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Case Report

Creation of Enteral Shortcuts as a Therapeutic Option in Children With Chronic Idiopathic Intestinal Pseudoobstruction

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INTRODUCTION

Chronic idiopathic intestinal pseudoobstruction (CIIP) is a severe motility disorder in which the symptoms and signs of intestinal obstruction occur without any obvious mechanical cause (1). Many patients have a long history of diagnostic procedures and often have undergone multiple surgical procedures before being diagnosed with CIIP (2). There is no established therapeutic regimen. Multidisciplinary management involving prokinetic drugs and home parenteral nutrition seems to be the most appropriate way to care for patients with CIIP (3). The role of surgery in CIIP has been controversial. Decompression stomas have been considered useful (4), whereas removal of parts of the gut is mostly seen as a last option (5). We present the case of a male 15-year-old patient with CIIP in whom side-to-side enteroanastomoses were performed to ease intestinal transport. In combination with home parenteral nutrition, he continuously gained weight after surgery and his quality of life improved markedly.

CASE HISTORY

The male patient first presented at our clinic in December 2004 with a history of recurrent ileus, massive abdominal distension, and weight loss. On admission, he did not tolerate any oral nutrition.

His abdominal problems had begun in the second month of life with chronic constipation and failure to thrive. Continuous weight loss and megacolon led to colostomy at 1 year of age followed by resection of the transverse, descending, and sigmoid colon under the suspected diagnosis of Hirschsprung's disease. However, after surgery he was still suffering from constipation alternating with episodes of severe diarrhea, which led to malnutrition and repeated hospitalization. At 12 years of age, the remainder of the colon was resected and a coecorectal anastomosis was performed. Stools were more fluid afterwards, but bowel movements did not improve significantly. The situation exacerbated at age 15 and the patient consulted our clinic.

On admission, the nearly moribund patient weighed 32 kg and measured 152 cm, both values below the third percentile. His bowel movements were changing from no emptying over several days to massive fluid-to-mushy stools with enteral fluid losses of up to 4 L/day. Sonography and abdominal x-ray revealed massively dilated, thin-walled, small bowel loops with little peristalsis, especially in the proximal parts (Figure 1). Rectal suction biopsy revealed normal ganglia and rectal manometry showed normal relaxation of the internal sphincter. Unexpectedly, endoscopic biopsies from the duodenum showed shortened villi as well as an increased number of intraepithelial lymphocytes, presenting similarly to the histological picture of florid coeliac disease. However, anti-endomysial antibodies and anti-tissue transglutaminase antibodies were negative. The patient also did not have the human leukocyte antigen types usually associated with coeliac disease. Anti-enterocyte antibodies were negative, providing no evidence for autoimmune enteropathy.

Laparotomy was performed and revealed massive intestinal adhesions. The bowel was extremely distended, thin walled, and filled with many liters of fluid. We performed an extensive adhesiolysis and took several full-thickness biopsies. A 20-cm part of the small bowel that appeared stenotic was resected.

Standard haemotoxylin and eosin staining of the full-thickness biopsies showed normal architecture of

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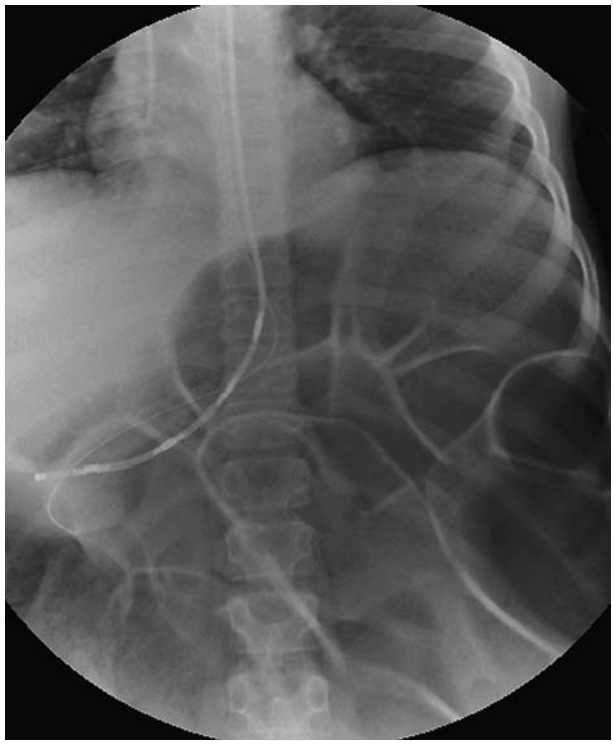

FIG. 1. Fluoroscopy showing massively dilated bowel loops.

the submucosal and the myenteric nerve plexus, but immunostaining for α-smooth muscle actin (α-SMA) showed absent α-SMA immunoreactivity in the circular muscle layer, whereas it was clearly positive in the longitudinal muscle layer and muscularis mucosae as well as around vessels (Figure 2). Acetylcholinesterase activity was markedly increased. Interstitial cells of Cajal showed normal staining and distribution.

After surgery the patient continued to experience massive gastric reflux and did not tolerate oral feedings. Prokinetic therapy with erythromycin did not improve the patient's condition. An upper gastrointestinal contrast study indicated an extrinsic obstruction of the duodenum. This led to repeat laparotomy 10 days after the first laparotomy. During this laparotomy, the suspected duodenal obstruction was attributed to passage problems over the vertebral column due to impaired bowel motility. A side-to-side duodenojejunostomy and 2 additional side-to-side enteroanastomoses between small bowel loops were performed to create intestinal shortcuts to ease intestinal transport.

