Multi-Disciplinary Intervention for Intussusception in the Setting of Peutz-Jeghers Syndrome: A Case Report

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BACKGROUND

Peutz-Jeghers syndrome (PJS) is a rare autosomal dominant condition with varying characteristics, among the most common being multiple enteric hamartomas, dark macules of the lips, and increased risk of cancer, namely gastric. Many patients remain asymptomatic from PJS, however complications such as obstruction, abdominal pain, rectal bleeding, and rarely intussusception can occur. Intussusception and its treatment options in the setting of PJS is poorly characterized in the literature. Here we describe the multi-specialty management of a patient with recurrent intussusception secondary to PJS.

CASE

Our patient is a 39-year-old female with a history of PJS who presented with colicky abdominal pain. Her work-up including CT imaging revealed intussusception secondary to large distal jejunal polyps. Her initial episodes were self-limited. Eventually she progressed, requiring endoscopic intervention by gastroenterology (GI) involving the snaring of several extremely large polyps. She later developed worsening abdominal pain secondary jejunal intussusception that was not amenable to endoscopic intervention. She was taken to the OR with colorectal surgery in collaboration with gastroenterology for exploratory laparotomy along with concurrent endoscopy. After reduction, a jejunal enterotomy was made and GI advanced a colonoscope proximally to remove multiple large polyps with a snare. An additional polyp with a 3cm base was identified that was too large for snare polypectomy. An enterotomy was made so that a stapler could be introduced. This was fired across the base of the polyp without any narrowing of the lumen. The enterotomy was closed primarily and no bowel resection was required.

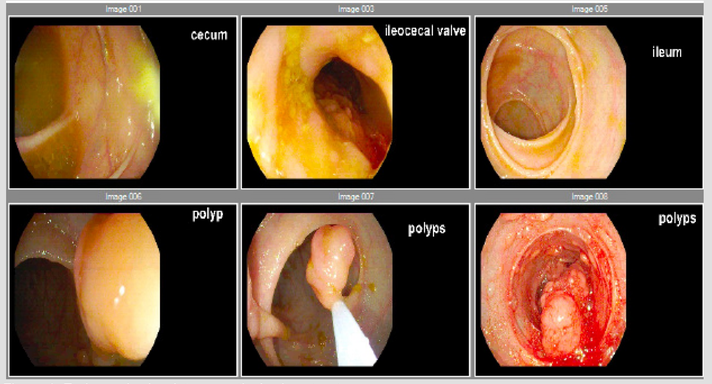


Figure 1: Endoscopic view intraoperatively via enterotomy

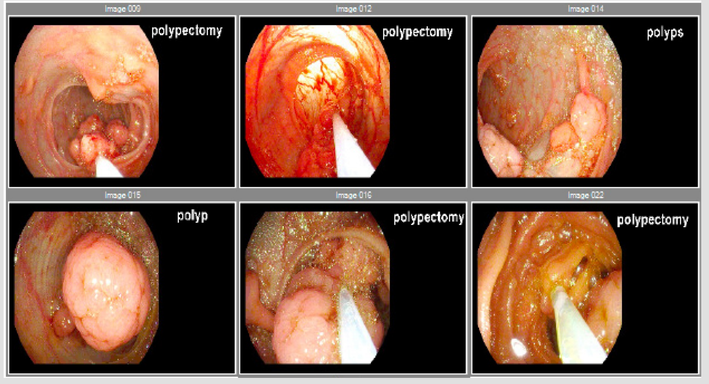


Figure 1: Endoscopic view of polyps and polypectomy

DISCUSSION

Intussusception resulting from PJS occurs in all age groups. Though not well characterized in the literature, the majority require surgical intervention. A combined approach involving exploratory laparotomy and intra-operative endoscopy has been safely described in the literature in a few case reports. This approach is advantageous because it allows for reduction of intussusception, direct inspection of bowel perfusion and a minimally invasive, bowel preserving way of removing lead point polyps. Without the aid of enteroscopy, a segmental bowel resection would be required in a patient that will likely require multiple future interventions. Intussusception in the setting of PJS is rarely reported and specific guidelines for treatment are not well defined. A multi-specialty collaborative intervention involving exploratory laparotomy and intra-operative endoscopy offers a safe and feasible intervention for patients with endoscopically unresectable polyps causing intussusception

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