Ileocolic Intussusception in Infancy is not Always Idiopathic – Adenomyoma as the Leading Point

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Patient: Female, 2-month-old
Final Diagnosis: Intussusception
Symptoms: Distention • ileus • vomiting
Clinical Procedure: Radiographic imaging • ultrasonography
Specialty: Surgery
Objective: Rare disease
Background: Intussusception is a frequent abdominal emergency in infancy, requiring immediate diagnosis and therapeutic intervention. In approximately 90% of cases, intussusception seems to be idiopathic. There has been a reported association of intussusception with lymphoid hyperplasia of Peyer patches in the terminal ileum, possibly acting as the triggering factor. Clinical presentation varies substantially, while the etiology seems idiopathic in most reported cases.
Case Report: This case describes a previously healthy 2-month-old girl who presented with an episode of non-bilious vomiting and deterioration during the 12 hours preceding the visit. Abdominal ultrasonography revealed the typical target sign in the right iliac fossa, without visible peristalsis, confirming the diagnosis of intussusception. Failure of non-surgical reduction led to emergency laparotomy with the working diagnosis of intussusception due to Meckel’s diverticulum. However, laparotomy instead revealed an adenomyoma of the small intestine, a rare benign tumor-like lesion, as the pathological lead point.
Conclusions: This case is interesting for 2 distinct reasons. Not only does it underline the need to maintain a high index of suspicion for triggering factors, even in patients within atypically affected age groups, but it also adds to the remarkably limited selection of reported adenomyomas of the small intestine acting as the pathological lead point for intussusception. In this case report, we aspire to emphasize that especially in patients outside the most affected age group, pediatric surgeons should remain aware of the possibility of adenomyoma as a pathological lead point.

Keywords: Adenomyoma • Intussusception • Gastrointestinal Diseases • Digestive System Surgical Procedures

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Background

Intussusception is defined as the condition in which part of the intestine folds into the adjacent part, with the most prominent type occurring in the ileocolic region and accounting for 90% of the cases [1,2]. Intussusception constitutes the second most frequent cause of acute abdominal distress in children, following appendicitis, and is the first cause of small bowel obstruction in infants, typically appearing between 3 months and 3 years of age, with a peak incidence between 4 and 9 months of age [2]. Intussusception is a medical emergency. Besides bowel obstruction, it impedes normal blood flow as the related mesentery is dragged within the intussusceptum. This can lead to venous congestion and edema. Owing to the latter, ischemia may arise, ultimately resulting in bowel necrosis, perforation, peritonitis, and sepsis [3,4].

In approximately 90% of cases, intussusception seems to be idiopathic. However, there has been a reported association with lymphoid hyperplasia of Peyer patches in the terminal ileum, which possibly acts as the triggering factor [2,5]. The influence of viral factors is thought to play a role, since intussusception often occurs in a seasonal pattern, after a respiratory or gastrointestinal tract infection, mainly with adenovirus or rotavirus, because of lymph node hypertrophy [6]. Among the wide variety of reported lead points, the most common is Meckel’s diverticulum, followed by intestinal duplication and polyps [7].

Patients typically present with intermittent, acute, colicky abdominal pain, vomiting, redcurrant jelly stools, and a palpable sausage-shaped abdominal mass. However, the clinical manifestation of intussusception may vary significantly, including non-specific symptoms. Ultrasonography is the method of choice for diagnosis, having both high sensitivity and specificity [1,2,8,9].

Herein, we describe a rare case in which adenomyoma of the small intestine served as the pathological lead point for intussusception in an unusually affected age group.

Case Report

A preterm 2-month-old girl (premenstrual age) was referred to the on-duty hospital with a documented 24-hour history of lethargy and reduced feeding. The patient’s history included a 25-day hospitalization at the neonatal intensive care unit due to preterm birth at 32 weeks. Her current state before admission, accompanied by an episode of non-bilious vomiting, had deteriorated over the previous 12 hours. However, there were no reported incidents of bloody or mucus-containing feces and no abdominal mass was palpated. In addition to a negative SARS-CoV-2 rapid antigen test, potential infectious diseases in the family were excluded.

