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【 CASE REPORT 】

Metachronous Rupture of Pancreatoduodenal Artery Aneurysm with Median Arcuate Ligament Syndrome: A Case Report and Review of 11 Cases

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Abstract:
Median arcuate ligament syndrome (MALS) involves narrowing of the celiac artery root from MAL compression, leading to pancreatoduodenal artery aneurysm (PDAA) due to increased retrograde blood flow from the superior mesenteric artery into the PDA. We encountered a case in which coil embolization was performed for PDAA rupture due to MALS. Four years later, a second PDAA occurred and ruptured, necessitating coil reembolization. There have been no reports of recurrence during long-term follow-up after PDAA treatment in patients with MALS. We herein report a rare case of metachronous PDAA rupture in the context of MALS. The relevant literature and 11 PDAA/MALS cases are discussed.

Key words: median arcuate ligament syndrome (MALS), pancreatoduodenal artery aneurysm (PDAA), coil embolization

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Introduction

Median arcuate ligament syndrome (MALS) is attributed to extrinsic compression of the vasculature and perineural ganglia. Its diagnosis is primarily made through computed tomography (CT), magnetic resonance imaging, and angiography. Treatment includes surgical dissection of the MAL and angioplasty of the celiac artery; however, no definition or standardized treatment approach has been established (1).

Pancreatoduodenal artery aneurysms (PDAAs) are rare. Visceral aneurysms are estimated to occur in 0.1-0.2% of the population. Splenic aneurysms are the most common type, accounting for 60% of cases, whereas PDAA is reported in 2-16% of cases (2, 3). PDAA has been associated with MALS, pancreatitis, and atherosclerosis. MALS is considered to be the most common cause and is thought to be induced by hemodynamic stress. Abdominal pain is the most frequently reported symptom in patients with PDAA rupture. Contrast-enhanced CT is a useful diagnostic tool for differentiating PDAA from acute pancreatitis (4). The mortality rate for ruptured cases is reported to be 10-21% or higher (5). Given the high mortality rate, prompt treatment should be considered upon the diagnosis. However, standardized treatment strategies have not yet been established for PDAA associated with MALS, similar to the lack of consensus regarding the treatment of MALS alone (6).

Short-term recurrence of visceral aneurysms after PDAA treatment in MALS has been reported (7, 8). We herein report a rare case of PDAA rupture in a patient with MALS that occurred four years after an initial rupture. Eleven cases of PDAA in the context of MALS encountered in the last 11 years at our hospital were reviewed and discussed.

Case Report

Previous episodes

A 50-year-old man visited a medical clinic approximately 4 years prior to the most recent admission because of a sud-

