Laparoscopic treatment of intestinal intussusception in Peutz-Jeghers syndrome: Case report and review of literature

Sayuri HASEGAWA, Takafumi SEKKA, Jinichi SOEDA, Kazuhiro ISHIZU, Eisuke ITO, Seiji MORITA and Hiroyasu MAKUUCHI*

Department of Surgery, Tokai University Hachioji Hospital
** Department of Surgery, Tokai University school of Medicine*

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A 47-year-old woman presented with an abdominal mass and nausea. Abdominal ultrasound and computed tomography (CT) showed a sausage-shaped mass with invagination. One polyp that appeared to exceed 3 cm was found in the sigmoid colon. Laparoscopy confirmed an intussuseption mass, and the intussusception was dissected by hand-assisted laparoscopy (HALS). The sigmoid colon was also mobilized to the site of the small incision and resected. Generally, we believe enterectomy including polyps should be avoided as much as possible in Peutz-Jeghers syndrome (PJS) because poly-surgery may lead to short bowel syndrome. In addition, PJS patients often undergo multiple surgery, and therefore dense intra-abdominal adhesions are seen at subsequent laparotomy, which makes surgery increasingly difficult with repeated operations. Laparoscopic-assisted surgery seems beneficial, as in the present case.

Key words: Peutz-Jeghers syndrome, intussusception, laparoscopic treatment

INTRODUCTION

Peutz-Jeghers syndrome (PJS) is a rare autosomal dominant disease with incomplete penetrance, characterized by perioral pigmentation and gastrointestinal hamartomatous polyps (mostly in the jejunoileum, but also in the stomach and colon) [1]. These polyps can grow to a very large size, and combined with their pedunculated nature, result in recurrent intussusception. In a Japanese series, 47% of patients suffered from intussception requiring operation [2]. Ideally, preservation of intestinal length is important in patients with Peutz-Jeghers because recurrence is seen in up to 10% of cases, and multiple resections could lead to the short bowel syndrome [3, 4]. However, recurrence of intussusception due to polyps is common in Peutz-Jeghers, and a combined approach of laparoscopy and endoscopy to reduce the need for multiple laparotomies may avoid untoward problems in the future. Laparoscopy is a minimally invasive approach that deals with the acute problem and may prevent adhesion formation.

CASE REPORT

The patient was a 46-year-old woman who had been treated for recurrent abdominal pain at another clinic but was referred to our hospital because of no improvement. Past history included appendectomy at age 10 years, partial resection of the small intestine for intussusception at age 13 when she was diagnosed as having PJS, and resection of the left ovary for an ovarian cyst at age 27. Physical examination showed pigmentation on her lips and oral mucosa. The abdomen was soft, and an elastic-soft tumor about 4 cm in size was palpable to the left of the umbilicus. Laboratory studies showed no abnormal findings. Abdominal ultrasound revealed a looping intestinal tract due to dilatation on the left of the umbilicus. In the lumen of the intestinal tract, a similar enteric tract structure was further noted, indicating intussusception (Fig. 1). Similarly, abdominal CT also demonstrated an intussusception, and many polyps in the lumen of the intestinal tract (Fig. 2). Contrast radiography of the small intestine showed immobile polyps of various sizes in the 3rd portion of the duodenum (Fig. 3a). The stacked coin sign was seen at the site consistent with a mass (Fig. 3b). Contrast medium flowed slowly toward the anal

