

Idiopathic Proximal Hemimegacolon in an Adult Woman

Jung Won Noh, MD, Poong-Lyul Rhee, MD*, Seo Young Son, MD, Chang Soo Ok, MD, Gayeon Lee, MD, and Byung-Hoon Min, MD

Department of Medicine, Samsung Medical Center, Sungkyunkwan University School of Medicine, Seoul, Korea

Idiopathic proximal hemimegacolon is a disorder characterized by bowel dilatation proximal to the splenic flexure. It is a very rare and therefore a poorly understood clinical entity. This report describes a case of idiopathic proximal hemimegacolon in a 44-year-old woman. The patient suffered from 2 episodes of constipation and bowel dilatation over 4 years and was successfully treated by medical therapy.

(J Neurogastroenterol Motil 2010;16:203-206)

Key Words

Idiopathic megacolon, Splenic flexure, Medical futility

Introduction

Idiopathic megacolon is a rare and poorly understood clinical condition that usually presents as refractory abdominal distension and constipation. It includes idiopathic megarectum, idiopathic megacolon, or a combination of these disorders. Among them, idiopathic megacolon without megarectum is the most uncommon disorder. In this case, we experienced the idiopathic megacolon with bowel dilatation only proximal to the splenic flexure.

Case Report

The patient was a 44-year-old woman. One month before being referred to our hospital, abdominal distension and con-

stipation developed and the severity of both increased with time. She was admitted to a local hospital and diagnosed as having megacolon. The patient could not defecate and the abdominal pain increased despite fasting and frequent enemas. Because of the intractable symptoms, she was referred to our emergency unit for further investigation. The physical examination during the obstructive episodes revealed marked abdominal distension with hypoactive bowel sounds. There was no palpable abdominal mass or signs of peritoneal irritation. The patient denied a history of metabolic, neurological, cardiovascular or pulmonary disease, previous abdominal or pelvic surgery, or a history of abdominal cancer, inflammatory disease or trauma. The patient was not taking any medications. The laboratory testing showed a normal hemoglobin, hematocrit, white blood cell count, blood glucose, creatinine, and coagulation profile. The serum electrolytes were also normal except hypokalemia was present with a level of 2.7

Received: January 14th, 2010 Accepted: March 18th, 2010

© This is an Open Access article distributed under the terms of the Creative Commons Attribution Non-Commercial License (<http://creativecommons.org/licenses/by-nc/3.0>) which permits unrestricted non-commercial use, distribution, and reproduction in any medium, provided the original work is properly cited.

*Correspondence: Poong-Lyul Rhee, MD

Department of Medicine, Samsung Medical Center, Sungkyunkwan University School of Medicine, 50 Irwon-dong, Gangnam-gu, Seoul 135-710, Korea

Tel: +82-2-3410-3409, Fax: +82-2-3410-6983, E-mail: pl.rhee@samsung.com

Financial support: None.

Conflicts of interest: None.

mmol/L (normal range 3.5-5.1 mmol/L). The thoracic imaging was normal. Abdominal simple radiographs showed signs of marked dilatation of the colon (Fig. 1A) especially in the cecum and transverse colon. The abdominal computed tomography (CT) scan showed bowel distension, and the transition zone was detected at the splenic flexure (Fig. 2). However, mechanical obstruction was not detected. Sigmoidoscopy was performed and no obstructive lesions or mucosal changes were noted. Samples taken from multiple random mucosal biopsies also revealed normal findings. Anorectal manometry showed a normal resting anal sphincter pressure, normal squeezing pressure during maximal voluntary effort, rectal sensory threshold, maximal tolerance volume (120 mL), and rectoanal inhibitory reflex was present. The colon transit time was delayed (total 135.6 hours, right colon 66 hours, left colon 56.4 hours, and rectosigmoid colon 13.2 hours). Based on these findings the patient was diagnosed with idiopathic

proximal hemimegacolon. First, the hypokalemia was corrected to between 3.8 mmol/L to 4.9 mmol/L. However, the abdominal distension and constipation were not improved. On the 19th day of hospitalization, a small amount of loose stool was passed and the abdominal pain was relieved. Bisacodyl suppositories and bisacodyl/docusate were started. On the 22nd day of hospitalization, the frequency of loose stools increased up to 3-4 times/day. Oral feeding was started on the 27th day of hospitalization after checking the CT endoscopy (Fig. 2B). The finding was marked dilatation of ascending and transverse colon at the level of distal transverse colon without obstructive lesion. The maximal diameter of dilated colon was 15 cm. The patient was discharged on the 44th day of hospitalization on oral mosapride, itopride, domperidone, and lactitol (Fig. 1B). After discharge, she did not have additional symptoms even after the medication was discontinued. However, 4 years later, abdominal distension