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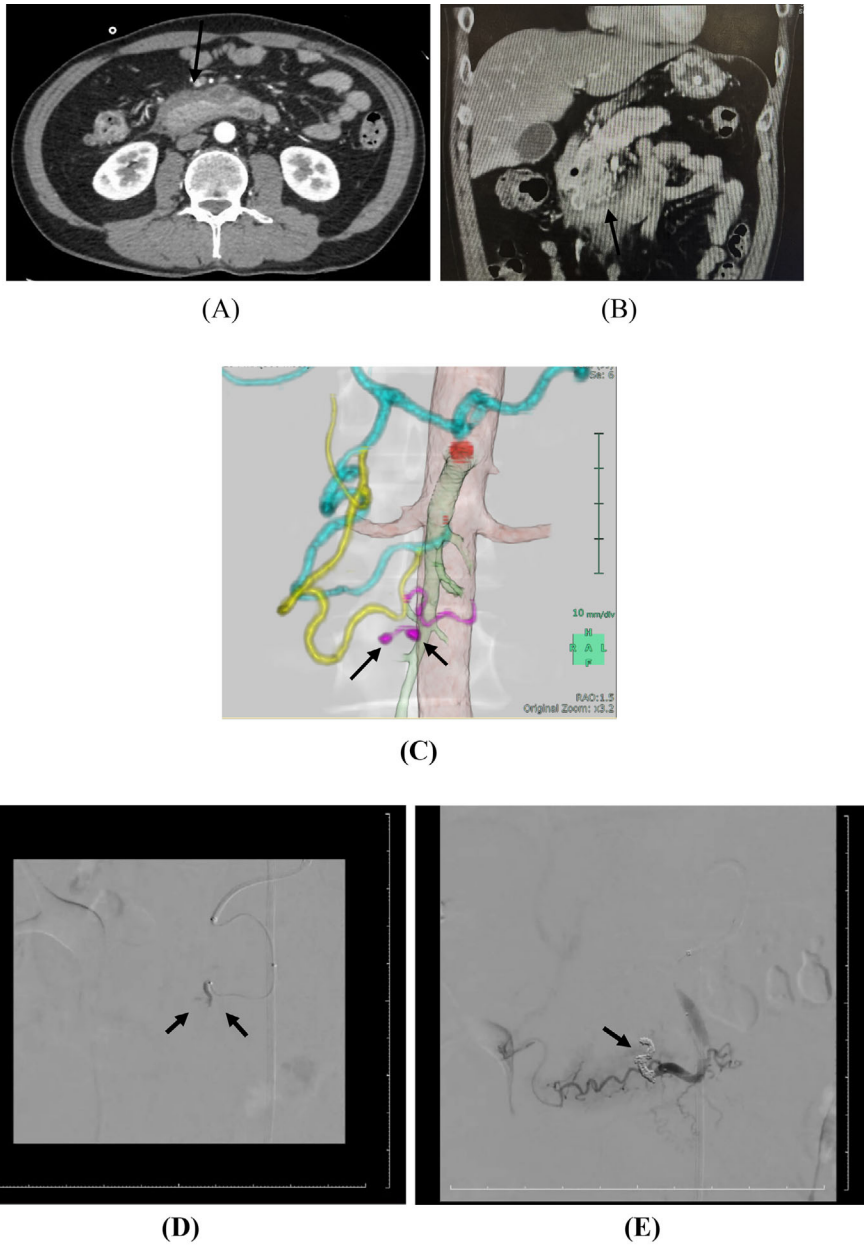

Figure 1. **(A-E) Contrast-enhanced computed tomography and an angiogram on admission with the first episode occurring approximately four years prior to the more recent episode.**

den onset of epigastric pain, with subsequent vomiting and difficulty with oral intake 1 week later. CT revealed a 5-cm cystic lesion around the third portion of the duodenum, which was causing obstruction. The patient was then transferred to our hospital for a further examination. Contrast-enhanced CT revealed two small aneurysms in the branches of the anterior inferior pancreaticoduodenal artery, which were in communication with the first jejunal artery. These findings also included stenosis at the origin of the celiac artery, leading to a diagnosis of retroperitoneal hematoma due to rupture of the PDAA associated with MALS. Abdominal angiography revealed similar findings, and arterial coil embolization was performed. After treatment, no rebleeding occurred, and the hematoma shrank; the patient was then able to resume oral ingestion and was subsequently discharged from the hospital (Fig. 1).

Present illness

Approximately 4 years later, the then 54-year-old patient presented to an outside medical clinic complaining of the same symptoms as those of the previous admission. CT performed at the clinic revealed retroperitoneal hemorrhaging, and the patient was transferred to our hospital. Upon admission, his vital signs were stable, and laboratory data were unremarkable (Table 1).

Contrast-enhanced CT showed an 8-cm hematoma in the retroperitoneum of the pancreatic head and third portion of the duodenum. In addition, the vessel wall of the anterior superior pancreaticoduodenal artery (ASPDA) was irregular and a pseudoaneurysm was suspected. Abdominal angiography revealed a protrusion in the ASPDA, which appeared to be a pseudoaneurysm, leading to the diagnosis of PDAA

Table 1. Laboratory Results of the Case Report Patient.

	X-4 years	X years
Peripheral blood cell		
WBC	7.8	13.5 $\times 10^3$/μL
RBC	558	468 $\times 10^4$/μL
Hb	15.8	13.7 g/dL
Plt	45.9	24.5 $\times 10^4$/μL
Blood chemistry		
Total protein	7.7	7 g/dL
Alb	4.0	3.9 g/dL
T-Bil	2.0	0.8 mg/dL
D-Bil	1.0	0.2 mg/dL
AST	145	24 U/L
ALT	375	24 U/L
LD	194	U/L
ALP	362	168 U/L
γ-GTP	118	22 U/L
Amylase	115	123 U/L
Lipase	108	77 U/L
BUN	15.1	12.5 mg/dL
Cre	0.95	0.78 mg/dL
CRP	0.55	0.5 mg/dL

WBC: white blood cell, RBC: red blood cell, Hb: hemoglobin, Plt: platelets, Alb: albumin, T-Bil: total bilirubin, D-Bil: direct bilirubin, AST: aspartate transferase, ALT: alanine transaminase, LD: lactate dehydrogenase, ALP: alkaline phosphatase, γ-GTP: gamma-glutamyl transpeptidase, BUN: blood urea nitrogen, Cre: creatinine, CRP: C-reactive protein

rupture.

