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Case Report

Distal pancreatectomy for pancreatic arteriovenous malformation: report of a case

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Arteriovenous malformation (AVM) of the pancreas is an uncommon disease, which can cause an abdominal pain. This disease is characterized by a tangled vascular network, including the whole or part of the pancreas, resulting in portal hypertension by forming a shunt of the pancreas arteries to drain directly into the portal venous system. This study presents a case that was suspected as AVM of the pancreas by preoperative contrast-enhanced computed tomography scan (CT). A 49-year-old male patient had several episodes of abdominal discomfort associated with dyspepsia for 4 days. Magnetic resonance imaging showed enhancement of the conglomeration about 1.5 cm size in diameter in the pancreas. Selective angiography showed the proliferation of a vascular network in the pancreas and an early visualization of the portal vein during the arterial phase. Distal pancreatectomy with splenectomy was done. Histology of the pancreas showed AVM, with enzymatic fat necrosis extending to the capsule of the pancreas. The patient recovered successfully without postoperative complications. Surgical resection of pancreas is the definitive treatment for symptomatic AVM. **(Ann Hepatobiliary Pancreat Surg 2017;21:172-175)**

Key Word: Pancreatic arteriovenous malformation

INTRODUCTION

Arteriovenous malformation (AVM) can occur in any gastrointestinal (GI) tract, but it is extremely rare in pancreas. Abnormal tangled arteries and veins in the pancreas form a direct drainage called shunt, which can cause portal hypertension. The incidence of pancreatic arteriovenous malformation (PAVM) remains low. Fewer than 200 cases have been reported since Halpern reported the first case in 1968.[1] In recent years, cases of PAVM have increased and it is believed that the development of image modalities raised the diagnostic rates. PAVM can be asymptomatic, but some patients present abdominal pain, terry stool, and GI bleeding. Herein, this research presents a case that was suspected as PAVM by preoperative computed tomography (CT) scan. Through this case, the researchers elucidated radiological, surgical, and histopathological characteristics of PAVM, in order to understand the disease in various angles.

CASE

A 49-year-old male, who was in good health prior, visited a local clinic for epigastric pain. The patient had dyspepsia associated with this pain for 4 days. The CT scan showed a mass like lesion in the pancreas tail. He was transferred to our institute for further evaluation. He had no significant medical history. Initial laboratory investigations showed a slightly elevated white cell count of 10.77×10^3/L (reference range $4.7\text{-}9.6\times10^3$/L). Abdominal plain film was normal.

A contrast enhanced CT demonstrated a high-density lesion of about 1.5 cm size in diameter with tortuous vascular in the tail of pancreas (Fig. 1A). An MRI showed intermediate signal intensity lesion of the pancreatic tail without distal pancreas ductal dilatation (Fig. 1B). Selective angiography showed increased vascularity with staining in the middle portion of the splenic artery. The great pancreas artery was hypertrophied and the vascularity was increased at the pancreas level. Early venous

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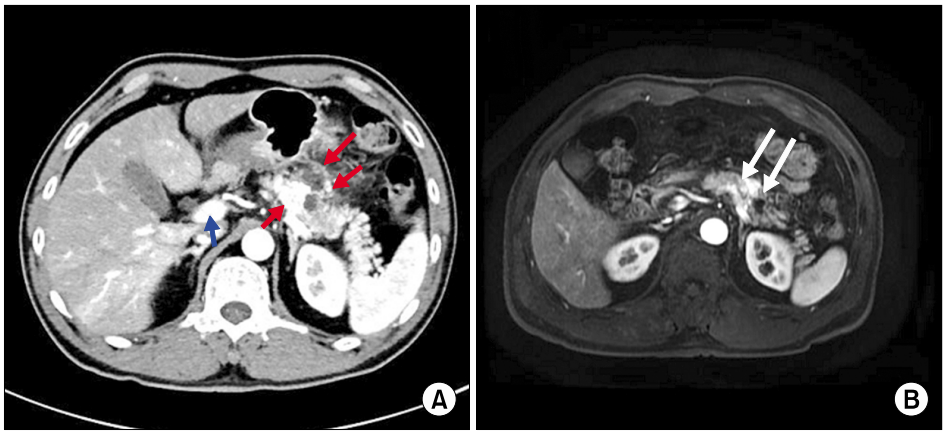

Fig. 1. Arterial phase contrast-enhanced computed tomography showed high density lesions of about 1.5-2.0 cm in diameter with tortuous vascular structure. Fluid collection was located at the distal portion of the vascular structure, with fat stranding around it (A, red arrows). The supplying artery is not clear, but early drainage is seen to the portal vein during the arterial phase (A, blue arrows). Magnetic resonance imaging showed intermediate a signal intensity lesion with cystic portion of the pancreatic body, without distal pancreatic duct dilatation (B, white arrows).