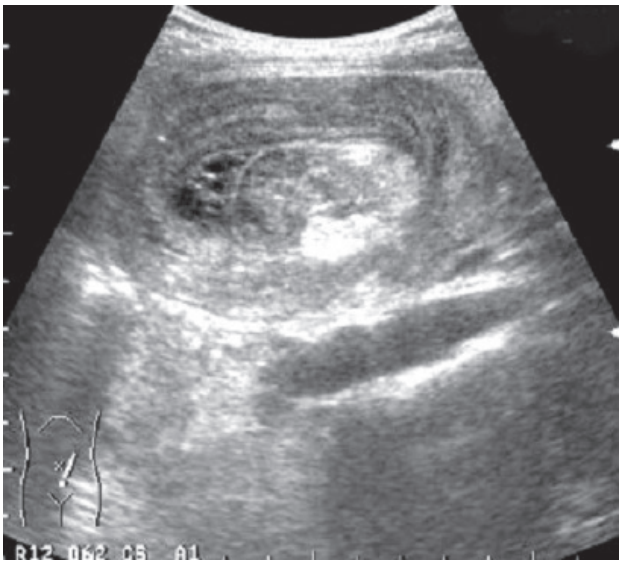

Fig. 1 Ultrasonographic examination revealed a tumor with high and low echogenicity which showed the target sign.

Sayuri HASEGAWA, Department of Surgery, Tokai University Hachioji Hospital, 1838 Ishikawacho, Hachioji, Tokyo 192-0032, Japan Tel: +81-426-39-1111 Fax: +81-426-39-1112 E-mail: sayuri-m@muc.biglobe.ne.jp

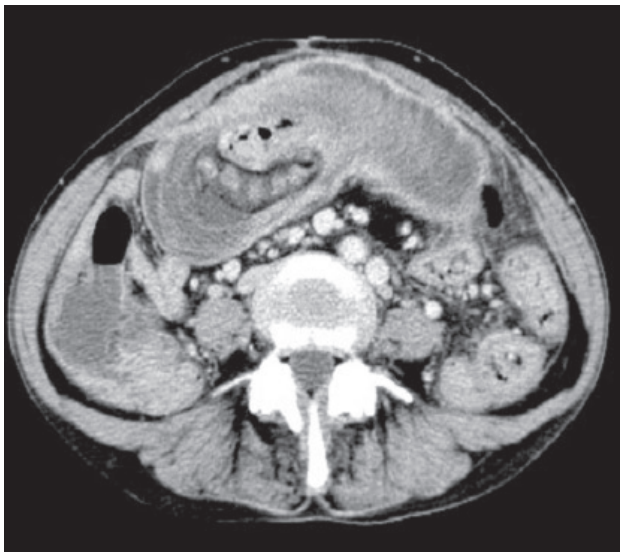

Fig. 2 Computed tomography revealed a circular mass with the target sign, and the lesion seemed to be an intussusception.

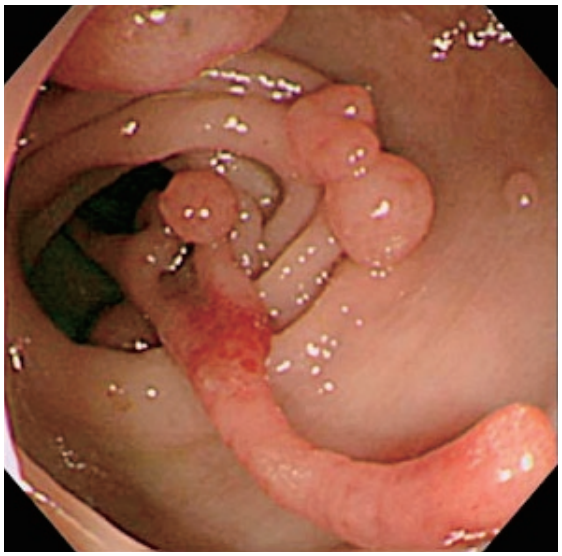

Fig. 4 Colonoscopy showed scattered polyps in the entire colon.

