CASE REPORT

A Case of Recurrent Acute Pancreatitis due to Pancreatic Arteriovenous Malformation

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Pancreatic arteriovenous malformation (AVM) is an extremely rare condition with various clinical manifestations. We report herein a case of recurrent acute pancreatitis due to pancreatic AVM in a 49-year-old man. This patient presented with epigastric pain that had developed after consuming alcohol 2 days prior to admission. Serum amylase and lipase levels were elevated and computed tomography revealed focal low-attenuation lesions with peripancreatic infiltrations in the pancreatic tail and multiple collateral vessels around the low-attenuation lesions. He was diagnosed with acute pancreatitis and pancreatic AVM. Although he had stopped drinking after the first attack of acute pancreatitis, his pancreatitis recurred twice within 3 months. He underwent a distal pancreatectomy after the third attack of acute pancreatitis. He was free of symptoms for 2 years after the pancreatectomy. (*Gut Liver* 2010;4:135-139)

Key Words: Pancreatitis; Pancreatic arteriovenous malformation

INTRODUCTION

Pancreatic arteriovenous malformation (AVM) is a rare condition defined as a tumorous formation or vascular anomaly built up via an aberrant bypass anastomosis of the arterial and venous systems in the pancreas.¹ Most patients with pancreatic AVM are asymptomatic. However, some present with symptoms varying from gastrointestinal bleeding, abdominal pain, jaundice and more.

We report a case of recurrent acute pancreatitis due to pancreatic arteriovenous malformation in a 49-year-old Korean man, which was successfully treated by surgical resection. We also review the literature concerning pancreatic AVM.

CASE REPORT

A 49-year-old man was admitted to our hospital due to severe epigastric pain with radiation to the back for past two days. He had been experiencing intermittent epigastric discomfort for previous 2 months. He was a social drinker and had taken a drink or two the night before the onset of symptoms. He also complained of nausea and vomiting. His past medical history included hypertension and diabetes mellitus for which he had been taking oral medications. He had no history of operation or trauma. No relevant family history, including diabetes and pancreatitis, was found.

His blood pressure and pulse rate were 182/92 mm Hg and 62/min and his body temperature was 36.8°C. His abdomen was soft and flat with decreased bowel sounds. He had severe epigastric tenderness without guarding or rebound tenderness. The liver and spleen were not enlarged and there was no palpable abdominal mass or ascites. Laboratory studies revealed a white blood cell count of 9,730/mm³, a hemoglobin concentration of 16.6 g/dL, and a platelet count of 234,000/mm³. Liver tests were as follow: cholesterol 220 mg/dL, total bilirubin 1.0 mg/dL, glucose 112 mg/dL, alkaline phosphatase 73

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Fig. 1. Arterial-phase computed tomography (CT) revealed tortuous tubular structures with marked enhancement in and around the pancreatic tail, and early opacification of the splenic and portal veins, findings that are suggestive of pancreatic arteriovenous malformation. The focal low-attenuation lesion in the pancreatic tail with peripancreatic fat infiltration was compatible with acute pancreatitis.

IU/L, AST 29 IU/L, ALT 49 IU/L, total protein 7.7 g/dL, and albumin 5.0 g/dL. Amylase and lipase were elevated to 486 U/L and 7,580 U/L respectively. Hemoglobin A1c was 7.2%. Serum electrolytes, coagulation, and hepatitis studies were within normal range. Computed tomography (CT) revealed focal low attenuated lesions with peripancreatic infiltrations in the pancreatic tail and multiple collateral vessels around the low attenuated lesions. These findings suggested focal acute pancreatitis and pancreatic AVM accompanying anatomical variation of intra-abdominal artery (Fig. 1). Anatomical variations of intra-abdominal arteries were detected. The hepatic artery replaced right hepatic artery and middle hepatic artery from superior mesenteric artery and left hepatic artery from left gastric artery. The arch of Rioloan was prominent probably due to the stenosis of superior mesenteric artery orifice. Upper endoscopy showed normal esophagus and stomach. The abdominal pain disappeared gradually on conservative management. The patient refused to proceed

Fig. 2. Angiography showing the vascular network (black arrow) with several tributary branches from the celiac trunk and superior mesenteric artery (SMA) in the pancreatic tail area, and early filling of the splenic and portal veins (white arrow). (A) Celiac angiography. (B) SMA angiography.

Fig. 3. (A) Histologic sections showed a dilatation of the peripancreatic vessels with thrombosis. (B) Irregularly enlarged angiodysplastic vessels with thick- and thin-walled vascular channels were evident (H&E stain, $\times 10$).

with further evaluations such as angiography and endoscopic retrograde cholangiography despite physicians' recommendations. He was subsequently discharged following a gradual recovery of symptoms including the abdominal pain.

