



Spontaneous Pancreatic Pseudocyst-Superior Mesenteric Vein Fistula

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ABSTRACT

Pancreatic pseudocyst formation is a common complication of chronic pancreatitis. Rarely, a fistula develops between the pseudocyst and the portal venous system. We present a case of a 50-year-old man who was found to have a pancreatic pseudocyst-superior mesenteric vein fistula after being evaluated for several months of abdominal pain and weight loss. The patient was treated with endoscopic stenting of the pancreatic duct along with early enteral nutrition and suppressive antibiotics, which resulted in improvement in his condition. This case report highlights clinical presentation and the complexity of treatment of this rare diagnosis.

KEYWORDS: pancreatic pseudocyst; chronic pancreatitis; pancreatic pseudocyst-portal venous fistula

INTRODUCTION

While pancreatic pseudocysts frequently complicate acute and chronic pancreatitis, the development of a fistulous connection between pancreatic pseudocysts and the portal venous system is very uncommon.^{1,2} The paucity of reported cases of pancreatic pseudocyst-portal venous (PP-PV) system fistulas limits the ability to evaluate the efficacy of diagnostic tools and treatments of this highly morbid condition. In this report, we describe a pancreatic pseudocyst-superior mesenteric vein (PP-SMV) fistula to highlight diagnostic and therapeutic considerations and challenges.

CASE REPORT

A 50-year-old man with alcohol use disorder presented with 6 months of fatigue, 15-pound weight loss, and abdominal pain. Initial history and workup were consistent with alcohol-induced pancreatitis. Contrast-enhanced computed tomography (CT) demonstrated diffuse portal venous thrombosis and a pancreatic pseudocyst with concern for a possible fistulous connection to the portal venous system (Figure 1). Magnetic resonance cholangiopancreatography (MRCP) demonstrated a 2.1 cm in diameter area of walled-off necrosis within the pancreatic head, which communicated with an 8.5 × 3.1 × 2.6 cm retroperitoneal pseudocyst (Figure 2). The pseudocyst also communicated through fistulous connections with the SMV and left portal vein (Figure 3). Endoscopic retrograde cholangiopancreatography (ERCP) with ventral pancreatic injection resulted in extravasation near the genu (Figure 4). Sphincterotomy was performed, and a 5 French × 9 cm stent was placed. The patient experienced relief in his abdominal pain after the ERCP. Anticoagulation for his extensive thromboses was deferred because of the high risk of bleeding in the presence of the fistula. He could not tolerate adequate oral nutrition, so nasojejunal feeds were started early in his course.

The post-ERCP course of the patient was complicated by septic shock secondary to *Clostridium perfringens* and *Haemophilus parainfluenzae* bacteremia. He improved with antibiotics. His course was further complicated by recurrent secondary bacterial ascites that also resolved with antibiotics and bilateral pleural effusions that briefly required chest tube placement. The case was discussed with interventional radiology, but further interventions were not performed because it was believed that the bleeding risk of any procedure, such as cystogastrostomy, was too high and he was improving without intervention. He was discharged to a rehabilitation facility with stable interval abdominal imaging on suppressive oral antibiotics. The patient had 2 repeat admissions

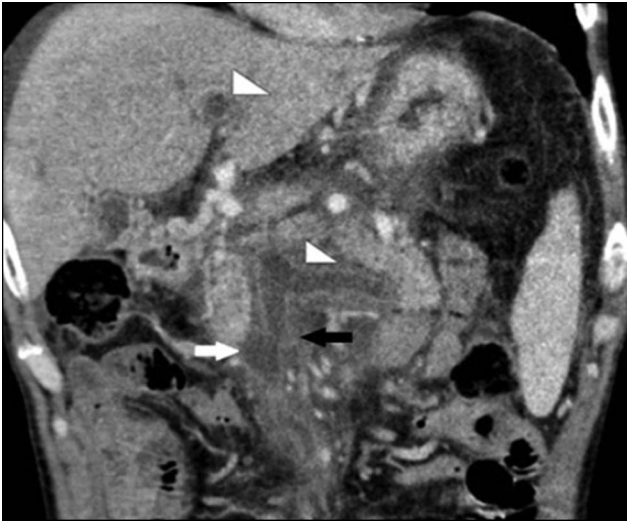


Figure 1. Coronal view of a contrast-enhanced CT scan demonstrating the inferior component of the pseudocyst (white arrow) fistulizing a branch of the SMV near the pancreatic head (black arrow). White wedges indicate the proximal SMV and left portal vein. CT, computed tomography; SMV, superior mesenteric vein.