On physical examination, the patient presented as mildly febrile with a temperature of 37.3°C, with mild abdominal flatulence, diffuse abdominal tenderness, and slightly decreased bowel sounds. The general status was lethargic, alternating with periods of agitation and irritability. Both respiratory and cardiac assessments were normal. A complete blood count was obtained, reflecting a combination of anemia and leukocytosis, with the results as follows:

- White blood cell count: 15 650 cells/μl;
- Neutrophil count: 57.7%, 9040 cells/μl;
- Lymphocyte count: 31.2%, 4880 cells/μl;
- Red blood cell (RBC) count: 3.91×10⁶ cells/μl;
- Hemoglobin, 10.4 g/dl, hematocrit 29.8%, mean corpuscular volume 73.7 fl, mean corpuscular hemoglobin 26.6 pg, and RBC distribution width, 12.2%;
- The stool test for occult blood was negative.

This constellation of findings raised the suspicion of intussusception, prompting imaging studies.

Ultrasonography and Abdominal Radiography

Abdominal ultrasonography revealed the typical target sign in the right iliac fossa, without visible peristalsis. In addition, an upright abdominal radiograph was notable for a positive step-ladder sign, with multiple air-fluid levels, a finding compatible with the diagnosis of intussusception. On account of persistent intussusception, as confirmed by an ultrasound conducted 1 hour later, the girl was transferred to our pediatric surgery department for further evaluation and treatment.

Initially, having no evidence of peritonitis or bowel perforation at clinical examination, our first therapeutic approach was an ultrasound-guided hydrostatic reduction. Unfortunately, because of inadequate nonoperative reduction, the patient was taken to the operating room and underwent laparotomy, which established the diagnosis of ileocolic intussusception, with Meckel’s diverticulum being the suspected lead point at the terminal ileum (Figure 1). Surgical reduction of the intussusception was performed.

The surgical reduction involved segmental resection of the ischemic bowel and of the leading point, which was thought to be Meckel’s diverticulum, because of its macroscopic morphology, its position – terminal ileum, and its referred bibliographic frequency. An end-to-end anastomosis was performed, along with a prophylactic appendectomy. Intrapertitoneal fluid was drawn for gram staining and cultures, which were negative for bacterial infection. Specimens of the small intestine and the appendix were sent for histological examination.

Figure 1

Adenomyoma manifesting as pediatric ileocolic intussusception

Blevrakis E. et al:
Interestingly, the biopsy results did not confirm the initial diagnosis of Meckel’s diverticulum as the pathological lead point of the intussusception. A profoundly less common entity was identified histologically and was regarded as the triggering factor for intussusception. More specifically, considering the small intestine specimen, the histopathologist described intramural glandular formations lined with a single layer of cuboidal epithelium without cellular atypia (immunochemistry results; CK7+, CK20-). A fraction of these formations showed cystic dilatation. Smooth muscle bundles were recognized among the glandular structures, confirmed by positive desmin immunohistochemical staining. Finally, biopsy results indicated mucosal ischemia, being intramural in places, obviously a result of intussusception. The aforementioned findings were consistent with the diagnosis of adenomyoma of the small intestine, arising from ectopic pancreatic tissue (Figure 2), which according to the histopathology results, was totally removed from the healthy tissue.

Postoperatively, after being sufficiently weaned from mechanical ventilation, the infant followed a steady course toward non-complicated recovery. The infant was hemodynamically stable, with blood pressure ranging between 70-95/40-55 mmHg and heart rate between 125-170 bpm. Clinical findings of gastrointestinal examination revealed abdominal flatulence, with present bowel sounds and passage of stools. Parenteral nutrition was initiated and on postoperative day 2, enteral feeding was started. Stress ulcer prophylaxis with esomeprazole was also provided. Antibiotic coverage consisted of intravenous piperacillin/tazobactam, metronidazole, vancomycin, and amikacin. The infant’s clinical status over the next few days was noticeably improved. On day 7 postoperatively, the patient was discharged in excellent clinical condition, with all inflammatory markers completely restored to within the normal range. A followup with ultrasonography was planned for 3 months after the discharge.