Initially, the gastric reflux continued to be massive (>2 L/day). Additionally, the patient produced 2 to 3 L of stool every day. Referring to a case report of a patient with CIIP and coeliac disease who had similar gastric reflux, intravenous therapy with prednisolone $2\,mg \cdot kg^{-1} \cdot d^{-1}$ was started (11). Cortisone therapy rapidly decreased the gastric reflux and the patient was able to start eating. However, the daily amount of stool continued to be very high (up to 4–6 L/day). During the following weeks, daily stool volumes decreased gradually to approximately 1500 mL/day.

After almost three months the patient was discharged in good condition into home care with an additional home parenteral nutrition of $30\,kcal \cdot kg^{-1} \cdot d^{-1}$. His body weight at the time of discharge was 37.2 kg. After discharge, the patient's condition remained stable and he developed well. He produced stools up to 5 times per day with changing consistency, which he regulated

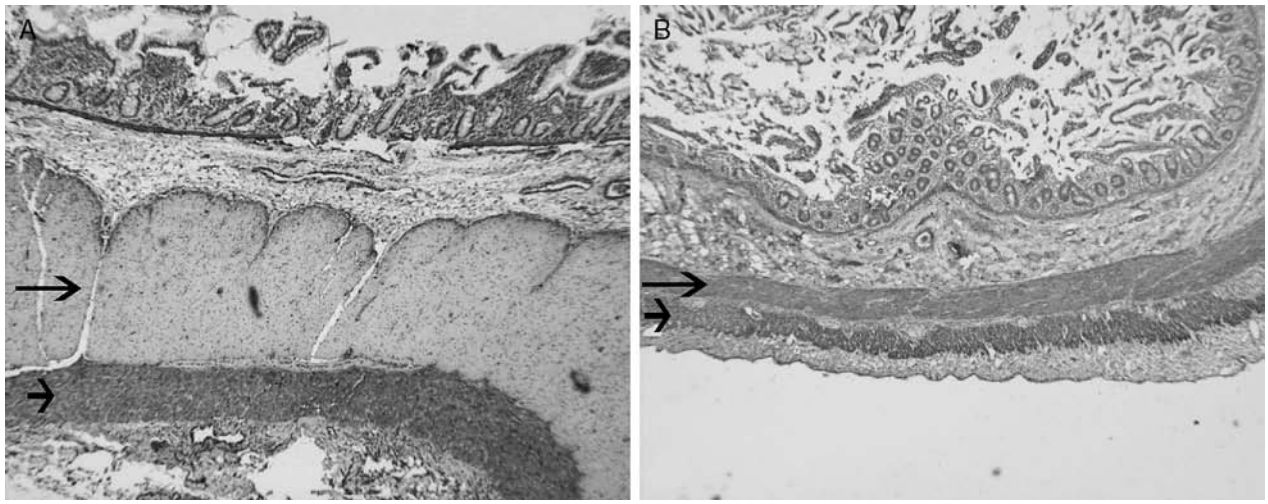

FIG. 2. α-SMA immunohistochemistry of a biopsy of small bowel of the patient (A) showed absent α-SMA staining in the hypertrophic circular muscle layer (long arrow). The longitudinal muscle layer (short arrow) showed normal immunostaining. B, Normal human control: jejunum stained for α-SMA.

individually by eating pureed carrots when he had mushy stools and applying clysters when constipated. He eats with good appetite and is not on a diet. Fourteen months after surgery his weight was 42.2 kg.

DISCUSSION

CIIP is a severe disease that implies a number of diagnostic and therapeutic problems. Our patient was misdiagnosed with Hirschsprung's disease in early childhood, which we were able to exclude by rectal manometry and rectal suction biopsies. Nevertheless, we had difficulty distinguishing CIIP from a severe form of coeliac disease because the histological findings of the duodenum mirrored the picture of florid coeliac disease. Coeliac disease can present with intestinal pseudoobstruction (6). However, other than the histology and the responsiveness of gastric reflux on cortisone therapy, no findings indicated coeliac disease. We set the patient on a gluten-free diet for some months, which he self-terminated without any adverse effects on his condition.

The diagnosis of CIIP was finally confirmed by radiological findings and histopathology. α-SMA deficiency in the jejunal circular muscle layer, as found in our patient, was first described in 1992 (7). α-SMA immunoreactivity was reported to be absent or partially missing in 24% to 58% of patients with CIIP (8). However, the usefulness of α-SMA deficiency as a possible diagnostic marker for myopathic types of CIIP has been questioned recently (2).

Operative therapy is established in the treatment of CIIP ranging from decompression stomas, ''venting'' enterostomy, and loop enterostomy (9,10) to small intestinal transplantation (11). Removing hugely dilated bowel loops and stenotic parts may be indicated to improve intestinal transport (12).

Our patient underwent 2 laparotomies during his stay in our institution. The multiple adhesions that were dissected during the first procedure may have been a cofactor in the exacerbation of his condition before admission. Because this first procedure did not sufficiently improve the condition of the patient and prokinetic therapy also did not help, we decided to perform multiple anastomoses to ease intestinal transport. At the time of this surgery, it was an experimental salvage procedure. In our opinion, the main functional mechanism of this procedure is the creation of an internal pop-off mechanism, which enables bowel contents to bypass atonic bowel segments. Therefore, it must be a prerequisite that at least parts of the bowel are functioning, which was the case in our patient. With this procedure, enterostomies with their associated hygienic, social, and psychological problems have been avoided. Further extended resection of the gut was not necessary, saving the patient from nonreversible loss of intestinal surface. In our patient quality of life was significantly improved by this procedure in combination with home parenteral nutrition. Based on the history of our patient we consider the creation of multiple enteroanastomoses a valuable therapeutic option in some children with CIIP when conservative treatment has failed.

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