CASE REPORT

Pancreatic Duct – Portal Vein Fistula in Acute on Chronic Alcoholic Pancreatitis: A Case Report and Literature Review

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Abstract

Chronic pancreatitis and significant alcohol use are common conditions and known risk factors for the development of pancreatic duct-portal vein (PD-PV) fistulas with a risk of subsequent liver necrosis accompanied by significant morbidity and mortality.

PD-PV fistulas can occur due to disruption of the PD and erosion of pancreatic enzymes into the wall of the PV. This is facilitated by the presence of a large pseudocyst (PC) in the vicinity of the PV, eventually creating a connection between the two structures.

Ductal anatomy should be evaluated with contrast enhanced CT and/or MRI when there is high clinical suspicion and a complex fluid density is visualized in the Portal vein on initial imaging. This will increase the odds of early identification of fistulization and subsequent planning for further management which is typically endoscopic or surgical depending on the severity of the presentation.

Introduction

Pancreatic duct-Portal Vein (PD–PV) fistula caused by direct erosion of the PD into the portal venous system can be a complication of pancreatitis with known risk factors of significant alcohol use, and pseudocyst formation. There are 18 cases reported in the literature which have been previously summarized.[1] Of these reports, 16 patients with PD-PV fistula had pancreatic pseudocyst associated with the disease.

Case Report:

A 56-year-old morbidly obese patient with a history of chronic pancreatitis and alcohol abuse presented to the emergency department with abdominal pain. Initial lab values included hemoglobin 8.6 gm/dL, platelets 117 K/mm3, albumin 2.2 g/dL, INR 1.5, total bilirubin 1.8 mg/dL, ALT 57 U/L, AST 215 U/L, alkaline phosphatase 820 U/L, and lipase 82 U/L. Abdominal ultrasound showed a heterogeneous liver mass and absent flow within the portal and hepatic veins. Contrast-enhanced CT was consistent with abdominal ultrasound findings of the liver mass, portal vein thrombosis (PVT), chronic pancreatitis, and splenomegaly. The main PV diameter was 3 cm. CT-guided liver biopsy was arranged during this admission and revealed micro and macro steatosis, and a benign cystic lesion. The patient was diagnosed with PVT, started on anticoagulation, and discharged home with an INR of 2.8.

Two weeks later, the patient's condition worsened and she was readmitted with INR above 12, albumin 1.6 mg/dL, lipase 1850 U/L. Repeat CT revealed significant expansion of the liver mass and PVT with occlusive thrombus extending into the Superior Mesenteric vein (SMV) with the main PV diameter measuring 4.8 cm. There was a 7-millimeter calcified stone visible in the posterior intrahepatic biliary system. She received FFP and was transferred to a larger hospital for further evaluation.

Subsequent arterial and venous phase post-contrast images revealed massive enlargement of the PV and diffuse enhancement of the PV walls extending to the SMV and the splenic vein (SV). Two calcifications were evident at the bifurcation of the right main PV and SMV representing pancreatic PD-PV fistulization. The pancreas was atrophic with scattered coarse calcifications and a retained calculus within the main PD at the level of the neck. Contrast- enhanced MRI ruled out other extra-ductal causes of the suspected fistula and confirmed the severe expansion of the portal venous system with no central enhancement. She was transferred to a tertiary care center for further evaluation and management.

Etiology and Differential

Pancreatic fistulas can occur due to disruption of the PD as a complication of chronic pancreatitis[2, 3] or as a result of trauma, and/or pancreatic surgery. Significant alcohol use and chronic pancreatitis are known risks factors for the development of pancreatic fistulas. Most of the patients with PD–PV fistulas have had a pseudocyst in the head of the pancreas in close proximity to the PV.[1, 2] Pancreatic enzymes play a significant role in the mechanism of fistula development. It is

believed that activated pancreatic enzymes can cause erosion of the PV wall stimulating inflammatory mediators leading to thrombus formation, followed by breakdown of the thrombus and subsequent filling of the PV with pancreatic secretions.[4] Patients with activated pancreatic enzymes within the portal venous system can have a wide range of manifestations ranging from vague abdominal pain to symptoms associated with diffuse liquefactive necrosis, which is one of the most severe complications of PD–PV fistula. This complication can present in one of the following scenarios: absence of portal venous blood supply or direct injury when activated pancreatic enzymes reach the systemic circulation.[5-8] In this scenario, the patients may often have amylase levels above 6000 U/L. The differential of PVT in this setting also includes the following: mass effect from a large pseudocyst or tumor, or hepatic hilar lymphadenopathy; however, these are not generally causes of fistulization.[9]

