Adult Hirschsprung’s Disease: Report of Two Cases

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Abstract

Adult Hirschsprung’s disease is rarely encountered in clinical practice. Surgery in these patients should preferably be staged in the form of an initial decompression colostomy and subsequent definitive operation. Two cases recently seen by us are presented, with a brief review of the subject.

Key words: Adult Hirschsprung’s disease, adult megacolon

Introduction

It is believed that only about 10% of patients with Hirschsprung’s disease remain undiagnosed during the neonatal period and become symptomatic in adult age. Individual experience with adult Hirschsprung’s disease is limited. In a review of 23 years’ experience at St Mark’s Hospital, only 26 proved cases were seen. We report two cases of adult Hirschsprung’s disease seen during the last two years (1985-86).

Case Reports

Case 1: A 16-year-old girl presented with a history of long standing constipation and a gradually increasing abdominal lump. Examination revealed an average built patient with a mobile, firm mass in the lower abdomen. The routine investigations were normal. With a preoperative diagnosis of an ovarian cyst a laparotomy was done. The sigmoid colon and upper rectum were greatly dilated and thinned. Both ovaries were normal. A right transverse colostomy was done. Postoperative colonogram (Fig 1) showed a narrowed rectum with proximal dilatation of the sigmoid colon. Pull thickness transanal rectal biopsy revealed absence of ganglion cells, confirming the diagnosis of Hirschsprung’s disease. Four weeks after the initial surgery, definitive surgery in the form of anterior resection with colorectal anastomosis was done. She recovered uneventfully. The colostomy was closed six weeks later and the patient is doing well on follow up.

Case 2: An 18-year-old male presented with constipation since childhood and recurrent episodes of large gut obstruction. On examination the abdomen was distended. There was no palpable lump. Plain X-ray of the abdomen showed a dilated left colon. Laparotomy revealed a dilated sigmoid colon and upper rectum. A right transverse colostomy was done. A full thickness rectal biopsy showed absence of ganglia on histopathological examination. Colonogram showed a dilated sigmoid colon and rectum proximal to a narrowed segment (Fig 2). Four weeks later, a modified Duhamel’s operation was done after adequate gut preparation. The colostomy was subsequently closed. The patient is on regular follow up and is well.

Discussion

Megacolon is an uncommon condition in which the bowel is persistently of increased diameter and is always associated with long-standing constipation. The diagnosis of Hirschsprung’s disease rests on the demonstration of aganglionosis in a full thickness...
rectal biopsy. This biopsy should be taken from just above the anorectal ring as the distribution of ganglia above this level is variable. Suction biopsy has also been reported to give good results. Barium enema characteristically shows a narrowed segment with proximal dilated colon. The narrowed portion is best seen on lateral view, which should be taken after removal of the enema catheter tip which can cause undesirable anatomic distortion of the diseased segment.

Operative procedures applicable for adult megacolon are the same as for the neonatal variety. Good results are achieved with these procedures depending on the experience and familiarity of the surgeon with a particular operation. Swenson's operation gives good long-term results but also has a higher rate of complications. There is a risk of impotence in males because of the pelvic dissection involved in this operation. Daukamel's operation and its modifications are probably the most suitable for adult megacolon. The procedure has no risk of impotence as there is no pelvic dissection involved. The operation also compensates for any discrepancy in size of the lumen between the distal and proximal rectal segments. The difference in size between the dilated and narrowed bowel is much more marked in adult megacolon as compared to the pediatric variety. Adult megacolon thus ideally needs a preliminary decompression colostomy, one stage definitive operation being both difficult and risky. In patients where the colon is thinned because of prolonged distension, as seen in our first case, a subtotal colectomy with colorectal anastomosis has been recommended.11

References
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