



Duodenojejunal intussusception secondary to a duodenal duplication cyst manifested by abdominal pain and obstructive jaundice in a 40-year-old female: A case report

¹Sosas, SCR

²Tan, DLU

³Bermas, J

Abstract

Introduction: Enteric duplication cyst is a rare congenital anomaly of the digestive tract, affecting 0.2% of children and 5-6% of adults, occurring in 1 in 4,500 births. Intussusception is uncommon in adults, accounting for less than 5% of cases, and is found in 1% of bowel obstruction patients. Clinical symptoms in adults can differ from the typical pediatric presentation.

Case: A 40-year-old female was hospitalized with epigastric pain and vomiting, which began 12 hours prior to admission. She experienced crampy pain, escalating to severe pain radiating to the right upper quadrant, along with 12 episodes of non-bilious vomiting. She had a previous history of acute cholecystitis and no known comorbidities. Upon admission, her blood pressure was elevated, and she had icteric sclerae and a tender right upper quadrant with a mass in the epigastrium. Laboratory findings showed leukocytosis, hypokalemia, hypoalbuminemia, and hyperbilirubinemia. A computed tomography of the whole abdomen with contrast revealed duodenojejunal intussusception with biliary obstruction, along with a duodenal duplication cyst measuring 4.2 x 5.8 cm, acting as the lead point, with invagination of part of the pancreatic head into the intussusception. She was managed with decompression, medications, and intravenous antibiotics. After three days, she underwent exploratory laparotomy, pancreatoduodenectomy, segmental resection of the jejunum, and anastomosis procedures. Histopathology confirmed the duodenal duplication cyst, showing intestinal-type mucosa lining exhibiting ischemic necrosis with no atypia or malignant tumor cells. The patient tolerated the procedure well, and her symptoms resolved. She was discharged on the 14th hospital day in stable condition.

Conclusion: Adult duodenojejunal intussusception is a rare disease that is difficult to diagnose due to its nonspecific symptoms and is possible in cases of duplication cysts which can act as a lead point, such as in our patient. Therefore, a high index of suspicion and imaging plays an important role in the diagnosis of a duplication cyst with intussusception in adults, especially those presenting with abdominal pain, vomiting, and jaundice. A correct and timely diagnosis is needed to prevent various complications including bowel infarction and sepsis.

Keywords: Duplication cyst, Adult intussusception

Correspondence:
Steven Clyde R. Sosas, MD
sosassteven@gmail.com

Introduction

Enteric duplication cyst is an uncommon congenital abnormality of the digestive tract affecting 0.2% of children and occurs in 1 of 4,500 births.¹ It has a 5-6% prevalence in adults and can occur at any level from the mouth to the anus with duodenal duplication cyst being the least common location among small bowel cysts and accounting for 4% of all gastrointestinal tract duplications, with estimated prevalence of less than 1 per 100,000 live births.² Duodenal duplication cysts can have an array of vague symptoms, often misleading the clinical picture, thereby posing a diagnostic challenge for clinicians. Its complication depends on the size and location of the cyst with larger-sized cysts resulting in compressive symptoms including pancreatitis, jaundice, malnutrition, and weight loss.³

Intussusception, a possible complication of a duodenal duplication cyst, is rare in adults compared to children which only accounts for <5% of all cases of adult intussusception and is found in 1% of patients with bowel obstruction. Moreover, adult intussusception differs from pediatric patients in terms of presentation, cause, diagnosis, and treatment.⁴

Duodenal duplication cyst is a diagnostic challenge and should not be overlooked due to its possible serious complications. Herein, we present the case of a duodenal cyst in a 40-year-old Filipino woman causing duodenojejunal intussusception and jaundice.

Objectives

General Objective

To report a case of duodenojejunal intussusception secondary to a duodenal duplication cyst in a 40-year-old female presenting with abdominal pain and obstructive jaundice

Specific Objectives

1. Elaborate on the prevalence of intussusception and duplication cysts in adults
2. Discuss the presentation and etiology of intussusception in adults
3. Discuss the diagnosis and management of intussusception and duplication cysts

Case Presentation

A 40-year-old female was hospitalized for epigastric pain and vomiting. Twelve hours prior to admission, the patient had sudden onset postprandial epigastric pain, non-radiating, crampy in character, with a pain score of 5/10. No consult was done, or medication taken. An hour prior to admission, epigastric pain persisted, now with a pain score of 10/10 and radiating to the right upper quadrant. This was associated with 12 episodes of intermittent non-bilious vomiting amounting to $\frac{1}{2}$ to 1 cup per episode made of previously ingested food. This was not associated with changes in stool caliber, urine color, flatus, bowel movements, fever, hematemesis, diarrhea, or constipation. Abdominal mass and enlargement, weight loss, icteric sclerae, and jaundice were not observed by the patient or her family. She was previously diagnosed with acute cholecystitis managed medically and noted to have resolved. She has no known comorbidities or previous surgeries. She has a family history of liver and colon cancer on the maternal side.

