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CASE REPORT

A case of chronic pancreatitis exacerbation associated with pancreatic arteriovenous malformation: a case report and literature review

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Abstract

A 60-year-old man with an unruptured cerebral aneurysm and family history of moyamoya disease was admitted to our hospital with epigastric pain since the previous day. Serum levels of pancreatic enzyme were elevated and abdominal contrast-enhanced computed tomography showed localized enlargement of the pancreatic tail in the arterial phase and revealed numerous areas of fine mesh-like vascular hyperplasia consistent with an enlarged pancreatic tail. We diagnosed pancreatic arteriovenous malformation (P-AVM) with acute pancreatitis. Furthermore, in the pancreatic body, endoscopic ultrasonography showed lobularity (honeycombing type) and hyperechoic foci (non-shadowing), which suggests chronic pancreatitis. Acute management was performed with conservative treatment including administration of replacement fluids and proteolytic enzyme inhibitor. Distal pancreatectomy for P-AVM was performed because P-AVM is associated with acute pancreatitis recurrence, development of portal hypertension, progression of chronic pancreatitis, and refractory duodenal bleeding. Histological findings on the resected specimens revealed the anastomosis of abnormal arteries and veins, which suggested P-AVM. In addition, inflammation accompanied by fat necrosis due to ischemic infarction in the pancreatic tail, which suggested acute pancreatitis, and mild fibrosis in the pancreatic body, which suggested chronic pancreatitis, were shown. Although P-AVM is associated with various complications, symptomatic P-AVM should be considered a chronic and progressive disease.

Keywords Pancreatic arteriovenous malformation · Acute pancreatitis · Chronic pancreatitis

Introduction

Pancreatic arteriovenous malformation (P-AVM), first reported by Halpern et al. [1], is defined as a disease that causes tumor formation or blood flow abnormality due to abnormal short-circuit anastomoses of the arterial and portal systems in the pancreas. P-AVM is a clinically important disease as it can cause gastrointestinal bleeding, acute pancreatitis, and refractory portal hypertension.

Recently, the number of reports of P-AVM has increased due to the development of various imaging modalities [2–4]. However, there are few reports of P-AVM associated with acute pancreatitis [3, 5–11], and it remains a rare pathology. Furthermore, the pathologies associated with P-AVM, such as its chronic effects on the pancreas, its association with other vascular diseases, and its modification when acute pancreatitis coexists, remain unknown.

Here, we report a case of a 60-year-old man with an unruptured cerebral aneurysm, family history of moyamoya disease, and early chronic pancreatitis, who was diagnosed with P-AVM with acute pancreatitis.

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Case report

A 60-year-old man with hypertension, dyslipidemia, and an unruptured cerebral aneurysm was admitted to our hospital due to increasing epigastric pain since the previous day's dinner. He described intermittent pain from the umbilicus to the back since his 20s, but he did not visit a hospital because this pain had always disappeared spontaneously.

His medications included amlodipine basilate, pravastatin sodium, and aspirin, and his blood pressure and dyslipidemia were well controlled. He had a history of cerebral infarction, ablation for Wolff–Parkinson–White syndrome, and percutaneous coronary intervention (stent placement) for unstable angina. He had no history of acute pancreatitis or abdominal trauma history. His alcohol consumption was 350 mL beer per day (< 20 g/day) and he smoked 20 cigarettes per day for 40 years. His son and grandson had moyamoya disease, and there was no family history of vascular disease in his wife's genealogy.

On examination, the patient's height was 164.2 cm, his weight was 60 kg, his body mass index was 22.3 kg/m^2, his pulse rate was 80 beats per minute and regular, his blood pressure was 153/90 mmHg, his temperature was 36.5 °C, and his respiratory rate was 10 breaths per minute. There were no recurrent nasal bleeding episodes or peripheral vasodilation of the lips, pharynx, fingers, or nose. Moreover, there were no surgical or trauma scars present on the abdomen, and the abdomen was soft with mild distention. Intestinal peristaltic sounds were slightly decreased. There was rebound tenderness around the umbilicus, but no muscular defense.

