

Peutz Jegher's Syndrome (Gastro-intestinal Polyposis) and Its Complications

Pages with reference to book, From 154 To 155

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Abstract

A rare case of multiple jejunal polyps, presenting as an acute abdominal emergency with complications of intussusception and gangrene is discussed. The relevant features of Peutz Jegher's syndrome and the surgical management of its complications are highlighted (JPMA 35 : 154, 1985).

Case Report

A male, aged 30 years, born and bred in Peshawar was admitted on May 5, 1984 as an emergency; his complaints were of intestinal colic associated with vomiting and constipation. For the past one year patient gave a history of recurrent intestinal colic with the passage of blood per rectum off and on. On examination, he was of dark-complexion with, pigmentation of the oral mucous membrane and heavily pigmented skin of the palms and soles. Abdominal examination revealed a distended and tense abdomen with two palpable lumps in the left hypochondrium. X-Ray abdomen showed the pattern of intestinal obstruction. Fig. 1 Polyp in the small bowel.

Operative Findings

Emergency laparotomy through a left paramedian incision revealed an intussuscepted mass in the small bowel, the jejunum invaginating the distal ileum. The intussusception was reduced and a polyp found in the invaginating jejunal part removed. Post-operatively the patient's condition worsened, with signs of peritonitis and the passage of blood per rectum. On May 9, a second laparotomy, through a mid-line incision, revealed a gangrenous portion of the jejunum measuring approximately 60 cm. About 70 cm. of proximal jejunum was viable and the whole of the ileum appeared normal. The proximal viable jejunum was dilated and contained a number of polyps, varying in shape and size, from full fledged well pedunculated to more sessile polyps adherent to and causing serosal indentations externally (Fig .1 and 2).

