Pancreatic arteriovenous malformation (AVM) is a rare condition defined as a tumorous formation or vascular anomaly arising from an aberrant bypass anastomosis of the arterial and venous systems in the pancreas (1). Esophageal varix bleeding by portal venous hypertension is the most common complication of pancreatic AVM. Duodenal ulcer bleeding or bleeding from the pancreatic AVM itself are other causes of bleeding associated with this disease entity. Pancreatic AVM presenting as retroperitoneal bleeding is an extremely rare clinical condition and is often misdiagnosed as a pseudoaneurysm. Treatment may include ligation of the afferent artery, embolization of feeding vessels, portacaval shunting, or surgical resection of the affected part or the entire affected organ (2).

We report a case of pancreatic arteriovenous malformation presenting mesenteric and peripancreatic hemorrhages. A 62-year-old man presented to the emergency department of our hospital with acute periumbilical pain. Diffuse mesenteric and peripancreatic hemorrhages and an approximately 1 cm aneurysmal sac were observed on contrast-enhanced computed tomography. Celiac and superior mesenteric angiography revealed pancreatic arteriovenous malformation, which was then treated with transcatheter arterial embolization using n-butyl-2-cyanoacrylate. Complete obliteration of pancreatic arteriovenous malformation was evident on a follow-up computed tomography performed 2 months later.

Index terms
Arteriovenous Malformation
Pancreas
Hemorrhage
Therapeutic Embolization

CASE REPORT

A 62-year-old man presented to the emergency department of our hospital with acute abdominal pain. He had no medical history of alcohol abuse and no history of trauma. He experienced sudden-onset of severe periumbilical pain during defecation the morning on the day of admission. His vital signs were stable. Physical examination revealed mild periumbilical and rebound tenderness. Laboratory data showed a mild increase in white blood cell count (11600/µL) and increased lactate dehydrogenase level (938 IU/L). His hemoglobin, alanine aminotransferase, aspartate aminotransferase, amylase, and lipase levels were normal. A dynamic abdominal computed tomography (CT) scan showed diffuse hemorrhagic infiltration of the mesenteric root and peri-pancreatic area with compression of the superior mesenteric vein. In addition, a 1-cm nodular enhancing lesion was found.
adjacent to the superior mesenteric artery (SMA) and dorso-medial to the pancreatic head, which was prospectively diagnosed as a pseudoaneurysm of the mesenteric artery with mesenteric and peripancreatic hemorrhages (Fig. 1A). He was referred to the intervention department for further evaluation. Selective superior mesenteric arteriography revealed a nodular nidus at the pancreatic head that was fed by a small branch of the inferior pancreaticoduodenal artery with a drainage vein to the upper superior mesenteric vein and early visualization of the portal vein (Fig. 1B, C). Another feeder of AVM was the dorsal pancreatic artery, branching from the proximal common hepatic artery just distal to the splenic artery bifurcation on celiac arteriogram (Fig. 1D, E). These findings were consistent with a pancreatic AVM. Owing to the relatively large extent of the hemorrhage and the presence of only two feeding arteries, endovascular treatment was considered to avoid invasive surgical excision. The feeding artery via the small branch of the inferior pancreaticoduodenal artery was easily super selected with a 2.4-Fr microcatheter (Progreat, Terumo, Tokyo, Japan) through the SMA approach. However, the other feeding artery via the dorsal pancreatic artery was successfully catheterized retrogradely through the pancreaticoduodenal arcade, after a failed attempt through the celiac approach due to the very acute origin of the target artery. The feeding arteries were embolized with injection of NBCA (Histoacryl, B. Braun, Melsungen, Germany) diluted 1:4 (first artery) and 1:2 (second artery) with iodized oil (Lipiodol, Guerbet, Aulnay-Sous-Bois, France). A post-procedure angiography revealed complete exclusion of the feeding arteries, nidus, and aberrant draining vein (Fig. 1F; G). The post-procedure course was uneventful, and the patient's symptoms abated after the procedure. A follow-up CT scan obtained 2 months after the procedure showed complete disappearance of the pancreatic AVM and mesenteric hemorrhage (Fig. 1H).

**DISCUSSION**

Pancreatic AVM is a rare condition defined as a tumorous formation or vascular anomaly arising from an aberrant bypass anastomosis of the arterial and venous systems in the pancreas (1). Halperrn et al. (1) first reported a patient with pancreatic AVM associated with Rendu-Osler-Weber disease in 1968. Pancreatic AVM is classified as a congenital anomaly thought to be caused by failure of the regulatory sphincteric mechanism of the arteriole-capillary junction, resulting in unrestricted overflow of arterial blood into the capillary bed, or by acquired disease complicated by trauma, inflammation, or tumor.

