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A case of single incision laparoscopic total colectomy for intestinal neuronal dysplasia type B



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ABSTRACT

INTRODUCTION: Intestinal neuronal dysplasia type B (IND-B) is an infrequent disease of the submucosal plexus of intestine manifesting chronic intestinal obstruction or severe chronic constipation. IND is rarely reported in adult patients.

PRESENTATION OF A CASE: The present study reports on the case of a 36 year-old woman suffering from longstanding chronic constipation and who was diagnosed with severe constipation in more than 20 years. Although she began to take a large amount of stimulant laxatives, such as “senna” and “bisacodyl”, constipation symptoms did not improve, she was admitted to our hospital. It was diagnosed with refractory constipation of the medication treatment-resistance, total colectomy with ileorectal anastomosis by single incision laparoscopic surgery (SILS) was performed. The final pathological diagnosis was IND-B.

DISCUSSION: Refractory constipation after medical treatment is often seen in young generation. SILS has benefits of better cosmesis, reduced morbidity, reduced postoperative pain, and reduced length of hospital stay.

CONCLUSION: For the patients with refractory constipation associated with neuropathy such as IND, total colectomy by SILS was very effective.

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1. Introduction

Chronic constipation is a common problem all over the world and is usually treated by medication, and surgical treatment has been indicated in case of refractory constipation. Constipation can result from several factors, like person's life style such as low fiber diet, inadequate fluid intake, consumption of some drugs, slow colonic motility, fecal evacuation disorders and combination of above [1]. As a cause of refractory constipation, it mentions malformations of the enteric nervous system. Intestinal neuronal dysplasia (IND), Hirschsprung disease (HD), hypoganglionosis, and ganglioneuromatosis constitute a group of malformations of the enteric nervous system so called intestinal dysganglionosis [2].

Nowadays, IND can be divided into two distinct subtypes. The type A of IND (IND-A) presents mostly in the neonatal period, which

comprises 5% of all IND cases and is characterized by congenital aplasia or hypoplasia of the sympathetic innervation, whereas the type B of IND (IND-B) is found in adults as well as in children, which comprises 95% of all IND cases, is characterized by hyperplasia of the parasympathetic submucosal and myenteric plexus [3,4]. Intestinal neuronal dysplasia usually presents in childhood and there have been a few reports in adults. Especially in IND-B cases series, refractory constipation has been reported as the commonest clinical presentation [5,6].

In the present report, we describe our experience of an adult patient with longstanding idiopathic constipation due to IND-B, who underwent single incision laparoscopic (SILS) total colectomy. The work has been reported in line with the SCARE criteria [7].

2. Presentation of case

A 36-year-old female patient presented to our department complaining of longstanding constipation during 20 years. She had received various treatments including some supplements and various laxatives. However, those treatments were ineffective, her symptoms worsened year by year. Eventually, she had taken over

Abbreviations: IND, intestinal neuronal dysplasia; SILS, single incision laparoscopic surgery.

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Fig. 1. Barium-enema showing no obvious mechanical obstruction.

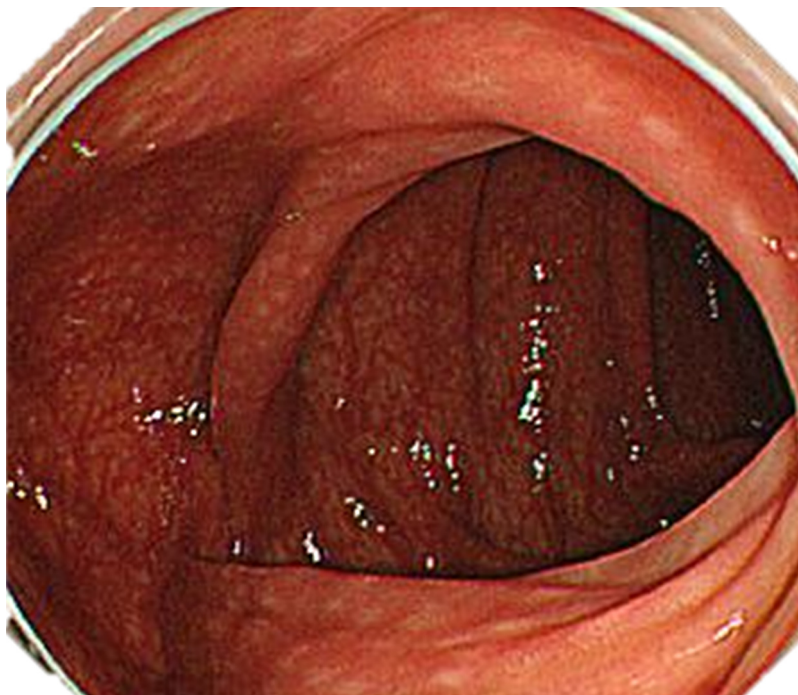


Fig. 2. Colonoscopic examination shows melanosis coli, and reveals no mechanical obstruction.

