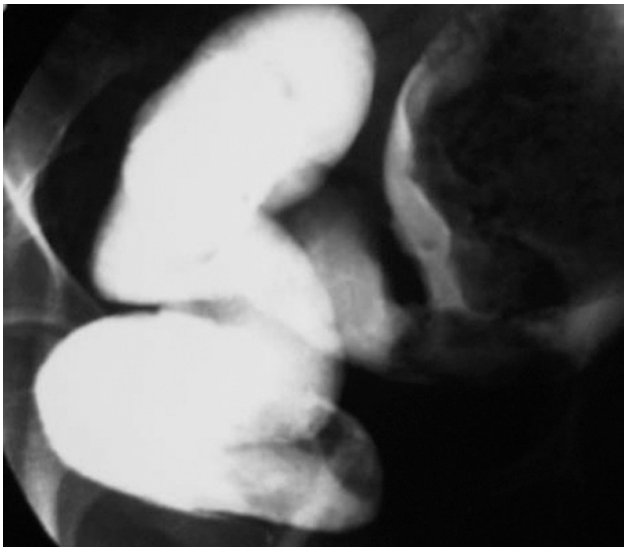

Figure 2. Single contrast barium enema demonstrating normal rectum and grossly dilated stool-filled sigmoid colon

up to the level of sacral promontory. Ileorectal anastomosis was performed. There were no complications during post-operative period. The patient was having 3 to 4 bowel movements in the early post-operative period; she was discharged from hospital on day 6 after surgery.

The patient was personally interviewed (T.P.) 1 year, 3 years and 5 years after the operation. She is extremely satisfied with operation, with significant improvement in the quality of life from before the operation. She has 3 to 4 bowel movements per day and no bowel movements overnight. She maintains perfect continence.

Discussion

HD is usually diagnosed till the age of 5 years (about 95%), mostly in the newborn period [1–3]. The term "adult HD" is controversial, because most often it's defined as disease in patients older than 10 years [4, 5] although some other authors think that the term can be used only when patients are 18 or 19 years old [1, 6]. The incidence of adult HD is unknown, but could represent 2% of chronic refractory constipation [3, 7].

In rare cases patients with Hirschsprung's disease do not present for medical care until adulthood, especially if they have "milder" disease. There is a hypothesis that the disease is "mild" because the proximal colon is hypertrophied to compensate for the distal obstructed aganglionic rectum [1]. The disease is diagnosed only when a dilatation and functional decompensation of the innervated colon occurs and the constipation develops. Symptoms are also masked by using laxatives and enemas [1–2]. There have been some cases when presentation of the disease was in adult life [5, 8]. So it is still unclear whether adult HD is due to failure of diagnosis during childhood years or whether it is due to the late presentation of disease [2, 9].

Surgical options in adult Hirschprung's disease have recently been extensively reviewed by Doodnath and Puri [2]. They found 490 reported cases from 1950 to 2009. The mainstay of treatment in the reported cases is a pull-through surgery, with Duhamel procedure being performed in 231 (47.2%) patients; 49 (10%) patients had the Swenson procedure, 40 (8.2%) patients had the Soave procedure.

Colectomy was performed in only 10 cases (2%). In our case the procedure was chosen because the colon was extremely dilated, it was not possible to evacuate the stool by mechanical cleansing and the patient preferred to avoid even a temporary stoma. Excellent clinical outcome after three years justifies the choice of operation. We think that high-pressure small bowel is able to propel liquid contents through the rectum which in turn, is able to empty. Possibly, the level of rectal dysfunction is less in HD patients who reach adult age than in childhood HD cases, where rectal emptying is more dysfunctional.

Conclusion

AHD must always be considered in patients with megacolon who have had severe chronic constipation since birth. Subtotal colectomy with ileorectal anastomosis is one of the possible surgical alternatives in such patients.

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