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Case Report

A case of recurrent intussusception in young adult female: Peutz Jegher syndrome

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ABSTRACT

Peutz Jegher syndrome (PJS) is an autosomal dominant inherited polyposis syndrome, in which multiple characteristic polyps occur in the gastro-intestinal tract associated with mucocutaneous pigmentation especially of vermillion border of the lips. Small bowel obstruction is presenting complaint in half of the cases-intussusception due to polyp being lead point is the high risk for obstruction. A 21 year old female patient with presented to emergency department with complaining of intermittent episodes of abdominal pain and multiple episodes of billious vomiting since 7 days. Patient had history of exploratory laparotomy 10 years back for similar complaints. On examination, melanocytic pigmentation present over vermillion border and palmar aspect. Enhanced computed tomography reveals telescoping of the distal jejunum to proximal ileal loops s/o jejuno-ileal intussusception with ileo-ileal intussusception were noted. On surgical decision, patient again underwent exploratory laparotomy which revealed intussusception with non-viable small bowel segment were found, Small bowel resection was performed and jejuno-ileal anastomosis was done. Patient follow up was maintained to go with colonoscopic evaluation. In conclusion, recurrent intussusception is a rare cause of acute abdomen in adults. A high index of suspicion and appropriate investigations are helpful in prompt diagnosis. Thorough evaluation intra operative and then regular periodic follow up is required in such cases for early detection of disease before complication occurs.

Keywords: Recurrent intussusception, Peutz Jegher syndrome, Colonic polyp, Lead point

INTRODUCTION

Peutz Jegher syndrome (PJS) is an autosomal-dominant hereditary inheritance which is associated with development of gastro intestinal polyps.[1] PJS affects around 1 in 8300 to 2,00,000 births.[2] Peutz Jegher was first reported by Peutz in 1921 and subsequently detailed explanation by Jegher, McKusick and katz in 1949.[3-5] This syndrome has association of gastrointestinal polyps with mucocutaneous pigmentation and familial incidence.[6]

In syndrome, gastrointestinal mucosa develops hamartomatous polyp which occur most commonly in the small bowel, colon and rectum (>90% of cases) and less commonly in the stomach or urinary tract. Hamartoma are associated with intussusception, bledding, anemia and obstruction.[7,8]

Here, we reported the uncommon case of recurrent intussuception in young female with previous history of bowel resection with presentation of jejunoileal, ileoileal intussusception due to PJS.

CASE REPORT

21 year old female patient came to emergency department with chief complaint of intermittent episodes of colicky abdominal pain with multiple episodes of billious

vomiting since 7 days and had similiar episodes before admission which resolved spontaneously. Patient gives H/o similiar complaints 10 years back for which she underwent exploratory laparotomy with gangrenous jejunal segement resection and jejuno-ileal anastomosis done.

On examination patient was having pulse rate 130/min and BP 100/60 mm Hg (tachycardiac and hypotensive). On physical examination, she had melanocytic pigmentation over palmar aspect of hand and oral cavity (Figure 1). Per abdominal examination shows, distended abdomen with diffuse tenderness all over abdomen and sluggish bowel sounds. Digital rectal examination revealed normal limits. Blood examination Hb 10.5 mg/dl with TLC 9500 ccmm, X-ray abdomen showed multiple dilated air fluid levels. CECT A+P s/o. There is telescoping of jejunum with adjacent mesentery and mesenteric vessels into proximal ileal loops s/o jejuno-ileal intussuscption. There is another short segment telescoping of the distal ascending colon with transverse colon along with adjacent mesocolon s/o colo-colic intussusception.

Short segement telescoping of proximal ileum into mid ileum with adjacent mesentery and mesenteric vessels s/o ileo-ileal intussuscption. The patient underwent emergency exploratory laparotomy, which reveals multiple (3) intussusception-jejunoileal, ileo-ileal and colo-colic. There is abrupt cutoff seen at 150 cm from DJ junction with intususception segement involving polyp as lead point, distal to it bowel loops appears collapsed with compromised vascularity. A segemnt of 25 cm found non-viable and decision of resection was made.