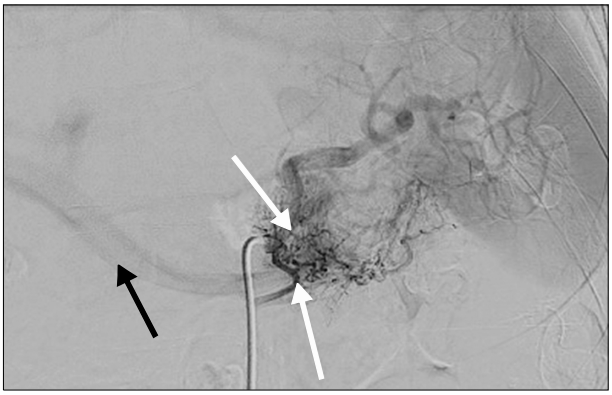

Fig. 2. Celiac angiography showed that the great pancreatic artery was hypertrophied, the vascularity increased at the pancreas level (white arrows), and the early venous draining was observed (black arrow).

drainage to the portal system was observed, suggesting arteriovenous malformation of the pancreas (Fig. 2).

Intraoperative examination revealed no abnormality of the surrounding vessels of the pancreas. The researchers performed distal pancreatectomy with a splenectomy. Duration of the operation was 2 hours 33 minutes and the estimated blood loss was less than 50 ml. Serial sections of the resected specimens showed dilated and tortuous vessels in the pancreas tail (Fig. 3). Histology of the pancreas showed AVM with enzymatic fat necrosis, extending to the capsule of the pancreas. The patient recovered successfully without complications and discharged on postoperative day 8. There was no evidence of recurrence for 8 months (Fig. 4).

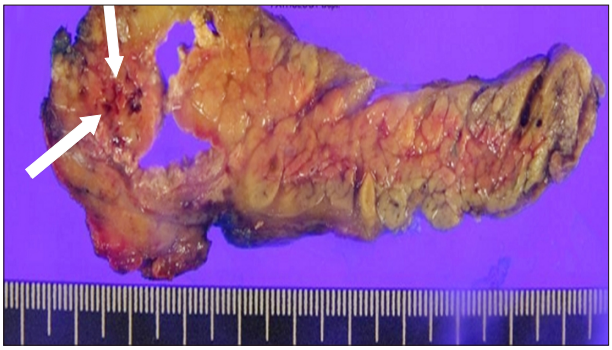

Fig. 3. Serial sections of the resected specimens showed dilated and tortuous vessels (white arrows) in the pancreatic body.

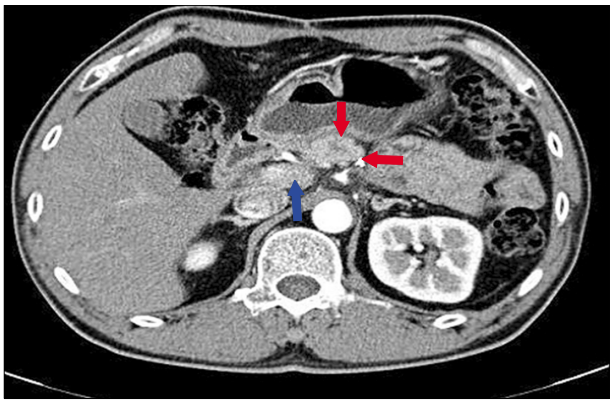

Fig. 4. Post-operation follow-up computed tomography scan after 8 months, showed that the remaining pancreas had no evidence of recurrence (red arrows). There was no evidence of early drainage to the portal vein during arterial phase (blue arrow).

DISCUSSION

PAVM is defined as an abnormally tangled arteries and veins in the pancreas forming a shunt, which can cause portal hypertension. It was first described by Halpern in 1968.[1] Many hypotheses have explained the origin of AVM. The loss of the regulatory sphincter mechanism at the arteriolar-capillary junction results in an overflow of arterial blood stream into the capillary bed forms a shunt.[2] PAVM is either congenital or acquired. Congenital PAVM can be explained as the malformation of pancreas during the embryonic period. Ventral pancreas and dorsal pancreas arise from the digestive organs, corresponding to the primary duodenum by the 4th week of gestation. The ventral and dorsal pancreas fuse after the duodenal tube has been rotated from the end of 5th week of gestation. At 8th week of gestation, the pancreas is entrapped in the duodenum. At the 10th week of gestation, the Oddi sphincter is formed and recanalization onsets on 11th week of gestation. Since this complex fusion starts, various embryological abnormalities and diseases may occur in this process (Fig. 5). Acquired PAVM can be caused by pancreatitis, tumors, and traumas. Most of the cases were considered to be congenital, including Osler-Weber-Rendu disease.[3]

