



Gastric Outlet Obstruction in Peutz-Jegher Syndrome

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Abstract

Peutz-Jeghers Syndrome (PJS) is a rare polyposis syndrome commonly affecting the small bowel with recurrent obstruction due to polyp related intussusception. Although the stomach is the second most common site for PJS polyps, gastric outlet obstruction due to gastro-gastric intussusception is an extremely rare phenomenon. In this report we share our experience of a case of an acute gastric outlet obstruction, due to a large distal gastric polypoid mass, in a 26 year old female and present our approach to the management of this rare occurrence.

Introduction

Peutz-Jeghers Syndrome (PJS) is a rare autosomal dominant polyposis syndrome characterized by the presence of mucocutaneous pigmentation and hamartomatous polyps in the Gastrointestinal tract (GI) tract. The most frequent sites for polyps in PJS are small bowel (50%), stomach (36%) and colon (21%). The commonest polyp related complication is intussusception, which occurs in the small bowel in approximately 95% of the cases, resulting in Small Bowel Obstruction (SBO) [1]. Colon is the second most common site of intussusception in PJS. Although stomach is the second commonest site for PJS polyps, intussusception causing Gastric Outlet Obstruction (GOO) is a rare phenomenon with only a few cases described in literature. Here we report a 26-year-old female patient who presented acutely with GOO from gastro-gastric intussusception extending into duodenum due to a large pre-pyloric polypoid mass acting as a lead point. She underwent an emergency laparotomy and limited segmental gastrectomy. In the UK and Europe, the popular method of reconstruction after partial or total gastrectomy is with a Roux-en-Y (R-Y) configuration; however, in this case we performed a gastro-gastric anastomosis. This report highlights: a rare complication of PJS, the factors that need to be taken into consideration in the management of such cases, and our rationale for the reconstruction type.

Case Presentation

A 26-year-old woman presented with a two week history of upper abdominal pain and non-bilious vomiting. Otherwise, she did not have any other GI or systemic symptoms. Her past medical history included a diagnosis of Peutz-Jegher syndrome and three acute admissions with small bowel obstruction due to polyp-related intussusception. As a result, she underwent a laparotomy and segmental small bowel resection on each of the previous admissions at the ages of 17, 23 and 25 years. She was known to have polyposis-related chronic anemia, for which she was on iron supplements, but no other drug history. On examination, she was tachycardic but the rest of her hemodynamic parameters were within normal limits. On palpation, there was a tender mass in the upper abdomen but no peritonism. A midline laparotomy scar was visible. There were no signs of upper or lower GI bleeds on digital rectal examination. Laboratory profile was unremarkable except for the following abnormalities:

Hemoglobin - 80 g/L
Platelet count - $497 \times 10^9/L$
Albumin - 12 g/L
C-reactive protein level - 99.8 mg/L
Lactate level - 0.4 mmol/L

An intravenous contrast enhanced Computed Tomography (CT) scan revealed distal gastro-duodenal intussusception with dilated fluid-filled stomach proximally and multiple polyps throughout the stomach (Figure 1). After initial assessment and resuscitation with IV fluids and analgesia, her case was discussed urgently within the departmental multidisciplinary meeting after which she underwent a laparotomy. Intraoperatively, true gastro-duodenal intussusception was

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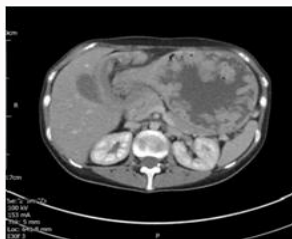


Figure 1: CT image demonstrating multiple polypoid lesions in the stomach and distal gastric intussusception into the duodenum.



Figure 2: Intraoperative photographs of gastro-gastric intussusception. (a) Invagination of the distal stomach into the duodenum (b) Pre-antral polypoid lesion acting as a lead point.

confirmed with invagination of the antrum and pylorus extending into the duodenum due to a large pre-pyloric polypoid mass acting as a lead point (Figure 2). Intra-operatively, the intussusception was reduced manually, a limited segmental gastrectomy involving most of the gastric antrum was carried out and a gastro-gastric anastomosis was performed. In the immediate peri-operative period, she was commenced on total parenteral nutrition via a central venous line in order to optimize her nutritional status during her recovery. She made an uneventful recovery and with input from the nutrition and dietetics team she was commenced on oral fluids on the third post-operative day, building up gradually over the course of her stay. She was discharged on the 7th post-operative day. At her 4 week review in clinic she had recovered well from her surgery and had resumed her normal oral intake. Her weight had been stable (Body mass index 19.5). Therefore, she was discharged back to the primary team caring for her PJS and surveillance.

Discussion

PJS is a rare condition with an incidence of 1 in 200,000 live births. It is caused by a mutation in the *STK11 (LKB1)* gene on chromosome 19p13. Those affected have hamartomatous polyps which can occur anywhere in the GI tract but the commonest sites are small bowel, stomach and colon. Extra-GI polyps have also been described in gallbladder, bronchi, urinary bladder and ureters [2]. Additionally, individuals with PJS have a significantly increased lifetime risk of gastro-oesophageal, small bowel, colonic and (in women) breast cancer. The risk of malignancy is independent of the polyp and whether these polyps undergo malignant transformation is disputed in current published literature [1]. The risk of the aforementioned PJS-associated malignancies is clearly of concern in the long term. This is significant particularly after the age of 50 where the risk of all cancers rises from 2% at age 20 to 31% at age 50 [3]. Indeed, current surveillance programs aim to identify any malignant changes early for a timely management. However, the commonest complication, throughout the lifetime of those affected with PJS, is SBO due to a polyp-related intussusception necessitating laparotomy in as many as 68% by the age of 18 years [1,2]. Moreover, repeat laparotomy

