Idiopathic Ileo-Ileal Intussusception In Older Children: A Case Report And Brief Literature Review

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Citation

Abstract
Small bowel intussusceptions are much less common than the ileocolic type. The unusual age group and variable presentation contribute to the frequent delay in diagnosis with increased morbidity and mortality. Majority of cases of isolated ileoileal intussusception are associated with a pathological lead point. Idiopathic ileoileal intussusceptions are rare, especially in older children. To our knowledge, this is the first reported case of antegrade ileoileal intussusception in older school-age children without an apparent predisposing cause.

INTRODUCTION
Intussusception is a disease primarily of infants and toddlers, with peak incidence occurring between 5-9 months. Only 10-20% of cases occur after 2 years of age (1). The incidence of the condition in school aged children has been reported to be 1-6% (2). 90% of intussusception in paediatric age range are ileocolic, ileocecal, or ileoileocolic in nature. Isolated small bowel intussusception (SBI), on the other hand, is rare. It is frequently associated with pathological lead points. Here we present a case of a 12 year old boy with surgically proven ileo-ileal intussusception without any specific discernible cause.

CASE HISTORY
A previously healthy 12-year old boy presented to the Emergency Department with a 3-day history of vomiting and gradually worsening, cramping right iliac fossa pain. He denied fever, dysuria, or weight loss. He reported a similar episode lasting 24 hours one month prior to this, which resolved spontaneously. His past medical and family history were otherwise negative.

On examination, he was apyrexic, but mildly dehydrated. Physical examination was significant for right iliac fossa tenderness with positive rebound tenderness. No palpable mass was found.

His white cell count was $12.9 \times 10^9/l$, with neutrophils of $10.7\times10^9/l$, and a raised C-reactive protein of 146 mg/l. The impression was that of acute appendicitis. He was given intravenous fluids and prepared for theatre. A plain abdominal X-ray or Ultrasound Scan (USS) was not performed on the clinical suspicion of acute appendicitis, as per the standards of care in our region, which rely on the clinical suspicion for the diagnosis of acute appendicitis in this age group.

An emergency laparotomy was later done on the same evening, using Mcburney’s incision. The appendix appeared grossly normal, with significant amount of free serosanguinous fluid. The procedure revealed an ileoileal intussusception, apex which was at the ileocecal junction, the intussusception was irreducible with retrograde manual pressure. There was no evidence of hypertrophied Peyer’s patches, or enlarged mesenteric lymph nodes. An ileocecal resection with side-to-side anastomosis was carried out. The postoperative recovery was uneventful.

The surgical specimen included a 4cm caecum, 1.5cm ascending colon, and a loop of terminal ileum measuring 24.5 cm. Macroscopically, the caecum appeared unremarkable. The terminal ileum was dilated to a maximum of 4.5cm over a length of 17cm. A length of small bowel measuring 21cm was intussuscepted, with no mass lesion identified. On opening, the mucosa of the small bowel was featureless and appeared haemorrhagic. The mucosal surface of the intussuscepted section was ulcerated and congested, with no polyp or tumour found. Histological evaluation of the specimen confirmed haemorrhagic infarction with focal preservation of the mucosa, consistent with intussusception. Microscopically, the appendix showed mild acute appendicitis.
DISCUSSION

Ileocolic intussusception is one of the most common causes of an acute abdominal emergency in children. Seventy percent of cases occur in the first year of life, with its incidence declining rapidly thereafter to <2% in 10-15 year olds. Intussusception confined to small bowel, however, is unusual. It accounts for 1-10% of all cases of childhood intussusception, but up to 50% of cases in older children. Amongst the SBI, ileoileal group constitute the bulk and is generally found in patients ranging between 2-20 years of age, with median age of 10 years.

Unlike ileocolic intussusceptions, small bowel intussusceptions is frequently associated with pathological lead points (in >29% of patients) or occurs postoperatively. Furthermore, after 2 years of age, pathological lead points are found in one third of patients, whereas toddler and younger children are more likely to have idiopathic intussusception. Commonly described pathological entities include Meckel’s diverticula, polyps, lymphomas, intestinal duplication, submucosal haemorrhage with Henoch-Schonlein purpura, and cystic fibrosis. We reviewed the available literature with similarities to this case. In previous studies, two patients older than 2 years old with confirmed ileoileal intussusceptions had no identifiable pathologic lead point. The only other case report of idiopathic intussusceptions in a teenage boy was that of a caecalocolic intussusception. These, together with the present case, highlight that SBI is an important cause of abdominal pain in this population and should be ruled out using appropriate investigations. Speculated factors predisposing to idiopathic SBI are: (i) swelling of small bowel wall, (ii) abnormal gastrointestinal motility, and (iii) scars or adhesions of the bowel from previous insult e.g. prior surgery. The common association of ileoileic intussusceptions and lymphoid hyperplasia may lend support to the above speculation. It is thought that the acute appendicitis was a separate pathological entity from the intussusception in our case report.

SBI usually presents with vomiting and colicky abdominal pain, which can mimic many conditions. The classic clinical features of palpable abdominal mass and bloody stools are rare in SBI. Moreover, presentation of SBI is typically subacute and therefore difficult to diagnose preoperatively. Telescoping of a segment of the intestine into the adjacent bowel loop can cause mechanical obstruction and ischemia. Early diagnosis and prompt treatment are of utmost importance in childhood intussusception in order to lessen discomfort and to avoid bowel ischemia or gangrene. We recommend that a diagnostic ultrasound should be conducted in all school-age children presenting with acute abdominal pain. Suspicious features of SBI on ultrasonography include a 2-3cm doughnut-like lesion in the right lower abdomen quadrant or paraumbilical region. The length of the intussusceptions may be helpful in determining the necessity of surgery. It has been reported that an intussusception length >3.5cm is a sensitive and specific predictor of the need for surgical intervention, as compared to those that will resolve spontaneously.

CONCLUSION

The diagnosis of intussusception in older children is challenging because of a varied clinical presentation and a wide differential diagnosis. Small bowel intussusception particularly, is easily overlooked clinically. This scenario highlights that idiopathic small bowel intussusception and acute appendicitis can occur simultaneously and that plain X-ray and ultrasonography are important to identify cases of intussusception in older children presenting with acute abdominal pain prior to surgery.

References

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