Early diagnosis and surgical management in adult intussusceptions caused by ectopic pancreas

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ABSTRACT

Adult intussusception comprises of only 5% of all intussusception cases with majority occurring in children and accounts for only 1–5% cases of bowel obstruction in adults. Ectopic pancreas is an infrequent anomaly defined as pancreatic tissue that lacks anatomical or vascular communication with the normal body of the pancreas. When reported it is mostly found in the stomach and small intestines but seldom in the colon. We present a case of a 31-year-old male who presented with symptoms of an acute abdomen which was diagnosed as an intussusception of an unknown etiology on a contrast-enhanced computed tomography (CT). The patient was resuscitated and immediately operated on and underwent a right hemicolectomy and eventually an end ileostomy formation. No cause for the intussusception was apparent intra-operatively but histological examination of the resected bowel specimen demonstrated ectopic pancreas. The patient had a very short recovery time without incident which may be attributed to his age and absence of co-morbidities. He was discharged on a follow-up plan to discuss the reversal. The patient went on to have reversal of his stoma after six months of the initial surgery with a very good outcome.

INTRODUCTION

Intussusception is defined as the invagination of a segment of bowel within an immediately adjacent segment and almost invariably occurs from proximal to distal. It is most frequent in children with incidence peaking at 5–10 months of age, becoming less common above two years and is predominantly rare in adults. Adult intussusception represents just 5% of all cases of intussusception and accounts for only 1–5% of cases of adult bowel obstruction [1–3]. Colo-colic type intussusception has been found to be more common in adults [4]. Unlike children, in whom around 90% of cases are idiopathic, adult intussusception is usually secondary to an underlying pathology, such as polyps, Meckel’s diverticulum, strictures, benign neoplasms, or carcinomas [3]. The presentation is mostly insidious in adults, with nonspecific symptoms but can lead to intestinal obstruction and ischemia of the advancing bowel. The primary management in children with uncomplicated intussusception consists of nonoperative reduction with air or barium enema. Adults, however, will typically require surgical intervention to identify the underlying pathology.
Ectopic pancreas is defined as pancreatic tissue that lacks anatomical or vascular communication with the normal body of the pancreas [5]. It is an infrequent congenital anomaly with an incidence of 0.55–13.7% on autopsy series [6, 7]. It has been reported to be most commonly located in the stomach (25–38% of cases), the duodenum (17–36% of cases), and the jejunum (15–22% of cases) although seldom described in other locations, such as Meckel’s diverticulum, colon, gallbladder, umbilicus, fallopian tube, mediastinum, spleen, liver and, as in our case, the ileum [8–10].

A literature review of 528 cases identified the prevalence of ileal heterotopic pancreas to be just 0.2% [11]. Ectopic pancreas (EP) usually presents in the form of small yellow nodules varying in size from 1 to 5 mm, thus can be misdiagnosed as a lipoma as reported on the primary CT scan in our patient’s case. In descending order of occurrence, the involved histologic layers are the submucosa, muscularis propria, and serosa [12]. They are classified according to the Heinrich classification system. Clinically significant lesions tend to be larger than 1.5 cm and involve or are adjacent to the mucosa [13].

Ectopic pancreas in the small intestine is typically benign and therefore most cases are asymptomatic, being discovered incidentally during endoscopy or surgery for another presentation or at autopsy. When symptomatic, patients can present with bleeding, pancreatitis and rarely symptoms associated with malignant transformation or, as in our case, bowel obstruction due to intussusception [8, 14–17]. In some cases it has led to death, as reported in literature [18].

We describe a rare case of an adult presenting with intussusception secondary to EP.

CASE REPORT

A 31-year-old male with no significant past medical history or previous abdominal operations presented with a 1-day history of severe lower abdominal pain, multiple episodes of vomiting, and loose stools. On examination, he had generalized abdominal tenderness with peritonism and absent of bowel sounds. Blood tests on admission displayed a raised white cell count (WCC) of 16 × 10^9/L, C-reactive protein (CRP) of <0.6 mg/L, and lactate of 6. A contrast-enhanced CT abdomen and pelvis was performed on a suspicion of acute abdomen caused by bowel ischemia. However, it revealed ileo-colic intussusception (Figures 1 and 2). A 26 mm fat density opacity representing a possible lipoma as a lead point.

