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Case Report

Total Colonic Aganglionosis, a Rare Case of Hirschsprung's Disease

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Abstract

Introduction: The condition, known as total colonic aganglionosis (TCA), is a rare variant of Hirschsprung's (HSCR) disease, which affects the entire colon with aganglionosis. Incontinence poses the primary challenge after a total colectomy. This case report highlights an unusual instance of TCA that resulted in a total colectomy for a 15-month-old patient, employing a modified technique that enhanced fecal continence.

Case Report and Discussion: A 9-month-old male patient was referred to our department with a diagnosis of HSCR for further treatment. From the patient's history, an inability to feed, bilious vomiting, and failure to expel meconium were reported on the 1st day of life. On the 21st day of life, exploratory laparotomy was performed, revealing a stenosis 6 - 7 cm from the ileocecal valve. Full-thickness biopsies from the ileum, transverse, and descending colon were sent to pathology. Appendectomy was performed, and the appendix was also sent for pathology. A loop ileostomy was created 10 cm from the ileocecal valve. The pathology report indicated a total lack of ganglion cells in each segment. At the age of 15 months, the primary surgical restoration was performed through a total colectomy with ileorectal anastomosis, retaining a small segment of aganglionic section to prevent incontinence. Prophylactic loop ileostomy reconstruction took place after rapid biopsies were sent during surgery. Ileostomy closure occurred after 3 months.

Conclusion: In such cases of HSCR urgent initial assessment with ileostomy is required. Keeping a small segment of aganglionic bowel could be a solution to the problem of incontinence.

Keywords: *Total Colonic Aganglionosis (TCA); Hirschsprung's Disease (HSCR); Quality of Life (QoL)*

Abbreviations

TCA: Total Colonic Aganglionosis; HSCR: Hirschsprung's Disease; QoL: Quality of Life; IAA: Ileoanal Anastomosis

Introduction

The condition known as total colonic aganglionosis (TCA) is a rare variant of Hirschsprung's disease (HSCR) characterized by absence of ganglion cells throughout the entire colon. Quality of life (QoL) is a critical outcome for patients following surgical intervention.

Constipation, fecal incontinence, and the presence of diarrhea are three of the most significant factors in studies comparing the quality of life among patients with TCA treated using the Duhamel, Suave or a combination of surgical techniques, based on international quality of life questionnaires for patients with HSCR.

Case Presentation

Male, 9 months old, referred with a diagnosis of total colonic aganglionosis (TCA) for further treatment.

Patient history: Feeding difficulty, bilious vomiting and no passage of meconium on the 1st day of life. On the 21st day of life exploratory laparotomy performed revealing a stricture 6 - 7 cm from the ileocecal valve. Full-thickness biopsies were taken from: The ileum, the transverse colon, the descending colon and the appendix. All the segments were sent for histopathological examination. A loop ileostomy was performed 10 cm from the ileocecal valve. The histopathological report showed a complete absence of ganglion cells in all sections.

The main surgical procedure, consisting of a total colectomy (ileorectal anastomosis), preserving a small segment of aganglionic bowel to prevent fecal incontinence, was performed at the age of 15 months. Continence gradually improved until the age of 18 months, when the ileostomy closure was performed.

Results

One year postoperatively, the toddler demonstrates satisfactory fecal continence without episodes of enterocolitis so far, diarrhea or perianal irritation. The number of bowel movements ranges between 4 and 6 per day. The patient's quality of life which was our primary objective, exceeded our expectations.

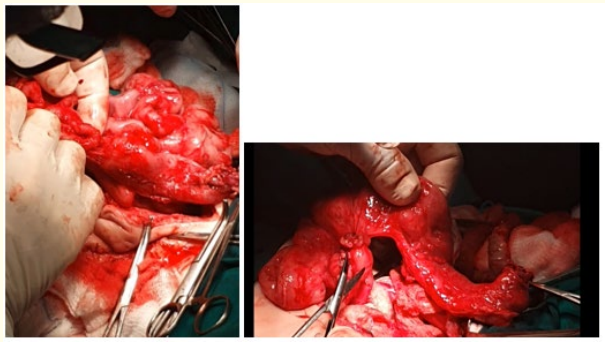

Image 1 and 2: *Stages of the total colectomy.*

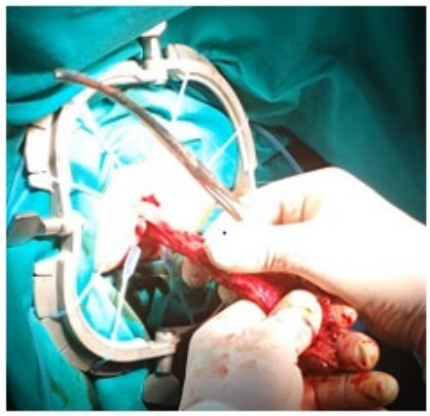

Image 3: *Transanal end stage for the anastomosis.*

Discussion

The fundamental idea behind treating Hirschsprung's condition is to restore intestinal continuity by resecting the aganglionic segment. The aganglionic zone is lengthy, and excision results in a decrease of absorptive surface, making TCA challenging to control with or without small bowel involvement. The external diversion, known as a caecostomy or ileostomy, leads in dangerous outcomes because of substantial diversion-related fluid and electrolyte loss and nutritional issues caused by the small intestine's variable length loss. It is very challenging to carry out a one-stage total colectomy on a newborn [1]. While there are reports of success with each technique, none has been shown to be better than the others and all have limitations. The long-term results of these individuals showed a significant morbidity related with the adoption of large aganglionic bowel lengths to optimize fluid absorption. In order to control ileostomy diarrhea, Shermata and Meller suggested a 10 cm ileocolostomy using the right colon [2].

The vast majority of reconstructions in the Nordic region (76%) involved IAA, either with or without a J pouch. Similar to other reports on surgical methods for Hirschsprung's disease [3,4]. no appreciable variations between the various surgical procedures were observed in the early or long-term complications. TCA management at tertiary centers is deemed appropriate and safe due to the low rate of major early complications requiring surgical intervention (Clavien-Dindo grade IIIb), the low need for repeat reconstruction due to residual aganglionosis (approximately 9%), and the lack of life-threatening complications [5-7]. Early problems were prevented when a protective stoma was present during reconstruction; patients without a covering stoma experienced all three of the anastomotic leaks that occurred during reconstruction.

Conclusion

After a successful TCA reconstruction, persistent intestinal conditions such as enterocolitis, diarrhea and perianal irritation are still frequent long-term issues. Postoperative follow-up should pay more attention to the dietary and psychosocial status of the condition because it presents lifetime challenges.

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