However, 7 weeks later, he was re-admitted with severe epigastric pain, although he had quit drinking after the first attack of acute pancreatitis. He was again diagnosed with acute pancreatitis. He recovered uneventfully with conservative management and refused further evaluation. However upon the third attack of pancreatitis which took place after 5 weeks, he agreed to proceed with invasive work-ups for further management for pancreatic AVM. Endoscopic retrograde cholangiography showed normal pancreatic duct. Selective angiography revealed abundant small abnormal vascular structures and early draining veins in pancreatic tail (Fig. 2). These findings were compatible with pancreatic AVM. Therefore, we recommended surgical treatment. The patient underwent distal pancreatectomy. Intraoperatively, the distal portion of the pancreas was diffuse hard, but there were no discrete pancreatic mass and exposed abnormal vessels.

Histologic sections showed a dilatation of peripancreatic vessels with thrombosis (Fig. 3A). Microscopic examination of the resected specimen showed tortuous dilated vessels which had thick and thin walls with focal intimal thickening and thrombosis. Moreover, edema and inflammatory cell infiltration, and necrosis were evident, confirming the presence of acute pancreatitis (Fig. 3B). The patient was discharged 35 days after the operation.

He tolerated well without any complaints of abdominal pain for 2-years of follow-up.

DISCUSSION

Pancreatic AVM was first reported in 1968 by Halpern *et al.*¹ Pancreatic AVM is an extremely rare condition and the true prevalence of disease is still unknown.^{2,3} However, the number of reported cases is recently increasing owing to the widespread use of imaging techniques. Pancreatic AVM can be classified into two types, congenital and acquired.⁴ In most cases, pancreatic AVM is considered a congenital anomaly arising from an anomalous differentiation in the rudimentary plexus of primordial blood vessels, as is Rendu-Osler-Weber syndrome. Only a few reported cases are considered acquired, which are usually secondary to inflammation, trauma or tumor.^{5,6}

Most patients with pancreatic AVM are presumed to be asymptomatic. In previous reports, clinical symptoms included varying combinations of gastrointestinal bleeding (47%), abdominal pain (41%), jaundice (14%), etc.² The main symptom of pancreatic AVM is gastrointestinal bleeding associated portal hypertension.⁷ To our knowledge, only 9 cases of pancreatitis associated pancreatic AVM have been reported in the literature to this point.⁷⁻¹⁴ The clinical characteristics of these patients are summarized in Table 1. Both congenital and secondary types of either acute or chronic pancreatitis associated pancreatic AVM were included in these reports.⁷⁻¹⁴ The majority of

Table 1. Clinical Characteristics of Previously Reported Case of Pancreatitis with Pancreatic Arteriovenous Malformation

Authors	Year	Age/Sex	ROW	Chief manifestation	Localization	Treatment
Chuang <i>et al.</i> ⁹	1977	56/M	Yes	Esophageal variceal bleeding Chronic pancreatitis (chronic alcoholics)	Body, tail	Distal pancreatectomy
Gomes <i>et al.</i> ⁸	1983	52/M	Yes	Recurrent GI bleeding Recurrent acute pancreatitis	Head	Embolization Pancreaticoduodenectomy
Ohtani <i>et al.</i> ¹⁰	1992	60/M		Acute pancreatitis Pancreaticovenous fistula	Tail	Distal pancreatectomy
Abe <i>et al.</i> ¹¹	1998	48/M		Acute pancreatitis	Body, tail	Pancreatoduodenectomy
Uda <i>et al.</i> ¹²	1999	48/M		Acute pancreatitis accompanying bleeding from bile duct and duodenal ulcer	Head	Preoperative embolization and pancreaticoduodenectomy
Koito <i>et al.</i> ¹³	2001	63/M		Acute pancreatitis	Body, tail	Distal pancreatectomy
Aida <i>et al.</i> ¹⁴	2002	54/M		Duodenal bleeding Recurrent acute pancreatitis	Head	Pancreatoduodenectomy
Kanno <i>et al.</i> ⁷	2006	44/M		Abdominal pain Acute pancreatitis	Body	Distal pancreatectomy
Kanno <i>et al.</i> ⁷	2006	49/M		Recurrent acute pancreatitis	Head	Pancreaticoduodenectomy

ROW, Rendu-Osler-Weber syndrome; GI, gastrointestinal.

the patients suffered from gastrointestinal bleeding and pancreatitis either simultaneously or independently. Prior to our case there was only one reported case of recurrent acute pancreatitis associated with pancreatic AVM which presented without gastrointestinal bleeding or ROW syndrome.⁷ Moreover, this is the first report of pancreatic AVM accompanying anatomical variations of intra-abdominal arteries.