The patient was resuscitated with intravenous fluids, antibiotics, and analgesia, and was taken to theatre for an emergency laparotomy within 3 hours of presentation. Intra-operatively, 50 cm of terminal ileum was found intussuscepted into 10 cm of ascending colon (Figure 3). Manual reduction was attempted without success. So, a right hemicolectomy was performed. Initially a defunctioning double barrel stoma formation was planned but not performed at this time due to significant edema of the bowel. After discussion, consensus was for a re-look laparotomy once the patient had stabilized with a view for stoma formation as the ideal approach. Approximately 24 cm of ileum and 17 cm of colon including cecum, ascending and proximal transverse colon were resected and a temporary primary anastomosis was formed. The abdomen was closed by a vacuum-assisted closure dressing. No visible or palpable mass was identified intra-operatively but histological examination of specimen concluded a focal ectopic pancreas. However, grading of the ectopic pancreatic tissue had not been done histologically because of the patchy nature of the tissue.

The patient was initially treated in the surgical intensive recovery unit post-operatively then stepped down to the ward. He required rectus sheath infusion and morphine patient controlled analgesia. The patient was treated with intravenous antibiotics and parenteral nutrition.

Figure 1: Target sign in intravenous contrast-enhanced CT coronal view.

Figure 2: Target sign in intravenous contrast-enhanced CT cross-sectional view.
Table 1: A summary of all reported cases of adult intussusception caused by ectopic pancreas in literature

| Title | Author | Symptoms | Duration | Imaging | Operation | Distance from IC valve | Site of HP | Layer of HP | Size of HP | Heinrich classification | Type of intussusception | Previous abdominal surgery | Treatment | Post-op | Reference |
|-------|--------|----------|----------|---------|-----------|------------------------|-----------|-------------|-----------|------------------------|--------------------------|---------------------------|-----------|---------|-----------|
| The role of laparoscopy in the management of a diagnostic dilemma: Jejunal ectopic pancreas developing into jejunojejunal intussusception | Giordano et al. | Nausea, intermittent abdominal pain | 2 days | CT scan shows a specific thickening of jejunal with partial contrastographic enhancement near the left colon, with a modest dilatation of the lumen of small bowel upstream of the injury | Diagnostic laparoscopy | Jejunum—mass of 4 cm in diameter was discovered on the intestinal serosal surface of the antimesenteric side | Type 2 | Jejunojejunal | n/a | | | Resection of intussusception + mass | (1198 Giordano et al., 2017) |
| Jejunal small ectopic pancreas developing into jejunojejunal intussusception: A rare cause of ileus | Shoji et al. | Nausea, intermittent abdominal pain | Several months | Plain abdominal radiography showed some intestinal gas and fluid levels. Abdominal CT scan demonstrated a target sign suggesting bowel intussusception and dilated small bowel. Subsequent jejuno-jejunography using a naso-jejunal tube showed an oval-shaped mass 15 mm in diameter with a smooth surface in the jejunum, which suggested a submucosal tumor (SMT), and edematous mucosa around the mass | Laparotomy | Jejunum | 3 | Jejunojejunal | n/a | | | Segmental resection of the ileum with ileoileostomy was completed. An enterotomy confirmed the presence of a pedunculated nodule (60 mm by 18 mm) with fatty tissue inside | (1200 Hirasaki et al., 2009) |
| Adult intussusception and gastrointestinal bleeding due to an isolated heterotopic pancreas | Wu et al. | Intermittent abdominal pain, nausea, vomiting | 1 month | Plain abdominal film showed a dilated small bowel and associated air fluid levels indicative of a small-bowel obstruction. Computed tomography scans of the abdomen revealed an ileal intussusception. A nodule with an abundant fatty component was noted in the computed tomography scan (Figure 1), which included several strips of high density inside, and was identified at the proximal end of the intussusception | Laparotomy | Ileum | n/a | Ileum | 6 × 1.8 cm | n/a | Ileocecal | n/a | Unremarkable | (1207 Wu et al., 2013) |
| Heterotopic pancreas: A rare cause of ileal intussusception | Monier et al. | Episodes of melena + constipation | 1 year | Computed tomography (CT) revealed a large circumferential lesion measuring approximately 8.0 × 1.8 cm involving the terminal ileum, which acted as a leading point to an ileo-ileal intussusception (Figure 1A–C). There were also other small multiple satellite lesions | CT enterography | Ileum | Submucosal | Ileum | n/a | Ileocecal | n/a | Resection of the segment containing the submucosal lesion was carried out with side-to-side anastomosis. | (1208 Monier, 2014) |
| Adult Intussusception caused by heterotopic pancreas | Karki et al. | Nausea, intermittent abdominal pain | 10 days | Radiographs of the chest and abdomen and abdominal ultrasound study showed no abnormalities. Computed tomography (CT) of the abdomen was arranged as intermittent bowel obstruction was highly suspected due to her clinical signs and symptoms. Non-contrast enhanced CT revealed wall and mucosal fold thickening, luminal dilatation in a segment of small intestinal loops about the jejunal level over the left side of the lower abdomen, and soft tissue mass in the bowel lumen (Figure 1). A targetshaped lesion consisting of multiple concentric rings was found on the left side on contrast-enhanced CT (Figure 2) | Laparotomy | Ileum | n/a | Serosal surface of antimesenteric side | 6.6 × 1.6 cm | Acinar glands, ductules, islets | Jejunojejunal | n/a | | Short segment of intussusceptions was reduced manually, a yellowish lipomatous mass (6.6 × 2.2 × 1.6 cm) was discovered on the serosal surface of the antimesenteric side (Figure 3). Segmental resection of the jejunum including the mass and end-to-end anastomosis was performed | (1216 Kok, 2007) |
Table 1: (Continued)

