

Pancreatic Pseudocyst-Portal Vein Fistula: Serial Imaging and Clinical Follow-up from Pseudocyst to Fistula

췌장의 가성낭종-문맥간 누공: 가성낭종에서 누공 형성까지의 영상 및 임상 소견의 연속적인 과정

Keum Nahn Jee, MD

Department of Radiology, Dankook University Hospital, Dankook University College of Medicine, Cheonan, Korea

Pancreatic pseudocyst-portal vein fistula is an extremely rare complication of pancreatitis. Only 18 such cases have been previously reported in the medical literature. However, a serial process from pancreatic pseudocyst to fistula formation has not been described. The serial clinical and radiological findings in a 52-year-old chronic alcoholic male patient with fistula between pancreatic pseudocyst and main portal vein are presented.

Index terms

Alcoholic Pancreatitis
 Pancreatic Pseudocyst
 Portal Vein
 Fistula

INTRODUCTION

The pancreatic pseudocyst is a very common complication of acute or chronic pancreatitis, usually by the latter and can also be caused by pancreatic trauma or surgery (1-3). The incidence of pseudocysts in chronic pancreatitis is 20-40% (1). Increased incidence of complications correlates directly with the duration of the pseudocyst and a 12% mortality rate was reported in untreated complicated pancreatic pseudocysts of more than 13 weeks (4). Communication of pancreatic pseudocysts with the pancreatic duct can be identified in 40-69% and pseudocysts containing active enzymes can cause potentially lethal complications (1, 4). Complications related to pancreatic pseudocysts include infection, hemorrhage, intraperitoneal rupture, fistula to adjacent organs, disseminated fat necrosis, arthritis, medullary bone necrosis, and vascular complications such as pseudoaneurysm, splenic vein obstruction, portal vein thrombosis or hypertension (1-5).

Received February 10, 2015; Accepted February 24, 2015

Corresponding author: Keum Nahn Jee, MD
 Department of Radiology, Dankook University Hospital,
 Dankook University College of Medicine,
 201 Manghyang-ro, Dongnam-gu, Cheonan 330-715,
 Korea.
 Tel. 82-41-550-6921 Fax. 82-41-552-9674
 E-mail: jkn1303@dreamwiz.com

This is an Open Access article distributed under the terms of the Creative Commons Attribution Non-Commercial License (<http://creativecommons.org/licenses/by-nc/3.0>) which permits unrestricted non-commercial use, distribution, and reproduction in any medium, provided the original work is properly cited.

A pancreatic fistula is an unexpected communication between the pancreas and the surrounding organs and can occur primarily as a result of trauma, pancreatic surgery and chronic pancreatitis. Pancreatic fistula usually associates with pancreatic pseudocyst and mostly communicates with skin and can also be communicated with other internal organs or spaces (2-5).

Pancreatic-portal vein fistula is exceedingly rare and has been previously reported only 18 cases in medical literature written in English. And no reports, to date, have described the serial process of pancreatic pseudocyst-portal vein fistula. A case of pancreatic pseudocyst-portal vein fistula through clinical and radiological evaluation is presented.

CASE REPORT

A 52-year-old man with previous history of recurrent alcoholic pancreatitis for three years was admitted to our hospital for recently developed severe epigastric pain. Contrast-enhanced ab-

dominal CT scan after symptom improvement showed a small cystic lesion in the pancreatic head and a few small fluid collections at pancreaticoduodenal groove and periduodenal area suggesting pseudocysts (Fig. 1A).

The follow-up CT imaging in eight months after the initial CT scan showed increased size of the pseudocyst in the pancreatic head with containing slight high-attenuating debris or blood clots and compression of adjacent main portal vein with short segmental thrombus of the proximal superior mesenteric vein (SMV) (Fig. 1B). He had the clinical symptoms and laboratory findings exacerbation of chronic pancreatitis. At that time, endoscopic retrograde pancreatography (ERP) did not reveal a leakage of pancreatic juice.

The third abdominal CT scan, after two months from the sec-

ond CT scan, showed that the pseudocyst in the pancreatic head much decreased in size measuring less than 1 cm, and that mesenteric venous thrombosis remained and expanded into the main portal vein (Fig. 1C) and intrahepatic portal vein in the right posterior segment. At the time he had completely normalized laboratory findings including prothrombin time and activated partial thromboplastin time and fully recovered from symptoms of pancreatitis.