is required in nearly half of the cases within 5 years of their first presentation. Other polyp related complications include GI bleeding, anemia and abdominal pains; commonly presenting in the second or third decade of life [4]. Obstruction due to intussusception can compromise perfusion of the involved segment leading to ischemia and necrosis requiring a laparotomy and resection. Intussusception within small bowel occurs more frequently in the jejunum compared to ileum. However, more proximal polyps rarely present with obstruction, such as in our patient's case who was admitted with gastric outlet obstruction. Recurrent episodes of intussusception make subsequent laparotomies more complicated and, in the long run, risks loss of bowel length. Hence the rationale for surveillance from an early age, as young as 8-years-old, in asymptomatic cases with known PJS. According to current European guidelines for children and adults with PJS, upper GI endoscopy, colonoscopy and Video Capsule Endoscopy (VCE), or Magnetic Resonance Enteroclysis (MRE) are the recommended means of surveillance [2]. The role of endoscopic assessment during surveillance is not limited to detection of polyps but can also be used as therapeutic procedures. Endoscopic polypectomy can be performed successfully in the majority of cases without any significant complications, thus minimizing the risks of SBO and the associated undesirable consequences of an acute presentation [5,6]. Polyp size is an important factor in determining further management, as polyps larger than 15 mm are associated with an increased risk of intussusception [7]. Current guidelines do not recommend endoscopic reduction of polyp-related intussusception or polypectomy in an emergency setting despite reports of successful removal of larger polyps in the elective non emergency setting. This is due to the risks associated with the procedure itself, as well as risk of unrecognized ischemic bowel. Endoscopy in this setting can be used only for localizing a polyp for a more targeted surgery and intra-operative identification and management of any previously unknown polyps. Therefore, operative approach using enterotomy and polypectomy is the mainstay of management of SBO due to PJS polyps. The aim is to preserve bowel length if there is no evidence of irreversible ischemic changes. In our patient's case, emergency presentation with GOO due to a large polypoid mass as a lead point resulted in a non-salvageable distal stomach necessitating resection. The stomach is the second most frequent site for polyps in PJS; however, it is rare for such polyps to cause Gastric Outlet Obstruction (GOO). Only a few cases of GOO have been described in the literature so far [8-15]. To the best of our knowledge this is the first case to have required a gastric resection. Current European practice for reconstruction after gastric resection is commonly with a R-Y configuration whereas a Billroth I (BI) gastro-duodenostomy or limited segmental resections are more popular in East Asian countries [16]. The best reconstruction method has been the subject of various studies as each method of reconstruction has its advantages and disadvantages. In this case report, we describe the rationale for performing a segmental resection instead of a formal gastrectomy with BI or Roux-en-Y reconstruction.

- Preserving the normal passage of the food bolus into duodenum maintains the physiological continuity of the GI tract which is particularly important in young individuals with PJS to minimize metabolic complications.

- A segmental gastrectomy is technically simple and quick to perform which is advantageous in an emergency setting. An end-to-end stapled gastro-gastric anastomosis is safe with very low risk of stricture, bleeding or dehiscence.

- Preserving the pylorus maintains the normal gastric physiology, as well as preserving the endoscopic access to the duodenum and proximal jejunum for ongoing surveillance.
- Avoiding a jejunojejunostomy that could become the site of future intussusceptions.
- Avoiding the bypass of any length of duodenum and small bowel thus reducing absorbing surface.
- Avoiding the risk of internal herniation through the mesenteric and Petersen's defect and therefore the possibility of bowel ischemia and massive loss of small bowel length.

The main drawback of a BI reconstruction is dumping syndrome and bile reflux. Acid and bile reflux gastritis and oesophagitis can significantly impair the quality of life in the long run as well as potentially cause malignant changes [17]. The Roux-en-Y reconstruction presents fewer functional side effects, as the reflux into the gastric remnant is a rare event and dumping syndrome can be easier to manage with diet changes [17]. In an emergency setting when patients are nutritionally compromised and more hemodynamically unstable the risk of anastomotic leaks is higher [18]. There is also a risk of small bowel intussusception at the jejunojejunostomy. In addition to the peri-operative complications, there is also a risk of Roux limb stasis, internal herniation and nutritional complications, which is of concern in our patient's case due to her previous bowel resections and chronic anemia at the time of presentation. A Roux-en-Y reconstruction involves the by-pass of at least 80 cm of proximal jejunum and prevents the endoscopic access to the duodenum. Hence patients with PJS need careful evaluation for management of their acute presentation. In cases with known PJS, information from previous surveillance and surgical history is crucial in the management of short and long term goals, as well as informed decision making with equal patient involvement. One of the important aims of surveillance is effective and early identification of polyps to reduce the risk of emergency admissions in order to preserve bowel length in the long run. Presentations with acute SBO should be managed with as minimal bowel resection as possible, such as enterotomy, polypectomy or limited resections.

Conclusion

PJS is a rare polyposis syndrome which requires lifelong surveillance. Gastric outlet obstruction in PJS is an even rarer event; however, the principles of management remain the same as any enteric intussusception – preservation of the gastrointestinal tract by limited resection and consideration for future surveillance. Due to the rarity of GOO in PJS, this report also adds to the limited pool of previously published knowledge on this particular complication of PJS and serves as a platform for further discussion.

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