Despite all the imaging modalities available, the identification of PD–PV fistula is difficult. Bland PVT due to cirrhosis, hypercoagulable state, or extrinsic compression of the PV should be taken into consideration when a low attenuation PV is seen on imaging.

Imaging Findings

Patients with PD–PV fistula have absent flow and complex fluid within the PV on initial ultrasound. On previous reports, contrast- enhanced CT demonstrated low attenuation thrombus with non-enhancing PV in most of the patients; however, it is unlikely to reveal the fistula. Cross-sectional MRI can evidence fluid signal intensity in the PV, adjacent pseudocysts, and a hyperintense signal in the PD–PV fistula tract. ERCP is diagnostic when the identified fistula extends from the PD; however, a stricture or a stone in the main PD duct will limit its use. In that case, there will be incomplete opacification of the duct. Percutaneous trans-hepatic portography (PTHP) is diagnostic if a fistula is identified during contrast injection. It allows direct access to the portal venous system, visualization of the tract from the origin to the PV, evaluation of the extent of the disease, and the extraction of portal fluid for analysis.[10]

On the other hand, patients with bland thrombosis would demonstrate absent portal flow and thrombus within the PV on ultrasound; non-enhancing hypodense PV on cross sectional CT; an isointense PV on T1, and increased PV signal on T2 of MRI. Stasis of flow without fistula is appreciated on percutaneous hepatic portography. Compression of the extrahepatic bile ducts by dilatated and congested paracholedochal veins of Petren and epicholedochal venous plexus of Saint is characteristic of non-cirrhotic portal biliopathy and can be revealed instead of thrombosis on ERCP.[11] Patients with thrombosis due to extrinsic compression from tumor or lymphadenopathy may evidence absent portal flow, narrowing of the vein at the site of the compression, and increased flow velocity on initial ultrasound. An enhancing or non-enhancing mass in the hilum or within the PV and increased PV

signal can be seen on post-contrast MRI. PTHP provides precise anatomic evaluation and may show an external compression defect. Ultimately, patients with chronic thrombosis develop periportal collateral vessels within the hepatic hilum which may be observed on different imaging modalities.