| Title | Author | Symptoms | Duration | Imaging | Operation | Distance from IC valve | Site of HP | Layer of HP | Size of HP | Heinrich classification | Type of intussusception | Previous abdominal surgery | Treatment | Post-op | Reference |
|-------|--------|----------|----------|---------|-----------|------------------------|-----------|------------|-----------|-------------------------|--------------------------|--------------------------|-----------|---------|-----------|
| Adult Intussusception caused by inverted Meckel's diverticulum containing mesenteric heterotopic pancreas and smooth muscle bundles | Lee et al. | Intermittent abdominal pain + haematochezia + malaena | 2 months | An endoscopic study was unable to locate the site of the bleeding. An abdominal and pelvic computed tomography (CT) scan showed a pedunculated intraluminal mass with central heterogeneous fat attenuation | Mini-laparotomy | Ileum | A nodular lesion with the adipose tissue sandwiched between the continuous lining of the proper muscle layers (Figure 2A). Microscopic examination revealed that the bulbous tip lesion was covered by the full thickness of the intestinal wall and had deep ulcerations (Figure 2C). The mucosa of the tip contained non-dysplastic epithelial glands. Focal heterotopic antral-type gastric tissue was also present (Figures 2C and 3A). Interestingly, ectopic pancreatic tissue and smooth muscle bundles were located within the entrapped mesenteric fat | 5.1 × 3.0 × 2.8 cm | Type 1 | Ileal | Appendectomy 10 years ago | Segmental resection | n/a | (1217 Lee, 2017) |
| Intussusception caused by a heterotopic pancreas. Case report and literature review | Chandra et al. | Intermittent abdominal pain, constipation, alternating bowel habit | 3 years | Ultrasound examination revealed a loop of abnormal bowel in the pelvis extending to the right iliac fossa over which it was noted that the patient experienced marked tenderness from the ultrasound probe. It contained concentric rings of high and low echogenicity, highly suggestive of a small bowel intussusception. No proximal dilatation was noted | Laparotomy n/a | Jejunum n/a | 60 × 40 × 35 mm | Acinar + ducts | Jejunojejunal n/a | n/a | (1220 Chandra, 2004) |
| Heterotopic pancreas as a leading point for small-bowel intussusception in a pregnant woman. | Gurbulak et al. | 31 weeks gravid, abdominal pain, bile-stained vomit | 4 days | Computed tomography of the abdomen revealed “target lesions” suggestive of a small bowel intussusception and free-fluid in the abdominal cavity | Laparotomy 80 cm + e-section | Ileum n/a | n/a | Acinar + ducts | Ileal | n/a | Laparotomy + c-section | (1222 Gurbulak, 2007) |
| Ileocolic intussusception due to ileal ectopic pancreas with abundant fat tissue mimicking lipoma | Chuang et al. | Intermittent abdominal pain, episodic vomiting | 6 months | The plain abdominal film showed dilated small bowel, and computed tomography (CT) scan of the abdomen and pelvis showed dilatation of the small bowel and intussusception of the small bowel into itself, a finding which suggested intussusception (Figure 1). Moreover, a nodule with an abundant fatty component was identified at the proximal end of the intussusception | Laparotomy n/a | Ileum n/a | n/a | Acinar + ducts | Ileal | Unremarkable recovery for patient + baby | Segmental resection of the ileum with ileoileostomy was completed | (1223 Chuang, 2010) |
Table 1: (Continued)