Discussion

Intussusception is among the most common causes of acute abdominal distress and intestinal obstruction in children younger than 3 years old. The classic clinical triad, comprising intermittent abdominal pain, redcurrant jelly stool, and a sausage-shaped abdominal mass, is found in less than 25% of cases [10]. Thus, it is crucial to highlight the broad spectrum of non-specific symptoms and the necessity to maintain a high index of suspicion, including in cases with a potentially atypical presentation, especially in children within the most affected age group. Non-stereotypical symptoms that may suggest intussusception include diarrhea, crying, lethargy, altered consciousness, sepsis, shock, and syncope [1].

In 2021, Kelley-Quon et al published a systematic review that presented an evidence-based algorithm for the therapeutic
approach to intussusception. According to this algorithm, ultrasound-guided enema reduction is a well-established therapy for intussusception, provided that the patient is stable, without evidence of bowel perforation or peritonitis. In the case of partial reduction, repetition of the enema for up to 4 attempts may lead to a successful reduction. Four hours of observation following non-surgical reduction is advisable to check for symptom remission and ensure a tolerated oral regimen. If this procedure fails, or in cases of emergency, operative management, either with manual reduction or with bowel resection and end-to-end anastomosis, is mandatory [11]. Longer duration of symptoms and older age at presentation are positive predictors of the presence of a lead point that predisposes the patient to the need for bowel resection [12]. A recent meta-analysis suggests that air enema is similar in effectiveness to liquid enema in reducing intussusception [13]. Early referral to experienced pediatric surgery departments is critical for reducing the number of cases that require surgical reduction [1].

Overall, our case demonstrates a 2-month-old female patient with a lethargic general status and an episode of vomiting. From an epidemiological point of view, since the highest prevalence of intussusception is noted in ages ranging from 3 months to 3 years and with idiopathic intussusception being the leading etiology, the histological results reporting adenomyoma of the small intestine as the triggering factor for intussusception were assuredly surprising.

Adenomyoma in the gastrointestinal tract is a rare benign tumor-like lesion which is not related to the development of malignancies in later life [5,11]. According to the reviewed English bibliography there are only 15 known cases of adenomyoma resulting in intussusception [14]. The pathogenesis of adenomyoma is not quite clear yet. Adenomyoma is considered a form of epithelial hamartoma or pancreatic heterotopia; thus, magnetic resonance imaging is of no use in this case. Histologically, glandular structures surrounded by interlacing smooth muscle fibers, primarily occupying the submucosa, are its most distinctive features. It predominantly occurs in the pyloric region of the stomach, with the second most common location being the small intestine, usually in the periamplullary area. Reports of jejunal and ileal adenomyoma also exist. The symptoms depend on the location of the lesion and the age of presentation [15]. The age of diagnosis covers an extensive spectrum, ranging from newborns to the elderly. It presents a bimodal peak incidence, attributed to the fact that if asymptomatic, adenomyomas of the small intestine will probably only be discovered during autopsy or surgery for another condition later in life [16]. In pediatric patients, intussusception is the most common reported complication of adenomyoma of the small intestine, as with our patient [17–22]. It is of great importance that no myxomatous tissue is left behind after treatment, because the presence of such residual tissue increases the risk of relapse [23,24]. Adenomyoma of the small intestine arising in Meckel’s diverticulum has also been reported [25]. Intriguingly enough, our initial diagnosis after laparotomy was Meckel’s diverticulum leading to intussusception. Our case further strengthens the association between adenomyoma of the small intestine and intussusception in children. The apparent rarity of this benign condition might be intensified by insufficient reporting of these cases and misdiagnosis [14].

Kazuki et al describe 15 known cases of intussusception, in which adenomyoma was found to be the lead point of intussusception in 11 cases (1 jejunum and 10 ileum) and the other 4 were caused by myxopthelial hamartoma (1 Meckel’s diverticulum, 3 ileum) [14]. Hence, we aim to draw attention to this condition as a pathological lead point of intussusception.

Conclusions

Adenomyoma of the small intestine is a rare pathological lead point of intussusception. Yet, it should not be ignored. In this case report, we aspire to emphasize that, especially in patients in the most affected age group, pediatric surgeons should remain aware of alternative pathological lead points.

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References: