

INTUSSUSCEPTION SECONDARY TO ISOLATED HETEROTOPIC PANCREAS OF THE ILEUM: CASE REPORT AND REVIEW OF THE LITERATURE

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Intussusception in the pediatric age group is usually idiopathic in origin, and in a small number of patients ranging from 2% to 12%, a pathological lesion as a leading point is identifiable.¹⁻⁴ Of the variety of pathological lesions identified as leading points for intussusception, Meckel's diverticulum is the most common, and very rarely, isolated heterotopic pancreas of the ileum.⁵ This is a case report of intussusception in an infant caused by an isolated heterotopic pancreas of the ileum. The literature on the subject is also reviewed.

Case Report

An 11-month-old male infant was admitted to the hospital because of abdominal pain, fever and vomiting of two hours' duration. The infant had had diarrhea the day prior to admission, and while in hospital, he had passed one bloody stool. Clinically, he was mildly dehydrated, and no masses could be felt on abdominal examination. The possibility of intussusception was raised, and this was confirmed by an abdominal ultrasound. A contrast enema revealed intussusception as far as the splenic flexure, and was partially reduced under fluoroscopic control.

The patient had a laparotomy through a transverse, muscle-splitting incision in the right iliac fossa. The intussusception was reduced manually, and while examining the small intestine, a small polyp was found in the distal ileum. This was excised via a small enterotomy, which was closed transversely. Postoperatively, the patient did well and was discharged home on the sixth postoperative day. Histology of the resected polyp (Figures 1 and 2) revealed a 1 cm polypoid tissue covered with a flat, severely inflamed and ulcerated intestinal mucosa. In the underlying stroma were abundant glandular ductal structures resembling those in the pancreas and surrounded by slightly irregular thin-walled blood vessels. Among the glandular structures were streaks of smooth muscle tissue as well as lymphatic hyperplasia at the margins.

FIGURE 1. Photomicrograph showing submucosally located pancreatic ducts in the wall of the ileum (large arrow). Note the ileal mucosa (small arrow) (10x).

FIGURE 2. Photomicrograph showing pancreatic ducts (20x) (large arrow). Note also the lymphoid tissue in the ileal wall (small arrow).

Discussion

The incidence of a localized pathological leading point for intussusception varies from 2% - 12% in large series. Ong and Beasley in a review of 630 episodes of intussusception found a pathological leading point in 60 episodes (9.5%).⁴ Issa et al. found a leading point in 8.2%

TABLE 1. *Clinical summary of children with isolated heterotopic pancreas of the ileum as a leading point for intussusception.*

Authors	Year	Age	Sex	Type of intussusception	Site of heterotopic pancreas
Carleton & Ackerbawn ¹⁰	1976	9 years	M	Ileo-ileal	Submucosal
Pan LC ¹⁸	1988	Not mentioned	Not mentioned	Not mentioned	Not mentioned
Erdener et al ¹¹	1993	8 months	M	Ileo-ileal	Submucosal
Abel et al ⁸	1999	16 months	M	Ileo-ileal	Serosa
		16 months	M	Ileo-colic	Serosa
Scholz, Loff, Wirth ¹²	2000	11 years	F	Recurrent ileo-ileal	Polypoid mass
Hamada et al ⁵	2000	14 months	M	Ileo-ileal	Serosa
		2 months	M	Ileo-ileal	Submucosal
		3 years & 4 months	M	Recurrent ileo-ileal	Intermuscular
Present case	-	11 months	M	Ileo-colic	Submucosal

of 233 patients with 244 intussusceptions,⁶ while Ein found 31 patients (5.5%) with a leading point among 569 children with intussusception.²