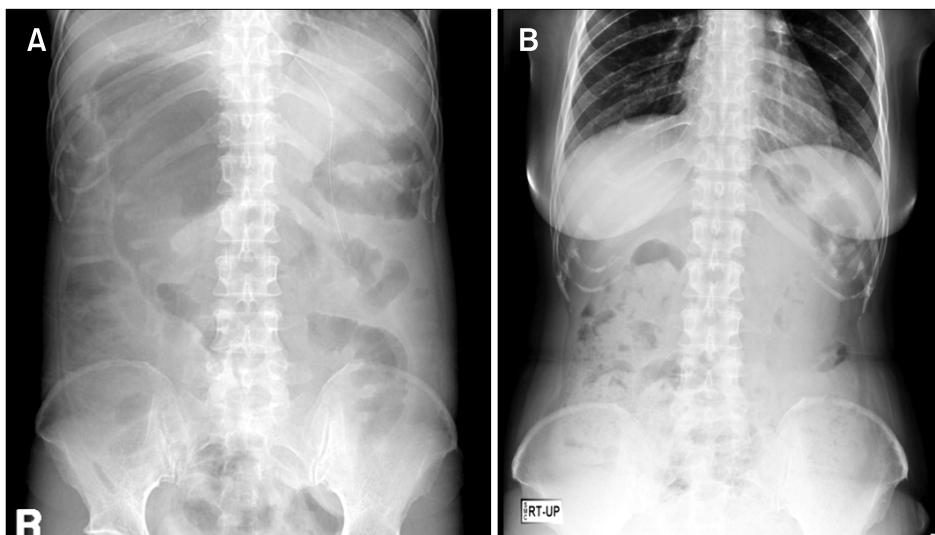


Figure 1. Abdomen supine X-ray. (A) Severely dilated proximal transverse colon. (B) Improvement of dilated colon after medical treatment.

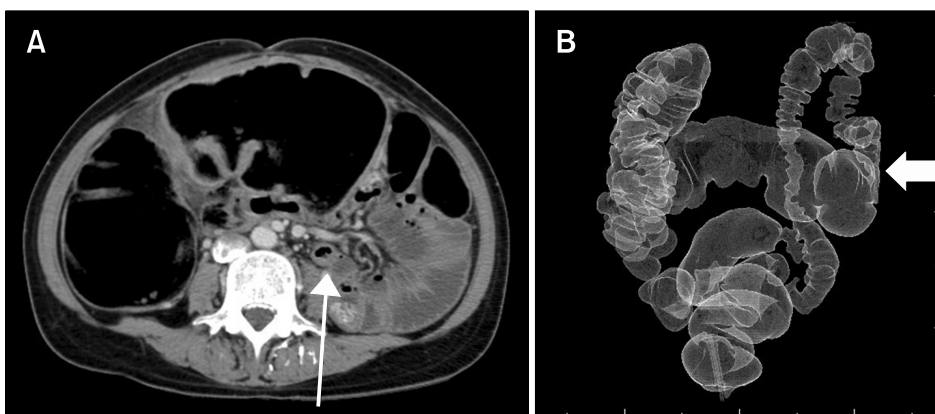


Figure 2. Abdomen pelvis computed tomography (CT) scan (A) and CT endoscopy (B) shows dilatation of the ascending and transverse colon. The arrows indicate the transition zone at the level of the splenic flexure with no evidence of mechanical obstruction.

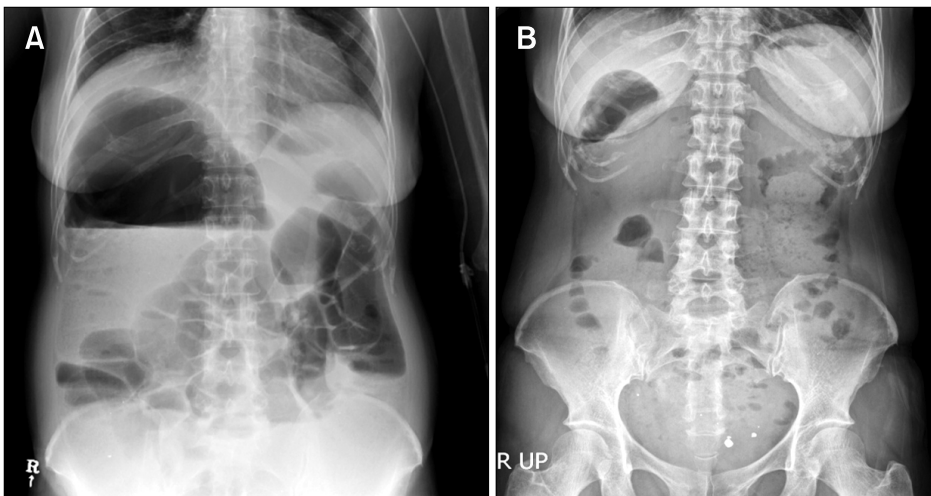


Figure 3. Abdomen erect X-ray. (A) The recurrence of hemimegacolon at the same site. (B) Improvement of recurrent hemimegacolon after medical treatment.



