

Multiple intussusceptions as primary manifestation of Peutz-Jeghers syndrome: Report of a case

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Abstract

Background: Peutz-Jeghers syndrome is a rare hereditary disorder characterized by hamartomatous polyps in the gastrointestinal tract and typical pigment lesions. It is a rare cause of multiple intussusceptions. Previous studies on Peutz-Jeghers syndrome reported only one case of multiple intussusceptions. We describe a case of appendiceal and multiple small intestine intussusceptions presenting as peritonitis in a patient with Peutz-Jeghers syndrome.

Case Presentation: A 11-year-old girl presented with an 1 day history of a sharp, non-radiating periumbilical pain. She underwent surgery with the diagnosis of peritonitis. Intraoperative findings included appendiceal and multiple small intestine intussusceptions. The final pathological evaluation of the specimen confirmed the diagnosis of Peutz-Jeghers syndrome. Conclusion: Multiple intussusceptions may occur as the primary manifestation of Peutz-Jeghers syndrome. Because of its complications, in view of the presence of multiple polyps, early intervention is strongly recommended. © 2009 by Center of Excellence for Pediatrics, Children's Medical Center, Tehran University of Medical Sciences, All rights reserved.

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Author keywords

Appendiceal intussusceptions; Hamartomatous polyps; Multiple intussusceptions; Peutz-Jeghers syndrome

Indexed Keywords

EMTREE medical terms: abdominal distension; abdominal pain; adolescent; amenorrhea; article; case report; clinical feature; digestive system examination; disease duration; female; fever; hemicolectomy; human; intestine intussusception; leukocyte count; nausea; neutrophilia; pain; peritonitis; Peutz Jeghers syndrome; umbilical pain; vomiting

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