On admission, the patient had elevated blood pressure of 130/90 with other vital signs noted to be stable. She had icteric sclerae and dry mucous membranes. The abdominal exam showed a non-distended, tympanic, soft abdomen with normoactive bowel sounds and a 2 x 2 cm nonmobile, non-pulsatile, non-tender mass over the epigastrium with well-defined borders and dull upon percussion. There was negative succussion splash, shifting dullness and fluid wave test. The right upper quadrant of the abdomen was tender with a positive Murphy's sign. The rest of the physical exams were unremarkable. At this point, peptic ulcer disease,

pancreatitis, small bowel obstruction, and both gastric and hepatobiliary malignancies were considered. She was placed on nothing per os, and a nasogastric tube was inserted for decompression. Medical therapy was initiated with 40 mg of intravenous (IV) omeprazole once daily and a single dose of 30 mg IV ketorolac for pain relief. She was also given 10 mg of metoclopramide IV every 8 hours and 10 mg of hyoscine n-butylbromide every 6 hours for nausea and vomiting. Empiric antibiotics were started with 400 mg of ciprofloxacin IV every 12 hours and 500 mg of metronidazole IV every 8 hours.

On the 3rd day of admission, the patient was scheduled for exploratory laparotomy, pancreateoduodenectomy, segmental resection of jejunum, hepaticojejunostomy, pancreaticojejunostomy, and gastrojejunostomy. Intraoperative findings during duodenotomy revealed an endophytic

mass with smooth mucosa on the medial wall of the second portion of the duodenum. Final histopathology of the duodenal mass revealed a duodenal duplication cyst showing intestinal-type mucosa lining exhibiting ischemic necrosis with no atypia or malignant tumor cells noted (Figure 5). Sections from the pancreas, duodenum, and gallbladder were unremarkable while sections from the peripancreatic fat showed few lymph nodes with no atypical cells noted.

Postoperatively, patient noted to be hypokalemic which was corrected by 3 cycles of potassium chloride 10 meqs and lung atelectasis prevented with incentive spirometry. There was no other complication during and after the operation and diet was gradually resumed and tolerated. Resolution of abdominal pain, vomiting, and jaundice noted postoperatively. The patient was discharged on the 14th hospital day in stable condition.

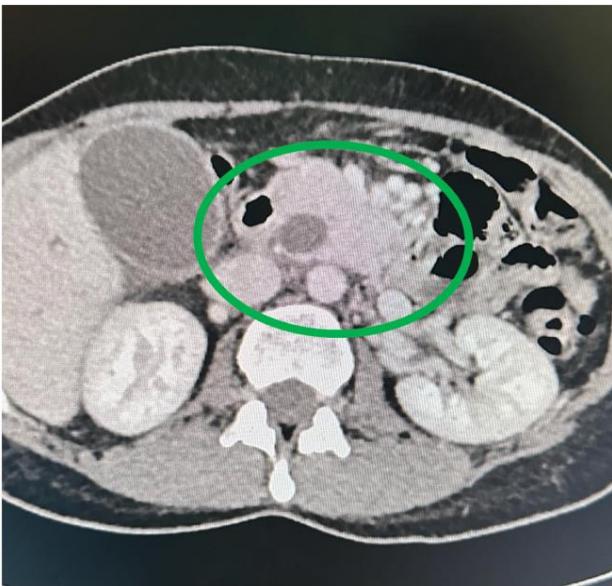


Figure 1: CT Scan of whole abdomen with contrast. Axial view showing a 4.2 x 5.8 cm duodenal duplication cyst with duodenal intussusception and invagination of the pancreatic head into the intussusception



Figure 2: CT scan of whole abdomen with contrast. Coronal view showing narrowing of the distal common bile duct and resultant hepatic biliary obstruction secondary to intussusception