The pathophysiology of both pancreatic AVM and pancreatitis are not fully understood yet. The mechanisms of pancreatitis arising from pancreatic AVM may be classified into three types: i) by bleeding from pancreatic AVM extending into the pancreatic duct,^{10,15} ii) by ischemic injury due to the pancreatic AVM "stealing" flow from normal surrounding parenchyma like brain AVM,⁷ iii) by pancreatic duct compression from pancreatic AVM. In this case, ischemia of the pancreas by the pancreatic AVM appeared to induce pancreatitis, because bleeding from pancreatic AVM into the pancreatic duct was not observed, and the resected specimens had normal margin and a dilatation of peripancreatic vessels with thrombosis. Endoscopic retrograde cholangiography revealed normal pancreatic duct without ductal obstruction.

Some reports described that pancreatic AVM can not only be the result but also the cause of pancreatitis.^{3,7,16-21} Pancreatitis can cause vascular damage including thrombosis or erosion of intrapancreatic and peripancreatic vessels by the direct effect of pancreatic enzymes.^{5,22} A case with a pancreatic pseudocyst eroding into both vascular and ductal structures, producing a pancreatic duct arteriovenous fistula was reported.⁵ In this case, pancreatic AVM and acute pancreatitis were revealed at the same time and this patient had no history of trauma or tumor. Therefore, this case is probably congenital disease, but not a secondary change due to pancreatitis.

Diagnosis is usually confirmed by imaging because the symptoms are not specific and the incidence of this entity is very low.²³ Pancreatic AVM is first detected by cross-sectional imaging (CT or MRI) or color Doppler ultrasound, although angiography should be performed in all patients with these lesions to delineate the extent of the AVM and to facilitate a rational approach to treatment. The features of pancreatic AVM indentified by multiphase contrast-enhanced CT include conglomeration of strong nodular stains and early contrast filling of the portal vein during the arterial phase. CTs can identify feeding arteries and its anatomical relationship with adjacent structures.² The CT findings of this case were not only competent for diagnosing pancreatic AVM but also for the identification of vascular structures including feeding artery, draining vein and intra-abdominal arterial variations. Color Dop-

pler ultrasound and MRI are good diagnostic modalities for the diagnosis of pancreatic AVM.^{7,20} The angiography is a definitive diagnostic test. The angiographic findings of pancreatic AVM are characterized by dilated and tortuous feeding arteries, a racemose intra-pancreatic vascular network followed by a transient dense pancreatic stain, early venous filling (into the portal vein), and early disappearance of the pancreatic stain.⁹ The differential diagnosis of the angiographic finding includes pancreatitis and hyper-vascular tumor such as cystadenoma, islet cell tumor, and metastasis. The tumors frequently remain densely stained in the venous phase.^{6,9,23,24} In this case, the findings on CT and angiography were compatible with pancreatic AVM as stated above. Therefore we could conclude that this case had congenital AVM of the pancreas, leading to the development of acute pancreatitis.

In patients with symptomatic pancreatic AVM, considerations should be given both to the treatment pancreatic AVM itself and to the management of complications in order to prevent recurrence or other complications, such as portal hypertension, and gastrointestinal bleeding. Complete surgical resection is radical and effective for pancreatic AVM, among several alternative approaches to the treatment of symptomatic pancreatic AVM. Transarterial embolization (TAE), vascular ligation and partial resection have been employed as alternative treatments. However, pancreatic AVM often has multiple feeding arteries making it very difficult to embolize or ligate all of them and incomplete treatments of pancreatic AVM can result in recurrent events and the progression of portal hypertension.¹³ Some authors suggested preoperative TAE to reduce the risk of surgery by decreasing portal flow.²³ There were also reported cases that were treated by embolization of draining veins of AVM and radiation treatment for massive pancreatic AVM.^{16,25} Since this patient had no further attack of pancreatitis after surgery, we were able to reconfirm that the recurrent pancreatitis had resulted from pancreatic AVM.

The treatment of asymptomatic pancreatic AVM is controversial. Some authors recommend early surgical resection since it could progress to portal hypertension, leading to gastrointestinal bleeding and/or rupture of the esophageal varices.¹³ However, the natural course of asymptomatic pancreatic AVM is not elucidated yet. Also morbidity and mortality rates of surgical treatment of pancreatic AVM are relatively high.

In summary, we experienced a rare case of 49-year-old Korean male who had recurrent acute pancreatitis associated with pancreatic AVM, which was successfully treated by surgical treatment. Pancreatic AVM should be considered as another potential source of recurrent acute

pancreatitis.

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