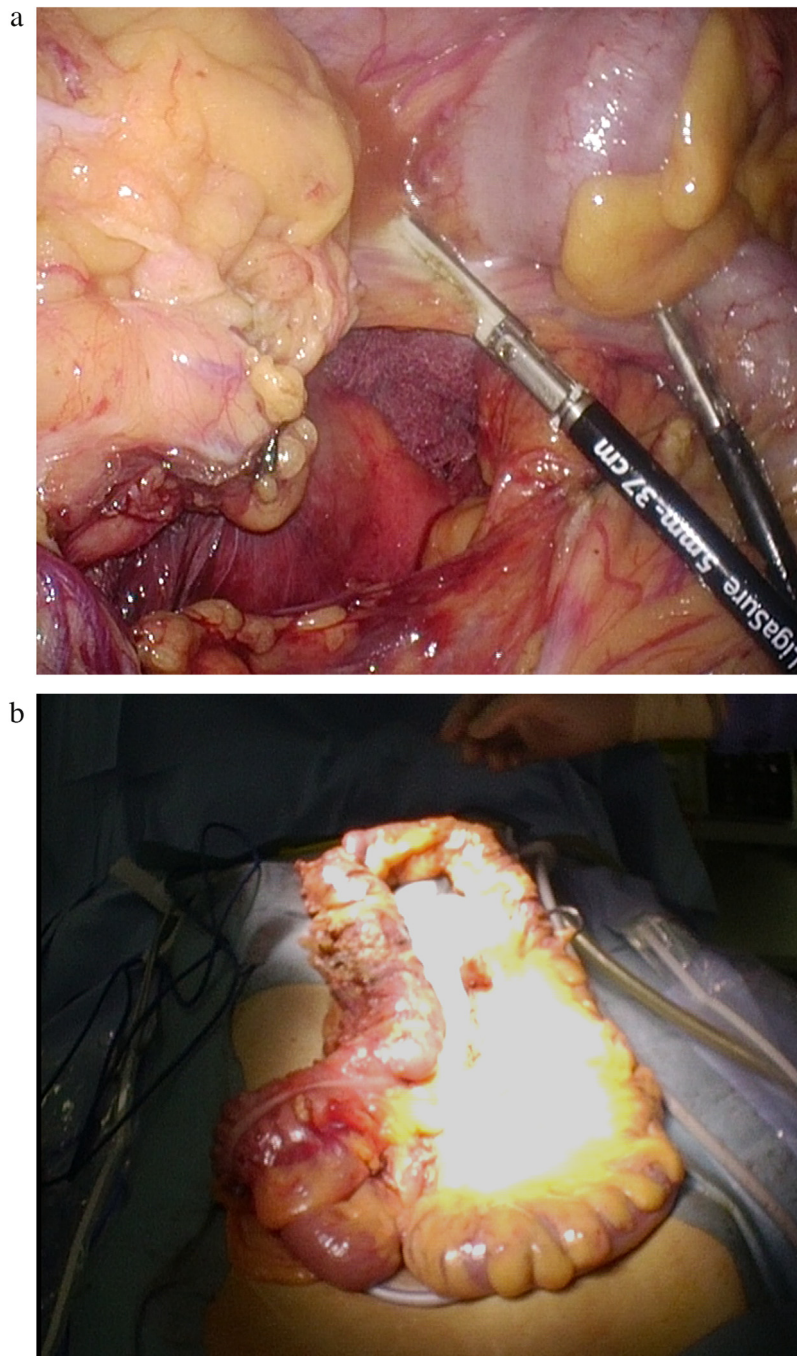


Fig. 3. a. Full mobilization of the splenic flexure. b. The specimen is pulled out from the single-port device.

