
Duodenal duplication with accessory papilla causing recurrent pancreatitis in a child: a case report

JOSÉ R. MUÑIZ-ARAGUNDE, MS*; MARÍA S. CORREA-RIVAS, MD†; HUMBERTO LUGO-VICENTE, MD‡

We discuss a 16-year-old male patient who presented with three episodes of recurrent pancreatitis within the last 6 months. Preoperative imaging studies suggested a choledochal cyst within the second portion of the duodenum. Patient was taken to surgery and the lesion was removed. Pathology examination of the cyst

revealed a duodenal duplication. The accessory pancreatic papilla entering the closed duplication cyst was the main cause of the pancreatitis in this child.

Key words: Cyst, Choledochal Cyst, Duodenum, Duplication, Pancreatitis, Malformation

A duodenal duplication cyst (DDC) is an extremely uncommon congenital malformation which in the majority of cases is diagnosed in the first two decades of life. The duodenum is an uncommon site for gastrointestinal duplication, being the ileum and esophagus the most common sites respectively (1). The clinical presentation usually includes obstructive symptoms, bleeding, pain, a palpable mass or distention, but pancreatitis may occur as well. To be classified as a DDC, the wall must be composed of smooth muscle, must be lined with gastrointestinal epithelium and must be adhered to a portion of the gastrointestinal tract (2). A case of a duodenal duplication with accessory pancreatic papilla causing recurrent pancreatitis in a child is presented here. Preoperative studies suggested the diagnosis of an intraduodenal choledochal cyst, also known as choledochocele.

Case Report

A 16-year-old Hispanic boy has been admitted to our hospital three times within the last six months complaining of severe upper abdominal pain and vomiting. Each attack lasted a few days and was accompanied by an elevation of serum amylase and lipase. On his last admission, the

physical examination demonstrated a soft abdomen with tenderness in the upper quadrants without any palpable masses. Chemistry profile revealed an amylase level of 118 U/L (NR 17 - 93 U/L), and a lipase level of 209 U/L (NR 3 - 68 U/L). A cell blood count disclosed a hemoglobin level of 14.9 g/dL (NR 14 - 18 g/dL), a white blood cell count of 7,400 /L (NR 4,000 - 10,000), and a platelet count of 164,000 (NR 140,000 - 400,000). The liver function tests showed a total serum bilirubin level of 0.8 mg/dL (NR 0.2-1.3 mg/dL), an aspartate aminotransferase level of 17 U/L (NR 11 - 35 U/L), alanine aminotransferase, 10 U/L (NR 5 - 54 U/L), and an alkaline phosphatase, 157 U/L (NR 39 - 117 U/L).

Endoscopic retrograde cholangiopancreatography (ERCP) performed after these episodes revealed a choledochal cyst within the second portion of the duodenum. Subsequent magnetic resonance cholangiopancreatography (MRCP) confirmed the diagnosis of the intraduodenal cyst or choledochocele, a rare variant of choledochal cyst classified as type III by Alonso-Lej (3). In view of these findings and recurrent symptoms, patient was taken to surgery.

Surgical Procedure

The abdomen was entered through a midline incision. After Kocker mobilization of the duodenum, a mass was palpable in the second portion. Through the cystic duct a cholangiogram demonstrated normal extrahepatic bile ducts and a filling defect in the duodenum (Figure 1). Lateral longitudinal duodenotomy revealed the intraluminal cystic mass fixed to the medial wall of the duodenum. The

From the *School of Medicine, †Department of Pathology and ‡Department of Surgery, University of Puerto Rico, Medical Sciences Campus.

Address correspondence to: Humberto Lugo-Vicente, MD, P.O. Box 10426, Caparra Heights Station, San Juan PR 00922. <titolugo@coqui.net>



Figure 1. Cholangiogram Demonstrating Normal Extrahepatic Bile Ducts and a Filling Defect in the Duodenum.

main papilla was identified between the wall of the cyst and the duodenum secreting bile (Figure 2). The cyst was opened and clear secretions with pancreatic stones were identified interiorly. Another accessory papilla was identified within the duodenal cyst secreting a clear fluid. We could not cannulate the small accessory papilla and irrigation through the cystic duct failed to produce