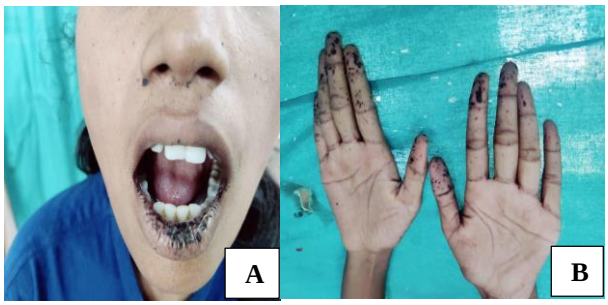

Figure 1: Melanin deposits over (a) vermillion border and (b) palmar aspect.

Non polyp segement margin were identified and anastomosis made between proximal jejunum-distal ileum (Figure 2). She was given broad spectrum antibiotics and analgesics for 7 days. Post-operatively patient had satisfactory progress, tolerate

ng oral feeding and was discharged on 14 th day. Resected bowel segment histopathology revealed polyp characteristic of PJS (Figure 3).

Further detailed family history non-contributory. Patient was kept on follow up- after 6 week colonoscopy was performed to look for polyposis which reveals rectosigmoid junction three polyps of size 1.5 cm, three diminutive polyps and polyp of size 2 cm with short stalk, descending colon-pedunculated polyp of size 3 cm was seen, at splenic flexure, a 3 cm pedunculated polyp seen, distal transverse colon two pedunculated polyps of size 2×1 cm seen, at hepatic flexure a polyp of size 3 cm was seen, distal ileum- multiple subcentimeter polyps are seen, colonic polyposis in K/c/o PJS (Figure 4).

Figure 2: Resected bowel segment showing polyp - lead point for intussusception.

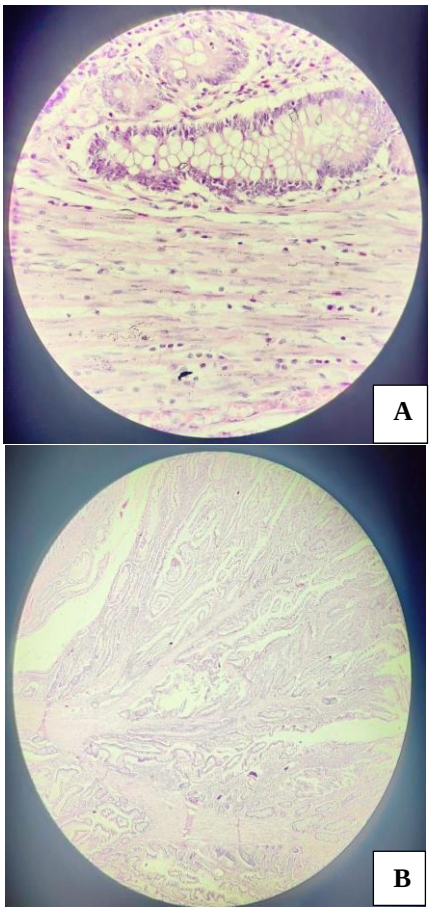

Figure 3: Histopathology slide showing hamartomatous polyp (characteristic of PJS).