| Title                                                                 | Author                     | Symptoms                        | Duration | Imaging                                                                 | Operation | Distance from IC valve | Site of HP | Layer of HP | Size of HP | Heinrich classification | Type of intussusception | Previous abdominal surgery | Treatment | Post-op | Reference |
|----------------------------------------------------------------------|----------------------------|---------------------------------|----------|-------------------------------------------------------------------------|-----------|------------------------|------------|-------------|------------|------------------------|--------------------------|----------------------------|------------|---------|-----------|
| Ectopic pancreas, intussusception, and a ruptured mesenteric band: An unusual association | Ganapathi et al.           | Abdominal pain, vomiting, diarrhea | acute    | Plain abdominal radiography showed one prominent loop of small bowel without clear evidence of obstruction. A CT scan of his abdomen 8 hours after admission showed features of ileoileal intussusception (Figure 1), with dilated small bowel loops proximally along with large amount of free fluid. There was also a suggestion of a large soft tissue mass within the pelvis (Figure 2) and intimately related to the intussusception. | Laparotomy | 30 cm                  | Ileum      | 30 mm       | Acini + ducts | ileal           | no                       | The band was ligated and 20 cm of intussuscepted small bowel was resected. Healthy intestine was anastomosed end-to-end. | Uneventful | (Ganapathi, 2011) |

Abbreviations: CT: Computed tomography; IC: Ileocecal; HP: Heterotopic pancreas
On day 8, the patient returned to the theatre for relook laparotomy, washout, closure of abdominal wall, and formation of end ileostomy. He recovered well in intensive treatment unit (ITU) post-operatively. He was extubated on day 9 and stepped down to the ward on day 11.

The patient’s symptoms slowly resolved and he recovered well. He was discharged on day 25 with follow-up arranged for review and discussion of stoma reversal. His stoma was reversed electively after six months with a very good outcome. He was discharged with a long-term follow-up plan.

DISCUSSION

Even if symptomatic, the pre-operative diagnosis of EP still remains challenging with imaging studies such as ultrasonography, CT, and endoscopy, not being specific as demonstrated in our case. Definitive diagnosis is reached with histopathology.

When EP has previously been located in the ileum causing intussusception, often a coexisting Meckel’s diverticulum has been noted, which is thought to exacerbate the ability of the EP to act as a lead point for intussusception [19, 20]. Isolated EP of the ileum causing intussusception without the presence of Meckel’s diverticulum, as reported here, is particularly rare. Cases of ileal pancreatic heterotopia causing intussusception has been described in children up to the age of 12 [21]. We provide a summary of all reported cases of adult intussusception caused by ectopic pancreas in literature (Table 1).

Manual reduction in our case was unsuccessful. Previous reviews recommend that the treatment of adult intussusception is resection of the intussusception mass without prior attempts to reduce it. The vast majority of adults with intussusception have an underlying pathology as the cause [22–24].

CONCLUSION

The role of laparoscopy in the management of intussusception has been described as an attractive option, especially in the emergency setting in hemodynamically stable patients with non-conclusive imaging. Although this may go on to require laparotomy in most adults and in children whose manual reduction fails, with confirmation of the diagnostic suspect of intussusception. It may entail smaller subsequent laparotomy incisions, shorter bowel manipulation time along with general reduction in post-operative hospital stay, and possible reduction in analgesia requirements, surgical site infections, cardiac respiratory complications, and post-operative mortality. However, in this case laparoscopy was not an option. Most of the times an adult has an underlying pathology for intussusception that will require proper exploration and resection. In some cases endoscopic approach has also been described as a safe and effective approach especially when found in upper gastrointestinal tract especially in the stomach.

