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Case report

Massive pancreatic pseudocyst with portal vein fistula: Case report and proposed treatment algorithm

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ABSTRACT

Pancreatic pseudocyst is a relatively common occurrence resulting from acute or chronic pancreatitis. However, a rare subset of these patients present with a pseudocyst fistulizing into the portal vein. We present the case of a 58 year-old woman with a rapidly expanding pancreatic pseudocyst with portal venous fistulization causing portal vein thrombosis, in addition to biliary and duodenal obstruction. The patient underwent surgical decompression with a cyst-gastrostomy and was well until one week post-operatively when she experienced massive gastrointestinal hemorrhage leading to her death. A review of the literature is presented and a treatment algorithm to manage patients with pancreatic pseudocyst to portal vein fistula is proposed.

Introduction

Pancreatic pseudocyst is a complication of acute and chronic pancreatitis. In rare cases, patients with pancreatic pseudocysts present with fistulae into the portal vein. Only 18 cases of this condition have been reported in the English literature to date [1–18]. The clinical presentation of pancreatic pseudocyst-portal vein fistula (PP-PV) varies, ranging from symptomatic abdominal pain [3,4,6,16,17] to portal hypertension due to thrombosis [8,10,11,14], systemic lipolysis [1,2,5,7,12], septic shock [9], and death [1,2,5,9]. We present a patient with a rapidly enlarging pancreatic pseudocyst with portal vein fistula causing biliary and duodenal obstruction who underwent cyst-gastrostomy. In addition to a review of the literature, a treatment algorithm is proposed.

Case report

The patient was a 58 year-old woman with a history of pancreatic divisum and hypothyroidism, who had undergone laparoscopic cholecystectomy for gallstone pancreatitis four years prior. She was subsequently admitted to an outside hospital for episodes of recurrent acute pancreatitis. Computed tomography (CT) and magnetic resonance imaging (MRI) revealed a new 6.7 × 4.2 × 8.2 cm pancreatic pseudocyst (Fig. 1). No surgical intervention was performed at the time because the cyst wall was deemed immature. The patient improved with conservative management and was subsequently discharged home. However, she returned to the outside hospital with recurrent abdominal pain, nausea and vomiting and imaging revealed that the pseudocyst had enlarged to 7.1 × 4.4 × 11.2 cm.

On transfer to our institution, physical exam was notable for mild jaundice and a tender, palpable mass in the mid-epigastric region. Laboratory test results demonstrated elevated alkaline phosphatase (AlkP: 997, normal values: 38–126 U/L), total bilirubin (T.bili: 5.2, normal values: 0.2–1.3 mg/dL), transaminases (AST/ALT: 133/120, normal values: 5–35/7–56 U/L), amylase (594, normal values: 30–110 U/L) and lipase (672, normal values: 7–60 U/L). MRI/magnetic resonance cholangiopancreatography (MRCP) of the abdomen revealed three pseudocysts with dimensions of 10 × 10 × 16 cm, 5.5 × 4.5 × 3.8 cm and 2.8 × 5.5 cm (Fig. 1). Although the pancreatic duct was seen in the tail of the pancreas, the head and body were obscured by the cysts. Subsequent volume

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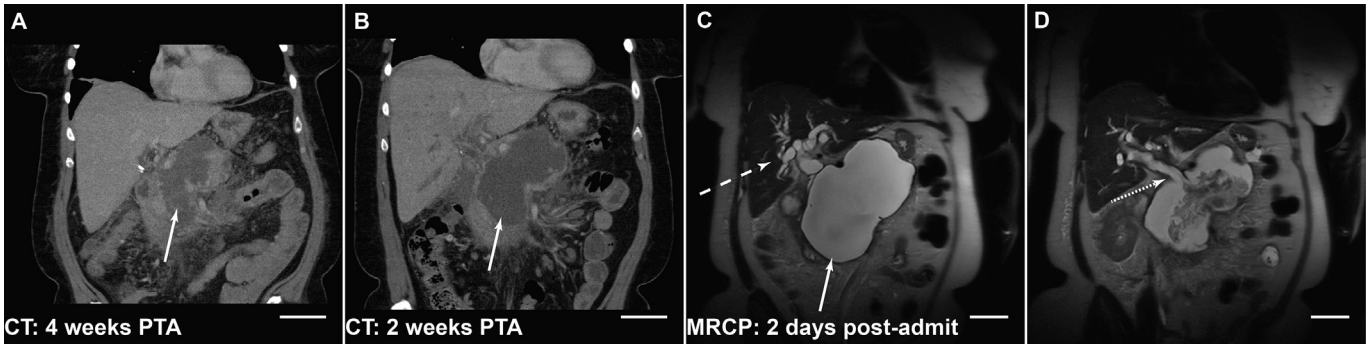

Fig. 1. Time evolution of the pancreatic pseudocyst. A) Computed tomography (CT) of the abdomen 4 weeks prior to current admission showed the pseudocyst in the epigastric region (white arrow). B) CT of the abdomen two weeks prior to current admission showed continued growth of the pseudocyst (white arrow). C) Magnetic resonance cholangiopancreatography (MRCP) showed continued enlargement of the pseudocyst (white arrow). The mass effect of the cyst has also caused biliary dilatation (dash arrow). D) MCRP at a posterior location compared to the slice shown in C). Connection of the cyst with the portal vein can be seen (dotted arrow). Debris found on exploration to be clotted blood is also seen inside the pseudocyst. (Scale bars = 5 cm).