The laboratory data at admission were as follows: white blood cell (WBC) count, 14,500/μL; platelet count (Plt), 256×10^3/μL; pancreatic amylase, 947 U/L; lipase, 2087 U/L; elastase1, 1542 ng/dL; blood urea nitrogen (BUN), 12.3 mg/dL; creatinine (Cr), 0.78 mg/dL; lactate dehydrogenase (LDH), 173 U/L; Ca 9.1 mg/dL; C-reactive protein (CRP), 0.09 mg/dL; carcinoembryonic antigen, 5.2 ng/mL; carbohydrate antigen 19–9, 4 U/mL; immunoglobulin G, 1212 mg/dL; and immunoglobulin G4, 66.4 mg/dL. There were no hepatobiliary enzyme or electrolyte abnormalities. Arterial blood gas analyses showed a base excess of 0.8 mmol/L and a partial pressure of oxygen of 79.3 Torr on ambient air. Abdominal ultrasonography revealed mild swelling of the localized pancreas but did not reveal gallbladder stones, choledocholithiasis, or neoplastic lesions. Abdominal computed tomography (CT) revealed mild swelling of the pancreatic tail, but apparent fluid collection around the pancreas and pancreatic calcification were not shown (Fig. 1a). Arterial phase-contrast-enhanced abdominal CT showed enhancement of the pancreatic tail and confirmed abundant small blood vessel hyperplasia from the splenic artery to the inner branch of the pancreas in a network structure. A pancreatic mass was not observed (Fig. 1b). Moreover, the splenic and portal veins were intensely contrasted early in the arterial phase (Fig. 1c, d). A volume-rendered three-dimensional CT of the arterial phase showed large numbers of dilated and tortuous arteries involving the pancreatic tail and splenic vein that were well-contrasted (Fig. 1e). Abdominal magnetic resonance imaging showed localized low-signal areas of the pancreatic body and tail in T1-weighted images, and dotted and linear low-signal areas at the same sites in T2-weighted images, which suggests flow void. Magnetic resonance cholangiopancreatography did not reveal dilation of the bile duct, pancreatobiliary maljunction, or expansion of the main pancreatic duct on the pancreatic tail. Magnetic resonance angiography revealed cerebral aneurysm, but not stenosis or occlusion at the terminal portion of the internal carotid artery and abnormal vascular networks in the vicinity of the occlusive or stenotic lesions. Endoscopic ultrasonography (EUS) revealed numerous point-like non-echoic regions in the pancreatic tail in B-mode (Fig. 2a). Furthermore, abundant blood flow signals at the same site (Fig. 2b) and turbulent flow of the splenic vein were observed on color Doppler. Lobularity (honeycombing type) and hyperechoic foci (non-shadowing), which are characteristic of early chronic pancreatitis, were found in the pancreatic body (Fig. 2c). Selective angiography of the celiac artery showed abnormal small vessel growth consistent with the CT findings of the pancreatic tail, and marked increases in splenic venous return. Moreover, the development of branching between the splenic and left gastric arteries and the common hepatic artery was observed (Fig. 3). Thus, we diagnosed P-AVM with acute pancreatitis. Based on the laboratory data at the time of admission, the patient did not have severe acute pancreatitis based on the severity criteria in Japan [12]. On admission, the patient fasted and received fluid replacements, proteolytic enzyme inhibitors, and antibiotics. The laboratory data 24 h after admission were as follows: WBC, 20,500/μL; Plt, 251×10^3/μL; pancreatic amylase, 1340 U/L; BUN, 8.8 mg/dL; Cr, 0.74 mg/dL; LDH, 196 U/L; Ca 9.1 mg/dL; and CRP, 4.84 mg/dL. Moreover, the severity reassessment within 24 h remained unchanged. The clinical course is shown in Fig. 4. Although pancreatitis was medically controlled, pancreatic tail resection was performed for P-AVM. Surgical findings showed fresh adhesions associated with acute pancreatitis, and extensive and strong adhesions within the posterior wall of the stomach and transverse colon mesentery as well as increased collateral circulation observed mainly in the AVM region of the pancreatic tail. Histological findings showed increased abnormal blood vessels with