The variety of pathological lesions identified as leading points in childhood intussusception include Meckel's diverticulum, which is the most common, intestinal polyps, duplication cysts, lymphoma and lymphosarcoma.^{1-3,4,7} In a large series of 60 patients with pathological leading points, Meckel's diverticulum was the cause in 27 (45%) of them.⁴ Puri and Guiney reviewed 292 consecutive children with intussusception, and 27 of them had identifiable intestinal lesions as a leading point. Among these, Meckel's diverticulum was the cause in 13 (48%) of them, while in 10 cases, intussusception was caused by a tumor of the small intestine.⁷ In rare isolated instances, heterotopic

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pancreas of the ileum acts as a leading point for intussusception.^{5,7-13}

Heterotopic pancreas is defined as pancreatic tissue that has neither anatomic nor vascular continuity with the normally located pancreas. It is not uncommon, and is found in 1%-2% of autopsies, and as an incidental finding in about 1 of every 500 upper abdominal operations.⁹ It occurs most commonly in the stomach, duodenum, and proximal parts of the jejunum, and in rare instances, at other sites including Meckel's diverticulum, ileum, colon, gallbladder, common bile duct, umbilicus, fallopian tubes, liver, spleen and mediastinum.^{13,14} Histologically, heterotopic pancreas is classified into 3 types: type I (all elements of the normal pancreatic tissue are present); type II (pancreatic tissue without islet cells); and type III (pancreatic ducts only).¹⁵ Our patient had type III heterotopic pancreas. Although, most patients with heterotopic pancreas are asymptomatic, various changes including pancreatitis, pseudocyst formation, inflammation, abscess formation, pancreatic adenoma and carcinoma have been reported in heterotopic pancreas.¹⁴⁻¹⁷

Heterotopic pancreas in the ileum is rare, and when seen, it is usually associated with Meckel's diverticulum. Isolated heterotopic pancreas of the ileum on the other hand is very rare, usually asymptomatic and discovered incidentally during surgery for other conditions, and very rarely as a leading point for intussusception.^{5,8,10-12} In the series of 292 consecutive children with intussusception by Puri and Guiney, none of the 27 patients identified with leading points had isolated heterotopic pancreas.⁷ In the review of 630 episodes of intussusception by Ong and Beasley, none of the 60 patients with pathological leading had isolated heterotopic pancreas.⁴ In a series of 134 children with intussusception, Hamada et al. found 8 (5.9%) with intestinal lesions as leading points, 3 with isolated heterotopic pancreas of the ileum.⁵ Pang et al. reviewed 32 histologically proven cases of heterotopic pancreas, of which 14 (44%) were symptomatic and 18 (56%) were asymptomatic or incidental. Six of the 14 symptomatic cases were in the ileum, and 4 caused intussusception, 2 in Meckel's diverticulum, 1 in a duplication cyst, and 1 as an isolated heterotopic pancreas of the ileum.¹⁸

In an extensive review of the literature, we found only nine cases (Table 1) of isolated heterotopic pancreas of the ileum as a leading point for intussusception in children.^{5,8,10,12,18} Their ages ranged from 2 months to 11 years, and in all except two, the intussusception was ileo-ileal, two of which were recurrent intussusception. Whereas idiopathic intussusception is usually seen in infants between 3 and 12 months of age, intussusceptions secondary to a pathological lesion on the other hand have a different age distribution. The relative frequency of a pathological lesion causing intussusception increases with age, but the first year remains the period with the highest frequency.^{2,4} Six of the nine children with isolated heterotopic pancreas of the

ileum causing intussusception were less than two years of age (Table 1).

Although there are a few instances in which an intussusception secondary to a leading point has been successfully reduced, most intussusceptions due to a pathological lead point are irreducible by barium enema.² With recent advances in ultrasonography, it is now possible to diagnose ileo-ileal intussusceptions pre-operatively, and to avoid delays, let patients undergo open reduction. It is also important to recognize heterotopic pancreas within the wall of the ileum after manual reduction of the intussusception by palpating the bowel. This is necessary for those patients with submucosal location of the heterotopic pancreas where the tumor may not be obvious from the outside, like it was in our patient. Simple excision is the treatment of choice. This will avoid recurrence of the intussusception as well as the risk of its subsequent clinical sequelae.

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