rendering of the images revealed that these cysts were in communication and formed a single cyst (Fig. 2). Direct communication between the main portal vein and pseudocyst was observed with similar signal intensity of the pseudocyst fluid and that within the thrombosed portal vein. Doppler ultrasound (DUS) demonstrated no blood flow in the portal vein (Fig. 3). Both intrahepatic and extrahepatic bile ducts were dilated to the level of the largest pseudocyst. An endoscopic retrograde cholangiopancreatography (ERCP) was performed, confirming external compression and obstruction. Distortion and narrowing of the duodenal lumen due to the pseudocyst prevented stent insertion during the ERCP procedure. An open cyst-gastrostomy was planned to relieve the external compression of both the biliary system and the duodenum.

Prior to surgery, worsening abdominal pain, rising white blood cell count and progressive jaundice were concerning for development of cholangitis. She underwent percutaneous transhepatic biliary catheter (PTC) drainage, with immediate improvement of her obstructive symptoms.

An open cyst-gastrostomy was performed. Brownish, turbid fluid was aspirated from the cyst and large amounts of old blood clot and necrotic debris were evacuated. Consistent with MRI and ultrasound findings, no evidence of active bleeding from the pseudocyst or compromise of the pseudocyst wall was noted. No attempt was made at a pancreatic resection due to the significant portal hypertension, inflammation, and mass effect of the giant pseudocyst.

Postoperatively, the patient's obstructive symptoms improved and her hemoglobin remained stable. Follow-up cholangiography via the PTC showed decreased mass effect of the pseudocyst and resolving biliary dilatation.

On postoperative day seven, the patient presented with sudden onset of hematochezia, hematemesis and hemodynamic instability resulting in immediate transfer to the intensive care unit. The patient was intubated for airway protection and an emergent esophagogastroduodenoscopy was performed which showed bright red blood in the stomach and blood clots and fibrotic material at the site of the cyst-gastrostomy. No active bleeding was identified in the stomach or duodenum. The patient required massive blood transfusion. Upon arrival to the interventional radiology suite for diagnosis and possible embolization, the patient experienced refractory hypotension and cardiac arrest. Despite emergency resuscitative efforts, the patient expired.

Post-mortem examination confirmed chronic pancreatitis with presence of a large pseudocyst. The portal vein had perforated into the pseudocyst, with over a liter of serosanguineous fluid and fresh blood clots present in the cyst, gastric lumen and bowel.

Discussion

Fistulization into the portal vein is a rare sequela of pancreatic pseudocyst formation. Eighteen cases of PP-PV have been reported to date in the literature [1–18] and are summarized in Table 1. While invasive procedures such as operative exploration [7,12], percutaneous [4] or endoscopic pancreatography [6,10,11,14,16–18] and angiography [3,4,8,10,16,18] have been used to diagnose this condition in the past, non-invasive imaging using CT, MRI or MRCP has recently been shown to be effective for diagnosis [13,15], and clearly demonstrated PP-PV in our patient. Although three dimensional rendering was necessary to demonstrate that the multiple pseudocysts initially seen on MRI were in fact connected and forming a single cyst. In the future, visually accurate [19,20] volume rendering may be helpful to both radiologists and surgeons for diagnosis and pre-operative planning.

The mechanism for fistulization remains poorly understood. However, it is believed that the high concentrations of pancreatic enzymes within the pseudocyst erode into adjacent structures [8]. Though fistulization has been hypothesized to occur after portal vein thrombosis (PVT) [8], several reports demonstrate pseudocyst fistulization in the absence of PVT [1,5,7,12]. Portal vein thrombosis may result from mass effect and compression by the pseudocyst along with associated peri-pseudocyst inflammation [21]. Alternatively, fistulization and release of digestive enzymes into a patent portal vein may directly cause intravascular thrombosis [15].

Serious complications of PP-PV noted in the literature include septic shock [9], systemic inflammation [15], lipolysis [1,2,5,7,12], hemorrhage [8], and death [1,2,5,9]. In particular, the presence of systemic lipolysis is associated with a high mortality rate. Clinically, patients present with arthralgia and purpuric nodules on the extremities. Biopsy of these nodules demonstrates subcutaneous lipolytic necrosis. Three of the five patients with systemic lipolysis documented by pathology died leading to an associated mortality rate of 60% [1,2,5]. In contrast, one mortality [9] was documented in the remaining 13 patients, for a mortality rate of 7.7%.

The cause of the systemic lipolysis is unclear [2,22]. It is hypothesized that the massive release of pancreatic enzymes into the systemic circulation through the fistula may be responsible. Systemic lipolysis only occurred in PP-PV patients with portal vein patency. This makes pathophysiologic sense as we hypothesize that systemic lipolysis requires acute decompression and rapid release of large amounts of pancreatic enzymes into the portal circulation.

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