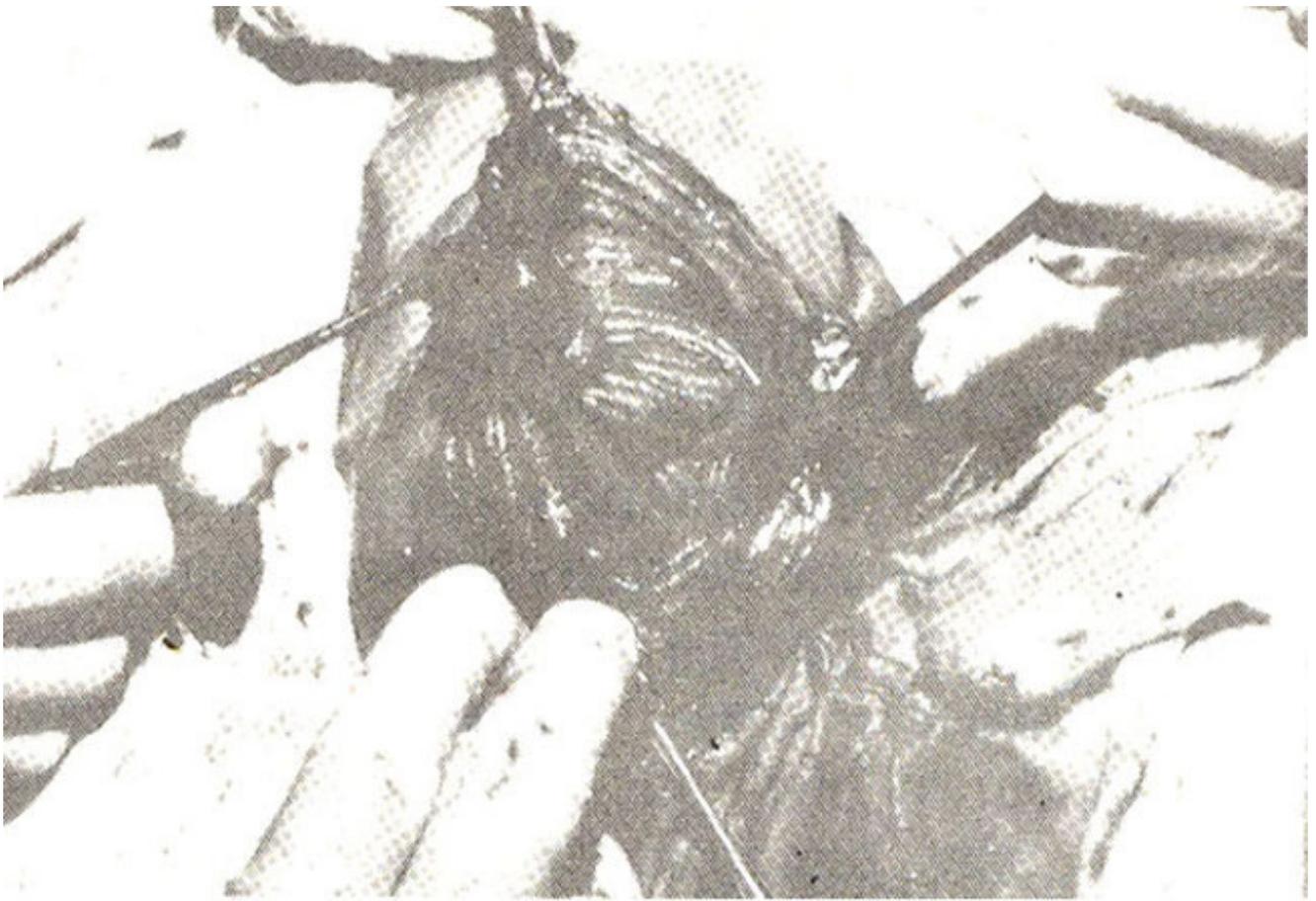


Fig. 1 Polyp in the small bowel.

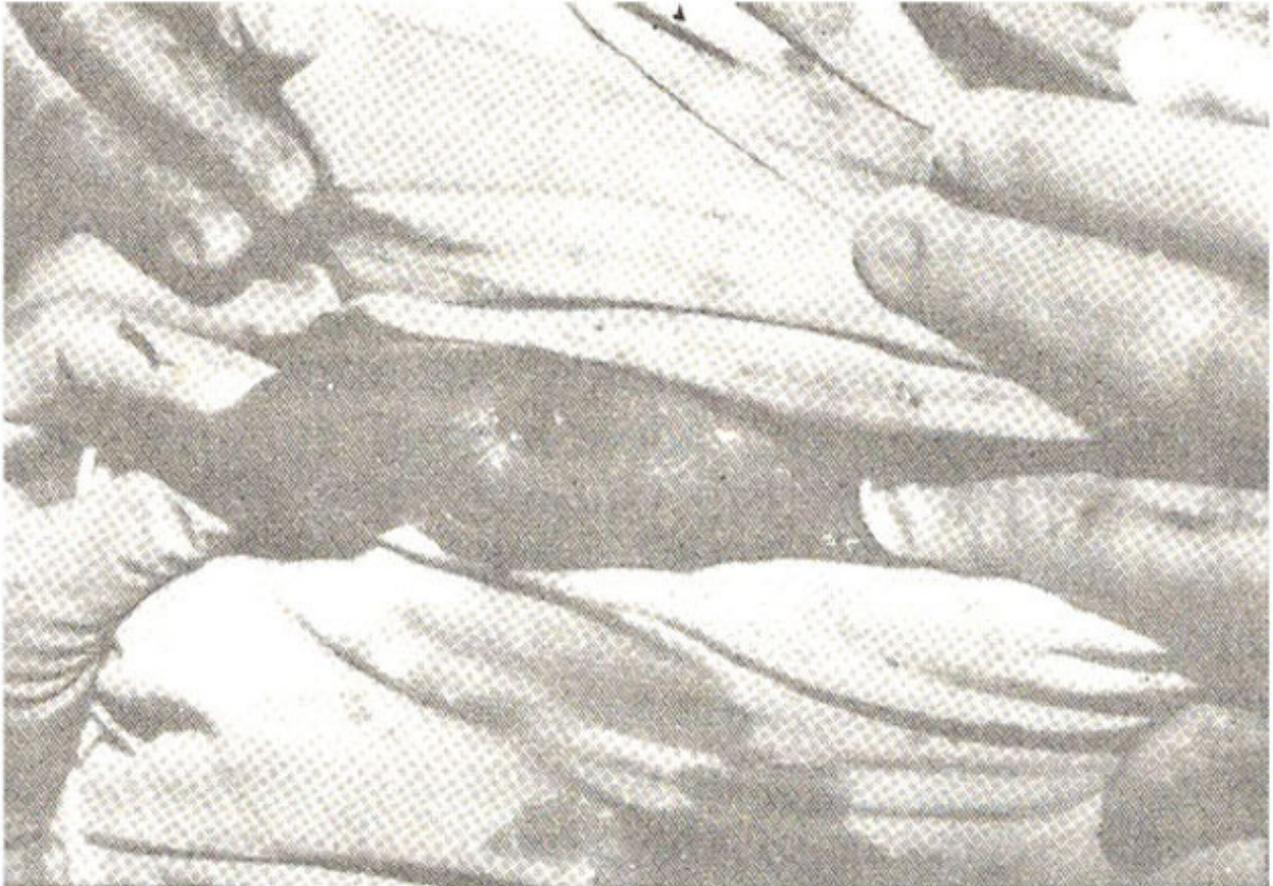


Fig. 2 Serosal indentation at the site of intraluminal polyp in Jejunum.

