Recurrent Intussusception Secondary to Multiple Peutz-jegher polyps: An Unusual Case with Review of Literature

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ABSTRACT

Peutz-Jegher polyps are hamartomas of the small bowel which occur as part of the Peutz-Jegher syndrome, a rare autosomal dominant condition characterized by gastrointestinal polyps and mucocutaneous melanotic pigmentation. Here we report the case of a young male suffering from this syndrome who presented with acute intestinal obstruction due to intussusception. Exploratory laparotomy was done which revealed jejuno-jejunal and jejuno-ileal intussusceptions for which resection and anastomosis was done. There were multiple intraluminal polyps in the jejunum which on histopathological examination, showed features of Peutz-Jegher Polyp. Patient behaved well in the post-operative period and is still on follow up with us.

Key words: Hamartoma, Intussusception, Intestinal obstruction, Polyps, Peutz-jegher syndrome.

INTRODUCTION

Peutz-Jegher Syndrome is an autosomal dominant condition characterized by the combination of hamartomatous polyps of the intestinal tract and hyperpigmentation of the buccal mucosa, lips and digits. The polyps may cause bleeding or intestinal obstruction (from intussusception). While the hamartomatous polyps themselves only have a small malignant potential, patients with this syndrome are at greater risk of developing carcinomas of the pancreas, liver, lungs, breast, ovaries, uterus, testicles and other organs.

Here we report a case of multiple Peutz-Jegher polyps leading to intussusception and presenting with small bowel obstruction as an emergency. Such a presentation is rare in the Indian subcontinent and due to scarcity of literature on Peutz-Jegher polyps syndrome in India we felt reporting this case may be a noteworthy contribution.

CASE REPORT

A 15 years old, male presented to the emergency department with complains of colicky abdominal pain and bilious vomiting along with history of abdominal distention and non-passage of flatus and stools for two days. There was a past history of surgery for similar complaints at the age of nine years. Records of the previous illness were not available but the parents were well aware that the child had intussusception which was surgically corrected. Since then the child had been asymptomatic. On physical examination, there were pigmented lesions on the lips and tongue (Figure 1). Abdomen was distended with tenderness in the lower abdomen. There was a mobile, tender mass palpable in the hypogastrium measuring 7 × 4 cm with smooth surface and well-defined margins. The mass was resonant on percussion. Per rectal examination was normal. Ultrasonography of the abdomen showed telescoping of gut at multiple levels suggestive of multiple intussusceptions. The patient underwent an emergency exploratory laparotomy after resuscitation with intravenous fluids and antibiotics. Intra operative findings revealed two small bowel intussusceptions (jejuno-jejunal and jejuno-ileal) with gangrene of the gut loops in the jejunole ileal intussusception (Figure 2). We reduced the jejunole-jejunal intussusception and resected the gangrenous distal segment with end to end jejuno-ileal anastomosis. On careful examination of the proximal bowel, there were multiple palpable polypoidal lesions present intraluminally in a segment of jejunum which was also resected and after enterotomy of this segment, multiple polyps were noticed (Figure 3). Post-operatively the patient recovered well and was discharged on 6th post-operative day. Histopathological examination of the resected bowel segment confirmed presence of multiple hamartomatous jejunal polyps (Figure 4).

DISCUSSION

Peutz-Jeghers Syndrome (PJS) is rare autosomal dominant disease characterized by hamartomatous polyps in gastrointestinal tracts and mucocutaneous
Bhatia et al.: Intussusception due to Peutz Jegher polyps

Table 1: Surveillance guidelines (American College of Gastroenterology)\(^\text{[9]}\)