Prompt arterial coil embolization was performed (Fig. 2). A double elementary diet tube was placed and enteral nutrition was initiated. The patient had repeated pseudoaneurysm ruptures, and MAL transection was performed.

Prior to ligamentotomy, selective celiac artery angiography showed retrograde contrast flow with no contrast in the gastroduodenal artery (GDA). Selective contrast of the superior mesenteric artery (SMA) revealed the GDA, and the common hepatic, splenic, and left gastric arteries were contrasted as well (Fig. 3).

Selective celiac artery angiography after ligamentotomy revealed progressive contrast in the GDA, with some dilation of the celiac artery root (Fig. 4). The celiac artery was more dilated after surgery than before surgery (Fig. 5).

Postoperatively, the patient had no abdominal pain, resumed eating and drinking, and was discharged from the hospital three days after surgery without any particular symptoms. The patient continued follow-up on an outpatient basis, and CT and blood tests were performed. To date, no recurrence has been observed for 22 months.

Discussion

MALS is the most common cause of PDAA formation, accounting for 175 of 258 reported PDAA cases (9). The mechanism by which MALS leads to PDAA formation is attributed to stress on the vessel wall as hepatic blood flow is maintained from the SMA through the pancreatic arcade (8).

Among visceral aneurysms, gastric and gastroepiploic artery aneurysms, GDAA, and PDAA are considered candidates for treatment because of their high risk of rupture, irrespective of their size. Coil embolization is the primary treatment recommended for both unruptured and ruptured aneurysms (10).

Although coil embolization for PDAA in the context of MALS is increasingly frequently recognized as the standard of care and primary treatment choice, subsequent interventions for MALS remain debatable.

To our knowledge, there are currently no established criteria for MAL resection as a primary treatment for MALS.

Previous research has included cases of PDAA rupture wherein stenting or ligamentotomy was performed along with successful coil embolization for MALS. In cases of unsuccessful transcatheter arterial embolization (TAE), some patients are referred for bypass surgery or other alternative procedures, such as covered stenting or stent-assisted coil embolization (8, 9).

Reportedly, some aneurysms disappear after MAL resection alone without other direct interventions; therefore, some studies have recommended MAL resection alone (11). However, following interventions for ruptured aneurysms, investigators have reported no new aneurysms or rebleeding even without MAL transection (12).

Considering the mechanism of aneurysm formation in PDAAs associated with MALS, reformation may occur if the MAL is not removed. However, to our knowledge, there have been no other reported cases of metachronous rupture of PDAA secondary to MALS.

We herein reported a case of PDAA secondary to MALS that was encountered at our hospital. Of the 11 cases of PDAA with MALS encountered over the past 11 years, 3 were unruptured, and 8 were ruptured (Table 2; Supplementary material 1). One unruptured case remained under observation, whereas the others underwent MAL resection without TAE.

One patient who underwent surgery (MAL resection and percutaneous transluminal angioplasty) exhibited multiple aneurysms in the pancreatic arcade and dorsal pancreatic arteries. However, coil embolization was not performed at the patient's request. In eight ruptured cases, TAE was performed with no failures; three of these cases underwent MAL resection, and the remaining five were followed up. Among the five patients without recurrence who were followed up, two died. One patient was initially diagnosed with subarachnoid hemorrhaging and a ruptured inferior PDAA but later died of septic shock due to a perforated duodenal ulcer during hospitalization. Another patient was admitted for stroke and was subsequently found to have a ruptured posterior superior PDAA. The patient developed obstructive cholangitis with a hematoma, which led to death. One pa-