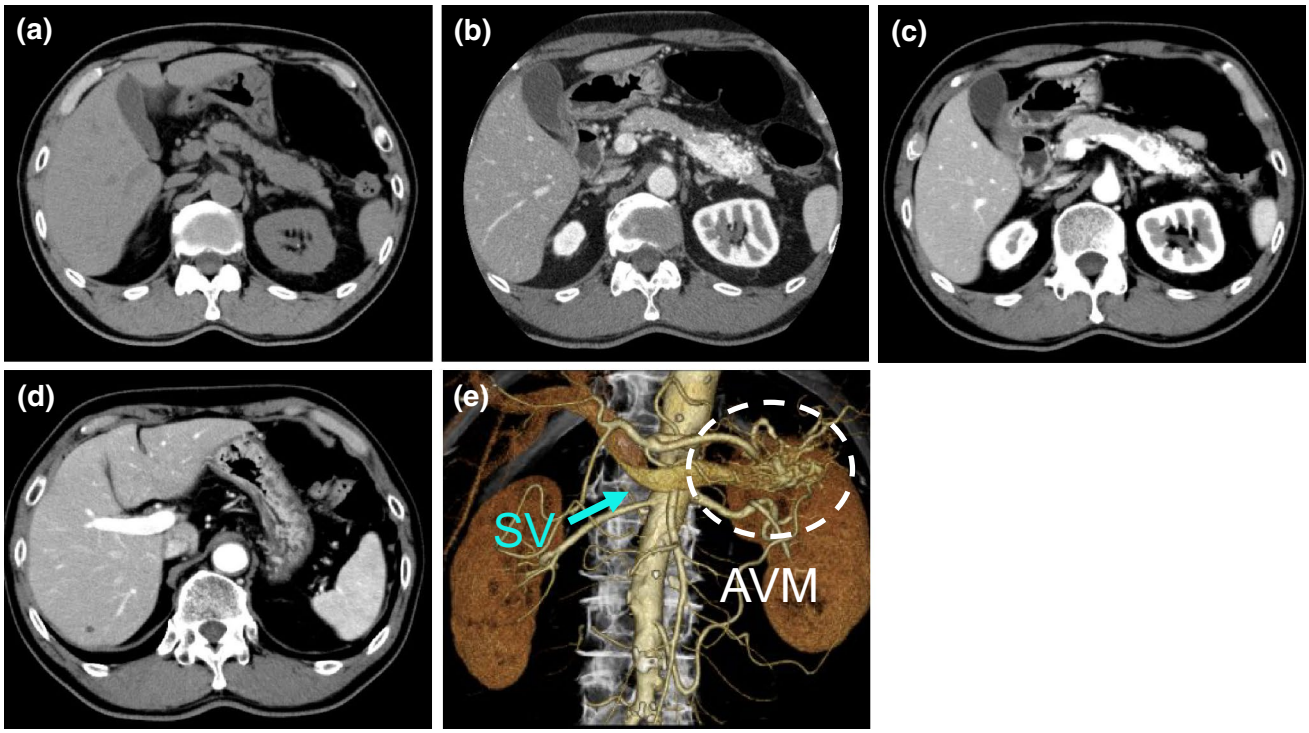

Fig. 1 Abdominal computed tomography (CT) scan. **a** Abdominal plain CT scan showing an enlarged pancreatic tail compared to the pancreatic head. No obvious fluid collection around the pancreas and pancreatic calcification was observed. **b** Arterial phase-contrast-enhanced CT scan reveals numerous areas of small blood vessel hyperplasia from the splenic artery to the inner branch of the pancreas in a network structure. **c** Arterial phase-contrast-enhanced CT scan showing a more intensely contrasted splenic vein, despite use of the arterial phase. **d** Arterial phase-contrast-enhanced CT scan showing a more intensely contrasted portal vein, despite use of the arterial phase. **e** Three-dimensional imaging showing abnormal blood vessel hyperplasia in the pancreatic tail, and a well-contrasted splenic vein (SV)

