

Peutz Jeghers syndrome with colonic and small bowel polyps

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Case Report

A 25-year old female from Vavuniya presented with a history of bleeding per rectum and episodes of severe colicky abdominal pain associated with vomiting of one year duration. She had undergone a laparotomy for symptoms suggestive of intestinal obstruction at the age of 11 years but medical records were not available. On examination, she had pigmentation of the lips and buccal mucosa. The abdominal examination was unremarkable except for a well healed midline laparotomy scar. Upper gastrointestinal endoscopy was normal up to the third part of duodenum but colonoscopy revealed two large sessile polyps in the distal transverse colon and three pedunculated and one sessile polyp in the sigmoid colon. The small bowel enema showed multiple filling defects suggestive of polyps in the proximal jejunum with a large polyp close to the duodenojejunal junction. A clinical diagnosis of Peutz Jeghers syndrome (PJS) was made.

All pedunculated colonic polyps were removed by diathermy snaring prior to surgery. Two weeks later a laparotomy was performed. Laparotomy revealed polyps in the proximal jejunum, transverse colon and sigmoid colon. There were two areas of jejunojejunal intussus-

ception (Figure 1), each containing a polyp at the apex. A 10 cm proximal jejunal loop containing multiple pedunculated and sessile polyps was excised (Figure 2). Rest of the jejunal polyps were excised via enterotomies. The sessile polyps in the transverse colon and the sigmoid colon were excised through multiple colotomies. Postoperative colonoscopies done at 3, 6 and 9 months did not reveal any recurrence of large bowel polyps. At present the patient is asymptomatic.

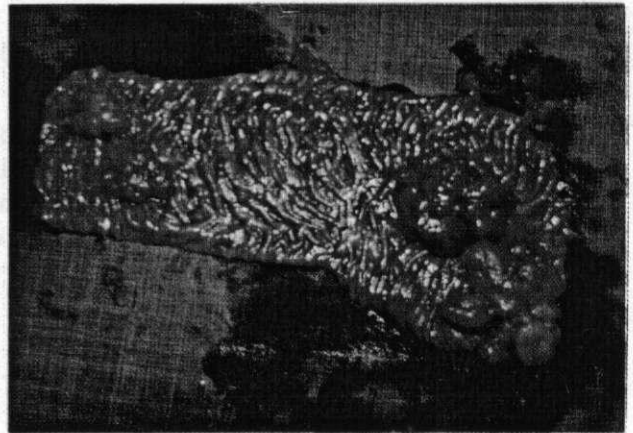


Figure 2. Resected segment of jejunum showing multiple polyps.



Figure 1. Two areas of jejuno-jejunal intussusception (indicated by the forceps).

Discussion

Peutz Jeghers syndrome is a disease manifested by a combination of muco-cutaneous melanin pigmentation and gastrointestinal polyposis. Melanosis can sometimes present on the digits and the perianal skin, but the characteristic sites are the lips and the buccal mucosa. Although polyps can occur anywhere in the gastrointestinal tract, the commonest site of polyposis is the small bowel (1). This patient presented to us with episodes of rectal bleeding and intermittent colicky abdominal pain associated with vomiting. Her abdominal pain and vomiting would have been due to the intussusceptions and her previous laparotomy may have been performed for a similar lesion.