Pancreatic Arteriovenous Malformation Combined with Portal Thrombosis

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Abstract

We encountered a case of portal vein thrombosis (PVT) after treatment for portal hypertension due to pancreatic arteriovenous malformation (PAVM). A 75-year-old man was admitted for the treatment of esophageal varices. Diffuse PAVM and aneurysm in the celiac and superior mesenteric arteries were detected via abdominal computed tomography and angiography. Although endoscopic sclerotherapy was performed, PVT was identified after the treatment and variceal bleeding continued. Autopsy was performed and the thrombus and malformation were pathologically confirmed. This case indicates that PVT can be associated with PAVM.

Key words: pancreatic arteriovenous malformation, portal vein thrombosis, abdominal aneurysm, endoscopic sclerotherapy

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Introduction

Pancreatic arteriovenous malformation (PAVM) is a rare disorder leading to gastrointestinal bleeding, but an optional treatment has not yet been established. Liver cirrhosis, hepatobiliary malignant tumor and pancreatitis are risk factors for portal vein thrombosis (PVT), and PVT sometimes combines with PAVM following sclerotherapy for gastroesophageal varices. However, there have been no reported cases that whereby PVT developed after the treatment of portal hypertension due to PAVM. Here, we describe the case of a 75-year-old male with PVT associated with PAVM.

Case Report

A 75-year-old man was referred and admitted to our hospital for the treatment of esophageal varices in August 2003. Two years previously, he was diagnosed with atrial fibrilla-

tion, and two months previously transcatheter ablation was carried out. In early July 2003, he noticed abdominal fullness and was diagnosed with ascites by a local doctor. He was hospitalized on July 18, 2003, because of hematemesis. Emergency endoscopic examination revealed bleeding esophageal varices and an endoscopic variceal ligation (EVL) was carried out. He had no history of alcohol abuse.

Physical examination revealed mild abdominal tenderness, no palpable mass, no bruits, and no telangiectasia of the skin or oral mucosa. Laboratory findings on admission are summarized in Table 1. Serum anti-HBs antigen and anti-HCV were negative, and the tumor markers of carcinoembryonic antigen (CEA) and carbohydrate antigen 19-9 (CA 19-9) were within normal ranges. Extreme hypervascularity of the entire pancreas was detected on dynamic CT, and multiple circular lesions appearing to be "signal-void" were shown on MRI T2-weighted images (Fig. 1A, B).

Angiography showed extensive arterial abnormality throughout the pancreas and early venous filling. An aneurysm of the splenic artery and bifurcation of the celiac ar-

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Table 1. Laboratory Findings on Admission

Hematology			Chemistry		
WBC	3,000	/ μ L	TP	6.9	g/dL
RBC	300	$\times 10^4$ / μ L	Albumin	3.8	g/dL
Hb	9.0	g/dL	T-Bil	0.7	mg/dL
Ht	28.7	%	AST	20	IU/L
PLT	24.2	$\times 10^3$ / μ L	ALT	17	IU/L
Coagulation			LDH	236	IU/L
APTT	36.8	Sec.	ChE	2.0	IU/mL
PT	>100	%	ALP	602	IU/L
Tumor markers			γ -GTP	117	IU/L
CEA	2.4	ng/ml	Amylase	49	IU/L
CA19-9	18	ng/ml	p-Amylase	24	IU/L
SPAN-1	12.1	U/ml	Lipase	13	IU/L
Virus markers			Trypsin	120	ng/mL
HBs-Ag	(-)		Glucose	93	mg/dL
HCV-Ab	(-)		CRP	0.10	mg/dL

pressure was 12 mmHg. Transcatheter embolization (TAE) was not examined because there were too many draining veins. He was treated with a diuretic drug, and endoscopic sclerotherapy was carried out for the esophageal varices. After treatment, he had continuous abdominal pain and portal venous thrombus (PVT) was identified by ultrasonography. Portal venous flow was weakly detected and no changes occurred in the laboratory data (Fig. 3). Total pancreatectomy and portal thrombectomy under abdominal operation were assessed, but he rejected any surgical therapy and was followed up by conservative therapy.

In October, he experienced recurrent gastrointestinal bleeding. Bleeding from esophageal varices was uncontrollable in spite of repeated EVL, and his general condition deteriorated. He died on October 27 and an autopsy was performed. Macroscopically, a honeycomb-like structure with collected vessels was shown in the pancreatic body, and an

Figure 1. (A) CT shows a round or tubular structure enhanced by bolus injection of contrast medium in the area of the pancreatic body and tail (arrowheads) and remarkably dilated splenic and portal veins. (B) Multiple circular lesions appear to be “signal-void” (arrowheads) by MRI T2-weighted image, which is characteristic of rapidly flowing blood on MRI.