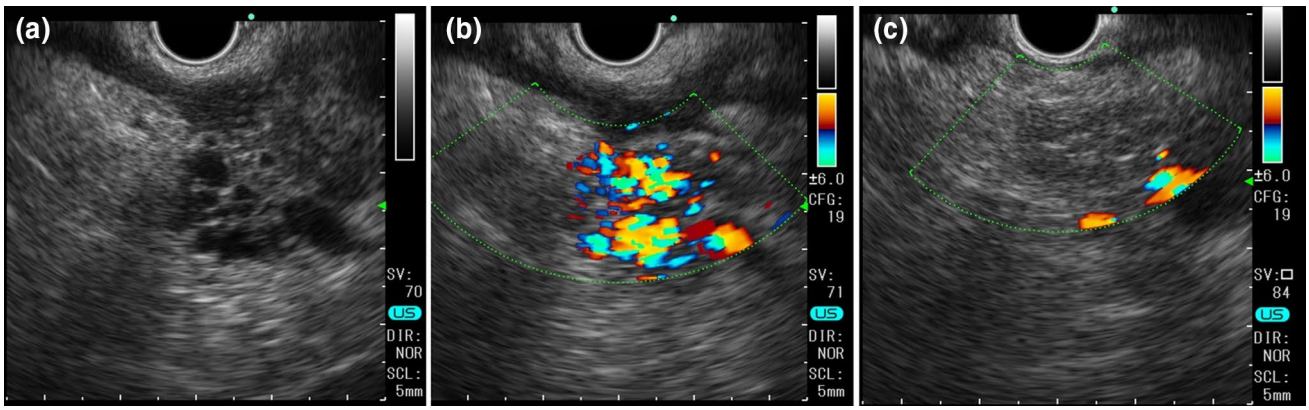

Fig. 2 Endoscopic ultrasound sonography (EUS). **a** B-mode EUS showing diffuse point-like echo-free areas in the pancreatic body and tail. **b** Color Doppler EUS showing pulsatile turbulent blood flow within the lesions in the point-like echo-free areas that diffusely appeared in the pancreatic body and tail. **c** B-mode EUS showing lobularity (honeycombing type) and hyperechoic foci (non-shadowing) in the pancreatic body

inappropriate intima and media inside the pancreas and the surrounding adipose tissue, with the anastomosis of the artery and vein, which was in line with AVM (Fig. 5a). On the pancreatic tail, fat necrosis accompanying arteries in which the intima and the media were thickened and the lumen narrowed was shown (Fig. 5b). Although invasion of foam cells and lymphocytes as well as fibrosis and parenchymal necrosis were observed, this inflammation

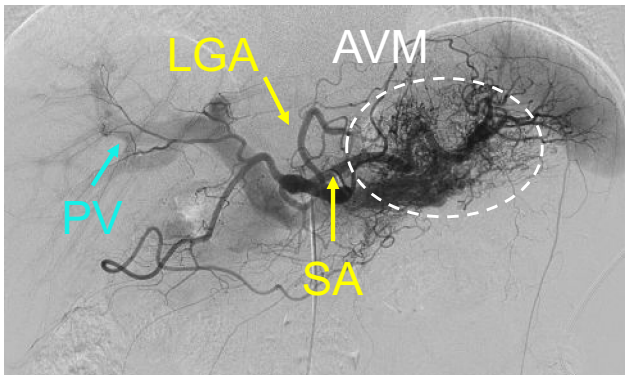

Fig. 3 Angiography. Angiography of the celiac artery showing abnormal small blood vessel growth in the pancreatic body and tail as well as splenic and left gastric artery branching from the origin of the common hepatic artery. The portal vein was contrasted despite the timing when the spleen was being contrasted. LGA, left gastric artery; SA, splenic artery; PV, portal vein; AVM, arteriovenous malformation