In nine months after the third CT scan, he had the fourth a CT scan due to postprandial epigastric pain for five days. The CT images showed increased size of the pseudocyst in the head of pancreas abutting into the dilated main portal vein filled with low-attenuating thrombus and periportal collateral vessels (Fig. 1D). His serum amylase and lipase levels were 1441 U/L (refer-

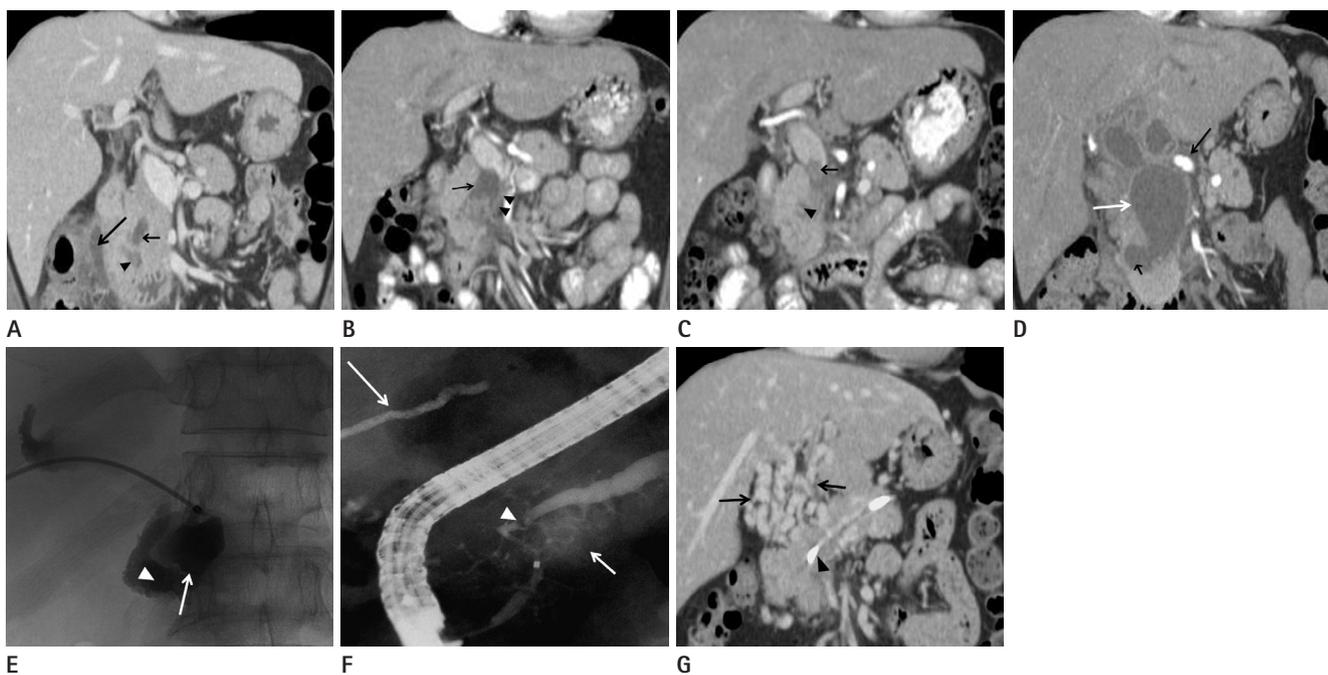


Fig. 1. Pancreatic pseudocyst-portal vein fistula in a 52-year-old chronic alcoholic male patient.

A. Contrast-enhanced CT scan with coronal reformation shows a small cystic lesion (short arrow) in the pancreatic head and a few small fluid collections at pancreaticoduodenal groove (arrowhead) and periduodenal area (long arrow).

B. Contrast-enhanced CT scan with coronal reformation shows increased size of the pseudocyst (arrow) in the pancreatic head with high attenuating debris or blood clot on dependent portion, compression of adjacent portal vein by the cystic lesion and short segmental thrombus in the proximal superior mesenteric vein (arrowheads).

C. Contrast-enhanced abdominal CT scan with coronal reformation shows a very small pseudocyst (arrowhead) in the head of pancreas and thrombus expansion into the distal main portal vein (arrow).

D. Contrast-enhanced CT scan with coronal reformation shows low attenuating pseudocyst (short black arrow) in the pancreatic head adjacent to the portal vein (white arrow) and surrounding periportal collateral vessels (long black arrow).

E. Percutaneous transhepatic portography shows contrast filled dilated main portal vein (arrow) and extravasation of contrast material into the pseudocyst (arrowhead).

