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Case Report

Jejuno-Jejunal Intussusception Secondary to Undiagnosed Hamartomatous Peutz Jegher's Polyp in A 9 Year-Old-Girl

Hassan RAA*, Hadi A, Yew CG and Ridzuan AS

Unit of Paediatric Surgery, Department of Surgery, Hospital Universiti Sains Malaysia, Kelantan, Malaysia

***Corresponding Author:** Hassan RAA, Unit of Paediatric Surgery, Department of Surgery, Hospital Universiti Sains Malaysia, Kelantan, Malaysia.

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Introduction

Jejuno-jejunal intussusception is very rare in children. In cases of Peutz Jegher Syndrome (PJS) hamartomatous polyp may act as a lead point to develop intussusceptions in an unusual segment of bowel. In cases of acute presentation with abdominal pain in a known case of PJS an intussusception have to be suspected and especially when mucocutaneous pigmentation is present.

Keywords: *Jejunojejunal; Intussusception; Peutz Jegher Syndrome; Paediatric; Children*

Case Report

A 9-year-old Malay girl presented with acute upper abdominal pain for 6 hours which was colicky in nature associated with non-bilious vomiting. There was no abdominal distension, constipation or loose stool. She was not known to have any medical illness. Clinically the child was in moderate to severe pain but afebrile. Abdominal examination revealed tenderness on deep palpation over epigastric region but there was no palpable mass. There were multiple small hyperpigmented mucocutaneous lesions around the lips and oral mucosa. Family history revealed that the child's father died at the age of 33 years due to metastatic lung cancer of unknown primary. He had similar mucocutaneous lesion. One of her paternal cousin had similar lesion.

Laboratory investigations showed leukocytosis (TWC: 13000), normal renal function and serum amylase. The abdominal x-ray was unremarkable. There was no evidence of bowel dilatation. Urgent abdominal ultrasound showed an elongated heterogenous soft tissue mass at epigastric region measuring 9.8 x 5.6 cm. Intussusception of bowel is suspected. Diagnostic and therapeutic ultrasound guided hydrostatic reduction under sedation attempted by radiologist but failed due to difficult interpretation. We proceed with CT Abdomen (Figure 1) and confirmed the diagnosis of jejuno-jejunonal intussusception.

Emergency laparotomy was performed. An irreducible gangrenous jejuno-jejunal intussusception segment about 25 cm in length (Figure 2). It was 50 cm distance from duodeno-jejunal junction. Gangrenous segment of bowel resected and primary end to end anastomosis done (Figure 3). Macroscopic pathological specimen revealed a polypoidal polyp measuring 3 x 3 cm as a lead point. Histopathological examination confirmed hamartomatous Peutz Jegher's polyp. Post operatively, patient recovered well and discharged 4 days after operation. Patient was seen at clinic 3 months post operatively with no complication.

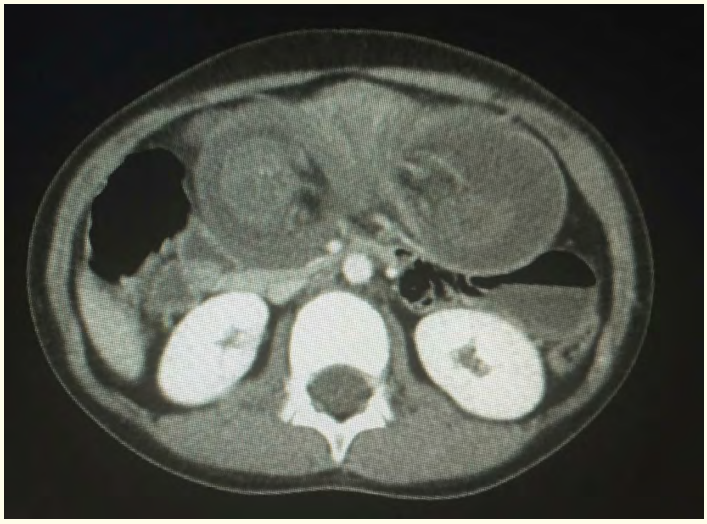

Figure 1: *CT Abdomen.*

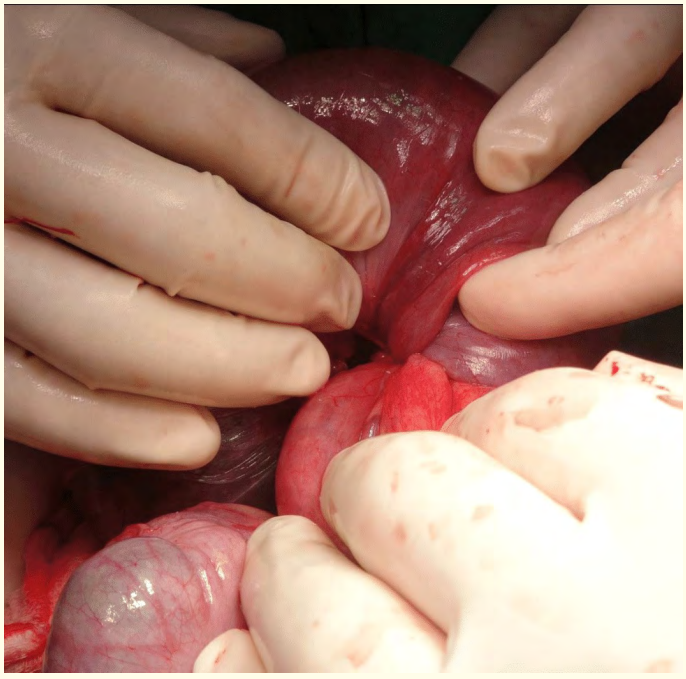

Figure 2: *Intraoperative Findings.*

Discussion

Peutz-Jeghers Syndrome (PJS) is rare autosomal dominance inheritance disease characterized by hamartomatous polyps in gastrointestinal tracts and mucocutaneous pigmentation [1]. It has an incidence of 1 in 150000 in North America and Western Europe [2]. These polyps may serve as pathological lead point for intussusception.

Intussusception is common in children. The incidence of intussusception varies with age but peaked between 3 and 36 months [3]. Presentation of above 3 years old as in the index case illustrated is uncommon. The yearly mean incidence of intussusception was 38, 31, and 26 cases per 100 000 live births in the first, second, and third year of life, respectively [4]. The preponderance of male to female ratio is (2:1) [5]. Ileocecal intussusception is the most commonest type seen while less is seen in jejuno-jejunal pattern [6]. Most intussusception are idiopathic in origin while only in 10% of cases the pathological leadpoint were identified [7]. Hamartomatous polyps of PJS as specific pathological leadpoint is rare [7].

Jejuno-jejunal intussusception in an undiagnosed hamartomatous Peutz Jegher's Polyp in older child may pose diagnostic dilemma. In general, paediatric population, presentation above 3 years old in childhood intussusception with jejunojejunal variety is uncommon. Although intussusception is a known manifestation of hamartomatous Peutz Jegher's small bowel polyps, this case report illustrates the challenges in diagnosis and decision making for surgery.

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