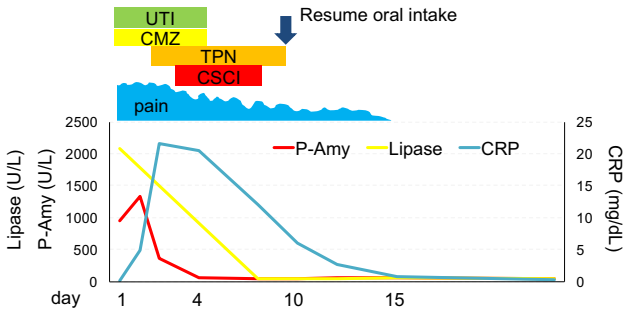

Fig. 4 Clinical course. The clinical course for a duration of 3 weeks after admission is shown. The patient was fasted upon admission and received fluid replacements, ulinastatin, and cefmetazole. On day 3, a continuous subcutaneous injection of fentanyl was started because of increasing abdominal pain. Although serum levels of CRP remained elevated, serum levels of pancreatic amylase normalized on day 4. The patient's abdominal pain gradually improved and the continuous subcutaneous injection of fentanyl was stopped on day 8. After resuming oral intake on day 10, pancreatic amylase levels did not increase and pancreatitis did not recur. UTI, ulinastatin; CMZ, cefmetazole; TPN, total parenteral nutrition; CSCI, continuous subcutaneous infusion of fentanyl

was relatively mild than this fat necrosis. In addition, the range of fat necrosis was relatively clear, and fat necrosis did not involve much in the pancreatic parenchyma. On the other hand, the parenchyma of the pancreatic body, which was compressed by P-AVM, was accompanied with mild atrophy (Fig. 5c) and fibrosis (Fig. 5d).

Discussion

We report the case of a 60-year-old man that was diagnosed with P-AVM associated with pancreatitis and might have factors of susceptibility to vascular diseases including AVM.

The origin of most P-AVM cases is congenital, stemming from a remnant of the fetal vascular system; however, P-AVM can also be acquired, involving etiologies including pancreatitis, trauma, and neoplasm [4]. Moreover, some cases are associated with hereditary hemorrhagic telangiectasia (HHT), an autosomal-dominant syndrome characterized by arteriovenous malformation of multiple organs and capillary dilation of the skin and mucosa [13]. As for etiology of P-AVM in our case, the etiology of P-AVM was thought to be congenital because of an absence in acquired factors, such as acute pancreatitis, apparent trauma, or neoplasms, as well as an absence of symptoms and a family history of HHT. However, surgical findings showed extensive and strong adhesions around the P-AVM, suggested that inflammation had been repeated. We could not deny acquired factor due to angiogenesis caused by recurrent mild idiopathic pancreatitis.

Previous reports have suggested that P-AVM sometimes causes acute pancreatitis. The mechanism underlying the development of pancreatitis associated with P-AVM is thought to be ischemia due to stealing [2, 9], bleeding into the pancreatic duct and the pancreatic parenchyma [6], and compression of the main pancreatic duct [14]. In the current case, the developed collateral circulation implicated that P-AVM had existed before the present pancreatitis occurred, and P-AVM was considered as one of the trigger of pancreatitis. In the histological findings, fat necrosis due to the vascular obstruction, caused by intimal hyperplasia of abnormal blood vessels and complicated abnormal array thickening of arterial media, were shown around the pancreatic tail. Furthermore, the range of fat necrosis was relatively clear and inflammation of the pancreatic parenchyma was mild. Therefore, it was thought that fat necrosis due to ischemic infarction caused spread of inflammation to pancreatic parenchyma and induced acute pancreatitis.

