CASE REPORT

Laparoscopic Management of Small Bowel Intussusception in a 16-Year-Old With Peutz-Jeghers Syndrome

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ABSTRACT

Introduction: Peutz-Jeghers is a rare autosomal dominant disorder characterized by hamartomatous polyps and discoloration of mucosal membranes. The polyps can occur anywhere in the gastrointestinal tract and can grow large enough to cause bowel obstructions.

Case Report: A 16-year-old male presented to the emergency department with signs and symptoms of an acute bowel obstruction. He had 2 days of abdominal pain, obstipation, and vomiting. He had a previous history of a colonoscopy with polypectomy at age 4, and hyperpigmentation of his mucous membranes.

Results: Computed tomographic (CT) scan revealed an intussusception of the small intestine. An exploratory laparoscopy found an intussusception of the mid jejunum. A laparoscopic-assisted small bowel resection was performed. Pathology showed a 5-cm polyp that acted as a lead point for the intussusception. Colonoscopy and upper endoscopy revealed 5 more polyps in the stomach and colon that were removed.

Conclusion: Small bowel obstructions can be managed successfully with minimally invasive approaches. The treatment of obstruction in these patients is to remove the offending hamartomatous polyp(s). The rest of the intestine needs to be examined and those polyps found should be removed. This can be done intraoperatively with laparoscopic-assisted enteroscopy and colonoscopy.

Key Words: Peutz-Jeghers, Intussusception, Laparoscopic, Hamartomatous, Polyp.

INTRODUCTION

Peutz-Jeghers syndrome (PJS) is a rare autosomal dominant condition characterized by hamartomatous polyps and mucocutaneous pigmentation of the lips, buccal mucosa, and digits.[1–4] Polyps can occur anywhere in the gastrointestinal tract and can grow large enough to cause bowel obstructions.

CASE REPORT

A 16-year-old male presented to the emergency department with signs and symptoms of an acute bowel obstruction. He had 2 days of abdominal pain, obstipation, and vomiting. He had a previous history of a colonoscopy with polypectomy at age 4, had hyperpigmentation of his mucous membranes, and his mother and maternal grandfather had a history of gastrointestinal polyps. Computed tomography revealed an intussusception of the small intestine. The patient underwent an exploratory laparoscopy and was found to have an intussusception of the mid jejunum **(Figure 1)**.

A laparoscopic-assisted small bowel resection was performed. Pathology showed a 5-cm polyp that had acted as a lead point for the intussusception **(Figure 2)**. The patient did well postoperatively. Colonoscopy and upper endoscopy revealed 5 more polyps in the stomach and colon. These were removed. The diagnosis of PJS was made from these findings. The patient had an uncomplicated postoperative course and was discharged home on postoperative day 3. He is one year out from surgery and has resumed his normal activities with no evidence of recurrence.

DISCUSSION

PJS was first described in 1921 by Peutz and subsequently elaborated upon by Jeghers in 1949.[2,3] Germline defects in the tumor suppressor gene serine/threonine kinase 11 (STK11) are implicated in this rare autosomal dominant inherited disease.[4] Its incidence is calculated in 1 in 200,000 liveborns, and its mean age of onset is 25.2 years.[7] The most common location of the hamartomatous polyps is the small bowel (78%), followed by the colon (42%), stomach (38%), and rectum (28%).[5] These polyps can

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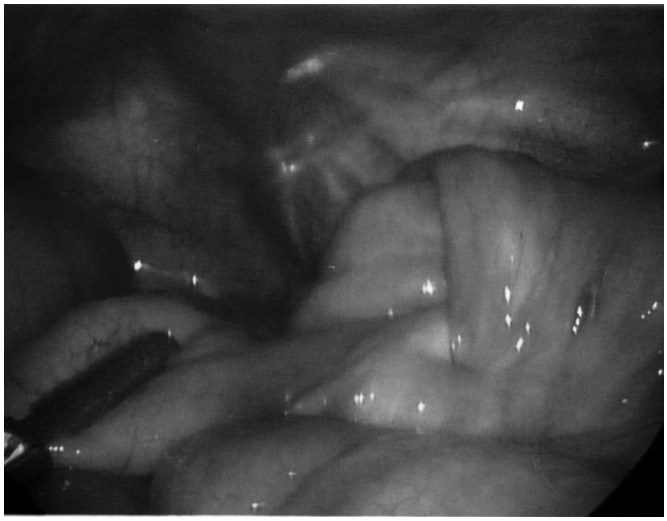

