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A Case of Ileocolic Intussusception in an Adult with Peutz-Jeghers Syndrome

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Abstract

Introductions: Peutz-Jeghers syndrome (PJS) is a rare autosomal dominant disorder characterized by hamartomatous polyps in the gastrointestinal tract and hyperpigmentation on the lips and oral cavity. Bowel obstruction, intussusception and bleeding are common complications in PJS patients. PJS patients also have an increased risk of gastrointestinal and extra-intestinal malignancies.

Case Description: A 32 years old male was brought to the emergency room due to suspected ileocolic intussusception. Ten years ago he had a history of laparotomy resection anastomosis of the bowel due to bowel polyps. Physical examination revealed multiple pigmented intraoral lesions. Abdominal examination showed mid-line laparotomy scar, distention with visible bowel movement. There was no palpable mass and no blood upon digital rectal examination. Computerized tomography of the abdomen demonstrated suspected ascending colon intussusception. Intra-operation; ileocolic intussusception was found with multiple polyps along the colon. Resection and stoma was done, with planned post-operative endoscopy via the stoma.

Conclusion: The standard treatment for intussusception in PJS patients is laparotomy bowel resection to remove the polyps causing the recurrent invaginations. It has been recommended that endoscopic polyps removal should be performed to avoid multiple surgical resections, which lead to short bowel syndrome. Due to increased risk of malignancies, regular screening of PJ patients is needed.

Keywords

Peutz-Jeghers Syndrome; Ileocolic intussusceptions; Polyps; Bowel obstruction

Introduction

Peutz-Jeghers syndrome (PJS) is a rare autosomal dominant disorder, with an estimated prevalence from 1 in 100000 people. It is characterized by hamartomatous polyps in the gastrointestinal tract and hyperpigmentation on the lips, oral cavity and nasal alae. Bowel obstruction, intussusception and bleeding are common complications in PJS patients [1-2]. PJS patients also have an increased risk of gastrointestinal and extra-intestinal malignancies [3].

PJS-associated polyps are found over 90% in the small intestine, followed by colon (53% of patients), stomach (49%) and rectum (32%). Well-planned polypectomy may prevent the need for repeated emergency surgery and extensive bowel resection due to intestinal complications such as intussusception; which may lead to short bowel syndrome [4]. Gastrointestinal polyps management and routine cancer screening is needed for early detection and surveillance to minimize the risk of malignancies. Small bowel intussusception has also reported in the literature [5].

Case Presentation

A 32 years old male was brought to the emergency room with symptoms of bowel obstruction since one week before hospital admission. Ten years ago he had a history of laparotomy resection anastomosis of the bowel due to bowel polyps. There was no pathological view from the operation. There was no endoscopic surveillance done before due to lack of pathologic review. Physical examination revealed multiple pigmented intraoral lesions (Figure 1). Abdominal examination showed mid-line laparotomy scar, distention with visible bowel movement. There was an increased bowel sound. There was no palpable abdominal mass and no blood upon digital rectal examination (Figure 1).

Laboratory investigations showed mild anemia, leukocytosis, hyponatremia and hypokalemia. Plain abdominal X-ray demonstrated bowel obstruction at the level of small bowel. Computerized tomography of the abdomen demonstrated suspected ileocoloascenden intussusception (Figure 2). Intra-operation; ileocoloascenden intussusception, 240 cm from ligament of Treitz was found. Bowel resection was done 5 cm proximal and distal of the intussusception (Figure 3). The resected bowel showed multiple

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