A CASE OF DOUBLE INTUSSUSCEPTIONS SECONDARY TO PEUTZ JEGHERS POLYPS IN A 38 YEARS OLD MALE

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ABSTRACT

Peutz Jeghers syndrome is characterized by mucocutaneous hyperpigmentation and gastrointestinal polyps leading to gastrointestinal bleeding and obstruction. Larger polyps can lead to intestinal intussusception. We report a case of 38 years old male presenting to the Emergency Department of Lady Reading Hospital Peshawar, with jejuno jejunal double intussusception.

Key Words: Double intussusception, Gastrointestinal polyps, Peutz Jeghers syndrome

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INTRODUCTION

Peutz Jeghers syndrome (PJS), an autosomal dominant hereditary disorder which was first described by a Dutch pediatrician Johannes Peutz in 1921 and in 1949 by Harold Jeghers. It has an incidence of 1 in 50 thousands to 2 million[1]. PJS is characterized by gastrointestinal polyps and mucocutaneous hyper-pigmentation, that occurs in 90% of the patients[2,3]. The prevalence of polyps in the gastrointestinal tract is said to be 32% in rectum, 48.6% in the stomach, 63.2% in colon and 64% in the small intestine[4]. Polyps larger than 15mm can result in intussusceptions[3]; a complication of PJS, 47% of which occur in the jejunum[1,3]. Jejunal polyps can result in gastrointestinal bleeding and obstruction on their own or due to intussusceptions[3]. PJS is also marked as a common cause of mortality[5]. In acute condition, the patient presents with abdominal pain along with mucocutaneous pigmentation[6]. As per author's knowledge, there are no reports of double jejuno-jejunal intussusceptions making the present case study a rare condition.

CASE PRESENTATION

A 38 years old male presented to the emergency department with the complaints of constipation, abdominal pain and per rectal bleeding for 2 days. Pain was of sudden onset, moderate and diffuse and more in the left iliac fossa. It was associated with vomiting, which was greenish in color and projectile. Per rectal bleeding was large in amount and dark red in color.

The patient had a history of haemorrhoidectomy 7 years back (record was not available). About 3 years back, he had experienced abdominal pain and bleeding per rectum for which colonoscopy was done. Colonoscopy findings were: multiple, large and small polyps in the sigmoid colon and rectum. Polypectomy was done for a large pedunculated polyp present in the sigmoid colon and the rest of polyps were not resected due to the patient's discomfort. The polyp was then sent for biopsy and reported as Puetz-Jegher's polyp for which the patient had a positive family history. Examination revealed an ill looking, pale and tachycardiac patient. Abdomen was distended, tense and tender, more on the left iliac fossa than the right with no bowel sounds. There was mucocutaneous hyper-pigmentation. Digital rectal examination was done. Inspection revealed external hemorrhoides, empty rectum and polyps felt by finger at 6cm from the anal verge. Finger was stained with dark red blood and anal tone was found to be normal.

In emergency department, he was started on IV fluids and IV antibiotics. Ultrasound abdomen and pelvis showed distended bowel loops with fecal material, no peristalsis and mild interloop ascites. Blood count showed anemia and leukocytosis; liver function tests were normal except for ALT which was 136 u/l; blood urea was 65 mg/dl and creatinine was 1.5 mg/dl. Sodium was low, i.e. 115 mmol/l and serum amylase was 36 u/l. The patient was shifted to the surgical ward where the patient had 3 units of blood transfusion for anemia due to ongoing bleeding per rectum.

The patient was further evaluated and a plan for CT abdomen with contrast was made. CT report showed a long segment of 18 cm of proximal jejunum is telescoping along with mesentery and the mesenteric vessels into the distal jejunum which was fluid filled and

dilated (forming pseudo kidney sign). Multiple enlarged lymph nodes were noted in the accompanying mesentery along with mesenteric vessels. Thus diagnosis of jejuno-jejunal intussusception was made as the case fulfilled the clinico-pathological diagnostic criteria given by WHO.

Patient was prepared for surgery. Exploratory laparotomy revealed jejuno-jejunal intussusception (double) with ischemic non-viable gut (four and a half feet length of jejunum) 4 inches distal from duodenojejunal junction. Resection of non-viable jejunum was done with the primary end to end anastomosis. Specimen was sent for histopathology. The results of histopathology showed 1 Puetz-Jegher's polyp and 1 hyperplastic polyp and rest of the polyps showed extensive infarction. No high grade dysplasia or invasive tumor was seen. The bowel wall showed extensive infarction and 12 reactive lymph nodes. The post-operative course was uneventful. Patient was stable and discharged on 5th post-operative day. Follow up in outpatient was uneventful.