70 tablets of stimulant laxatives per day. Basic laboratory data on admission were within normal range. Clinical examination revealed soft and flat abdomen, and bowel sound was normal. Systemic and neurologic diseases were excluded by medical history, physical and blood examination. Barium-enema examination and abdominal computed tomography scan showed elongated colon, but did not find any mechanical obstruction (Fig. 1). Colonoscopy showed melanosis in her entire colon, whereas did not reveal any mechanical obstruction (Fig. 2). After a preoperative consultation with the patient and her family, total colectomy with ileorectal anastomosis by SILS was chosen because of recurrence risk for constipation.

2.1. Operative technique

She was placed in the supine position, and a 3-cm “zigzag skin incision” [8] through the umbilicus was made. A lap protector and EZ Access (Hakko Co., Ltd., Tokyo, Japan) were placed through the incision. Three 5-mm ports were placed in EZ access mounted on the lap protector, and pneumoperitoneum was obtained. Almost all the procedures were performed with usual laparoscopic instruments and the operative procedures were much the same as in usual laparoscopic colectomy. To obtain better exposure of the operative field, we used percutaneous traction of vessel pedicles by suture thread needle in the medial approach. At first, we started

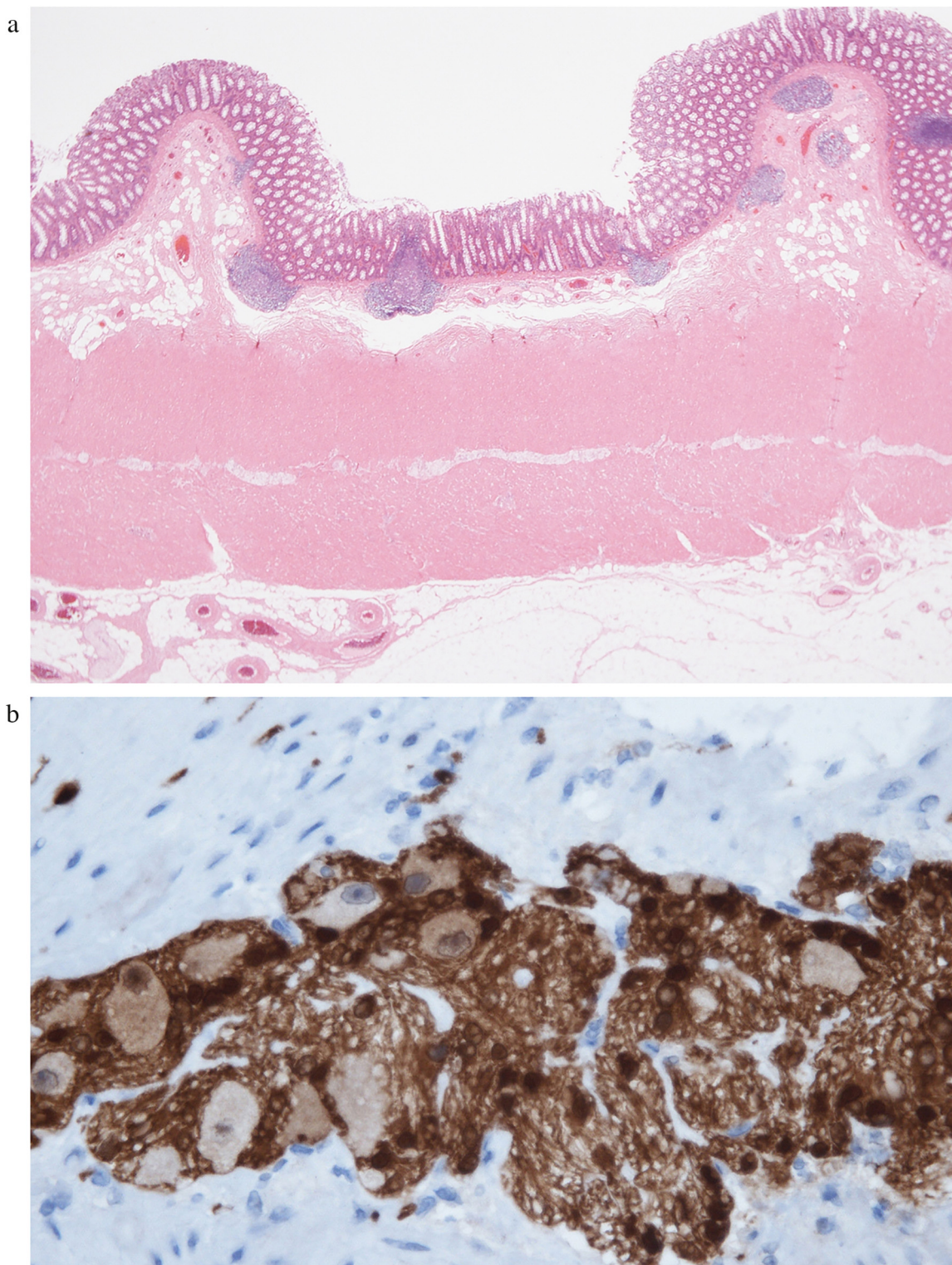


Fig. 4. a. A small number of isolated ganglion cells and giant submucosal ganglia are also detected in the submucosa. b. Protein S-100 is increased in giant ganglion cells.

