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A management case study of intussusception revealing by recurrent small bowel Peutz-jeghers syndrome

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Abstract

Introduction: Peutz-Jeghers syndrome (PJS) is a rare autosomal dominant disorder characterized by hyperpigmentation on the lips and oral cavity and gastrointestinal (GI) hamartomatous polyps. PJS patients usually present with acute complications such as bleeding, bowel obstruction and intussusception.

Presentation of case: We hereby report a case of a 25-year-old male, who presented with complaints of colicky abdominal pain since 4 days. On abdominal ex-amination, tenderness in the right iliac fossa and increased bowel sounds. Patient presented 3 months ago with bleeding pr and rectal mass. Ultra sound of the abdomen demonstrated suspected ileo-colic intussusception. The patient underwent colonoscopy showed pedunculated polyp and polypectomy from ileo-caecal region. The symptoms did not subside after 2 days, hence was taken up for a mid-line laparotomy that showed an ileo-colic and jejuno-jejunal intussusception. Reduction of this intussusception was successfully done with, multiple enterotomy and polypectomy. The specimen further confirmed the diagnosis of hamartomatous Peutz-Jeghers polyps with no malignancy. Physical examination revealed multiple pigmented spots on the oral cavity and palm. Thus, the diagnosis of PJS was made.

Discussion: PJS is a rare inherited disease that often remains undiagnosed for many years. Patients often presents with acute complications such as bleeding per rectum, intussusception or obstruction.

Conclusion: Early detection and identification of PJS in patients and their family members can improve the

prognosis, prevent complications. Patients should be on regular follow up once the diagnosis has been made.

Keywords: Syndrome, Prognosis, Hamartomatous.

Introduction

Peutz-Jeghers syndrome (PJS) is a rare autosomal dominant disorder characterized by hyperpigmentation on the lips and oral cavity and gastrointestinal hamartomatous polyps.[1] It can be diagnosed using clinical criteria which includes the presesnce of ≥2 Peutz-Jeghers polyps anywhere in GI tract, or at least one intestinal Peutz-Jeghers polyp with family history of PJS or classic muco-cutaneous pigmentation.[2] This disorder is seen uniformly in both genders and not diagnosed until late childhood or early adulthood when they present with some complications.[3,4]Acute intestinal intussusception is rare in adults, but one of the common complications of PJS. It represents 5 % of all intussusceptions and accounts for 1-5 % of all intestinal obstruction in adults.

Case Report

A 25-year-old male, who presented with complaints of colicky abdominal pain since 4 days. Her vitals are stable. Physical examination revealed multiple pigmented spots on the oral cavity and palm. On abdominal examination, tenderness in the right iliac fossa and increased bowel sounds. Patient presented 3 months ago with bleeding per rectum and rectal mass. Patient gives history of resection and anastomosis done in view of gangrenous bowel 10 years ago, was not evaluated further.

Ultra sound of the abdomen demonstrated suspected ileo-colic intussusception. The patient underwent colonoscopy showed pedunculated polyp and polypectomy from ileo-caecal region. The symptoms did not subside after 2 days, hence was taken up for a mid-line laparotomy that showed an ileo-colic and jejuno-jejunal intussusception. Reduction of this intussusception was successfully done with, multiple enterotomy and polypectomy.

The specimen was sent for histopathological evaluation. On gross examination, multiple grey white to grey brown polypoidal tissue. On microscopy, multiple polyps show polypoidal tissue lined by intestinal epithelium with characteristic arborizing network of connective tissue, smooth muscle, lamina propria and glands, suggesting hamartomatous Peutz-Jeghers polyps with no malignancy. Thus, the diagnosis of PJS was made.

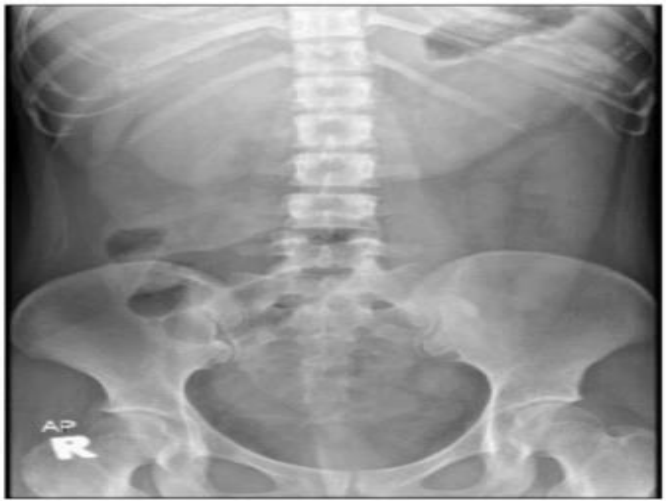

Figure 1: Demonstrated plain abdominal radiography with paucity of small bowel gas and absence of rectal gas

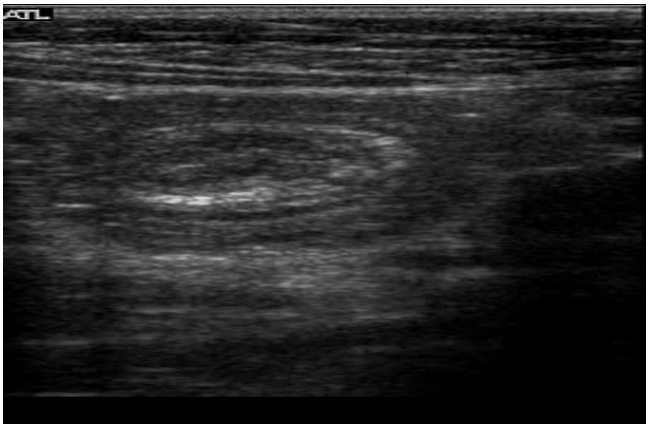

Figure 2: Ultrasound showing ileo-colic intussusception (target sign)