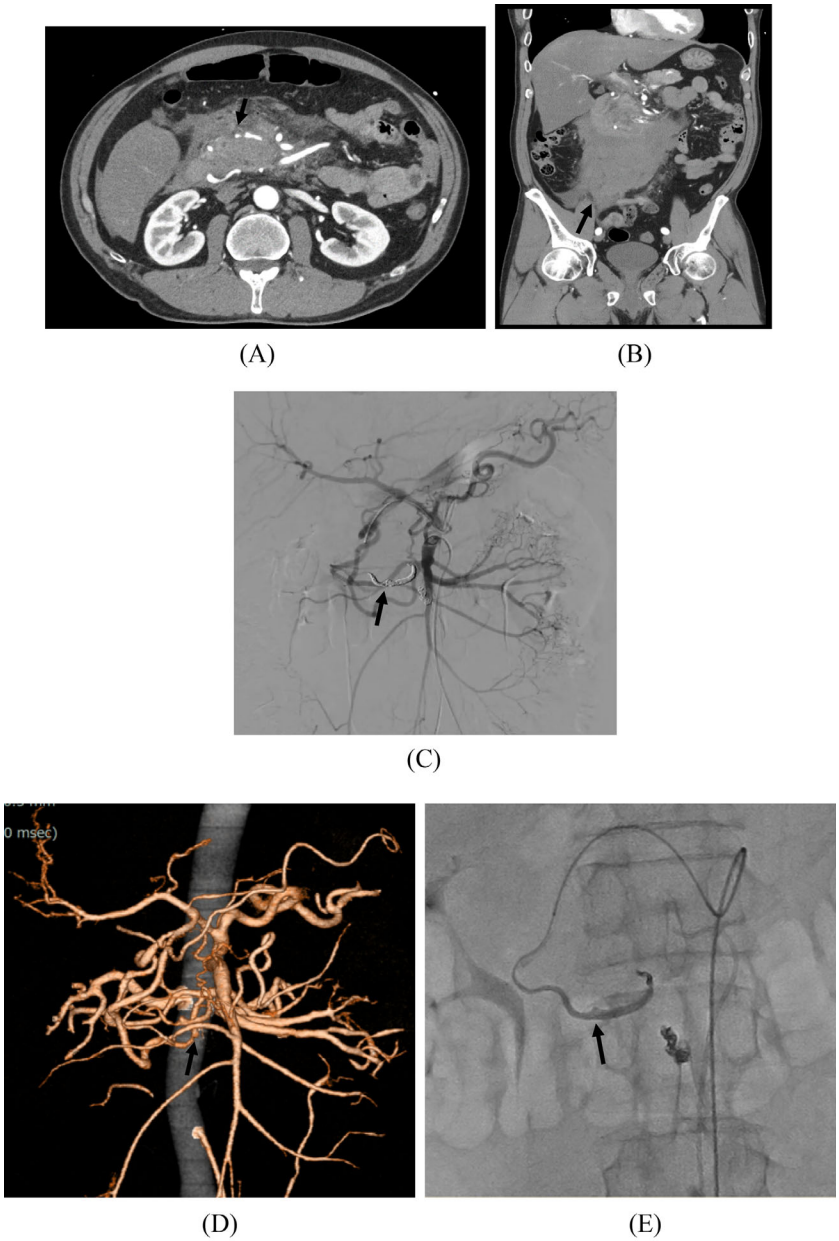

Figure 2. **(A-E) Contrast-enhanced computed tomography and an angiogram on current admission (approximately four years after the initial episode and admission).**