Figure 2. (A) Celiac arteriogram shows tortuous and dilated feeding arteries (pancreatoduodenal arteries, dorsal pancreatic artery, transverse pancreatic artery, and short gastric artery) which led to an extensive racemose intrapancreatic vascular network and early filling dilated splenic and portal veins. Aneurysm of the splenic artery at the bifurcation of the celiac artery (arrow) was also noted. (B) Superior mesenteric and middle colic arteriogram revealed an arteriovenous malformation of the pancreatic tail and early filling splenic vein. The splenic vein was opaque in the early phase.

tery were also observed (Fig. 2A, B). Hepatic vein wedge organized thrombus completely occupying the whole lumen

Figure 3. Ultrasonography showed the thrombus in the right branch of the portal vein (arrowheads). Weak portal venous flow was detected by color doppler imaging.

Figure 4. (A) Macroscopically, there was a honey-comb like structure with collected vessels in the pancreatic body, and an organized thrombosis occupying the whole lumen of the main portal vein, and the right and left branches of the portal vein, and superior mesenteric and splenic veins. (B) Microscopic analysis of the autopsy specimen revealed numerous dilated and tortuous vessels that cannot be morphologically differentiated as either arteries or veins in the pancreatic parenchyma. The pancreatic exocrine glands were atrophic, but without the presence of tumor or inflammation. This lesion was considered to be of congenital origin. (HE stain, $\times 20$)

of the main portal vein, and right and left branches of the portal vein, and superior mesenteric and splenic veins was noted (Fig. 4A). There were no findings of obstruction or ischemia in the hepatic vein and inferior vena cava. Microscopically, numerous dilated and tortuous vessels that could not be morphologically differentiated as either arteries or veins were noted in the pancreatic parenchyma (Fig. 4B). The pancreatic exocrine glands were atrophic without fibrosis, inflammation or cancer. No findings of cirrhosis or congestion were shown in the liver tissue.

Discussion

PAVM was first reported in 1968 by Halpern et al, and is defined as a tumorous formation or vascular anomaly which builds up via an aberrant bypass anastomosis of the arterial and venous systems in the pancreas (1). An increasing num-

ber of cases of PAVM are being diagnosed via improvements in imaging techniques, but it is still considerably rare in the world. We examined the references for the past 40 years by Medline and Igaku chuo zasshi, and summarize 90 cases of PAVM in previous reports in Table 2 (1-13). The average age at diagnosis was 52.5 years (range ; 7 months old to 73 years old), with a male predominance. PAVM is sometimes combined with liver diseases including liver cirrhosis and hepatocellular carcinoma. Clinically, gastrointestinal bleeding and abdominal pain due to portal hypertension are the main symptoms. In this case, gastrointestinal bleeding was critical, but no chronic liver disease was detected. In Western countries, congenital PAVM associated with Rendou-Osler-Weber (ROW) disease was previously reported (1, 5, 9). So far, there has only been one reported case of PAVM combined with intra-abdominal aneurysms (14), though PAVM is sometimes observed to be associated

Table 2. Summary of 90 PAVM Cases (%)

Backgrounds		Complications	
Mean age	52.5 y (7 mo - 73 y)	Varices	19 (21)
Male : Female	73 : 17	Liver cirrhosis	11 (12)
Incidence		Duodenal ulcer	15 (17)
Congenital	49 (54)	ROW disease	7 (8)
Acquired	5 (6)	Hepatoma	7 (8)
Unknown	36 (40)	Treatment	
Localization		Pancreatic resection	35 (39)
Head	37 (41)	Intestinal resection	6 (7)
Head - Body	9 (10)	Operation for portal hypertension	2 (2)
Body	9 (10)	TAE	15 (17)
Body - Tail	15 (17)	Arterial ligation	2 (2)
Tail	7 (8)	Conservative	22 (24)
Entire	13 (14)	Others	5 (6)
Symptoms			
GI bleeding	48 (54)		
Abdominal pain	27 (30)		
Abdominal discomfort	2 (2)		
Jaundice	4 (4)		
No symptom	14 (14)		
Others	4 (4)		

with ROW disease. However, there has been no such case in Japan, and PAVM in Japan might be different from that in other countries. This case was thought to be congenital because an acquired factor was not detected, but the mucocutaneous symptom suggesting ROW syndrome was absent. Recently, it was reported that transarterial embolization

(TAE) and transjugular intrahepatic portosystemic shunt (TIPS) are effective for PAVM (2, 3, 6), though surgical treatment is classically radical and effective for PAVM. In this case, it was thought that TAE would not be effective because there were too many drainage veins.

In such cases, the appearance of PVT may aggravate portal hypertension and result in uncontrollable variceal bleeding. PVT is usually generated based on some risk factors such as hepatic disorders, abdominal inflammation, malignancies, hypercoagulability, myeloproliferative syndrome and abdominal intervention (15), and is sometimes fatal when the esophageal or gastric varices are ruptured. But the etiology was unknown in nearly half of the patients (16). In the present case there was no such risk factor and the origin was not clarified. Although there are several case reports of PVT occurring after sclerotherapy (17), controlled studies have revealed that sclerotherapy does not increase the risk of PVT (18, 19).

Here, we presented a case of PVT following treatment for PAVM. It should be noted that PVT may occur following treatment for portal hypertension even in the absence of liver disease.

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