F. Endoscopic retrograde pancreatography shows dilatation of main pancreatic duct and a stricture (arrowhead) communicating with contrast filled pseudocyst (arrow). Glue embolization state of right portal vein is shown at superior side (long arrow).

G. Contrast-enhanced CT scan with coronal reformation shows extensive periportal collateral vessels (arrows), obliteration of main portal vein, disappeared pseudocyst in the pancreatic head and stent (arrowhead) in main pancreatic duct.

ence level 28–100) and 3422 U/L (reference level 13–60), respectively. But he had normal range of white blood cell count, C-reactive protein and various serum thrombogenic factors including protein C, protein S, antiphospholipid antibodies, anticardiolipin antibodies, and antithrombin III. The unusual appearance of imaging finding and discrepancy between imaging and clinical findings prompted further investigation of percutaneous transhepatic portography (PTP) the next day after the CT scan. Aspiration with removal of thrombus from the main portal vein and drainage of fluid were done through the right portal vein. The communication between the portal vein and the pseudocyst was confirmed by visualization of the pseudocyst after contrast injection into the portal vein (Fig. 1E). Dark brownish fluid from the portal vein revealed high amylase and lipase levels. Glue embolization for the right portal vein was done. Four days after the PTP, he underwent ERP and it showed dilatation of the main pancreatic duct and stricture of the duct communicating with the pseudocyst (Fig. 1F). The patient underwent endoscopic transpapillary pancreatic stent insertion. During the time, he was treated conservatively for relapsed chronic pancreatitis by fasting, fluid replacement, parenteral nutrition, and intermittent analgesics.

The contrast-enhanced CT scan in six months after the procedures showed complete resolution of the pseudocyst, extensive periportal collateral vessels and obliteration of main portal vein (Fig. 1G) and then the stent was removed. For 62 months after the removal of the stent, he has been doing well without any clinical evidence of recurrent pancreatitis.

DISCUSSION

An unusual complication of pancreas-portal vein fistula has been reported only 18 cases in medical literature written in English. They mentioned autopsy findings, diagnosis, treatment and review of the literature (2, 3, 6-10). But none of them described or demonstrated the serial process of the fistula formation.

The precise pathogenesis of pancreatic pseudocyst-portal vein fistula is still controversial. The proposed mechanism of pancreas-portal vein fistula involves uninhibited pancreatic enzymes within a pseudocyst causing erosion of the wall of the portal vein. The erosion of venous wall incites venous thrombosis and subsequent filling of the portal vein with pancreatic secretions

from the pseudocyst and break down of the thrombus (2). And these serial processes usually occur rapidly and might cause severe clinical manifestations (2, 6, 7).

However, in the presented case, the initial thrombosis of SMV was caused by compression of the main portal vein by the pseudocyst in pancreatic head and ERP did not show the leakage of pancreatic juice at the time. And mesenteric venous bland thrombosis progressed into portal vein even after decreased size of the pseudocyst in the pancreatic head without evidence of relapsed pancreatitis. Eleven months have taken from the initial appearance of mesenteric venous thrombosis to the fistula formation. During the period between the mesenteric venous thrombosis and the pseudocyst-portal vein fistula, there must have been an occurrence of complex fistulous connection of the pancreatic duct, the pseudocyst and the portal vein by exacerbation of chronic pancreatitis. Normal laboratory findings except hyperamylasemia and hyperlipasemia at the time of diagnosis of pseudocyst-portal vein fistula. Such laboratory findings could be explained by pancreatic pseudocyst-portal venous fistula itself without relapsed chronic pancreatitis.

In the presented case, the two processes of pancreatic duct-pseudocyst fistula and pseudocyst-portal vein fistula seemed to occur gradually. And it could be speculated that already existed portal vein bland thrombosis played a role for gradual progression of the process of the fistula formation.

In the review of the literature, clinical manifestations varied from mild symptoms managed by conservatively (3, 8) to very severe clinical manifestations needed surgery or with death (2, 6, 7, 10) and it is unclear why various severity of the disease state is presented in each patient.

The pancreas-portal vein fistula usually suggests fistula between a pancreatic pseudocyst and portal vein, under the prerequisite of active pancreatic enzymes in the pseudocyst or fistulous tract between the pancreatic duct and a pseudocyst (2, 3, 6, 8, 10). But direct fistula between the pancreatic duct and portal vein (7) and pancreatic ascites from disruption of the main pancreatic duct without pseudocysts (5, 9) were reported.