over the next few weeks because of complications, including secondary bacterial peritonitis, *Clostridium difficile* colitis, and intra-abdominal abscess. After he was discharged from the second readmission, the patient underwent an endoscopic ultrasound to assess for interval changes and potential drainage sites. The pseudocyst was considerably improved in size and no longer communicated with the pancreatic duct (Figure 5). The stent was removed, but the fluid collection was not accessible from the gastrointestinal lumen for drainage.

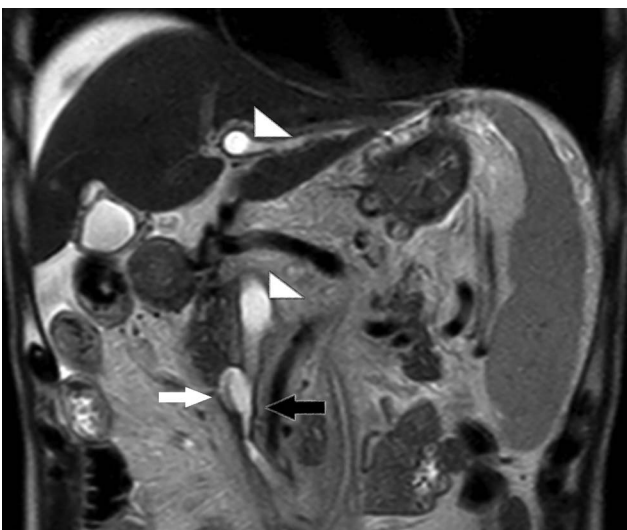


Figure 2. T2-weighted MR image demonstrating the inferior component of the pseudocyst (white arrow) fistulizing a branch of the SMV near the pancreatic head (black arrow). Fluid density and correlating MR T2 bright fluid signal noted in the partially visualized proximal SMV and left portal vein (white wedges). MR, magnetic resonance; SMV, superior mesenteric vein.



Figure 3. MRCP MIP reconstructions are used to emphasize fluid in a 3D image. The large pseudocyst is noted (white arrows) with a cystic component fistulizing a branch of the SMV (black arrow). Extension of the pseudocyst conforms to the extrahepatic and intrahepatic portal venous systems (white wedges). MRCP, magnetic resonance cholangiopancreatography; SMV, superior mesenteric vein.

DISCUSSION

Pancreatic pseudocysts are common complications of chronic pancreatitis, occurring in 20%–40% of cases.^{1,2} Fistula development between pancreatic pseudocysts and the portal venous system is exceedingly rare.³ This complication has been described more frequently in men and those with significant

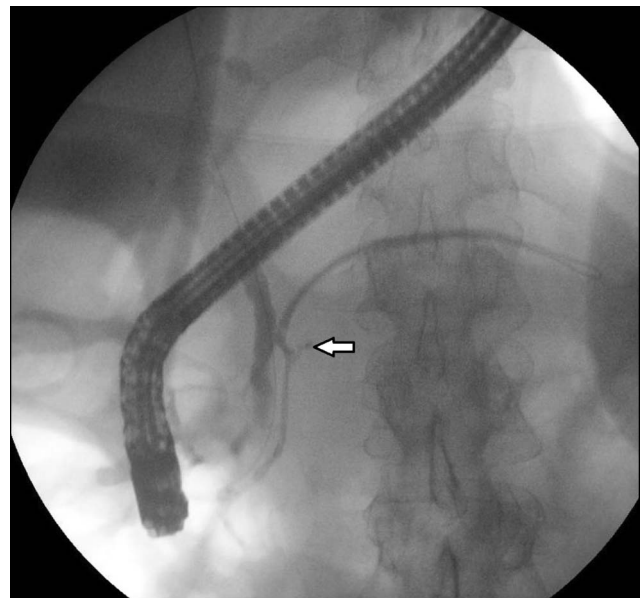


Figure 4. ERCP fluoroscopic image demonstrating cannulation of the pancreatic duct and extravasation of contrast near the genu of the pancreas (white arrow). ERCP, endoscopic retrograde cholangiopancreatography.