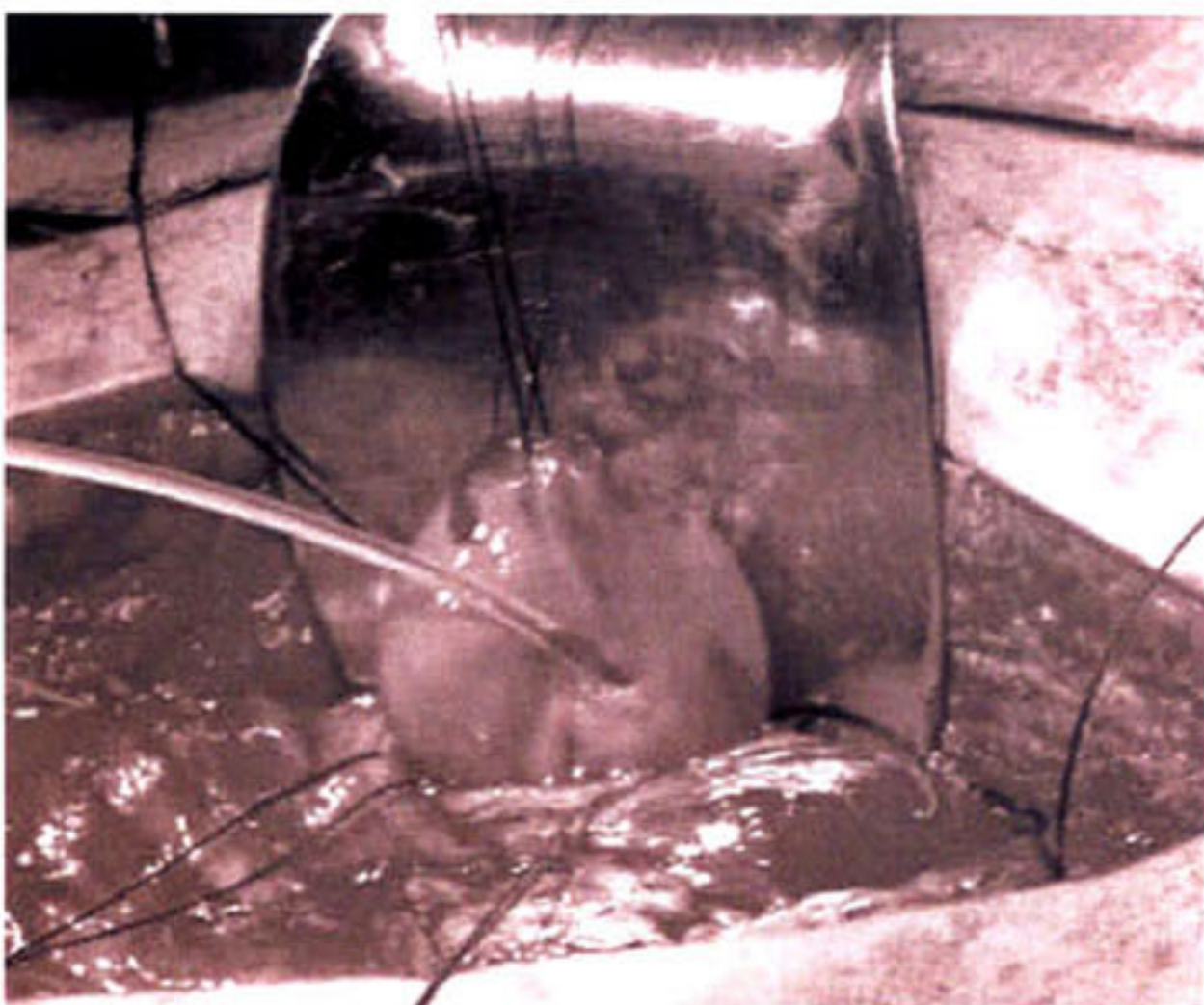


Figure 2. The Main Papilla, Cannulated With a Small Feeding Tube, is in the Medial Wall of the Duodenal Duplication Cyst With Bile Coming Through.

secretions through this papilla. The cyst wall was then removed and marsupialized taking great care not to disrupt the main or accessory papillas. The duodenum was closed transversely and the gallbladder removed. The child left the hospital five days after the procedure without complications. No recurrence of the pancreatitis has been documented during the past twelve months of follow-up.

Pathology findings

Tissue was received fixed in formalin, and processed and stained as routine with hematoxylin and eosin stains. The specimen consisted of a non opened gallbladder and two segments of tan rubbery tissue from the cyst wall. The gallbladder measured 6 x 3 cm and the separate segments 4 x 1.5 x 0.3 cm. Upon opening the gallbladder, it contained abundant green bile. The mucosa was green and velvety with no gross lesions or calculi. The separate segments were sectioned and submitted entirely. On microscopic examination, these segments revealed duodenal mucosa on both sides, and an edematous wall with intervening muscularis mucosa and muscularis