In our case, inflammation and mild fibrosis were histologically confirmed consistent with the region of pancreatic body where lobularity (honeycombing type) and hyperechoic foci (non-shadowing) were observed in EUS findings, suggested early chronic pancreatitis. The underlying etiology of present chronic pancreatitis was unlikely to be alcohol due to low consumption (< 20 g/day) [15], although a previous meta-analysis suggested that there is no obvious threshold value of alcohol consumption associated with increased risk for chronic pancreatitis [16].

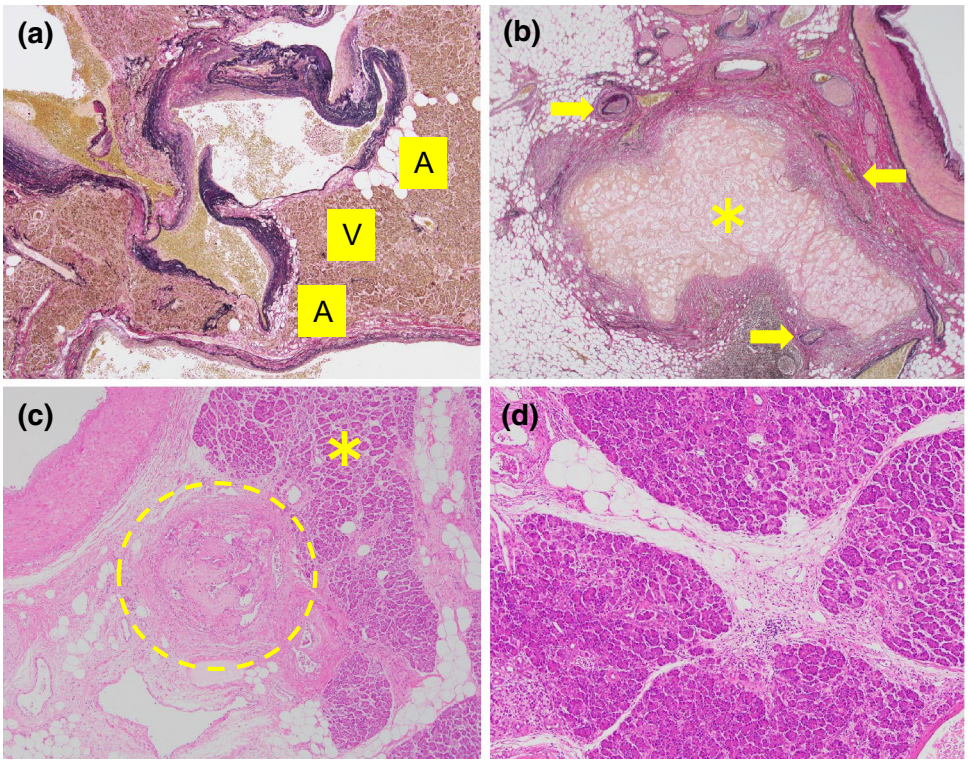

Fig. 5 Histology of pancreatic specimens. **a** Elastica van Gieson (EVG) staining (×40). An artery with thick tunica media and a vein without a medial membrane were conjugated. A, artery; V, vein. **b** EVG staining (×20). Fat necrosis was shown around the pancreatic tail (*). In addition, small arteries, in which the intima and the media were thickened and the lumen narrowed, were shown around fat necrosis (arrow). **c** Hematoxylin and eosin (HE) staining (×40). Pancreatic parenchyma was pressed by pancreatic arteriovenous malformation (broken line) and the atrophy of that region (*) was shown. **d** HE staining (×40). Mild fibrosis was observed in the pancreatic body

On the other hand, it was considered that compression or repeated inflammation by P-AVM were the causes of chronic pancreatitis. Compression by P-AVM, repeated inflammation from P-AVM or recurrent mild pancreatitis may play a similar role as persistent pathogen response to parenchymal injury or stress, caused fibrosis in chronic pancreatitis [17]. Furthermore, this repeated inflammation has a possibility to involve in fibrosis further develops, as in necrosis–fibrosis theory [18] and sentinel acute pancreatitis event theory [19]. Therefore, P-AVM may be a potential etiology of chronic pancreatitis.