Figure 5. EUS of an anechoic peripancreatic lesion without internal debris consistent with the continued presence of a pancreatic pseudocyst. EUS, endoscopic ultrasound.

alcohol use.^{1,4-6} Most cases develop in the context of chronic pancreatitis and less commonly during episodes of acute pancreatitis.^{4,7} The exact mechanism remains disputed. One possible explanation is that the thrombus develops first because of stasis from compression of the vein by the pseudocyst and the thrombus then acts as a nidus for the fistula to develop.^{1,3-6} It is also possible that the fistula develops first because of pancreatic enzymes destroying the pseudocyst and vein walls and inflammation from the pancreatic enzymes within the vein leads to thrombosis.^{1,3,5-8} Several cases that involved serial imaging documented the presence of fistula before thrombosis, suggesting that at least in some cases, the second theory seems more likely.^{4,8}

CT is a common initial imaging modality for patients with PP-PV fistulas. A review of 17 PP-PV fistula cases found that 14 of 17 patients were initially imaged with CT, which was diagnostic in only 1 of 14 cases. The remaining cases were diagnosed with MRI, percutaneous transhepatic portography, surgery, and autopsy.⁶ MRCP seems to be more sensitive than CT with one review study finding MRCP to be diagnostic in 4 of 4 patients when it was the initial imaging study. That same review evaluated 4 cases in which the initial imaging modality was CT and found it was diagnostic in 0 of 4 cases with three-fourths of cases diagnosed on subsequent MRCP and one-fourth diagnosed based on a repeat CT scan.⁹

Treatment options include conservative management, endoscopic treatment, and surgical resection. Conservative management may be appropriate in mild or asymptomatic cases because spontaneous resolution has been documented in these cases.^{3,10} Surgery has typically been reserved for severe presentations. A review of 13 surgically managed cases found that 6 of 13 patients died of postoperative complications, suggesting that surgery is high-risk.¹⁰ Endoscopic stenting of the pancreatic duct has been effective in certain cases and resulted in clinical improvement in the case described in this report.^{3,11,12}

A treatment algorithm proposed by Ng et al⁸ suggested conservative therapy with percutaneous drainage in cases in which the pseudocyst does not communicate with the pancreatic duct and endoscopic stenting in cases in which communication is present. Given the high rates of mortality in surgically managed cases, we agree with this approach for patients who can tolerate endoscopy. Optimal duration of stenting has not been investigated, but in this case, the stent was removed after 4 months when significant improvement in fluid collection was noted. Other aspects of treatment that warrant consideration include anticoagulation, nutritional support, and prophylactic antibiotics. We favor avoidance of therapeutic anticoagulation for associated thrombi because of bleeding risk, and we advocate for early enteral nutrition in individuals not tolerating oral intake.^{1,5} Infectious complications have been described in numerous PP-PV cases, and we suggest the use of prophylactic antibiotics as part of initial management.

In summary, fistula development between a pancreatic pseudocyst and the portal venous system is rare and life-threatening.^{4,5,7} Treatment options include conservative management, endoscopic stenting, or surgery. We presented a case in which a patient clinically improved with endoscopic stenting of the pancreatic duct and aggressive treatment of complications, including malnutrition and infection.

DISCLOSURES

Author contributions: S. Goble conducted chart review and literature review and drafted the initial manuscript. A. Ayoub provided content editing for the final manuscript. S. Boeke provided the figures along with interpretation of the figures. R. Matlock provided care for the patient on the consultant team and was available for consultation on manuscript development.

A. Malli provided care for the patient on the consultant team, reviewed the manuscript, supervised the project, and is the article guarantor.

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Informed Consent was obtained for this case report.

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