Figure 1. Laparoscopic image of intussusception in the mid jejunum.

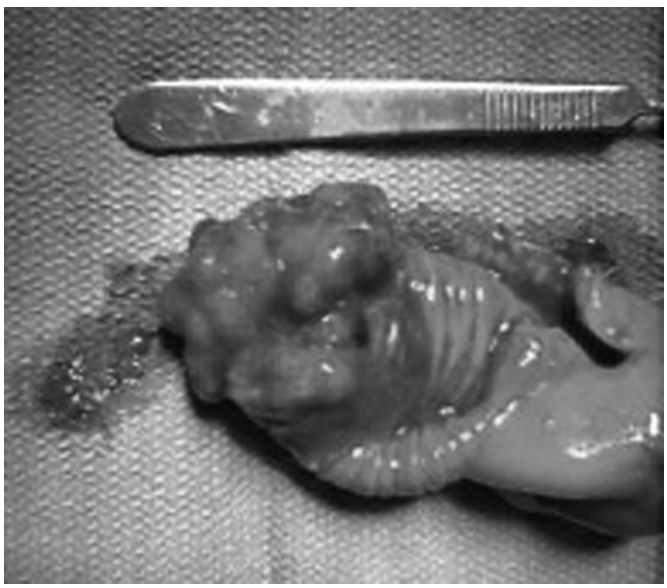

Figure 2. Jejunal polyp acting as lead point for intussusception.

cause obstruction in up to 43% of cases and rectal bleeding in up to 14% of patients.[1] The syndrome is associated with a 2% to 10% increased risk of cancer of the intestinal tract, from the stomach to the rectum. There is also an increased risk of extraintestinal malignancies, including cancer of the breast, ovary, cervix, fallopian tubes, thyroid, lung, gallbladder, bile ducts, pancreas, and testes.[4]

Small bowel obstructions can be managed successfully with minimally invasive approaches.[8] Treatment of obstruction in these patients is to remove the offending hamartomatous polyp. However, recurrence of intussusception episodes occurs in at least 10% of cases, resulting in repeated surgical intervention.[1] Thus, the rest of the intestine needs to be examined, and those polyps found should be removed. This can be done intraoperatively with laparoscopic-assisted enteroscopy and colonoscopy. Once the gastrointestinal tract has been cleared of polyps, the recommended interval of small bowel follow-through is from 2 years to 3 years. The presence of polyps larger than 1.5 cm in diameter mandates another complete gastrointestinal evaluation with endoscopic removal of polyps.[5] Patients should also be screened periodically for malignancies of the breast, cervix, ovary, testis, stomach, and pancreas.

To our knowledge, there are at least 2 other published case reports regarding the laparoscopic management of bowel obstructions in PJS.[1,6] The ideal way to remove a pedunculated polyp acting as a lead point is endoscopically. When this is not possible, laparoscopy offers a safe and effective method for surgical management with reduction of the intussusception and small bowel resection.

CONCLUSION

The diagnosis of PJS should be considered in patients presenting with a clinical picture of bowel obstruction and mucocutaneous hyperpigmentation. If the diagnosis is made preoperatively, optimal management should include laparoscopic treatment of the bowel obstruction and intraoperative enteroscopy. If the diagnosis is made after the operation, the patient needs complete evaluation of their gastrointestinal tract. Surveillance is also mandatory for these patients.

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