There were at least seven polyps in the proximal jejunum. No polyps were detected in the distal ileum and the large gut. Resection of the gangrenous portion with end-to-end anastomosis was performed and four readily accessible polyps in the viable proximal jejunum were removed. His further post-operative recovery was uneventful. Post-operative Barium enema and colonoscopy did not show any polyps in the large gut.

Histopathology

Histological sections showed branching fibromuscular tissue covered by small intestinal mucosa, which in some parts displayed mild cellular atypia. The growth pattern in some parts was adenomatous, while occasionally in other parts it was papillary.

Diagnosis: Polyp (Hemangioma of small intestine).

Discussion

Patient came from N.W.F.P. where the people are of fair-complexion. The most striking feature of the patient was his dark complexion, with muco-cutaneous pigmentation especially heavy pigmentation of the palms and soles. The patient said that his family was generally of dark-complexion and also that his brother was available for examination. The physical examination of his brother confirmed muco-cutaneous pigmentation especially of the cheeks and the nose forming a "butter fly" configuration. His brother refused to have Barium studies performed on him.

There are three features of Peutz Jegher's syndrome

- 1) Muco-cutaneous pigmentation;
- 2) Benign polyps occurring in any part of the gastro-intestinal tract but mainly in the jejunum; and
- 3) Autosomal dominant inheritance.

The melanotic pigmentation characteristically involves lips and oral mucosa, but can appear in frequently on the palms and soles and in the interdigital space¹. These cutaneous lesions may not be present at birth but appear during adolescence and may disappear with advancing age. The oral mucosal lesions are however present at birth and persist throughout life. The pigmentations are not painful, hairy or vascular. They are not potentially malignant. They have great diagnostic significance, to surgeons faced in the examination of such patients complaining of abdominal cramps, acute intestinal obstruction or unexplained G.I.T. bleeding. The second feature of this entity, namely intestinal polyposis, is rarely encountered in infants but appears during or adolescence. The adenomatoid tumors² are generally found in the jejunum, less frequently in the ileum, occasionally in the duodenum and rarely in the stomach or colon. Polyps of the Peutz Jegher's syndrome may be pedunculated or sessile and may vary in number and diameter in the same segment of the bowel. The surface is similar to that of mucosa from which they appear. Although there have been genuine reports of malignancy in Peutz Jegher's syndrome, most observers conclude it is not premalignant. In a study of 14 patients, Reid^{2,3} found a 2-3% risk of malignancy in Peutz Jegher's syndrome. Gastric and duodenal polyps are more likely to undergo malignant changes.

Acute intestinal obstruction from small bowel intussusception is the major complication requiring surgery. Bleeding from polyps is extremely common; it is usually mild, persistent or recurrent and results in iron deficiency anemia.

Less frequently massive gastro-intestinal bleeding may occur, particularly from a gastric polyp.

It was Jegher's who demonstrated that the 'hereditary peculiarity' of melanotic pigmentation was carried as a 'Mendelian dominant' characteristic. There were no skip generations and both sexes of all ethnic groups were equally affected. This indicates a high degree of penetrance.

Based on the combined experience of many authors, the approach to the management of Peutz Jegher's syndrome and its complications should be symptomatic. Prophylactic treatment has little role in the management of small intestinal polyps when detected for several reasons. Firstly, despite the frequent intestinal invagination of these polyps, the majority reduce spontaneously. Secondly, they have little malignant potential. Thirdly, it is impossible to predict location of other polyps or the recurrence of intussusception, as the polyps are known to progress in segmental spurts. Lastly, overzealous resection may lead to malabsorption and mortality. The exception to the above rule may be gastric or duodenal polyps, which could be a coincident premalignant adenoma.

Intestinal invaginations leading to intussusception and obstruction is a major complication of Peutz Jegher's syndrome. The operative procedure should be aimed at salvaging small bowel length in order to avoid subsequent short gut syndrome. Resection should be limited to gangrenous portions only, and if the involved segment is viable, the offending polyps should be excised by enterostomy in the healthy segment of the bowel. Although majority of the polyps may be pedunculated, some sessile polyps will require bowel resection. Simultaneous multiple intussusceptions requiring multiple bowel resections in a single procedure are possible.

Acknowledgement

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