Surgical treatment of P-AVM, which is considered curative, is often selected for symptomatic cases, particularly for acute pancreatitis [3, 7]. Transcatheter arterial embolization (TAE) and radiotherapy are sometimes performed in patients with impairments in their general condition [3, 4]. While treatment for asymptomatic P-AVM is controversial, early treatment has been recommended to prevent the development of refractory portal hypertension [2, 20]. In the present case, acute pancreatitis and abdominal pain were controlled using conservative management, but treatment of P-AVM was considered necessary because of a potential for the recurrence of acute pancreatitis, development of portal hypertension, progression of chronic pancreatitis, development of a refractory duodenal ulcer due to oral administration of aspirin, and ischemia. We selected surgical resection as the preferred treatment

Table 1 Clinical characteristics of P-AVM patients internationally

	Patients ($N=120$)
Age	54 (41–67)
Sex, *n* (%)[a]	
Male	101 (88%)
Female	13 (12%)
Ethnic group[b], *n* (%)	
Asia	92 (76.7%)
North America	13 (10.8%)
Europe	11 (9.2%)
South America	2 (1.7%)
Oceania	1 (0.8%)
Unknown	1 (0.8%)
Symptom, *n* (%)	
GI bleeding	43 (35.8%)
Abdominal pain	42 (35%)
Asymptomatic	19 (15.8%)
Other	5 (4.2%)
Unknown	11 (9.2%)

[a]Six cases did not have description about sex

[b]Asia; Japan (71 cases), Korea (16 cases), India (2 cases), Malaysia (1 case). Saudi Arabia (1 case), Turkey (1 case). North America; United State of America (12 cases), Canada (1 case). Europe; France (3 cases), Spain (2 cases), Italy (2 cases), Slovenia (1 case), Greece (1 case), Serubia (1 case), United Kingdum (1 case). South America; Argentine (1 case), Chile (1 case). Oceania; Australia (1 case)

Table 2 Location and treatment of P-AVM patients internationally

	Patients ($N=120$)
Location of P-AVM, *n* (%)	
Head	58 (48.3%)
Head and body	5 (4.2%)
Head and tail	10 (8.3%)
Body	2 (1.7%)
Body and tail	18 (15%)
Tail	8 (6.7%)
Whole	8 (6.7%)
Unknown	11 (9.1%)
Treatment, *n* (%)	
Surgery[a]	63 (52.5%)
TAE alone	14 (11.7%)
Radiation[b]	5 (4.1%)
TIPS[c]	2 (1.7%)
Ligature of artery	2 (1.7%)
Conservative management	21 (17.5%)
Unknown	13 (10.8%)

TAE, transcatheter arterial embolization; TIPS, transjugular intrahepatic portosystemic shunt

[a]Six patients had surgery after TAE

[b]One patients had radiation after TAE

[c]Two patients had TIPS after TAE

because it was difficult to embolize multiple feeders by TAE and the patient's general condition was good.