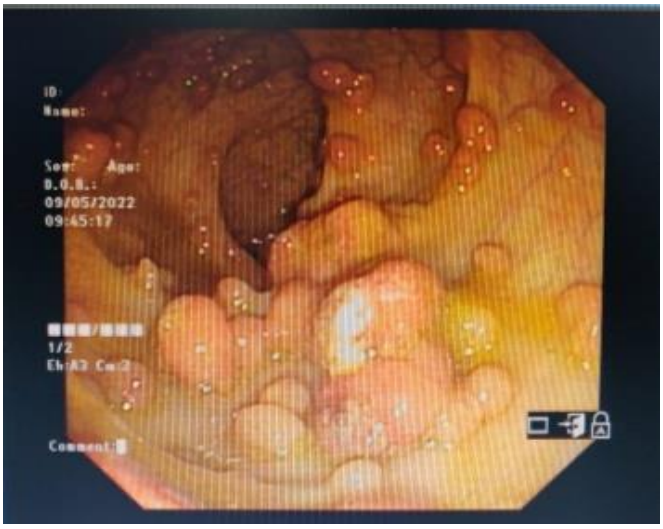

Figure 3: Upper GI scopy showing multiple hamartomatous polyp over the stomach

Figure 4: Gross examination of the ileocolic resection specimen.

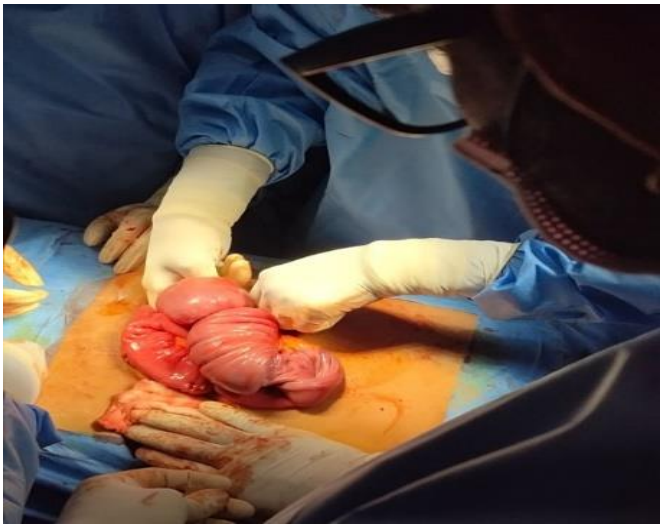

Figure 5: Ileo-colic intussusception

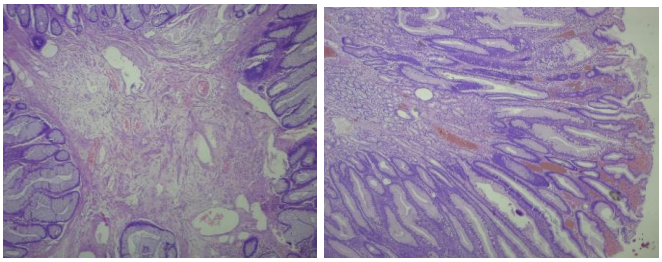

Figure 6 and 7: Multiple polyps show polypoidal tissue lined by intestinal epithelium with characteristic arborizing network

Discussion

PJS is a rare inherited disease that often remains undiagnosed for many years, reported to be 1 in 100,000 individuals. Patients with hamartomatous polyp(s) who also meet at least two of the following clinical criteria can be diagnosed with PJS: small bowel polyposis, labial melanin deposits, and a family history of the syndrome. The syndrome affects all racial groups and affects both men and women equally. Asymptomatic intervals interspersed with consequences including stomach pain, intussusception, which frequently results in intestinal obstruction, polyp extrusion via the rectum and bleeding which is frequently occult, characterise the clinical appearance of PJS. In half of the cases, small intestinal obstruction is the initial complaint, and relaparotomy due to polyp-induced complications happens often and may do so at relatively frequent intervals.

Patients usually present to hospital with complications as we see in our case patient presented with pain abdomen and diagnosed to have ileo-colic intussusception where the polyps were acting as lead point for intussusception. Earlier PJS was considered benign condition and was treated conservatively. Patient was kept on observation and underwent laparotomy when presented with complications, this resulted in multiple laparotomies hence pateints are recommended to undergo endoscopy

and polypectomy for polyps in the stomach or colon that are greater than 1 cm in size noted during endoscopic surveillance. Our patient underwent colonoscopy which confirmed ileo-colic intussusception which couldn't be reduced. Patient was posted for exploratory laparotomy, reduction of this intussusception was successfully done, bowel segments were found to be viable. Multiple enterotomy and polypectomy carried out to reduce the risk of intussusception in the future.

Conclusion

Early detection and identification of PJS in patients and their family member's as well as close cancer surveillance can improve the prognosis, prevent complications. Patients should be on regular follow up once the diagnosis has been made.

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