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Pediatric Intusseption caused by Peutz Jeghers Polyp: A Case Report

Authors
Vishnu Swaroop Shrivastava, Aarti Dhatwalia, Sarita Asotra, Rajkumar
Department of Pathology and surgery, Indira Gandhi Medical College, Shimla, Himachal Pradesh
Corresponding Author
Sarita Asotra
Professor, Department of Pathology, IGMC Shimla

Abstract
Small bowel intussusception is a rare yet significant clinical condition that can lead to various complications, particularly in the pediatric population. Here, we present a case report of a 6-year-old male child with jejunojejunal intussusception caused by multiple Peutz-Jeghers-type hamartomatous polyps in the small intestine.
The patient presented with persistent abdominal pain and vomiting, leading to the diagnosis of intussusception in the left hypochondrium. An exploratory laparotomy revealed jejunojejunal intussusception, which was manually reduced. Histopathological examination of the resected segment confirmed the presence of hamartomatous polyps with distinct histological features characteristic of Peutz-Jeghers polyps.
The distinction between Peutz-Jeghers syndrome and solitary Peutz-Jeghers-type hamartomatous polyps remains a subject of debate, especially in cases lacking mucocutaneous pigmentation and a positive family history. We discuss the complexities in defining this clinical entity and emphasize the significance of histopathological examination for accurate diagnosis.
Our case highlights the importance of considering rare pathologies in the absence of classical clinical features or positive family history, emphasizing the role of histopathology in confirming the diagnosis of Peutz-Jeghers-type polyps. Small bowel intussusception should prompt thorough evaluation, especially in pediatric cases, to prevent complications and ensure timely management.
Keywords: *Intussusception, Polyp, Peutz-Jeghers polyp.*

Introduction

Mucosa of the small intestine covers roughly 90% of the luminal surface area of the digestive system, small bowel polyps and tumors are relatively uncommon. The most frequently encountered small bowel polyps include adenomas in familial adenomatous polyposis (FAP) syndrome, as well as hamartomas associated with Peutz-Jeghers and Cronkhite-Canada syndromes. These lesions are identified as potential causes of bowel intussusception[1], a condition characterized by the telescoping of a proximal segment of the gastrointestinal tract into an adjacent distal segment[2]. Intussusception can manifest at any age, yet it is most prevalent among children aged between 5 and 10 months. It stands as the primary cause of bowel obstruction in

children and the second most common reason for acute abdomen in children, following appendicitis. Among the various polyps, Peutz-Jeghers syndrome is a rare, autosomal-dominant[3] disorder characterized by hamartomatous polyps found in any part of the alimentary tract and accompanied by mucocutaneous pigmentation[2]. A Peutz-Jeghers polyp lacking mucocutaneous pigmentation or a positive family history is defined as a case of solitary Peutz-Jeghers-type hamartomatous polyp. It has been suggested that this condition should be recognized as a distinct clinical entity separate from Peutz-Jeghers syndrome. A solitary Peutz-Jeghers-type hamartomatous polyp is histologically characterized by tree-like branching of smooth muscle fibers[4], with a core of smooth muscle, covered by mucosal tissue with near-normal appearance. These hamartomatous polyps can lead to intussusception of the intestines, with the ratio of occurrences in adults to children estimated at 1:20.

Here, we present a case report of a 6-year-old male child who presented with jejunojejunal intussusception caused by a Peutz-Jeghers-type polyp.

Case Report

A 6-year-old male child presented at the surgical department of IGMC Shimla with a complaint of abdominal pain persisting for one week. The pain intensified over the last two days, reaching a moderate severity and accompanied by episodes of vomiting. The patient's vital signs were found to be within normal limits. Upon physical examination, the abdomen was soft, and no palpable abdominal mass was detected. The patient's family history was unremarkable. Laboratory investigations revealed a hemoglobin level of 10.5g/dl, while other parameters, including liver function tests, kidney function tests, and C-reactive protein (CRP), fell within the normal range. Abdominal ultrasound revealed intussusception in the left hypochondrium along with mesenteric lymphadenopathy. Consequently, an exploratory laparotomy was performed, involving the manual reduction of jejunojejunal intussusception. The specimen was preserved in formalin and sent to the Department of Pathology at IGMC, Shimla for further examination.

Upon gross examination, the intestinal segment measured 3.5 cm in length, displaying a conglomerate complex of multiple polyps, with the largest measuring (2.5 x 2 x 1.5) cm (see Figure 1). The polypoidal segment was situated 1 cm away from one resection margin and 1.2 cm away from another. Microscopic examination revealed a hamartomatous polyp lined by benign intestinal epithelium, with the lamina propria exhibiting smooth muscle arborization, indicative of a Peutz-Jeghers polyp (see Figure 2).