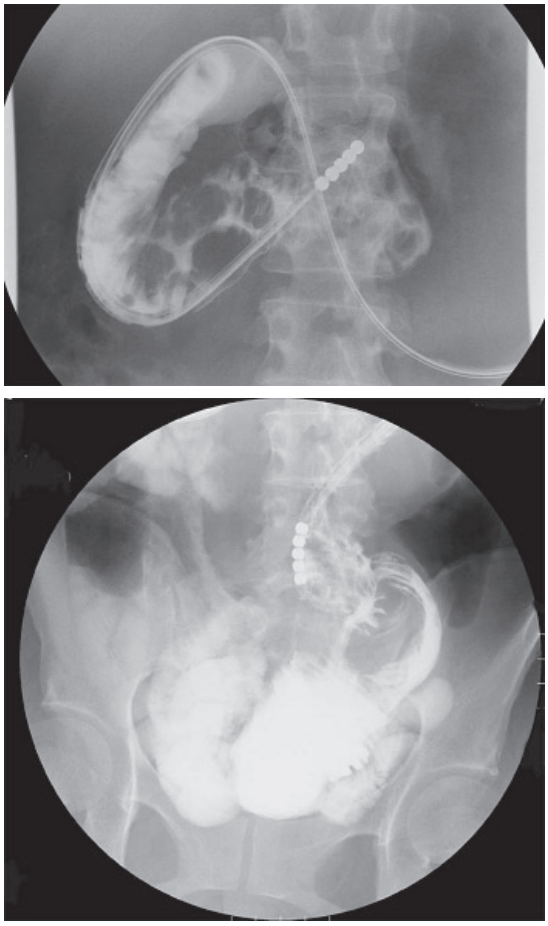

Fig. 3 **a**: Long-tube enterogram showed immobile polyps of various sizes in the 3rd portion of the duodenum.
b: Long-tube enterogram showed the classic "stacked coin" appearance of small intestinal intussusception.

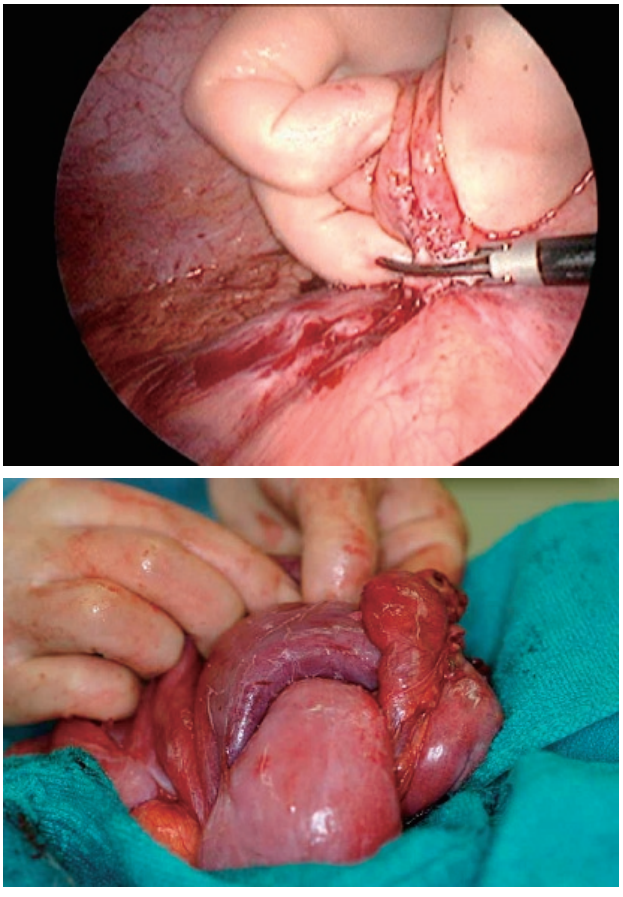

Fig. 5 **a**: Laparoscopic adhesiolysis of the intussuscepting segments.
b: Through a 4-cm mini-laparotomy, the intussuscepting segments were brought out extracorporeally.

side. From these findings, the diagnosis of small bowel intussusception due to gastrointestinal tract polyposis was made, and decompression was performed by placing an ileus tube. Elective operation was performed. Preoperative colonic search revealed scattered polyps in the entire colon (Fig. 4), and therefore we decided to excise any sigmoid colonic polyps greater than 2 cm simultaneously.