Figure 1: CT abdomen showing jejuno-jejunal intussusception (pseudo kidney sign)

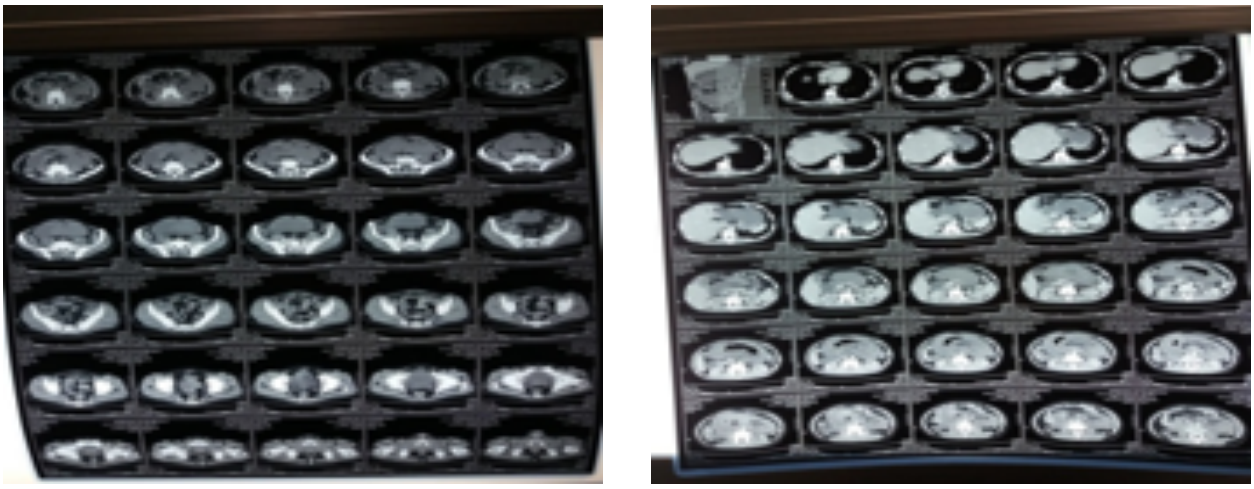
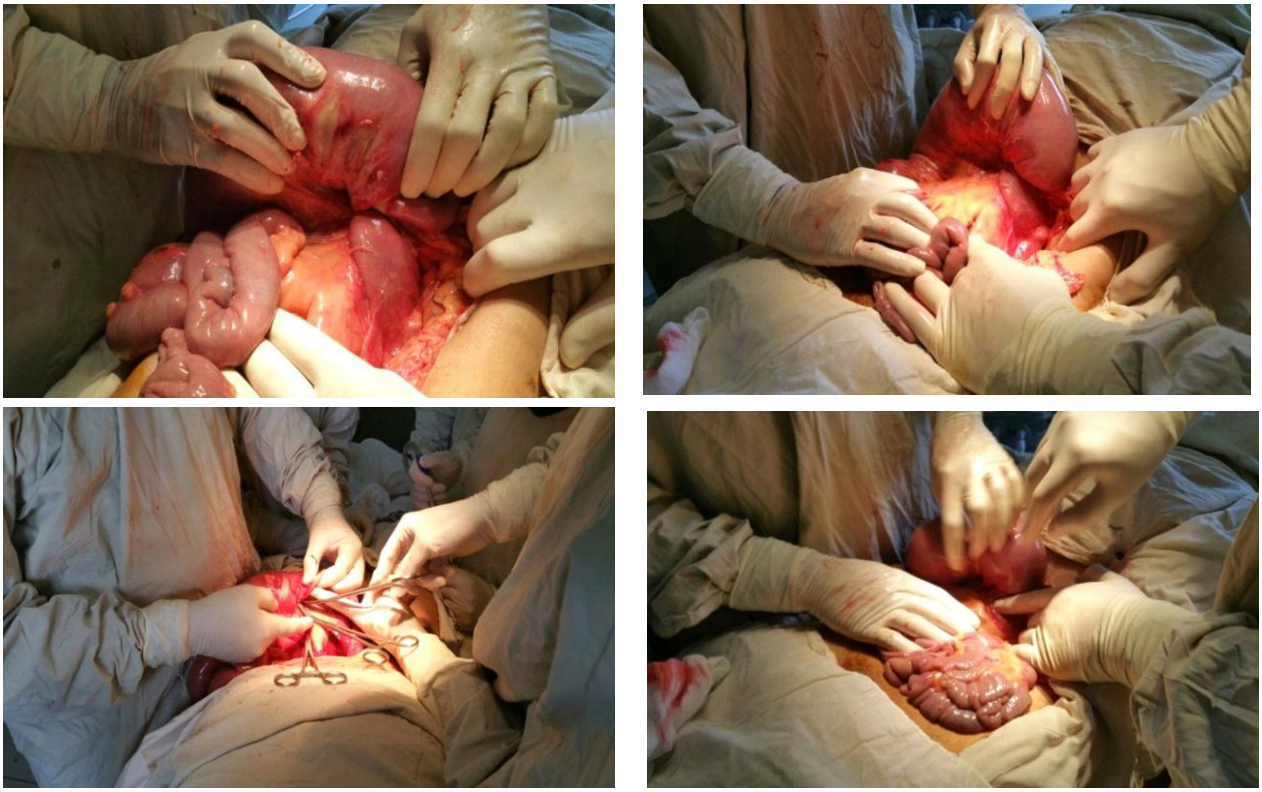