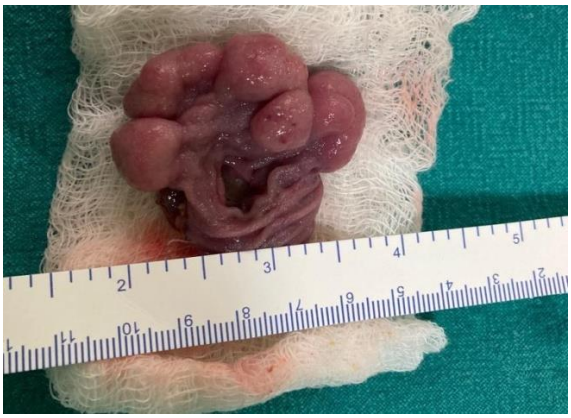

Figure 1. The intestinal segment showing conglomerate complex of multiple polyps

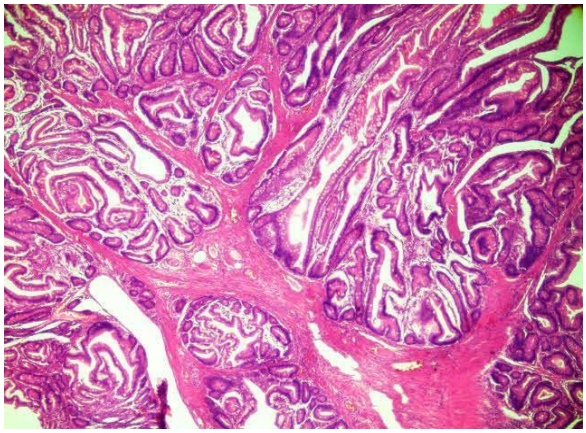

Figure 2. Microscopic examination showed hamartomatous polyp lined by benign intestinal epithelium with lamina propria showing arborization of smooth muscle.

Discussion

Intussusception is defined as the process where a section of the intestine slides into the lumen of the adjacent distal bowel, along with its mesenteric fold. This occurrence disrupts peristalsis, obstructs the normal flow of intestinal contents, compromises the mesenteric vascular flow in the affected bowel segment, and ultimately leads to intestinal obstruction and inflammation of the bowel wall. In our case, intussusception in the jejunum was attributed to the presence of multiple polyps in the small intestine of a 6-year-old male child. Notably, small bowel polyps can be categorized into various types, including hyperplastic polyps, lipomas, sporadic adenomas, familial adenomatous polyposis (FAP), familial juvenile polyposis, and those associated with Peutz-Jeghers syndrome.

Peutz-Jeghers syndrome (PJS) is characterized by the familial occurrence of gastrointestinal hamartomatous polyps along with mucocutaneous hyperpigmentation. The diagnosis of PJS can be established using the WHO criteria[5], which include three or more histologically confirmed Peutz-Jeghers type polyps (PJP), or any number of PJP with a family history of PJS, or characteristic, prominent mucocutaneous pigmentation with a family history of PJS, or any number of PJP and characteristic, prominent mucocutaneous pigmentation. Patients with PJS face an elevated risk of both gastrointestinal and extraintestinal malignancies. The gastrointestinal hamartomatous polyps in patients with PJS exhibit a distinctive histological appearance, with interdigitating smooth muscle fibres forming a characteristic branching tree pattern (arborization).

In our case, due to the absence of mucocutaneous pigmentation and a positive family history, the presentation was characterized as a solitary Peutz-Jeghers-type hamartomatous polyp. However, it remains unclear whether the presence of a solitary Peutz-Jeghers polyp in the intestine should be considered an incomplete form of Peutz-Jeghers syndrome or a distinct entity, particularly in the absence of clinical features and a positive family history.

Conclusion

In conclusion, small bowel intussusception, although relatively uncommon, remains a significant clinical entity that can lead to various complications, including bowel obstruction and inflammation. Our case report of a 6-year-old male child with jejunojejunal intussusception caused by multiple Peutz-Jeghers-type hamartomatous polyps in the small intestine highlights the importance of considering rare pathologies, especially in the absence of classical clinical features or positive family history.

The distinct histological characteristics of Peutz-Jeghers polyps, characterized by the presence of interdigitating smooth muscle fibres forming a branching tree pattern, emphasize the importance of histopathological examination in confirming the diagnosis. Additionally, the lack of mucocutaneous pigmentation and positive family history in our case underlines the complexity in distinguishing between an incomplete form of Peutz-Jeghers syndrome and a solitary Peutz-Jeghers-type hamartomatous polyp as a separate clinical entity[6].

Sources of Support- Nil

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