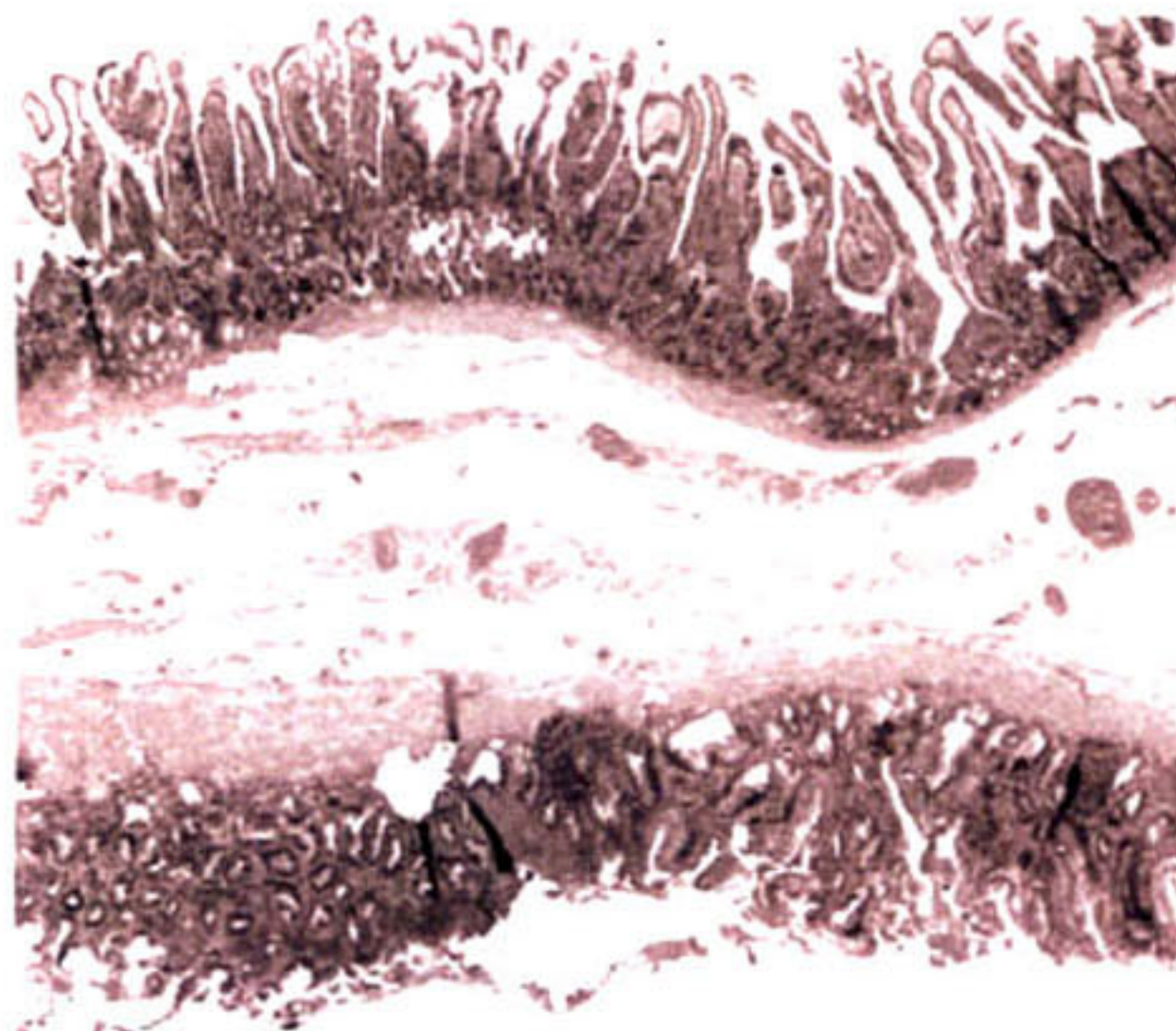


Figure 3. Note Duodenal Mucosa on Both Luminal Sides, Muscularis Mucosa and Muscularis Propria.

propria (Figure 3). Sections of the gallbladder revealed mild chronic and acute inflammation. The diagnosis of duodenal duplication was rendered with a gallbladder with mild acute and chronic infiltrates.

Discussion

Duplication cysts are hollow epithelial-lined, spherical or tubular structures that are attached to some portion of

the gastrointestinal tract, most frequently the distal ileum followed by the esophagus and duodenum (1,4). Duodenal duplication cysts account for only 4% to 12% of all intestinal duplications (2). The epithelial lining of the duplication cyst may be that of the adjacent bowel, and occasionally ectopic gastric or pancreatic tissue, or even respiratory epithelium may sometimes be present within it (4,5). Those cysts containing ectopic gastric mucosa may present with bleeding, ulceration or perforation (4,5). Physical examination and laboratory studies usually reveal no abnormalities. Sonography and computed tomography scan reveal a discrete, fluid-filled structure abutting the medial wall of the descending duodenum (6). These patients usually have symptoms of partial gastric outlet obstruction such as nausea, vomiting, epigastric pain, and abdominal distention (7). However, in this case the patient presented recurrent episodes of pancreatitis in which there was no family history of chronic pancreatitis. The most common causes of acute pancreatitis such as migrating gallstones obstructing the pancreatic duct and massive intake of alcohol are rare to find in pediatric patients.