Literature reviews have noted that 85-88% of patients are male and 77-78% of patients as Asian. The mean age was 50-51 years old.[4] PAVM symptoms are variable. They can also be asymptomatic. When symptoms are present, abdominal pain and GI bleeding are the most common symptoms. Duodenal ulceration and abdominal angina can also occur due to vascular steal phenomenon.[5] GI bleeding is thought to be caused by 1 of 4 mechanisms: rupture of PAVM vessels into the pancreatic duct; rupture of PAVM vessels into the common bile duct; rupture of gastroesophageal varices due to portal hypertension; and GI ulceration bleeding due to ischemic change caused by vascular steal phenomenon.[6]

Diagnosis is made with imaging such as CT, magnetic resonance imaging, Doppler ultrasound, and angiography. On dynamic CT scans, PAVM shows a complex and strong contrast enhancement and an early injection of contrast agent into the portal vein in arterial phase. Angiography is the most common requested investigation and helps in the diagnosis of PAVM. It also has an advantage of catheterization and helps to consider differential diagnoses. The arterial phase of angiography shows feeding arteries that are extended and twisted. After the pancreas gets deeply visualized for a while, a vascular network is seen inside the tumor and the portal system becomes visible.

Treatment with transarterial embolization (TAE), irradiation, transjugular intrahepatic portosystemic shunt and operation have been introduced as management of PAVM.[7] TAE is a valuable alternative to patients who cannot have an operation. Preoperative TAE can reduce bleeding during the operation and improve surgical outcomes. However, the results of TAE are unpredictable,

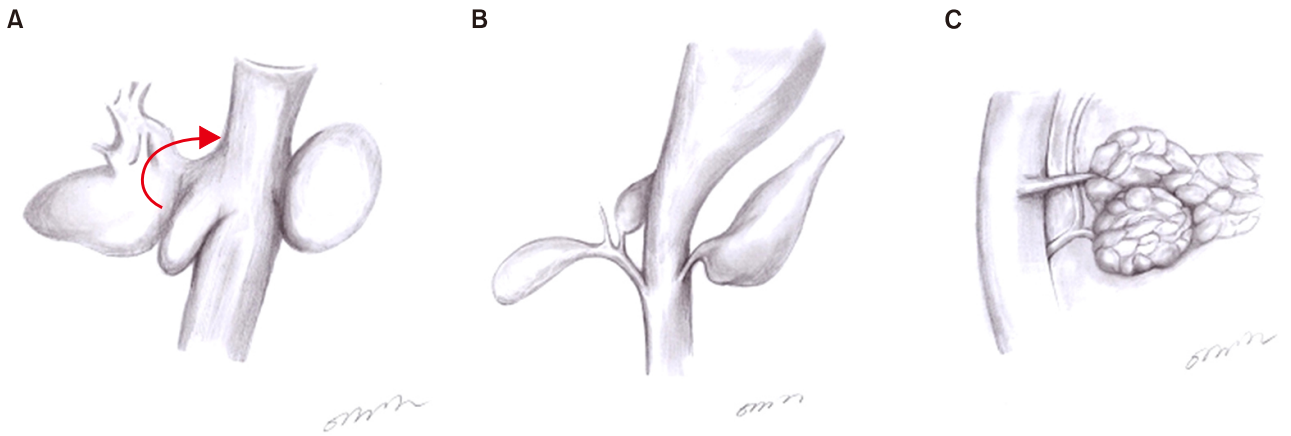

Fig. 5. Illustration of development of the pancreas during the embryonic period. Ventral and dorsal pancreas arise from the digestive organs at the 4th week of gestation (A). Ventral pancreas slowly rotates toward the dorsal pancreas (B). Ventral and dorsal pancreas fuse, after the duodenal tube has been rotated from the end of 5th week of gestation (C).

because it is very difficult to embolize every supplying artery. Recurrent bleeding after TAE has been reported up to 18-37% of patients, waiting for surgery.[8] Therefore, surgery is essential for patients who have experienced bleeding before operation.

Although PAVM is a rare disease, it is a highly treatable condition. Surgical resection of pancreas is a definitive treatment for symptomatic AVM. It prevents the development of portal hypertension. Portal hypertension is the most important prognostic factor for PAVM, because of the life-threatening GI bleeding. It also acts as a key factor for recurrence.[4] The procedure depends on the location of the lesion. When the body and tail of the pancreas are involved, the pancreas can be partially resected. There is risk of recurrence in the remaining pancreas after surgery. Thus, accurate resection and close observation with long term follow-up are required.

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