Figure 4. Abdomen simple X-ray shows improved hemimegacolon followed in the outpatient clinic.

and constipation occurred again. She was admitted to a local hospital for 20 days and sigmoidoscopic irrigation and enemas were performed there. However, the symptoms were not relieved. Therefore, she was referred to our emergency unit for further management. Severe abdominal distension was noted on physical examination and on the simple abdominal radiograph (Fig. 3A). The laboratory testing was normal except for ALT (119 U/L) and AST (141 U/L). The abdominal CT scan showed similar findings compared to the previous study. Bisacodyl/docusate was started on hospital day 2. By hospital day 8, loose stools were passed after a glycerin enema and the abdominal pain and dis-

tension were relieved. On hospital day 10, a large amount of loose stool was passed and magnesium oxide, mosapride, domperidone, and colonlyte were started. Stool passage was increased to 7-15 times/day. On hospital day 16, oral feeding was started and the ALT and AST returned to normal. On hospital day 25, the patient was discharged (Fig. 3B). Three months later, she visited the outpatient clinic and did not have abdominal distension or constipation without medication (Fig. 4).

Discussion

Megacolon can be observed in Hirschsprung's disease, with an infection (Chaga's disease) and in some disorders of the endocrine or central nervous system (including spinal trauma and senility). However, in the absence of an organic cause, it is referred to as idiopathic.^{1,2} Idiopathic megacolon is a heterogeneous disorder including idiopathic megarectum, idiopathic megacolon, or a combination of these disorders.³ Idiopathic proximal hemimegacolon is defined as megacolon only proximal to the splenic flexure.⁴ Idiopathic proximal hemimegacolon is a very rare and therefore a poorly understood clinical entity. The pathogenesis underlying idiopathic proximal hemimegacolon is currently unclear. The plausible explanations include abnormalities involving the extrinsic nerves, the enteric nerve plexus, or the intestinal smooth muscle, which could lead to gut dilatation and impaired motility.⁵ Anal manometry is clinically useful in a few patients with chronic constipation, although it has potential value for the exclusion of Hirschsprung's disease, due to the demonstration of internal sphincter relaxation in response to rectal distension.⁶ The clinical features of patients with idiopathic proximal hemi-

megacolon are similar to those reported in idiopathic megacolon without megarectum in previous studies. Patients with idiopathic megacolon are older than patients with idiopathic megarectum. In this case, the patient experienced her first episode at 44-years of age. Most patients present with constipation; however, documented bowel habits are very variable, from 5 times per day to only once every 3 months. Abdominal distension is more common than abdominal pain. The majority of patients have normal intelligence. Most patients have no significant family history. Our patient had abdominal distension and pain, normal intelligence, and no familial history. Most patients show a delayed colon transit time. Delay in the colon transit is most prominent in the right colon area. In the rectosigmoid area, it is normal.^{2-4,7,8} Our patient also showed delay in right colon transit. The abnormal AST/ALT levels were considered secondary to hepatocongestion induced by the megacolon. Patients with idiopathic megacolon are managed conservatively initially. The majority of patients can be successfully managed without surgery.² For shortening bowel transition time and symptom relief, the prokinetic drugs like mosapride and itopride could be coadministered.⁹ However, medical treatment may fail to alleviate symptoms in some patients,⁸ may be poorly tolerated, and must be continued lifelong to prevent the recurrence of symptoms.

Megacolon proximal to the splenic flexure is referred to as idiopathic proximal hemimegacolon. Here, we present the case of a 44-year-old woman with idiopathic proximal hemimegacolon

that had 2 episodes over 4 years. She was successfully managed with medical treatment only.

References

1. Ehrenpreis T. Megacolon and megarectum in older children and young adults. Classification and terminology. *Proc R Soc Med* 1967;60:799-801.
2. Gattuso JM, Kamm MA. Clinical features of idiopathic megarectum and idiopathic megacolon. *Gut* 1997;41:93-99.
3. Stabile G, Kamm MA, Hawley PR, Lennard-Jones JE. Colectomy for idiopathic megarectum and megacolon. *Gut* 1991;32:1538-1540.
4. Min BH, Son HJ, Kim JJ, Rhee JC, Lee SJ, Rhee PL. Idiopathic proximal hemimegacolon: radiologic findings and analyses of clinical and physiological characteristics. *Abdom Imaging Published Online First*: 22 Apr 2009. doi:10.1007/s00261-010-9606-8
5. Georgescu EF, Vasile I, Georgescu AC. Intestinal pseudo-obstruction-a rare condition with heterogeneous etiology and unpredictable outcome. A case report. *J Gastrointest Liver Dis* 2008;17:77-80.
6. Diamant NE, Kamm MA, Wald A, Whitehead WE. AGA technical review on anorectal testing techniques. *Gastroenterology* 1999;116:735-760.
7. Gattuso JM, Kamm MA, Talbot JC. Pathology of idiopathic megarectum and megacolon. *Gut* 1997;41:252-257.
8. O Súilleabháin CB, Anderson JH, McKee RF, Finlay IG. Strategy for the surgical management of patients with idiopathic megarectum and megacolon. *Br J Surg* 2001;88:1392-1396.
9. Mishima Y, Amano Y, Okita K, et al. Efficacy of prokinetic agents in improving bowel preparation for colonoscopy. *Digestion* 2008;77:166-172.