from the mobilization of the left side colon by medial approach. Followed by opening omental bursa, it took down the splenic flexure, and finished the full mobilization of the left side colon (Fig. 3a). Subsequently, it was working on the mobilization of the right side colon using a medial approach. After the division and ligation of the ileocolic vessels, blunt dissection was performed toward the hepatic flexure in upper layer of subperitoneal fascia and it finished the

mobilization of hepatic flexure by dissection of lateral attachment. Finally, the specimen was extracted through the umbilical port, and colon resection between ileum end and recto-sigmoid colon was extracorporeally achieved (Fig. 3b). Functional end to end ileorectal anastomosis was performed with staplers. The operation time was 187 min and intraoperative blood loss was only 10 ml.

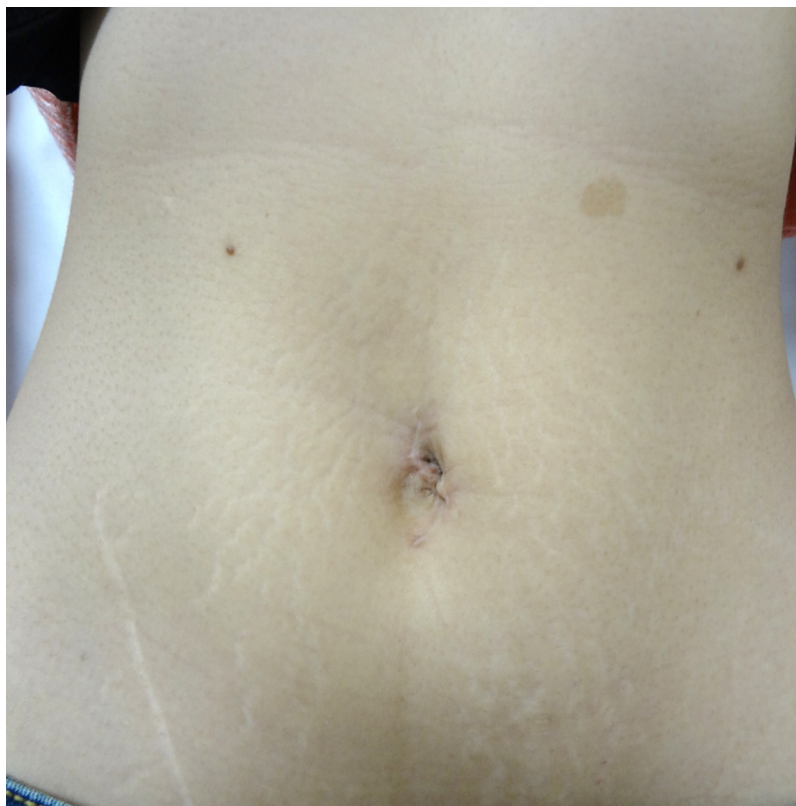


Fig. 5. Operative scar (postoperative 6 months).

2.2. Pathological findings

The pathological findings of the resected colon showed that the nerve bundles appeared hypertrophic, containing increased numbers of thickened disorganized axons in the intramuscular and submucosal nerve fascicles. Small number of isolated ganglion cells and giant submucosal ganglia were also detected in the submucosa (Fig. 4a). There was an increase of ganglionic nerve cells in the ganglion with approximately ten nerve cells per ganglion.

Immunohistochemical examination was applied by using antibodies against protein S-100, neuron-specific enolase, glial fibrillary acidic protein and synaptophysin. The positive for protein S-100 (Fig. 4b) and synaptophysin indicates an increase in ganglion cells, and glial fibrillary acidic protein meaning the increase of nerve plexus around blood vessels. Eventually, the diagnosis of IND-B was established.