In total, 119 cases of P-AVM have been reported in Pubmed from 1950 to 2017, based on searches using "arteriovenous malformation" and "pancreas" as keywords. The clinical features of 120 cases, including those of the current patient, are summarized in Tables 1 and 2. The median age at diagnosis was 52 years (range 7 months to 75 years) and 88% of patients were male. The number of reports from Asia was the largest, with a frequency of 76.7% (92/120). There were 71 cases in Japan and 16 cases in Korea, accounting for the majority of cases in Asia (87/92, 95%). This disease distribution is similar to that of moyamoya disease, which is common in Asia, particularly Japan and Korea [21]. In Japan and Korea, 90% [22] and 76% [23] of patients with moyamoya, respectively, have mutations in RNF 213 p.R 4810, a major founder mutation. Since the patient had a family history of moyamoya disease and similarity of disease distribution in our case, patients with P-AVM may have a founder mutation, which is similar to moyamoya disease. Gastrointestinal bleeding and abdominal pain were the major clinical symptoms that triggered diagnosis. Of the 120 patients described in the literature, 62 (52.5%) underwent surgical resection, 14 (11.7%) underwent TAE, and 21 (17.5%) underwent conservative medical treatment. Twenty-four cases were diagnosed with P-AVM during examination of their acute pancreatitis, as in our case. Among them, 15 cases that could be analyzed in detail in the English literature are summarized in Table 3. The median age was 54 years (range 41–75 years) and all patients were male. P-AVM was located in the head of pancreas in five patients, in the head and body in one patient, in the body in two patients, in the body and tail in two patients, in the tail in four patients, and in the whole pancreas in one patient. Of these 15 patients, 3 cases were described as having chronic

Table 3 Summary of cases diagnosed as P-AVM triggered by abdominal pain due to acute pancreatitis internationally

No.	References	Year	Ethnic	Age	Sex	Symptoms	Location	CP	Treatment
1	[5]	1982	USA	52	Male	Abdominal pain	Head	No	Embolization, PD
2	[6]	1992	Japan	60	Male	Left hypochondria	Tail	No	DP
3	[7]	2001	Japan	63	Male	Abdominal pain	Body and tail	No	DP
4	[8]	2002	Japan	54	Male	Upper abdominal pain	Head	No	PD
5	[2]	2006	Japan	46	Male	Epigastric pain	Head	+	PD
6	[2]	2006	Japan	44	Male	Epigastric pain	Body	+	DP
7	[3]	2012	Korea	64	Male	Epigastric pain	Entire	No	TP with portal vein resection
8	[3]	2012	Korea	45	Male	Epigastric pain	Body	No	MP
9	[3]	2012	Korea	56	Male	Epigastric pain	Tail	No	DP
10	[3]	2012	Korea	46	Male	Abdominal pain	Tail	No	Laparoscopic DP
11	[3]	2012	Korea	51	Male	Epigastric pain	Tail	No	DP
12	[9]	2015	Japan	41	Male	Epigastric pain	Head	No	PD
13	[10]	2015	France	56	Male	Upper abdominal pain	Head	No	TAE
14	[11]	2016	USA	54	Male	–	Head and body	No	TAE
15		2017	Japan	67	Male	Upper abdominal pain	Body and tail	+	DP

USA, United State of America; CP, Chronic pancreatitis; no, no description; PD, pancreatoduodenectomy; DP, distal pancreatectomy; TP, total pancreatectomy; MP, middle pancreatectomy

pancreatitis, and the remaining 12 cases were not assigned a diagnosis. Thirteen patients underwent surgical resection while two underwent TAE. TAE was chosen in these patients due to a severe case of pancreatitis with accompanying myocardial infarction [10] and due to the development of hemorrhagic shock because of massive gastrointestinal bleeding after treatment in the other patient [11]. The other cases of P-AVM with pancreatitis received curative surgical treatment.

In conclusion, we report a case of P-AVM with acute pancreatitis that contribute to chronic pancreatitis. Patients with symptomatic P-AVM should be considered a chronic and progressive disease, and require curative surgical treatment to avoid life-threatening pathology.

Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest.

Ethical standards All procedures followed have been performed in accordance with the ethical standards laid down in the 1964 Declaration of Helsinki and its later amendments.

Informed consent Informed consent was obtained from the patient for being included in the study.

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