

Rare presentation of Recurrent Multiple Intussusceptions in a case of Post Total Colectomy for Peutz-Jeghers Polyposis

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Abstract: Peutz-Jeghers Syndrome is rare hereditary polyposis condition. Intussusception is a known complication of Peutz-Jeghers Polyposis. Recurrent adult intussusception is a very rare condition in a post total colectomy case for Peutz-Jeghers Polyposis. We report a case of adult intussusception in a 24 year old male, 8 years post total colectomy and ileo-rectal anastomosis for intussusception due to Peutz-Jeghers Polyposis, who presented with features of intestinal obstruction. Exploratory laparotomy revealing multiple Jejunum-Jejunal intussusceptions and Ileo-Rectal intussusception.

Key Words: adult, intussusception, multiple, Peutz-Jeghers Polyp, recurrent.

I. Introduction

Peutz-Jeghers Syndrome (PJS) is a complex hereditary polyposis condition. It is a rare disorder which is characterized by typical pigmented perioral macules, pigmented spots in the buccal mucosa which are present in 90% of patients, and the presence of multiple hamartomatous polyps predominantly in the gastrointestinal tract. Polyps may occasionally be absent. Polyp sizes may vary from a few mm to 6 or 7 cm. Most of the patients have a characteristic clinical course of recurrent episodes of polyp induced bowel obstruction and bleeding. The disease affects males and females equally.

We report the case of a 24 year old male patient, 8 years post total colectomy and ileo-rectal anastomosis for intussusception caused due to multiple polyposis colon, who presented in Emergency department with features of intussusception. Although intussusception has been reported in PJS, literature reviews had shown that there are no cases reported for multiple intussusceptions following Total Colectomy in Peutz-Jeghers Polyposis. Therefore we report this quite rare case

II. Case Report

A 24 year old male patient presented in Emergency department with complains of left sided abdominal pain associated with passing bloody mucoid stools for past 3 days. Patient had undergone total colectomy and ileo-rectal anastomosis at the age of 17 for multiple polyposis when he presented with acute intestinal obstruction due to coli-colic intussusception. Patient has family history of Familial Adenomatous Polyposis. His mother was found to have same disease and died of its complication.

At the time of presentation in Emergency Department his B.P was 100/60 mm of Hg, heart rate was 120/min and saturation was maintained in 99% in room air. Physical examination revealed melanin pigmentation over lips, buccal mucosa and feet (Photo plates 1 & 2). His abdomen was distended, a tender palpable mass in umbilical region and left iliac fossa. Per rectal examination showed finger staining with bloody mucoid stool. His hemoglobin level was 9.3 g/dl, total count was 21500 /cc.mm, PT INR was 1.36, total bilirubin was 1.8 mg%, total protein was 5.5 g%, serum albumin was 3 g%, platelet count was 391000 and his renal parameters were normal. Emergency CECT Abdomen with Angiogram revealed long segment intussusceptions in left lower quadrant with loss of vascularity, jejunum-jejunal intussusceptions and left PUI calculus (Photo plate 3). Emergency exploratory laparotomy was done. Patient had ileo-rectal intussusception with gangrene ileum of 35 cm in length, multiple jejunal polyp, multiple jejunum-jejunal intussusceptions partially reduced and multiple mesenteric lymph nodes (Photo plates 4,5&6). He underwent resection of involved segment of ileum and end ileostomy and resection of involved segment of jejunum and jejunum-jejunal anastomosis.

Histopathology examination of specimen showed features of intussusceptions and gangrene of small intestine. HPE of polyps was consistent with a diagnosis of Peutz-Jeghers Polyp. HPE of lymph node showed reactive hyperplasia.

Post-operative recovery was uneventful and was planned for subsequent ileo-rectal anastomosis which the patient underwent after 3 months.

III. Discussion

Peutz-Jeghers Syndrome was described by Jan Peutz, a Dutch physician in 1921, who noted a relationship between the intestinal polyp and mucocutaneous macules in Dutch family. Later in 1949, Harold Jegher, an American physician gave a descriptive reports of Peutz-Jeghers Syndrome. The eponym Peutz-Jeghers syndrome was introduced by the radiologist Andre J. Bruwer in 1954. It's an autosomal dominant inherited disorder characterized by intestinal hamartomatous polyps associated with skin and mucosal melanin deposition [1]. The incidence of Peutz-Jeghers is approximately 1 in 120000 births [2].