The postoperative course was uneventful, and she was discharged on postoperative day 8. Her operative scar was small and very cosmetic (Fig. 5). She has remained asymptomatic with normal bowel motility during a 2-year follow-up.

3. Discussion

IND is a pathological condition, which belongs to the group of the gastrointestinal neuromuscular diseases and has been most recently included in the classification for this heterogeneous group of complex changes in the enteric nervous system [9,10]. Commonly, IND is associated with intestinal chronic constipation. Two forms of IND are recognized [3,4]. IND-A is extremely rare and patients with IND-A typically present in the neonatal period. In contrast, IND-B represents more than 95% of all IND cases, which is characterized by hyperplasia of the parasympathetic submucosal plexuses. Typical histological features of IND-B include hyperganglionosis, giant ganglia, ectopic ganglion cells and increased

acetylcholinesterase activity in the lamina propria and around the submucosal blood vessels. These pathological changes commonly involve in the distal colon. However, they can affect any segment of enteric nervous system and occur in different age groups ranging from newborns to adults [11]. The pathophysiology of chronic constipation and colon dysmotility in IND is still not clear. It is known that 5-hydroxytryptamine, which stimulates contraction of smooth muscle layers, is produced by neuroendocrine cells. Kobayashi et al. reported that an increase of neuroendocrine cells and morphological changes such as giant cells in patients with IND-B [12]. A possible explanation of colon dysmotility or chronic constipation may be an increased number of 5-hydroxytryptamine-positive cells in the mucosa of colon and rectum.

In the present case, the final diagnosis of IND-B was established. This case was a rare in the literature and very few cases worldwide. She had suffered from longstanding, refractory constipation during over 20 years. Conservative treatment for IND-B patients has been reported to be effective in 33–64% of the cases [13], whereas other authors report that IND is a less mild disease than generally thought and a more aggressive management should be concerned. Since colonic peristalsis in IND cases is impaired, surgery is often the appropriate treatment [14,15]. An important issue in the surgical treatment of IND-B cases is to specify the colonic segment that has dysganglionosis and a demonstrable innervation defect. In cases of entire colon involvement of IND changes such as the present case, a total colectomy should be the treatment choice. Even in cases of an isolated colonic segment affected by IND-B changes, it is difficult to determine if only part of the colon does not function properly rather than the entire colon [16]. Furthermore, segmental and total colectomy with ileosigmoid anastomosis frequently result in persistent or recurrent constipation, and up to 50% of patients who have undergone these procedures have required additional resection [17–19]. Therefore, we performed total colec-

tomy with ileorectal anastomosis by SILS as the surgical procedure in the present case.

Recently, attempts to further minimize trauma related to surgical procedures and improve their cosmetic effects, especially in young patients such as the present case, resulted in modification of the laparoscopic technique to a surgical procedure with a single, a small incision, most commonly in the umbilicus, called SILS [20,21]. Single incision laparoscopic surgery has emerged as a new generation of laparoscopic colorectal surgery that is akin to conventional surgery, with the added benefit of better cosmesis, reduced morbidity, reduced postoperative pain, and reduced length of hospital stay, along with minimal trocar related potential complications of abdominal trauma, vessel injury, wound infection, trocar site hernia, and port-recurrences [22]. Charudutt et al. described that a total colectomy by SILS is feasible and very comparable with multi-port laparoscopic colectomy in terms of length of hospital stay, operative time, and complication rates [23]. To our knowledge, this is the first report of total colectomy by SILS for a young patient with refractory chronic constipation due to IND-B. For refractory constipation patients, it is necessary to consider laparoscopic total colectomy such as SILS approach.

4. Conclusion

In the patients with refractory constipation, it is necessary to care in mind that some patients require surgical treatment. Especially for the patients with neuropathy such as IND changes, total colectomy with ileorectal anastomosis by SILS is feasible and effective.

Conflict of interest

None.

Funding

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Ethical approval

None.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Authors contribution

Taro Masuda, Takashi Nonaka, Toshiyuki Adachi, Makoto Hisanaga, Shigeki Nagayoshi, Takayuki Tokunaga, Ken Taniguchi, Hirokazu Kurohama, Masahiro Ito collaborate in medical care. Hikaru Fujioka looked over the manuscript.

Guarantor

Takashi Nonaka.

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