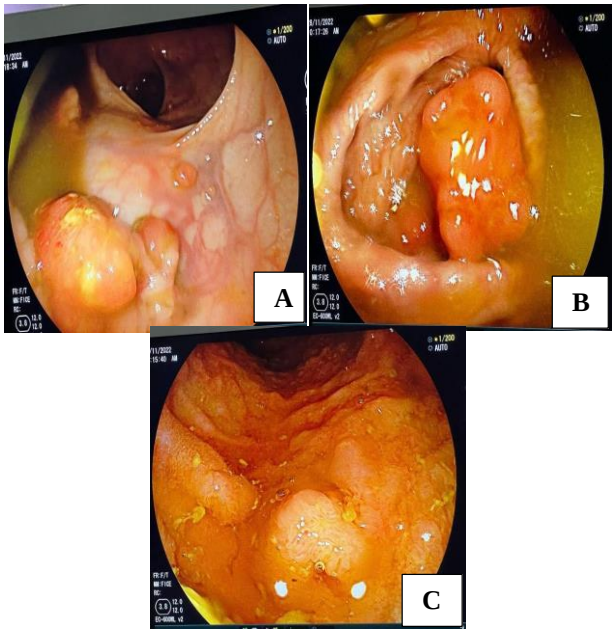

Figure 4: Showing colonoscopic view of polyps on f/u period.

DISCUSSION

PJS is an inherited polyposis syndrome in which multiple characteristic polyps occur in the gastrointestinal (GI) tract associated with mucocutaneous pigmentation, especially of the vermilion border of the lips. PJS is an autosomal dominant disease caused by a germline mutation in the STK11 (LKB1) gene.[8] The estimation of the population prevalence of PJS differs between studies, ranging from 1 in 8300 to 1 in 280,000 individuals. The probable prevalence is around 1 in 100,000 people.[2] The diagnosis of PJS can be made in patients with hamartomatous polyp(s) with at least two of the following clinical criteria also present: labial melanin deposits, Family history of the syndrome, small bowel polyposis The syndrome appears equally in males and females and is found in all racial groups. The clinical manifestation of PJS is abdominal pain with complications, such as abdominal pain, intussusception often leading to intestinal obstruction, polyp extrusion through the rectum, and bleeding, which is often occult.[7,9]

Small bowel obstruction is the presenting complaint in half of the cases, and relaparotomy polyp-induced complications occur commonly and might do so at quite short intervals. In addition to polyposis, previous studies have reported an increased risk of GI and extra-GI malignancies in PJS patients, compared with that of the general population. The PJS is associated with an increased risk of gastrointestinal and non-gastrointestinal malignancies.[7]A meta-analysis involving six studies and 210 patients showed a cumulative risk of 93% from 15 to 64 years for all types of malignancies.[9,10] Thus, the relative risk of an individual with PJS to present neoplasia in any region, compared with the general population, is up to 15 times higher. The most frequent neoplasm in patients with PJS is the colonic tumor (57%), followed by breast (45%), pancreas (36%), stomach (29%), ovary (21%), small intestine (13%), and uterus (10%) tumors.

Among gastrointestinal cancers, increased cancer risk was indicated for the colon, stomach, small intestine, and pancreas. Also, female PJS patients are at greater risk of gastrointestinal and gynecological cancers, ovarian cancer, cervical cancer, uterine cancer, and breast cancer. Meanwhile, the high risk of pulmonary cancer, renal cancer, prostatic cancer, bone cancer, and leukemia has also been reported. There is wide variability in cancer risk estimates, as reviewed in a recent meta-analysis study. PJS patients requires special surveillance that includes multiple organs, as it is associated with increased risk of cancer in many organs. Screening begins at 8 to 10 years of age with an evaluation of small bowel. If initial exam is normally, a repeat evaluation is recommended at 18 years of age and then at 2-3 year intervals.[11]

In our case, the patient was admitted to the hospital with recurrent intussusception. The exploratory laparotomy revealed intussusception (lead point being polyp) with non-viable small bowel segment. Small bowel resection was performed and side to side jejuno-ileal anastomosis done.

CONCLUSION

Recurrent intussusception is a rare cause of acute abdomen in adults. A high index of suspicion and appropriate investigations (USS, barium enema and CT scan) can result in prompt diagnosis. Unlike children 75% of cases are due to malignant tumours in the small bowel or colon. The extent of resection and operative technique depend upon the age of the patient, results of investigations (benign or malignant) and the length of the bowel involved. Through evaluation intra-op and post-op follow up is required in such case to prevent further progression of disease.

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