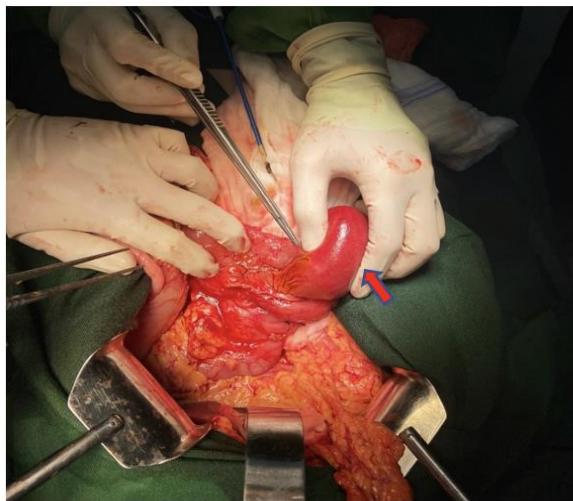


Figure 3. Intraoperative finding of a 45 x 45 x 35 mm duodenal cyst (red arrow) located at the 2nd part of the duodenum

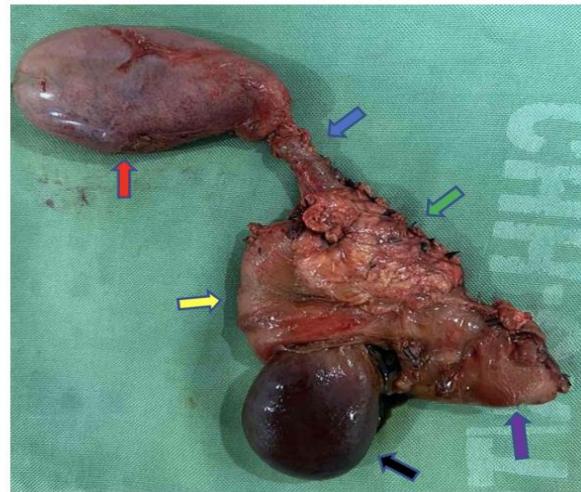


Figure 4. Postoperative specimen containing the gallbladder (red arrow), common bile duct (blue arrow), duodenum (yellow arrow), pancreatic head (green arrow), duodenal cyst (black arrow) and jejunum (purple arrow)

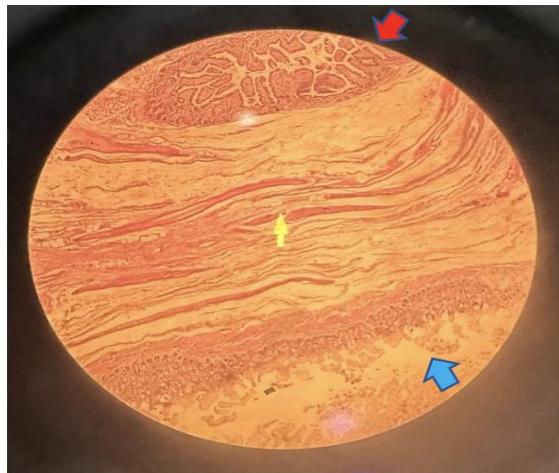


Figure 5. Specimen histology showing the duodenal lumen (red arrow), lamina propria (yellow arrow) and duplication cyst showing intestinal-type mucosa lining (blue arrow)

Case Discussion

Enteric duplication cysts (EDC) are rare congenital malformations formed during the embryonic development of the digestive tract with an incidence of 1 in 4500 births, found in 0.2% of all children, with a slight male predominance.³ The exact etiology of EDCs remains unknown although multiple theories have been postulated including errors of recanalization of the gut or incomplete twinning of the

alimentary tract. These can occur anywhere along the gastrointestinal tract starting from the mouth until the rectum and are most found in the ileum (33%), followed by the esophagus (20%), colon (13%), jejunum (10%), stomach (7%), and duodenum (5%).¹ Signs and symptoms are diverse and are mostly diagnosed in the first 2 years of life in more than 80% of children while it is rarely found in adults with a 5-6% prevalence.² In a

meta-analysis by Chen et al., the most common symptom is abdominal pain and nausea with vomiting in 80% and 42% of cases, respectively. In the same study, the most common complication is pancreatitis seen in 53% of patients while 21% of patients presented cholestasis, hepatitis, and other hepatobiliary involvement. Other unusual manifestations include weight loss (8.5%), gastrointestinal bleeding (6.4%), cyst infection (4.3%), and intussusception (4.3%).⁴