The cause of Peutz-Jeghers Syndrome (PJS) in most cases (66-94%) appears to be a germline mutation of the *STK11/LKB1* (serine/threonine kinase 11) tumor suppressor gene, located on band 19p13.3. [3]

A clinical diagnosis of PJS can be made when an individual has two or more of the following features, [6],

1. 2 or more PJS polyps of small intestine.
2. Characteristic pigmentation of mouth, lips, nose, eyes, genitalia, or fingers
3. Family history of PJS.

Affected patients during first 3 decades of life present with manifestation of gastro-intestinal bleeding as rectal bleeding and anemia and with recurrent abdominal pain caused by intestinal obstruction and/or intussusceptions [5, 6]. The sites which are most commonly affected by Peutz-Jeghers polyps in the gastrointestinal tract are the small bowel (64% of patients), colon (63.2%), stomach (48.6%) and rectum (32%) in decreasing frequency [7]. The incidence of polyp in small intestine is greatest in jejunum and progressively decreases in the ileum and duodenum [6]. The occurrence of adult intussusception is rare, and most commonly involves small intestine [8, 9, 10]. Whereas in case of children its common and large intestine is mostly affected [11]. The pedunculated nature of polyps, combined with large size to which it can grow can lead to recurrent intussusception in small bowel [2]. There is an increased risk of gastrointestinal and extra intestinal malignancies such as tumor breast, pancreas, lung and reproductive tract in PJS patients. They have 15 – fold increased risk of developing intestinal cancer compared with that of general population [2]. A histological evidence of a hamartomatous-adenomatous-carcinomatous evolution and a direct hamartoma–carcinoma sequence in PJS patients have been documented in the literature. In addition, PJS patients can have both adenomatous and hamartomatous polyps separately, especially in the large intestine, and a malignant transformation of a small bowel hamartoma to a leiomyosarcoma has been reported [2]

Multicentric study by Barussaud M, Regenet N, Briennon X, de Kerviler B, Pessaux P and Kohneh-Sharhi showed that CT associated/not associated with barium enema may be the most accurate modality for diagnosis of adult intussusception. A definitive surgical resection remains the recommended treatment in most cases of PJS which are associated with intussusception, obstruction or persistent intestinal bleeding [12].

Recent advances in genetic testing, magnetic resonance enterography, double ballooning endoscopy, IOE (combination of laparotomy with endoscopy) and capsule endoscopy, will all result in a timely diagnosis and management of patients with PJS. Any polyp more than 1.5 cm should be removed if possible, as it generally causes intussusception. Colon should be surveyed every 2 years endoscopically and should be screened for malignancies of GI tract [13]. Relatives should also be screened for polyposis coli.

In our case of Peutz-Jeghers syndrome, multiple intussusception (ileo-rectal and jejuno-jejunal) occurred 8 years after total colectomy which was done for colo-colic intussusception caused due to polyposis coli. This is a very unusual case and to the best of our knowledge, similar case has not yet been reported.