REFERENCES

1. Hirasaki S, Kubo M, Inoue A, Miyake Y, Oshiro H. Jejunal small ectopic pancreas developing into jejunojejunal intussusception: A rare cause of ileus. World J Gastroenterol 2009;15(31):3954–6.
2. Gupta RK, Agrawal CS, Yadav R, Bajracharya A, Sah PL. Intussusception in adults: Institutional review. Int J Surg 2011;9(1):91–5.
3. Marinis A, Yiallourou A, Samanides L, et al. Intussusception of the bowel in adults: A review. World J Gastroenterol 2009;15(4):407–11.
4. Winslet MC. Intestinal obstruction. In: Russel RCG, Williams NS, Bulstrode CJK, editors. Bailey & Love’s Short Practice of Surgery. 24ed. UK: CRC Press; 2004. p. 1194–7.
5. Mulholland KC, Wallace WD, Epanomeritakis E, Hall SR. Pseudocyst formation in gastric ectopic pancreas. JOP 2004;5(6):498–501.
6. Gokhale UA, Nanda A, Pillai R, Al-Layla D. Heterotopic pancreas in the stomach: A case report and a brief review of the literature. JOP 2010;11(3):255–7.
7. Lai EC, Tompkins RK. Heterotopic pancreas. Review of a 26 year experience. Am J Surg 1986;151(6):697–700.
8. Tanaka K, Tsumoda T, Eto T, et al. Diagnosis and management of heterotopic pancreas. Int Surg 1993;78(1):32–5.
9. Bromberg SH, Camilo Neto C, Borges AFA, Franco MIF, França LCM, Yamaguchi N. Pancreatic heterotopias: Clinicopathological analysis of 18 patients. [Article in Portuguese]. Rev Col Bras Cir 2010;37(6):413–9.
10. Ormarsson OT, Gudmundsdottir I, Márvik R. Diagnosis and treatment of gastric heterotopic pancreas. World J Surg 2006;30(9):1682–9.
11. Bruneton J. Aberrant Pancreases. Imaging of Gastrointestinal Tract Tumors. Berlin, Heidelberg: Springer; p. 68–72.
12. Gupta MK, Karlitz JJ, Raines DL, Florman SS, Lopez FA. Clinical case of the month. Heterotopic pancreas. J La State Med Soc 2010;162(6):310–3.
13. Armstrong CP, King PM, Dixon JM, Macleod IB. The clinical significance of heterotopic pancreas in the gastrointestinal tract. Br J Surg 1981;68(6):384–7.
14. Hirasaki S, Tanimizu M, Moriwaki T, Nasu J. Acute pancreatitis occurring in gastric aberrant pancreas treated with surgery and proved by histological examination. Intern Med 2005;44(11):1169–73.
15. Pang LC. Pancreatic heterotopia: A reappraisal and clinicopathologic analysis of 32 cases. South Med J 1988;81(10):1264–75.
16. Emerson L, Layfield LJ, Rohr LR, Dayton MT. Adenocarcinoma arising in association with gastric heterotopic pancreas: A case report and review of the literature. J Surg Oncol 2004;87(1):53–7.
17. Hsia CY, Wu CW, Lui WY. Heterotopic pancreas: A difficult diagnosis. J Clin Gastroenterol 1999;28(2):144–7.
18. Hida Y, Kitamura O, Shigeta A, Takatsu A, Yoshino Y, Ohtsuki M. Analysis of sudden death caused by intestinal obstruction. [Article in Japanese]. Nihon Hoigaku Zasshi 1997;51(6):423–9.
19. Chandra N, Campbell S, Gibson M, Reece-Smith H, Mee A. Intussusception caused by a heterotopic pancreas. JOP 2004;5(6):476–9.
20. Lee S, Cho SW. Adult intussusception caused by inverted Meckel’s diverticulum containing mesenteric heterotopic pancreas and smooth muscle bundles. J Pathol Transl Med 2017;51(1):96–8.
21. Sundaram J, Menon P, Kumar V, Rao KLN, Vaiphei K, Kakkar N. Isolated ileal pancreatic heterotopia causing intussusception with gangrene. Fetal Pediatr Pathol 2015;34(4):252–6.
22. Kia KF, Mony VK, Drongowski RA, et al. Laparoscopic vs open surgical approach for intussusception requiring operative intervention. J Pediatr Surg 2005;40(1):281–4.
23. Giordano A, Alemanno G, Bergamini C, Prosperi P, Bruscino A, Valeri A. The role of laparoscopy in the management of a diagnostic dilemma: Jejunal ectopic pancreas developing into jejunojejunal intussusception. Case Rep Surg 2017;2017:8452947.
24. Zhou Y, Zhou S, Shi Y, Zheng S, Liu B. Endoscopic submucosal dissection for gastric ectopic pancreas: A single-center experience. World J Surg Oncol 2019;17(1):69.

Author Contributions

Smriti Karki – Conception of the work, Design of the work, Acquisition of data, Analysis of data, Interpretation of data, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Hina Aziz – Acquisition of data, Analysis of data, Interpreting of data, Drafting the work, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Joseph Watfah – Conception of the work, Design of the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

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Conflict of Interest

Authors declare no conflict of interest.

Data Availability

All relevant data are within the paper and its Supporting Information files.

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