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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 polyps (Figure 4). Resection

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and ileo colon tranversum double barrel stoma was done, with planned post-operative endoscopy via the stoma. Histopathological examination confirmed the diagnosis of hamartomatous PJ-polyps with no malignancy (Figure 5). Future planning includes scheduled polypectomy via endoscopy and stoma closure.

Discussion

The incidence of Peutz-Jeghers syndrome (PJS) is reported to be 1 in 100000 individuals. The classic finding of PJS includes hamartomatous polyposis of the gastrointestinal tract and mucocutaneous hyperpigmentation on the lips, around the mouth, nostrils and buccal mucosa [1]. Patients with PJS often present with a history of intermittent abdominal pain due to small bowel intussusception caused by the polyps. They may reduced spontaneously or develop into bowel obstruction. PJ-related polyps may also ulcerate leading to acute blood loss and chronic anemia [2]. This patient had the classic PJS characterization of oral hyperpigmentation and hamartomatous GI polyposis. He had the complication of intussusception with bowel obstruction.

Figure 1: Hyperpigmentation of the lips, buccal mucosa and nasal alae.

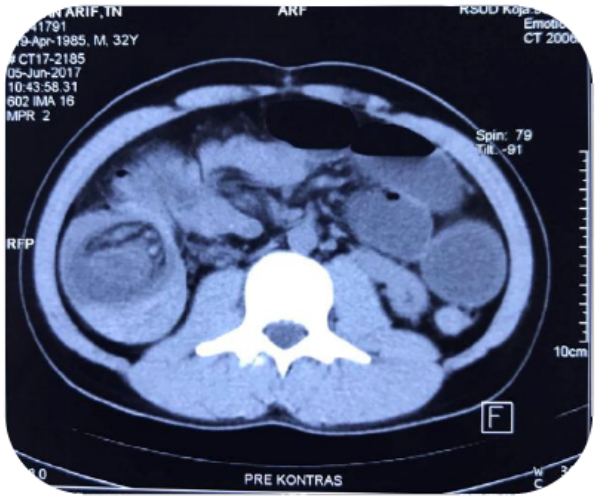

Figure 2: CT Scan showing ileocolic intussusception with "target sign".

Figure 3: Ileocolic Intussusception.

Figure 4: Multiple polyps inside of the resected bowel.

The diagnosis of intussusception was made based on this patient's complaint, clinical signs and symptoms as well as imaging studies. Computed tomography (CT) findings appear as a complex soft-tissue mass composed of a central intussusception, described as "target sign" [4]; which was seen in this patient. The standard of procedure for intussusception in PJS patients has been laparotomy bowel resection to remove the underlying polyps causing possible recurrent intussusceptions [5]. To avoid short bowel syndrome due to multiple surgical resection, endoscopic polyp removal has been

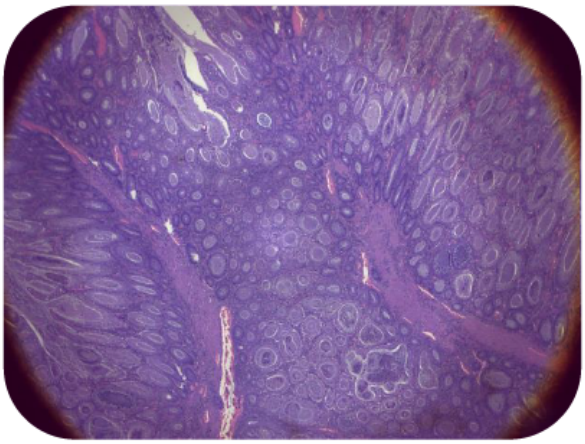

Figure 5: Histopathological examination of hamartomatous PJ- polyps with no malignancy.

recommended [6]. This can eliminate GI symptoms and prevent or postpone repeated abdominal surgeries.

PJ patients have an increased risk of both gastrointestinal and extra- intestinal malignancies, thus long-life regular screening is recommended [3,5]. Monitoring of hemoglobin levels, regular breast, gynecologic and testicular examination. Pelvic and pancreatic US screening may be considered. GI tract screening can be performed with upper and lower endoscopy [3-4]. This patient underwent laparotomy bowel resection and stoma with a planned upper endoscopy and lower endoscopy through the stoma for further diagnostic procedure including future molecular analysis.

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.

Conflict of Interest

The authors declare that there is no conflict of interest regarding the publication of this paper.

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