IV. Photo Plates



Photo plate 1) Pigmentations in oral mucosa



Photo plate 2) Pigmentations in the sole



Photo plate 3) CT Abdomen showing Ileo-Rectal intussusception

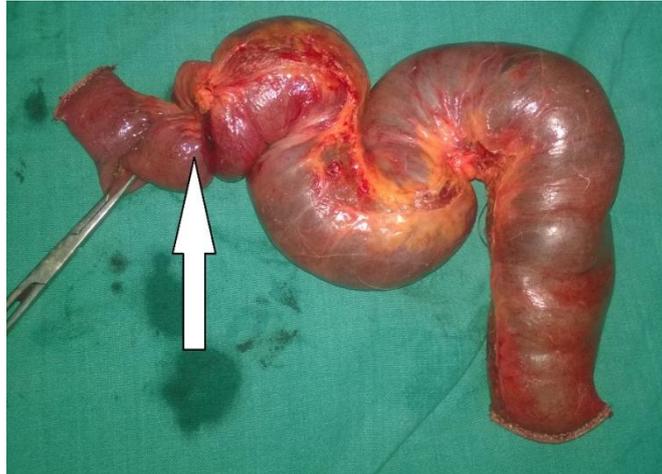


Photo plate 4) Resected segment of ileum showing ileo-rectal intussusception



Photo plate 5) Multiple polyps initiating lead points for intussusceptions

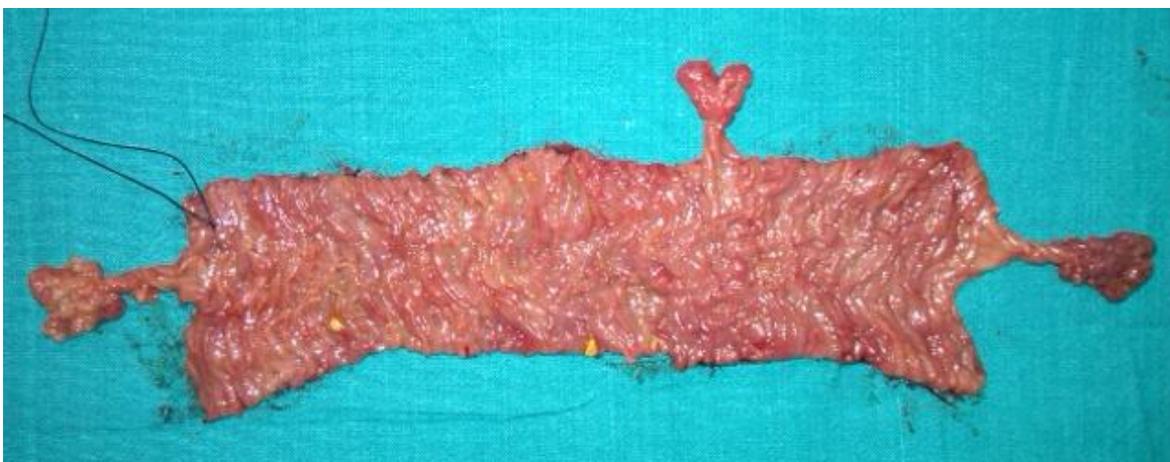


Photo plate 6) Resected segment of jejunum showing multiple pedunculated polyp

V. Conclusion

Recurrent Multiple Intussusception in a post total colectomy done for Peutz-Jeghers Syndrome and multiple polyposis is a very rare presentation. Such a post total colectomy patient coming with colicky abdominal pain should be evaluated with CT Abdomen, enteroscopy, Capsule Endoscopy etc. with a clinical suspicion of intussusception.

Routine surveillance with endoscopy will help in early detection of polyps and endoscopic resection is possible in an earlier stage. There by decreasing the need for bowel resection and subsequent complication of Short bowel syndrome.

References

- [1]. Cunningham JD, Vine AJ, Karch L, Aisenberg J. The role of laparoscopy in the management of intussusception in the Peutz-Jeghers syndrome: case report and review of the literature. *SurgLaparosc Endosc*1998 ; 8(1):17-20
- [2]. McGarrity TJ, Kulin HE, Zaino RJ. Peutz-Jeghers syndrome. *Am J Gastroenterol*. 2000;95:596-604.
- [3]. Sokmen HM, Nce AT, Bolukba C, Kilic G, Dalay R, Kurda OO . A Peutz - Jeghers syndrome case with iron deficiency anemia and jejuno - jejunal invagination. *Turk J Gastroente-rol* 2003; 14(1):78-82.
- [4]. Schreiberman IR, Baker M, Amos C, McGarrityTJ. The hamartomatous polyposis syndromes: a clinical and molecular review. *Am J Gastroenterol* 2005; 100(2):476-490
- [5]. Zbuk KM and Eng C. Hamartomatous polyposis syndromes. *Nat ClinPractGastroenterolHepatol* 2007; 4(9):492 – 502
- [6]. Gammon A, Jasperson K, Kohlmann W, Burt RW. Hamartomatous polyposis syndromes. *Best Pract Res ClinGastroenterol* 2009; 23(2):219 – 31
- [7]. Utsunomiya J, Gocho H, Miyanaga T, Hamaguchi E, Kashimure A. Peutz -Jeghers syndrome: its natural course and management. *Johns Hopkins Med J* 1975; 136(2):71- 82
- [8]. Laws HL and Alderate JS: Small bowel obstruction a review of 465 cases. *South Med J* 1976; 69:733- 734.
- [9]. Stewardson RD, Bombect CT, NyhersLM.Critical operative management of small bowel obstruction. *Ann Surg* 1978; 187:189 – 193
- [10]. Briggs DF, Arpathios J, Zollinger RW. Intussusception in adults. *Am J Surg* 1961; 101:109- 113
- [11]. Hinds R, Philp C, Hyer W, Fell JM. Complications of childhood Peutz - Jeghers syndrome: implications for pediatric screening. *J PediatrGastroenterolNutr* 2004; 39(2):219 - 20.
- [12]. Barussaud M, Regenet N, Briennon X, de Kerviler B, Pessaux P, Kohneh-Sharhi N, et al. Clinical spectrum and surgical approach of adult intussusceptions: a multicentric study. *Int J Colorectal Dis*. 2006; 21:834-39.
- [13]. Mahmoud Najjia, Rombeau John, Ross Howard M, Fry Robert D. Sabiston Textbook of Surgery. 17th ed. New Delhi: Elsevier Publishers; 2005. Colon and Rectum, In: Townsend MC, editor; pp. 1451-52.