Because there were three surgical scars, a camera port was first inserted to the right of the umbilicus, and 5-mm ports were inserted into three different sites. The abdominal tissue was densely adherent due

to three previous operations. Laparoscopic adhesiolysis of the intussuscepting segments was performed, and the sigmoid colon was also mobilized. Through a 4-cm mini-laparotomy, the intussuscepting segments and the sigmoid colon with polyps were brought out extracorporeally. Although extracorporeal reduction was performed employing Hutchinson's maneuver, the intussuscepting segments were excised, because areas with poor circulation were found (Fig. 5a & 5b). After making an incision through the intestine, the polyps in the sigmoid colon were excised. The histologic diagnosis of all polyps both in the intestine and sigmoid colon was hamartomas. The patient started taking meals on Day 5 and was discharged on Day 8. At two years postoperatively, she was symptom free.

DISCUSSION

Peutz-Jeghers syndrome is an unusual, inherited gastrointestinal (GI) hamartomatous polyposis syndrome that is associated with mucocutaneous pigmentation. The relationship of mucocutaneous pigmentation and intestional polyposis was first reported in 1921 by Peutz [5], who studied seven family members over three generations. Of Peutz's seven patients, seven had intestinal polyposis, four nasal polyposis, and one bladder polyps. After an initial report of two patients by Jeghers in 1944 [6], the definitive clinical description of PJS was written by Jeghers *et al*. in elegant detail in 1949 [7]. They outlined the clinical manifestations and outcome of 10 patients with mucocutaneous pigmentation and intestinal polyposis. The author also recognized it was inherited as a simple Mendelian dominant trait. The incidence has been estimated as one in 120,000 births [8].

The predominant clinical features of PJS are the result of GI polyposis. GI polyposis becomes clinically manifest early in life, with the age of diagnosis or death ranging between 9 and 39 years [7]. In a Japanese series of 222 patients with PJS, the average age of diagnosis in male patients was 22 years and in female patients, 26 years [2]. One-third of PJS patients will experience symptoms during their first decade of life, and 50-60% of patients will experience them before age 20 years [9].

The most common location of Peutz-Jeghers polyps is the small intestine (jejunum > ileum > duodenum), followed by the large intestine and stomach. In a collective series of 182 cases reported from the Mayo Clinic, 96% of patients had small bowel, 27% colon, 24% rectum, and 24% stomach polyps [10, 11]. The polyps grow to a very large size, and combined with their pedunculated nature, result in recurrent intussusception. In the Japanese series, 47% of patients suffered from intussusception requiring operation [2]. The polyps are hamartomas of smooth muscle that extends, tree-like, into the lamina propria. Usually broad-based, they vary in size from a few millimeters to several centimeters. Larger hamartomas often contain foci of adenomatous changes. Histological evidence of hamartomatous-adenomatous-carcinomatous evolution has been demonstrated for stomach, small bowel, and colorectal polyps in PJS [3, 12, 13]. While the origin of cancer within the GI tract of PJS patients is debated, patients with PJS are at significantly increased risk of developing cancer outside the GI tract - in the pancreas, breast, lung, uterus, and ovary [14].

The major abdominal symptoms are abdominal pain and GI bleeding. The treatment of intussusception varies according to the age of the individual. In adults, it is well known that a malignant lead point, usually metastatic, is the etiologic agent responsible for 46% to 50% of cases of intestinal invagination. The correct procedure for such cases is laparotomy and en bloc resection of the involved segment of bowel with its lymphatic drainage area [15]. In PJS patients, because of the recurrent nature of the polyps, reduction of the intussusception, followed by enterotomy and polypectomy or limited resection must be considered the procedure of choice, because cure is not possible and extensive resection is not indicated [16].

In this case, where there was a known history of hamartomatous polyps from PJS, there was no question but to reduce the intussusception and then remove the lead point polyp. This was done laparoscopically in an attempt to reduce postoperative pain, hasten patient recovery, and decrease the length of hospital stay. Ideally, preservation of intestinal length is important in patients with PJS, because recurrence is seen in up to 10% of cases, and multiple resections could lead to the short bowel syndrome [3, 4]. Laparoscopy is a minimally invasive approach that deals with the acute problem and may prevent adhesion formation and subsequent repeat laparotomy for small bowel obstruction. As intussusception in PJS has a considerable recurrence rate, upper/lower endoscopic and laparoscopic resection may be indicated in order to avoid multiple laparotomies and their complications.

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