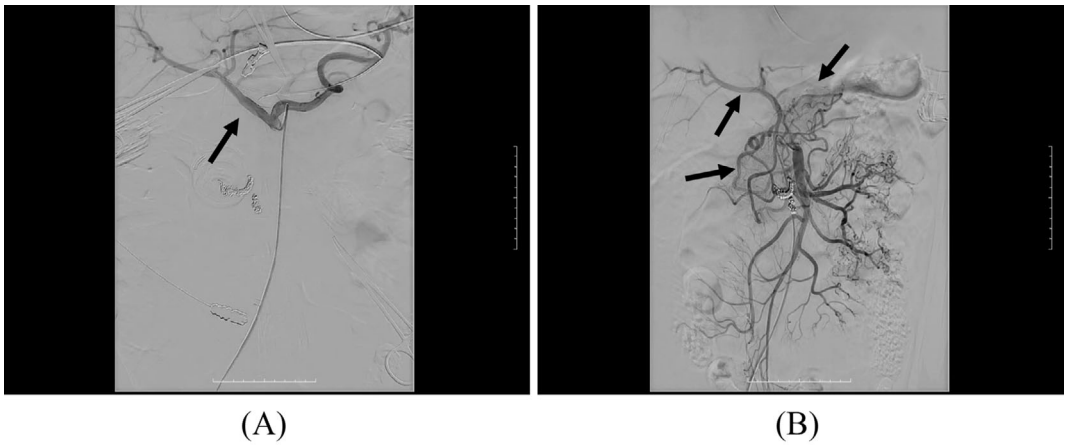

Figure 3. **(A) An angiogram via the celiac artery before ligamentotomy and (B) an angiogram via the superior mesenteric artery before ligamentotomy.**

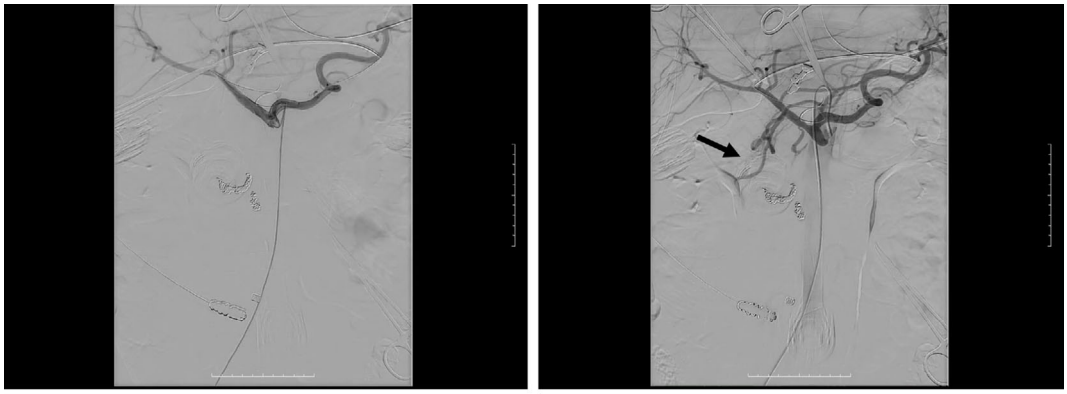

Figure 4. **(A) An angiogram via the celiac artery before ligamentotomy and (B) after ligamentotomy.**

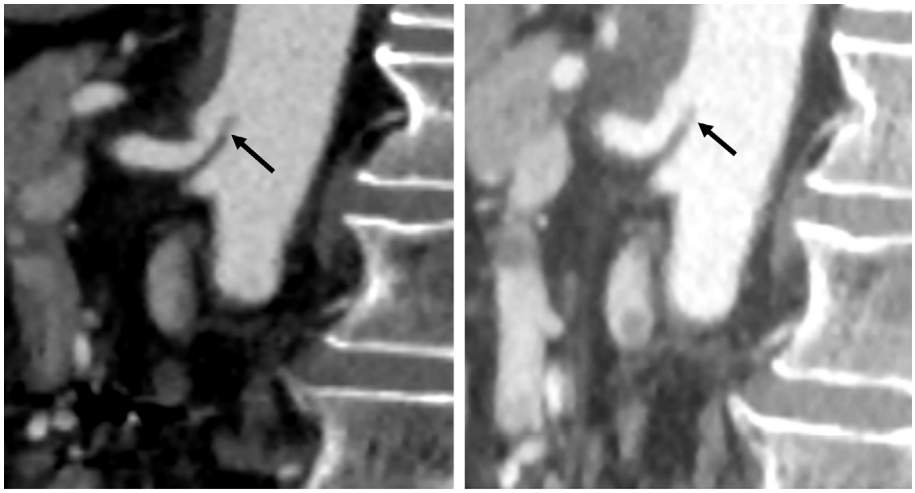

Figure 5. **(A) Contrast-enhanced computed tomography of the celiac artery before ligamentotomy and (B) after ligamentotomy.**

tient showed spontaneous improvement in celiac artery compression without any specific intervention after TAE. Three patients who underwent MAL resection are currently under observation.