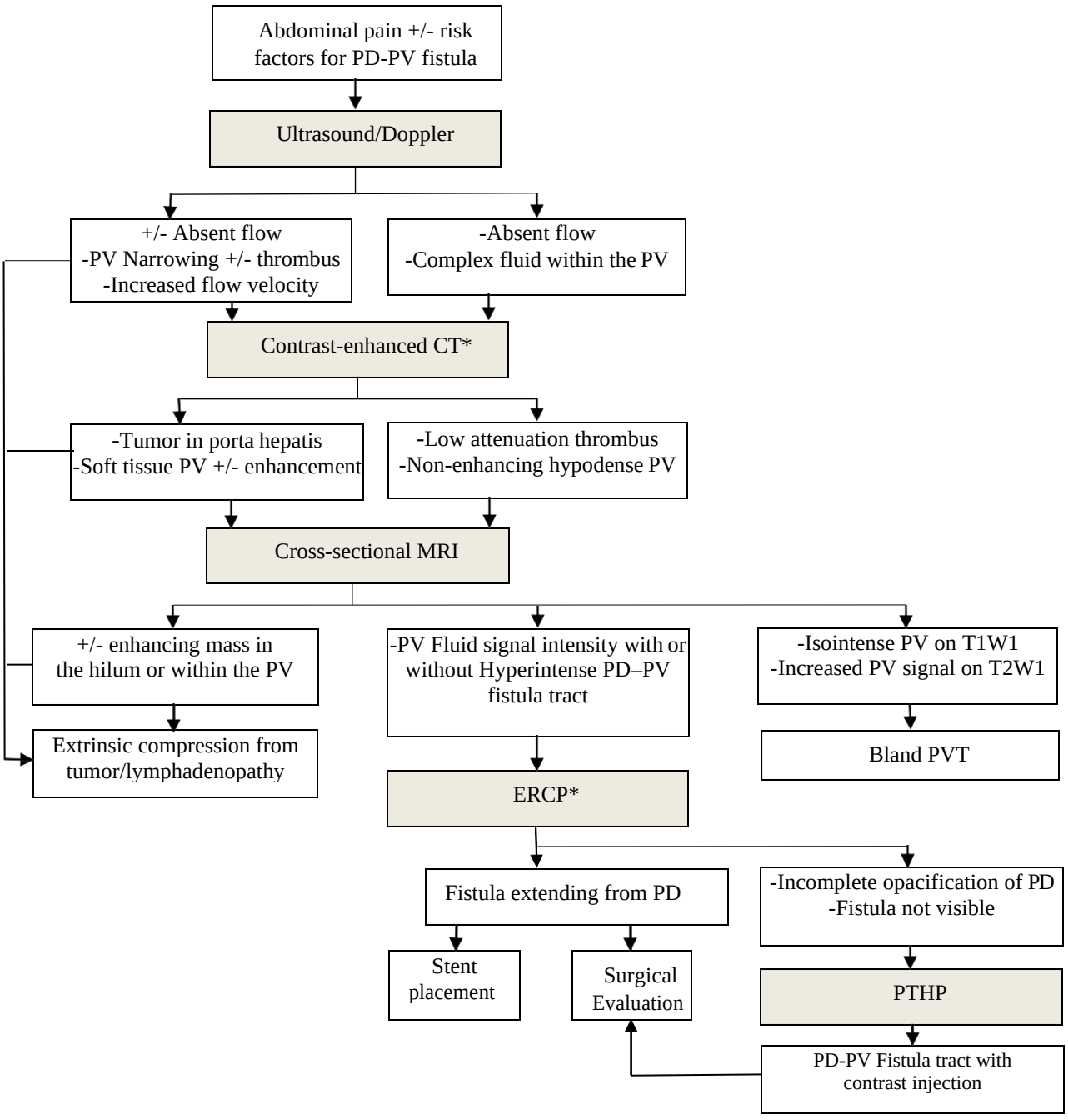
Treatment and Prognosis

Patients with PD–PV fistula must be evaluated on a case by case basis. Imaging assessment and early identification of PD–PV fistula is important for surgical planning. Early surgical intervention may be justified if the patient presents with or develops liquefactive necrosis as this carries a higher morbidity and mortality.[12-14] A minority of patients who are asymptomatic or have mild symptoms, improve with conservative management. Patients with more advance disease require ERCP with pancreatic stent placement or aggressive surgical treatment to establish pancreatic duct drainage when ERCP is not feasible. The surgical approach including portal vein plasty (for primary closure of the fistula) or pancreatectomy and pancreaticojejunostomy (for blocking the flow of pancreatic fluid) is considered definitive treatment. However, it carries a high morbidity and mortality risk and should only be attempted once the acute phase has subsided.[15] In one reported case, a pancreatic stent alone was used to treat the fistula successfully.[16]

Conclusions

PD-PV fistulization is a rare but potentially serious complication of pancreatitis with pseudocyst formation. It should be evaluated with contrast enhanced CT or MRI whenever initial imaging or clinical presentation are suggestive. ERCP and transhepatic portography can be effective in the visualization of pancreaticoportal fistulas when initial imaging fails to confirm the diagnosis [fig.1]. A surgical approach to management carries high morbidity and mortality risk. Management options can range from ERCP with pancreatic stent placement to surgery.

Figure 1. Proposed Diagnostic and Therapeutic Algorithm for Pancreatic Duct–Portal Vein Fistula



*Peri-portal collateral vessels and portal biliopathy from portal hypertension can be seen in contrast-enhanced CT and ERCP respectively.

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