The definitive diagnoses could be made by surgery, autopsy, or various imaging of CT, MR imaging, endoscopic retrograde cholangiopancreatography (ERCP) and PTP according to the previous reports. And two most useful imaging modalities are ERCP and PTP. ERCP does provide a dynamic view of pancre-

atic ductal system and depict the fistula. PTP can also show pseudocyst-portal vein fistula (2, 3, 8, 10). CT and MR cholangiopancreatography were rarely helpful for the definitive diagnosis and usually gave the supportive imaging findings of the fistula (2, 3, 6-10). The treatment of pseudocyst-portal vein fistula usually depends on the patient's clinical symptoms. A patient with asymptomatic or mild symptomatic can be treated conservatively. A patient with failure of conservative management or leakage of pancreatic juice associated with disruption of the main pancreatic duct on ERCP can be treated by endoscopic pancreatic stent insertion. And a patient with signs of sepsis or disseminated fat necrosis is usually surgically treated by pancreaticoenterostomy and pancreatectomy (2, 3, 6-10). Portal vein embolization as a treatment option for pseudocyst-portal vein fistula has not been reported until now.

Successful treatment by glue embolization of portal vein and pancreatic stent insertion for pseudocyst-pancreatic fistula were done without any complications in this case.

This is a case report about the exceedingly rare pancreatic pseudocyst-portal vein fistula, pointing out the demonstration of serial process from pseudocyst to fistula formation based on the clinical, radiological and pathophysiological correlations.

REFERENCES

1. Aghdassi A, Mayerle J, Kraft M, Sielenkämper AW, Heidecke CD, Lerch MM. Diagnosis and treatment of pancreatic pseudocysts in chronic pancreatitis. *Pancreas* 2008;36:105-112
2. Hammar AM, Sand J, Lumio J, Hirn M, Honkonen S, Tuominen L, et al. Pancreatic pseudocystportal vein fistula manifests as residivating oligoarthritis, subcutaneous, bursal and osseal necrosis: a case report and review of literature. *Hepatology* 2002;49:273-278
3. Yamamoto T, Hayakawa K, Kawakami S, Nishimura K, Katsuma Y, Hayashi N, et al. Rupture of a pancreatic pseudocyst into the portal venous system. *Abdom Imaging* 1999; 24:494-496
4. Bradley EL, Clements JL Jr, Gonzalez AC. The natural history of pancreatic pseudocysts: a unified concept of management. *Am J Surg* 1979;137:135-141
5. Kaman L, Behera A, Singh R, Katariya RN. Internal pancreatic fistulas with pancreatic ascites and pancreatic pleural effusions: recognition and management. *ANZ J Surg* 2001; 71:221-225
6. Lee SH, Bodensteiner D, Eisman S, Dixon AY, McGregor DH. Chronic relapsing pancreatitis with pseudocyst erosion into the portal vein and disseminated fat necrosis. *Am J Gastroenterol* 1985;80:452-458
7. Skarsgard ED, Ellison E, Quenville N. Spontaneous rupture of a pancreatic pseudocyst into the portal vein. *Can J Surg* 1995;38:459-463
8. Van Steenberg W, Ponette E. Pancreaticportal fistula: a rare complication of chronic pancreatitis. *Gastrointest Radiol* 1990;15:299-300
9. Yoon SE, Lee YH, Yoon KH, Choi CS, Kim HC, Chae KM. Spontaneous pancreatic pseudocyst-portal vein fistula presenting with pancreatic ascites: strength of MR cholangiopancreatography. *Br J Radiol* 2008;81:e13-e16
10. Brown A, Malden E, Kugelmas M, Kortz E. Diagnosis of pancreatic duct-portal vein fistula; a case report and review of the literature. *J Radiol Case Rep* 2014;8:31-38

췌장의 가성낭종-문맥간 누공: 가성낭종에서 누공 형성까지의 영상 및 임상 소견의 연속적인 과정

지금난

췌장의 가성낭종-문맥간 누공은 극히 드문 췌장염의 합병증이다. 이러한 증례는 이제까지 의학논문에서 18편이 보고되었다. 그러나 췌장의 가성낭종에서 누공이 형성되기까지의 연속적인 과정에 대한 보고는 없었다. 이 증례 보고는 52세의 만성 알콜 중독인 남성 환자에서 췌장의 가성낭종에서 누공이 형성되기까지의 과정에 대한 연속적인 임상적, 영상의학적 소견에 대한 보고이다.

단국대학교 의과대학 단국대학교병원 영상의학과