The reasons for considering and performing surgery for MALS in the aforementioned cases included a second rupture, as in the present case, multiple aneurysms, contrast from the SMA showing reflux into the GDA and splenic artery aneurysm, and a relatively localized site of stenosis at the root of the celiac artery. MAL transection alone did not improve the hemodynamics in some cases. Additional stenting or balloon dilation was necessary and resulted in hemodynamic improvement in three of the six cases, as confirmed by CT or angiography.

Many studies have reported that the long-term prognosis after MAL release is good (13, 14), and the previous case of rupture of a residual PDAA after MAL resection also occurred because coil embolization had not been performed before PDAA rupture.

Although this is the first case of recurrence during long-term follow-up after PDAA treatment in MALS, MAL resection after aneurysm treatment should be considered to prevent recurrence, given the high mortality rate.

Conclusion

Long-term follow-up is necessary for PDAA secondary to MALS because it can recur and rupture later in life. Owing to the possibility of re-rupture of the aneurysm, the decision to proceed with resection should be based on the individual risk assessment for each case.

Written informed consent was obtained from the patient.

The authors state that they have no Conflict of Interest (COI).

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Table 2. Review Summary of Case Patients' Demographic and Other Information.

Patient number	Age/ sex	Past medical history	Symptoms	Initial diagnosis	Diagnosis	TAE	Management for MALS	Follow-up	Recurrence
1	68M	Hypertension Appendectomy Sigmoid colon cancer	None	DPAA	DPAA	No	Operation + PTA	120 months	None
2	75F	Hyperlipidemia Rectal carcinoma Thyroid cancer	None	IPDAA	IPDAA	Yes	None	3 months	None
3	68M	Gastric cancer Prostate cancer Lumber hernia Appendicitis	None	GDAA, IPDAA	GDAA, IPDAA	No	Operation + PTA	Until 60 months	None
4	52M	None	Abdominal pain	Pancreatitis	MCA, IPDA rupture	Yes	None	24 months	LCAA 15 days after TAE
5	53M	Appendicitis	Abdominal pain	Retroperitoneal hematoma	IPDAA rupture	Yes	Operation	Until 12 months	None
6	74M	Stroke Diabetes mellitis Sigmoid colon cancer Abdominal incisional hernia Cholecystitis Prostate cancer	Fever, hematemesis	PSPDAA rupture	PSPDAA rupture	Yes	None	Died	
7	57M	Hypertension Sleep apnea syndrome	Abdominal pain	Pancreatitis, intra-abdominal hematoma	PIPDA rupture	Yes	None	36 months	None
8	66M	None	Abdominal pain	ASPDA rupture	ASPDA rupture	Yes	Operation + balloon	12 months	None
9	52M	Hepatitis B	Abdominal pain, confusion	Intra-abdominal hematoma	RGEAA rupture, multiple GDAA	Yes	Operation	1 month	None
10	62M	Hypertension Prostate cancer ADPKD Glaucoma Parkinson disease Brain artery aneurysm Diverticulitis	Altered level of consciousness	SAH, IPDAA rupture, ruptured duodenal ulcer	IPDAA, CHAA, SMAA, SpAA	Yes	None	Died	-
11	50M	None	Abdominal pain	Intra-abdominal hematoma	AIPDAA rupture	Yes	Operation after second rupture	6 months	ASPDA 4 years after initial rupture

M: male, F: female, DPAA: dorsal pancreatic artery aneurysm, PDAA: pancreatoduodenal artery aneurysm, MALS: median arcuate ligament syndrome, PTA: percutaneous transluminal angioplasty, IPDAA: inferior PDAA, GDAA: gastroduodenal artery aneurysm, MCA: middle colic artery, LCAA: left colic artery aneurysm, TAE: transcatheter arterial embolization, PSPDAA: posterior superior pancreatoduodenal artery aneurysm, PIPDA: posterior inferior pancreatoduodenal artery, ASPDA: anterior superior pancreatoduodenal artery, RGEAA: right gastroepiploic artery aneurysm, ADPKD: autosomal dominant polycystic kidney disease, SAH: subarachnoid hemorrhage, CHAA: common hepatic artery aneurysm, SMAA: superior mesenteric artery aneurysm, SpAA: splenic artery aneurysm: AIPDAA: anterior inferior pancreatoduodenal artery aneurysm

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