Intussusception is more common among the pediatric population and adult cases only account for <5% of all cases of intussusception and are found in 1% of patients with bowel obstruction.⁵ The most common locations involved in intussusception are at the junctions between mobile and fixed segments of the bowel, such as the freely moving ileum. Duodenojejunal intussusceptions are rarely encountered because of the fixation of a large portion of the duodenum to the retroperitoneum.⁶ In this case, the patient is a 40-year-old female with a duodenal duplication cyst acting as a lead point causing invagination of a segment of the small bowel lumen of the adjacent segment resulting in a duodenojejunal intussusception presenting as abdominal pain, vomiting, and jaundice. In obstructive jaundice, direct bilirubin is typically more elevated however, cases with predominant indirect hyperbilirubinemia may occur. This atypical presentation can be attributed to several mechanisms. In the early phase of biliary obstruction, indirect bilirubin may rise before sufficient direct bilirubin accumulates in the circulation. Concurrent hepatocellular dysfunction, such as ischemic or inflammatory injury, may also impair bilirubin conjugation, resulting in elevated indirect levels. Furthermore, partial or intermittent biliary obstruction may limit the backflow of direct bilirubin, thereby favoring a relative increase in indirect bilirubin.

The cause of intussusception may be intraluminal, mural, or extraluminal. Intraluminal mass can cause intussusception when it is pulled forward by peristalsis and drags the attached bowel wall segment with it. Mural causes occur in focal bowel walls that do not contract normally and is being rotated and kinked by adjacent bowel wall. Lastly, extraluminal factors like adhesions bind to one side of the bowel and cause focal

kinking leading to intussusception.⁶ In this case, the duodenal duplication cyst acts as an intraluminal mass pulled forward by peristalsis, dragging the proximal duodenum and pancreatic head with it.

As opposed to children, which most commonly present with acute symptoms and a triad of crampy abdominal pain, vomiting, and currant-jelly stools, adult intussusception often presents with nonspecific chronic or subacute symptoms, which may be related to the intussusception causing intermittent partial bowel obstruction.⁶ Moreover 90% of childhood intussusception cases are idiopathic but more than 90% of adult intussusception have distinct causes with neoplasm being the most common cause. Other non-idiopathic cases are usually caused by postoperative adhesions, Crohn's disease, infections, intestinal ulcers, and Meckel's diverticulum.⁷

Diagnosis of intussusception in adults is a challenge because of its relative infrequency and the often vague and variable clinical findings at presentation. Even with a complete past medical history, physical examination, and laboratory test, a correct pre-operative diagnosis of intussusception is still difficult and is noted to have been made in only 32-50% of cases.⁷ Useful radio-diagnostic instruments to help in diagnosing intussusception include abdominal X-ray, barium studies, abdominal ultrasound, and abdominal CT. The plain radiograph is usually the first imaging study done in intussusception where an "air crescent" sign of intraluminal air trapped between the walls of the intussusceptum and intussusciens can be seen. It can also be used to assess the degree of bowel obstruction and detect pneumoperitoneum/pneumatosis which is a sign of bowel compromise. The "coiled spring" sign can be seen in barium studies which are produced by contrast within the compressed lumen of the intussusceptum and within the space between the intussusceptum and intussusciens. Intussusception in adults is like those in the pediatric population when seen in an ultrasound. It appears as an outer hypoechoic rim representing the intussusciens and a central area of increased echogenicity representing the intussusceptum giving the classic appearance of a target or doughnut mass. The CT or MRI findings of intussusception are virtually pathognomonic and appear as a mass lesion,

representing a thickened segment of the bowel. Additional use of contrast appears as a rim between the intussusceptum and intussuscipiens similar to the coiled spring appearance seen in barium studies.⁶ In a series reported by Paredes et al., diagnostic studies used to assess 12 cases of adult intussusception included abdominal X-ray, ultrasound, CT, pre-operative colonoscopy, and opaque enema. Findings supported an order of diagnostic accuracy as follows: CT (8 out of 10 patients), followed by ultrasound (6 of 12 patients), followed by opaque enema (2 of 4 tests performed), and colonoscopy (2 of 5 tests performed); no mention was made of the accuracy of abdominal X-ray. Diagnostic accuracy of CT is as high as 58 to 100% in other reports. In our patient, a CT scan with contrast was done which showed telescoping of the duodenum into the proximal jejunum with the presence of a non-enhancing cystic lesion along the leading edge measuring approximately 4.2 x 5.8 cm with invagination of part of the pancreatic head into the intussuscipiens.⁷

In most cases, surgical exploration is the treatment of choice specifically in cases of acute abdomen, intussusception with a mass visible on imaging, and in colonic or ileocolic intussusception. Intussusception found in the colon is usually associated with neoplasm and attempts in reducing it pose a risk for the spread of malignant cells. A more conservative approach or serial clinic and imaging evaluation or attempt for reduction can be done in entero-enteric intussusceptions without lead point mass and short affected segment (< 3.5 cm).⁸ In this patient presenting with an intussusception with a visible mass on a CT scan, a surgical approach was done.

