A 56-Year-Old Woman With Severe Chronic Constipation: What Is the Underlying Pathology?

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A 56-year-old woman with chronic constipation visited Chung-Ang University Hospital. She had suffered from chronic constipation, abdominal pain and abdominal distention, and took prokinetics, bulking agents and osmotic laxatives for more than 5 years. Her symptoms became worse and she could defecate only 3 or 4 times per a month. She had no history of any specific medical disease or trauma. The abdomen was distended without palpable

mass. The laboratory findings were not remarkable.

The plain abdominal radiograph showed ileus with air-fluid levels (Fig. 1A). Abdominopelvic CT revealed a marked colonic distension, whereas the entire colon was well traced without a definite obstructive lesion (Fig. 1B). During the colonoscopy and double-contrast barium enema, there was neither an intra-luminal lesion nor narrowed segment which was suggestive of transi-

Figure 1. Abdominal X-ray and CT findings. (A) The plain abdominal radiograph shows ileus with multiple air-fluid levels. (B) Abdominopelvic CT reveals marked colonic distensions with abundant feces and fluid from cecum to sigmoid colon, whereas the entire colon is well traced without a definite obstructive lesion.

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tional zone. Colon transit time using radio-opaque markers was severely prolonged. The right, left, rectosigmoid and total colon transit time were 106.8, 21.6, 13.2 and 141.6 hours, respectively.

Figure 2. Rectoanal inhibitory reflex on anorectal manometry. Rapid rectal distention with balloon (30 mL) induces a transient increase in rectal pressure, followed by a more prolonged reduction in anal pressure due to relaxation of the internal anal sphincter.

Anorectal manometric findings were within normal range and the rectoanal inhibitory reflex was well preserved (Fig. 2).

She has taken large doses of laxatives for several months, and her symptoms improved temporally. However, the constipation aggravated again and her quality of life deteriorated progressively.

Since escalation of medication had not been effective for her constipation for many years, surgical treatment was chosen under the impression of chronic intestinal pseudo-obstruction. Total colectomy with ileorectal anastomosis was performed. Histologically, markedly decreased number of ganglion cells in the myenteric plexus was observed in the entire colon (Fig. 3) in which the pathologic findings confirmed hypoganglionosis.

Chronic intestinal pseudo-obstruction is a rare debilitating disorder characterized by obstructive symptoms without any lesion occluding the gut.^{1,2} Chronic idiopathic intestinal pseudo-obstruction (CIPO) is diagnosed when the underlying cause is not found in spite of the thorough medical examination.^{3,4}

The present case is a neuropathic type of CIPO, and demonstrates hypoganglionosis to be involved in medically intractable

Figure 3. Myenteric (Auerbach) plexus of the colon of the present case (A, B) and the control (C, D). (A) The number of ganglion cells in the present case is markedly reduced (arrow) compared to that in the control (H&E, $\times 200$). (B) Serial section stains with antibody to S-100 protein show that the ganglion cells are negative for staining (arrow) (Immunohistochemical stains for S-100, $\times 200$).

chronic constipation.

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