Figure 2: Per operative view of dilated duodenum and distal collapsed gut loops due to intussusception



DISCUSSION

Peutz-Jeghers syndrome, also known as hereditary intestinal polyposis syndrome, is associated with production of gastrointestinal benign hamartomatous polyps along with mucocutaneous pigmentation[7]. It appears due to the mutation of the STK11 gene, a tumor suppressor gene, located on band 19p13.33[8]. Thus patients with PJS are at higher risk to develop cancers of gastrointestinal tract, uterus, breast, ovaries and testicles[9]. The incidence of PJS is reported to be 1 in 8300 to 1 in 280,000, appearing equally in both the genders.

The reported prevalence of PJS is 1 in 0.1 million[10,11]. The clinico-pathological diagnostic criteria given by WHO for this rare disorder includes family history of PJS, mucocutaneous pigmentation, 3 or more polyps showing histological features similar to PJS and mucocutaneous pigmentation with any number of Peutz Jeghers polyps[12]. Clinically, patients with PJS present with 2 major symptoms, namely: hamartomatous polyps in the gastrointestinal tract and hyperpigmentation of digits, nostrils, eyes and peri and intra oral regions[8,10]. Polyps of the gastrointestinal tract can occur anywhere from stomach to rectum though they appear most commonly in the small intestine; the most common site being the jejunum[3]. This can lead to intestinal intussusception along with obstruction[8].

Intestinal intussusception is defined as "telescoping of one segment of bowel into another one". It is a rare disorder in adults and uncommon in children with an incidence of 1 in 1300 abdominal obstruction cases[13]. It appears due to unequal motility of adjacent intestinal segments. The proximal intestinal segment is labelled as intussusceptum and the distal as intussuscipient. Of all intussusceptions 5% appear in the adults and 1% of all obstructions occur in the bowel[14]. Intestinal intussusception occurs about 65-75% in gastric antrum, 3-8% in colon and ileo-colon, 17-21% in small bowel, 1% in duodenum, 1% in esophagus and 1% in gallbladder and jejunogastric region[15]. It results mostly due to intestinal polyps larger than 15 mm in diameter[16].

Intestinal double intussusception is a very rare condition in adults, the first case of which was reported in 2013 in a 46 years old woman[17]. Another case of jejunogastric intussusception with jejunojejunal intussusception in 2013 was a 55 years old woman[18]. El-Hage-Chehade et al[19] in 2013 reported another case of double ileoilial intussusception in a 26 years old male secondary to Burkitt's lymphoma. No other reports of double telescoping of intussusceptions are found in the literature[17]. Intussusception is a rare condition in adults while double intussusception is extremely rare[19]. Any gastrointestinal region can be affected by polyps and can recur. Thus the decision of amount of resection is challenging as patients can develop short bowel syndrome, a well-known complication of repeated resections, as the patient gets unstable due to malnourishment[8]. Diagnosis is done mostly in 2nd or 3rd decade of life with presentation of abdominal pain, bowel obstruction, intestinal intussusception, rectal prolapse of polyps with bleeding and anemia[11].

Standard treatment for PJS include laparotomy and resection of symptomatic gastrointestinal polyps that result in intussusception. Some patients might require surgical resection multiple times. Therefore endoscopy is recommended to remove all polyps so that short bowel syndrome can be avoided. Gold standard diagnostic criteria is the use of double balloon enteroscopy along with capsule enteroscopy[11]. Education regarding symptoms of intestinal obstruction and regular cancer surveillance should be provided to the patients[5].

CONCLUSION

Intestinal obstruction due to intussusception is a common presentation in pediatric population but Peutz Jeghers polyps syndrome leading to intussusception is a rare presentation. Most cases require surgical management. Thus in cases of intussusception bowel should be examined for intestinal polyps and PJPs.

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