In a meta-analysis by Hong et al., where 464 adult patients had undergone surgery for intussusception, it had a 22.1% pooled rate for postoperative complications. More than half of the reported complications were surgical site infections. Other complications included pulmonary atelectasis, pneumonia, pulmonary thromboembolism, deep vein thrombosis, ileus, gastrointestinal bleeding, acute tubular necrosis, and cardiac arrhythmia.⁹ None of these were observed in our patient postoperatively.

Conclusion

Adult duodenal intussusception is a rare disease that is difficult to diagnose due to its nonspecific symptoms and is possible in cases of duplication cysts which can act as a lead point, such as in our patient. Therefore, a high index of suspicion and imaging plays an important role in the diagnosis of a duplication cyst with intussusception in adults especially those presenting with abdominal pain, vomiting, and jaundice. A correct and timely diagnosis is needed to prevent various complications including bowel infarction and sepsis. This is best managed surgically, as conservative approaches carry a high risk of recurrence and complications. In this case, pancreateoduodenectomy with segmental jejunal resection successfully relieved obstruction, resolved symptoms, and prevented malignant transformation. Surgical intervention remains the definitive treatment, with postoperative recovery and long-term outcomes generally favorable when performed promptly.

Recommendations

1. Collect data to provide the prevalence of duodenal duplication cyst and intussusception in adults in the local setting.
2. Compare the incidence and outcome of surgical and nonsurgical management of intussusception in adults.

Patient Consent Statement

The patient consented to the publication of details about her medical condition and treatment in a case report. Her identity would remain confidential and anonymous. Her participation was completely voluntary.

References

1. Sangüesa Nebot C, Llorens Salvador R, Carazo Palacios E, Picó Aliaga S, Ibañez Pradas V. Enteric duplication cysts in children: varied presentations, varied imaging findings. *Insights Imaging*. 2018;9(6):1097-1106. doi:10.1007/s13244-018-0660-z
2. Gupta A, Chakaravarthi K, Pattnaik B, Kaman L. Duplication cyst of ileum presenting as acute intestinal obstruction in an adult. *BMJ Case Rep.* 2016;2016:bcr2016214775. Published 2016 Oct 6. doi:10.1136/bcr-2016-214775
3. Chen JJ, Lee HC, Yeung CY, Chan WT, Jiang CB, Sheu JC. Meta-analysis: the clinical features of the duodenal duplication cyst. *J Pediatr Surg*. 2010;45(8):1598-1606. doi:10.1016/j.jpedsurg.2010.01.010
4. Anand S, Aleem A. Duplication Cyst. [Updated 2022 Oct 24]. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2023 Jan. Available from: <https://www.ncbi.nlm.nih.gov/books/NBK564347/>
5. Lu T, Chng YM. Adult intussusception. *Perm J*. 2015;19(1):79-81. doi:10.7812/TPP/14-125
6. Ozer A, Sarkut P, Ozturk E, Yilmazlar T. Jejunoduodenal intussusception caused by a solitary polyp in a woman with Peutz-Jeghers syndrome: a case report. *J Med Case Rep*. 2014;8:13. Published 2014 Jan 8. doi:10.1186/1752-1947-8-13
7. Huang BY, Warshauer DM. Adult intussusception: diagnosis and clinical relevance. *Radiol Clin North Am*. 2003;41(6):1137-1151. doi:10.1016/s0033-8389(03)00116-7
8. Marsicovetere P, Ivatury SJ, White B, Holubar SD. Intestinal Intussusception: Etiology, Diagnosis, and Treatment. *Clin Colon Rectal Surg*. 2017;30(1):30-39. doi:10.1055/s-0036-1593429
9. Panzera F, Di Venere B, Rizzi M, et al. Bowel intussusception in adult: Prevalence, diagnostic tools and therapy. *World J Methodol*. 2021;11(3):81-87. Published 2021 May 20. doi:10.5662/wjm.v11.i3.81
10. Hong KD, Kim J, Ji W, Wexner SD. Adult intussusception: a systematic review and meta-analysis. *Tech Coloproctol*. 2019;23(4):315-324. doi:10.1007/s10151-019-01980-5