



JEJUNAL-JEJUNAL INTUSSUSCEPTIONS DUE TO PEUTZ-JEGHERS POLYPS : A RARE CASE REPORT AND REVIEW OF LITERATURE.

General Surgery

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ABSTRACT

Peutz-jeghers syndrome is characterized by perioral pigmented macules and pigmented spots in the buccal mucosa in 90% of the patients and multiple hamartomatous polyps in the gastrointestinal tract. A 14 year young girl presented to our accident and emergency with complaint of severe pain abdomen for two days and non-passage of stool and flatus for one day. On local examination there was generalized peritonitis. Since she was in acute abdomen, so taken up for emergency laparotomy after basic investigations. On through examinations of gut we found there was telescoping of 20 cm of jejunal segment into the distal jejunum starting nearly 50 cm distal to duodenojejunal junction. A polyp was also present nearly 30 cm distal to duodenojejunal junction.. Histopathological examination of the polyp was reported as peutz-jeghers polyp. Peutz-jeghers syndrome may present as intussusception and a complete history and examination may help in the diagnosis.

KEYWORDS

Peutz-Jeghers, Intussusception, Jejunal-jejunal, Polyp, Hamartomatous.

INTRODUCTION

It was first described by Y. Peutz in a family with skin pigmentation and polyps of small intestine in 1921 and H. Jeghers et al in 1949 (1) and consequently the name of Peutz-Jeghers syndrome (PJS) was adopted for this disorder. The risk of gastrointestinal and extragastrointestinal malignancies is 13 times and 9 times higher compared to general population (2). Although the risk of malignant transformation of hamartomatous polyps is rare but the overall risk of gastrointestinal and extragastrointestinal malignancies is increased (3). Here we present a very interesting case of peutz jeghers polyp in a 14 years young girl who presented with symptoms of intestinal obstruction and was diagnosed as a case of intussusception due to Peutz-Jeghers polyps.

CASE REPORT

A 14 year young girl presented to our accident and emergency with complaint of severe pain abdomen for two days and non-passage of stool and flatus for one day. There was history of 4 to 5 episodes of vomiting containing ingested food material. There was no history of similar complains in the past. There was no history of any gastrointestinal malignancy in the family. There was no hyper pigmentation of mucosa or skin. Abdomen was distended, generalised tenderness was present and bowel sounds were increased. There was a sausage shaped lump palpable in the left lumbar region which was mobile and was not moving with respiration. Ultrasonography and CECT abdomen revealed ileo-ileal intussusception with dilated proximal small bowel.

Patient was taken up for emergency laparotomy. There was telescoping of 20 cm of jejunal segment into the distal jejunum starting nearly 50 cm distal to duodenojejunal junction (Fig 1). A polyp was also present nearly 30 cm distal to duodenojejunal junction (Fig 2). Rest of the bowel inspected was found normal. The intussuscepted part was resected and end to end jejunal-jejunal anastomosis was done. Wedge resection of the segment containing polyp proximal to the intussusception and closure of enterotomy was done. Resected wedge segment showed presence of a pedunculated polyp of size 1 x 0.5 cm with verrucous surface. Cut section of the intussuscepted segment revealed presence of another polyp acting as lead point of intussusception and gangrene of the intussuscepted bowel (Fig 3). Postoperatively period was uneventful. Patient was allowed orally on fourth postoperative day and was discharged on 7th postoperative day. Histopathological examination of the polyp was reported as peutz jeghers polyp.



Fig 1- Jejunojejunal intussusceptions



Fig 2- Proximal jejunal polyp



Fig 3- Cut section showing polyp as lead point

DISCUSSION

Peutz-Jeghers syndrome was first described by Y. Peutz in 1921 as gastrointestinal familial polyposis with pigmentation in a Dutch family. In 1949, H. Jegher defined the relation between pigmented lesions, gastrointestinal polyposis and increased risk of carcinoma in 10 cases (4). Peutz-Jeghers Syndrome is an autosomal dominant disorder characterised with gastrointestinal polyps, melanotic macules and an increased cancer risk. These can present from early infancy to adulthood (5). It can have both familial (80%) and sporadic (25%) transmissions. The disease has an incidence of 1 in 30,000 to 1 in 1,20,000 live births as described (2). STK11/LKB germline mutation accounts for majority of cases whereas large deletions account for 30% of cases. Peutz-Jeghers polyps are hamartomatous polyps involving jejunum, colon, duodenum and stomach in decreasing frequency. The disease may cause complications like abdominal pain, anaemia and acute intussusception (1). In a study conducted by Utsunomiya et al including 222 patients with Peutz-Jeghers syndrome intussusception occurred in 46.9% of patients. Polyps occurred in stomach in 48.6%, small intestine in 64%, colon in 53.2% and rectum in 32% of patients (6). Polyps may cause intussusceptions of which some are relieved by themselves and other sometimes may cause obstruction. Along with polyps abnormal mucocutaneous pigmentation is also a characteristic of Peutz-Jeghers syndrome but less than 5% of the patients lack mucocutaneous pigmentation (3). Intussusception is observed in 47 to 69% of adult patients suffering from Peutz-Jegher syndrome and occurs mainly in the small bowel with polyps acting as lead point (7).

In patients without a family history of Peutz-Jeghers syndrome, a diagnosis of Peutz-Jeghers syndrome is made if either of the following are present: characteristic melanotic macules and one or more intestinal polyps with PJS-type histology, or two intestinal polyps with Peutz-Jeghers type histology (Mayo clinic 2010). According to Mayo clinic pigmentation often fades with puberty or may be very faint or absent in adults (8). Patients with suspicion of Peutz-Jeghers syndrome can be investigated by stool examination for occult blood, upper gastrointestinal endoscopy, colonoscopy and CT scan (9). A combination of surgical and endoscopic polypectomy is effective in management of polyps. Indications of surgery are intestinal bleeding, intussusceptions, intestinal obstruction and malignant polyps. As many polyps as possible should be removed so as to prevent the recurrence of complications (1). It is challenging to decide the amount of resection as polyps can occur in any region of entire gastrointestinal tract and the recurrence of the polyps is also common (9).

In our case patient neither had a family history of Peutz-Jeghers Syndrome nor did she have any hyperpigmentation of skin or mucocutaneous tissue. But the patient had two polyps in the jejunum with histological features of peutz-jeghers polyp. Hence the case satisfies the mayo clinic criteria for the diagnosis of peutz-jeghers syndrome. Our patient presented with jejunal-jejunal intussusceptions and required resection of the involved segment and anastomosis. Also another polyp present in the proximal jejunum was removed by wedge resection of the bowel with closure of the enterotomy.

CONCLUSION

Intussusception is one of the causes for intestinal obstruction in the paediatric age group. Peutz-jeghers syndrome may present as intussusception and a complete history and examination may help in the diagnosis. Early decision for exploratory laparotomy should be taken and thorough examination of bowel should be done to watch for presence of any other polyp.

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