<table>
<thead>
<tr>
<th>Site</th>
<th>Age to begin surveillance (years)</th>
<th>Surveillance interval (years)</th>
<th>Surveillance procedure and comments</th>
</tr>
</thead>
<tbody>
<tr>
<td>Colon</td>
<td>8, 18(^\text{a})</td>
<td>3</td>
<td>Colonoscopy(^\text{a})</td>
</tr>
<tr>
<td>Stomach</td>
<td>8, 18(^\text{a})</td>
<td>3</td>
<td>Esophagogastroduodenoscopy(^\text{a})</td>
</tr>
<tr>
<td>Small Bowel</td>
<td>8, 18(^\text{a})</td>
<td>3</td>
<td>Video capsule endoscopy(^\text{a})</td>
</tr>
<tr>
<td>Pancreas</td>
<td>30</td>
<td>1-2</td>
<td>MRCP or Endoscopic ultrasound</td>
</tr>
<tr>
<td>Breast</td>
<td>25</td>
<td>1</td>
<td>Annual self-exam starting age 18, annual breast MRI, and/or mammogram starting at age 25</td>
</tr>
<tr>
<td>Ovarian</td>
<td>25</td>
<td>1</td>
<td>Pelvic exam and pelvic or transvaginal ultrasound, CA-125 probably not helpful</td>
</tr>
<tr>
<td>Endometrial</td>
<td>25</td>
<td>1</td>
<td>Pelvic exam and pelvic or transvaginal ultrasound</td>
</tr>
<tr>
<td>Cervix</td>
<td>25</td>
<td>1</td>
<td>Pap smear</td>
</tr>
<tr>
<td>Testicular</td>
<td>Birth to teenage years</td>
<td>1</td>
<td>Testicular exam, ultrasound if abnormalities palpated or if feminization occurs; 10 to 20% of benign Sertoli cell tumors become malignant</td>
</tr>
<tr>
<td>Lung</td>
<td>-</td>
<td>-</td>
<td>Provide education about symptoms and smoking cessation.</td>
</tr>
</tbody>
</table>

\(^{\text{a}}\)Start at age 8 years; if polyps present, repeat every 3 years; if no polyps, repeat at age 18, then every 3 years, or earlier if symptoms occur.

Clinical picture of the syndrome includes two major components: mucocutaneous pigmentation and hamartomatous polyposis of the intestinal tract. The pigmented lesions are often seen on the lips, around
the mouth, eyes, nostrils, on the buccal mucosa; and sparsely on the fingers, soles of the feet, palms, anal area and intestinal mucosa. Gastrointestinal polyps are most commonly found in small intestine, but can occur anywhere from the stomach to rectum. PJ polyps may lead to intussusception, some of which may spontaneously reduce while others lead to obstruction. Peutz-Jeghers polypos can also ulcerate, leading to acute blood loss or chronic anemia. Fewer than 5% of patients with PJS lack the abnormal mucocutaneous pigmentation, and fewer than 5% of patients with the pigmentation have no PJ polyps. The diagnosis is more likely possible in pediatric patients as pigmented lesions may fade at puberty. In a case series reported by Suda et al., solitary Peutz-Jeghers type hamartomatous polypos presented without pigmented spots in 87% cases, involved only one organ in the gastrointestinal tract, and showed a predilection for males in 74% cases.

The diagnostic criteria for Peutz-Jeghers syndrome proposed by the Johns Hopkins Registry include histopathologically verified hamartomatous polypos with at least two of the following: Small-bowel location for polyposis, mucocutaneous melanotic pigmentation, and a family history of Peutz-Jeghers syndrome. Our patient had histopathologically verified hamartomatous polypos with two of the additional criteria i.e. small bowel polyposis and mucocutaneous pigmentation. The evolution of PJS during life exposes the patient to a predisposition to developing malignant neoplasias, mainly in the small bowel, caecum, rectum, as well as pancreatic adenocarcinomas. Other more rare carcinomas could occur in the breast, uterus, testicles and ovaries. The risk for cancer in these patients is 18 times higher in comparison with the general population, and higher the number of intestinal polyposis higher is the chance for malignant transformation in PJS.

As the polyps can develop at any region in the entire GI tract, their recurrence is quite common, making it a challenge for the surgeon to decide regarding the amount of resections that should be done. Short-bowel syndrome is a well-known complication of repeated resections making the patient quite unstable due to malnourishment. The goal of treatment is to clear as much polyp as enough, and to avoid resections. Consequently, minimally invasive approaches are recommended and some of the authors advocate both the surgical and endoscopic techniques at the same session that is called "clean sweep" technique, in which the surgery team examines the whole intestinal tract with gastro-duodenoscopy, enteroscopy, push endoscopy and remove/destroy the smaller polypos to achieve a polyp free intestinal mucosa. This technique reduces the further need for abdominal surgery and complications. Patients with Peutz-Jeghers Syndrome should be educated about the symptoms of intestinal obstruction and the need for cancer surveillance.

Table 1 lists the surveillance guidelines as recommended by American College of Gastroenterology.

### REFERENCES
