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PEUTZ-JEGHERS SYNDROME: AN UNUSUAL CAUSE OF RECURRENT INTUSSUSCEPTION IN A 7-YEAR-OLD BOY

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Introduction & Aim: Peutz-Jeghers syndrome (PJS) is an autosomal dominant inherited disorder characterized by intestinal hamartomatous polyps in association with mucocutaneous pigmentations. Although, intussusception has been reported as a well-known complication of PJS, recurrent intussusception as an alarming finding in a patient with normal gastrointestinal endoscopy is uncommon. A 7-year-old boy who had recurrent intussusception episodes and diagnosed as PJS with histopathologically after surgical excision of involved bowel segments is presented to discuss the clinical features and treatment options of recurrent intussusception as a presenting finding of PJS. We aimed to present this case to emphasize that patients with PJS may present with intussusception and hamartomatous polyps might be precancerous, in this syndrome.

Case Report: A 7-year-old boy was admitted to emergency with complaints of abdominal pain and vomiting lasting three days. He had a history of recurrent colicky abdominal pain for approximately six months. There was no history of hematemesis or melena. He had undergone upper gastrointestinal endoscopy and colonoscopy because of multiple hyperpigmented macules over only the lips and it is revealed normal endoscopic findings. His physical examination showed slight distension with generalized tenderness. According to physical examination findings and bowel obstruction, he underwent explorative laparotomy. A jejuno-jejunal intussusception was found 5 cm distal of the ligament of Treitz during surgical exploration. Intussusception could not reduce manually and a polypoid mass in the jejunal lumen was the leading point. The intussuscepting segment including the polyp was excised by small bowel resection, and end-to-end anastomosis was performed. Histopathological examination of the specimen revealed hamartomatous polyp and Peutz-Jeghers syndrome was confirmed histologically with low-grade dysplasia.

Conclusion: Peutz-Jeghers syndrome is a hereditary disease characterized with gastrointestinal polyposis and skin or mucosal pigmentation. Polypoid lesions are almost hamartomatous and are not usually precancerous. Our patient underwent upper and lower endoscopy with presumptive diagnosis of PJS before the clinical findings of intussusception. Endoscopic evaluation revealed normal findings without any polypoid lesion. Although the association of PJS and intussusception is well-known, this complication is rarely presenting finding for PJS. Patients with hyperpigmentous lip lesions with recurrent intussusception episodes may require surgical explorations not only relieving small bowel obstruction but also diagnose the underlying polypoid lesions. In conclusion, patients with PJS should be regularly and closely monitored in order to reduce the number of laparotomies and due to the increased risk of cancer.

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