A duodenal duplication cyst with an accessory papilla was the cause of the recurrent episodes of pancreatitis in our patient since the pancreatic secretions entering the duodenal cyst could not enter the gastrointestinal tract. This created an obstruction to the pancreatic flow, stasis, and the formation of pancreatic stones leading to symptoms of recurrent pancreatitis similar to previously reported cases of duodenal duplication cysts communicating with the pancreatic duct (8,9). The treatment for most duplication cysts is complete surgical excision (10), thus preventing further complications such as bleeding, perforation, and even malignancy arising from ectopic gastric mucosa (11).

In the case presented here, an ERCP and a MRCP were performed, and both studies suggested a choledochoceles type III in the second portion of the duodenum. The patient went to surgery with this pre-operative diagnosis. During surgery, the cyst was resected and sent to pathology to know its histology. Since pathology reported duodenal mucosa on the inside of the cyst, the diagnosis of a choledochal cyst was discarded and the post operative diagnosis of DDC was confirmed.

During removal of a duodenal duplication cyst, care must be taken not to injure the main or accessory papilla. It is recommended that an intraoperative cholangiogram through either the gallbladder or cystic duct be done to delineate the periampullary region before opening the duodenal wall.

Although duodenal duplication cysts have been treated by surgical procedures almost always, there are cases reported where this anatomical abnormality has been

managed by endoscopic resection (12). It has been shown that by using the O-ring ligation kit, the cyst can be resected making the DDC symptoms disappear, suggesting another useful way to manage this condition (13).

Resumen

Discutimos un paciente masculino de 16 años quien presentó tres episodios de pancreatitis recurrente en un período de 6 meses. Las imágenes radiográficas preoperatorias sugerían un quiste coledocal dentro de la segunda porción del duodeno. El paciente se llevó a cirugía y la lesión removida. La patología de este quiste mostró ser una duplicación duodenal. La papila pancreática accesoria que entraba a la duplicación duodenal fue la causa principal de los episodios de pancreatitis en este joven.

References

1. Taft DA, Hairston JT. Duplication of the alimentary tract. *Am Surg*. 1976; 42(7): 455-462.
2. Jo YC, Joo KR, Kim do H, Park JH, Suh JH, Kim YM, Nam CW, et al. Duodenal duplicated cyst manifested by acute pancreatitis and obstructive jaundice in an elderly man. *J Korean Med Sci* 2004; 19(4): 6004-07.
3. Alonso-lej F, Rever W, Passagno D. Congenital choledochal cyst, with a report of 2 and analysis of 94 cases. *Int Abstr Surg* 1959; 108: 1-30.
4. Gross RE, Holcomb GW, Farber S. Duplication of the alimentary tract. *Pediatrics* 1952; 9(4): 449-468.
5. Rubin RB, Saltzman JR, Zawacki JK, Khan A, Swanson R. Duodenal duplication cyst with massive gastrointestinal bleeding. *J Clin Gastroenterol* 1995; 21: 72-74.
6. Omojola MF, Hood IC, Stevenson GW. Calcified gastric duplication. *Gastrointest Radiol* 1980;15(3): 235-238.
7. Laraja RD, Rothenberg RE, Chapman J, et al. Foregut duplication cyst: a report of a case. *Am Surg* 1995; 61(9): 840-841.
8. Niehues R, Dietl KH, Bettendorf O, Domschke W, Pohle T. Duodenal duplication cyst mimicking pancreatic cyst in a patient with pancreatitis. *Gastrointestinal Endoscopy* 2005;62(1): 190-192.
9. Ogura Y, Kawarada Y, Mizumoto R. Duodenal duplication cyst communicating with an accessory pancreatic duct of Santorini. *Hepatogastroenterology* 1998; 45(23): 1613-1618.
10. Holcomb GW 3rd, Gheissari A, O'Neill JA Jr, Shorter NA, Bishop HC. Surgical management of alimentary tract duplications. *Ann Surg* 1989; 209(2):167-174.
11. Falk GL, Young CJ, Parer J. Adenocarcinoma arising in a duodenal cyst: a case report. *Aust N Z J Surg* 1991; 61(7): 551-553.
12. Sezgin O, Altiparmak E, Yilmaz U, Saritas U, Sahin B. Endoscopic management of a duodenal duplication cyst associated with biliary obstruction in an adult. *J Clin Gastroenterol* 2001; 32(4): 353-355.
13. Wada S, Higashizawa T, Tamada K, Tomiyama T, Ohashi A, Satoh Y, Sugano K, Nagai H. Endoscopic partial resection of a duodenal duplication cyst. *Endoscopy* 2001; 33(9): 808-810.