Pancreatic AVM has various clinical manifestations, which may be asymptomatic or include abdominal pain, pancreatitis, and/or gastrointestinal (GI) bleeding. Nishiyama et al. (2) reviewed the cases of 42 patients with pancreatic AVM. They reported that 50% of patients presented with life-threatening GI bleeding. The AVM was located in the pancreatic head in 56% of the patients, 31% of whom also had an extrapancreatic AVM (2). Makhoul et al. (3) classified the bleeding mechanisms of pancreatic AVM into two categories as follows: first, bleeding from a ruptured esophageal or gastric varix secondary to portal hypertension induced by the AVM; and second, hemorrhage from direct erosion of the AVM into the pancreatic duct or through the adjacent intestinal mucosa as a duodenal ulcer. However, pancreatic AVM presenting as retroperitoneal bleeding has been rarely documented. In the present case, the hemorrhage was present in the small bowel mesentery and peripancreatic retroperitoneal space. As the patient's pain developed acutely during defecation, we assume that this rare instance of retroperitoneal bleeding was the result of an abrupt increase in portal venous pressure induced by increased intraperitoneal pressure.

Doppler ultrasonography is a non-invasive and easily repeated modality that can be performed at a patient's bedside. Pancreatic AVM appears on Doppler ultrasonography as mosaic lesions composed of pulsatile waves, but ultrasonography has limitations in revealing the full extent of the lesion and its relation to adjacent organs. The characteristic CT findings of AVM include nodular enhancement and early appearance of the portal venous system. Although nodular enhancement and early appearance of the portal venous system were observed in 76.9% and 66.7% of patients, respectively, in one study (4), some cases demonstrated uncharacteristic imaging findings that led to misdiagnosis as a pseudoaneurysm or a true aneurysm, as in the present case. Magnetic resonance imaging (MRI) shows a tangle of tubular structures with a signal void on T2-weighted image for high blood flow and hyperintense enhancement of tubular structures on enhanced arterial phase, as a characteristic feature of AVM (5). However, MRI is not a widely used imaging modality for cases of acute bleeding. Dilated and tortuous feeding arteries, racemose
staining in the nidus, and early appearance of the portal venous system have been reported as characteristic angiographic findings of AVM (4).

The treatment methods for pancreatic AVM are surgery (57.1%); nonsurgical therapy, including embolization, irradiation, or a portovenous shunt (14.3%); and no treatment (28.6%) (2). Surgery is the least preferable treatment method because total extirpation of the affected organ, or at least the involved portion, is the only way to assure a complete cure and eliminate the possibility of recurrent bleeding. However, excision of the pan-

**Fig. 1.** Transcatheter arterial embolization of pancreatic arteriovenous malformation in a 62-year-old man presenting as retroperitoneal bleeding.

A. Portal-phase dynamic computed tomographic image shows an enhanced nodular vascular lesion with mesenteric and peripancreatic hemorrhages adjacent to the pancreatic head (arrowheads). The superior mesenteric vein is compressed by hematoma (arrow).

B. A selective superior mesenteric arteriogram shows a nodular nidus at the pancreatic head that is fed by a small branch of the inferior pancreaticoduodenal artery (arrow).

C. Delayed superior mesenteric arteriogram shows the tortuous drainage vein (arrowhead) to the superior mesenteric vein and early visualization of the portal vein (arrow).

D. A selective celiac arteriogram shows a second feeding artery through the dorsal pancreatic artery that originates from the proximal common hepatic artery just distal to the splenic artery bifurcation (arrow).

E. A selective dorsal pancreatic arteriogram through the pancreaticoduodenal arcade shows early visualization of the portal vein (arrow).

F. A post-procedural angiogram shows complete exclusion of the feeding arteries, nidus, and aberrant draining vein.

G. Indirect portogram after procedure shows patent portal venous flow despite spreading of several NBCA droplets in liver.

H. A computed tomographic scan obtained 2 months after the procedure, shows complete embolization of the pancreatic arteriovenous malformation and improved retroperitoneal hemorrhage.

NBCA = n-buty1-2-cyanoacrylate
Transcatheter Arterial Embolization of Pancreatic Arteriovenous Malformation Presenting as Retroperitoneal Bleeding

In conclusion, we report a rare case of pancreatic AVM presenting as retroperitoneal bleeding, which was successfully treated with TAE without any complications.

REFERENCES

췌장 동정맥기형에 의한 후복막강 출혈: 사례 보고

지승우1 · 강웅래1* · 김영환2

급성 복통으로 내원한 62세 남자의 췌장 동정맥기형에 의한 출혈을 경동맥색전술로 치료한 증례에 대해 보고하고자 한다.

조영증강 전산화단층촬영에서 장간막과 췌장주위의 출혈과 함께 1 cm 크기의 동맥류성 낭이 발견되었다. 복강동맥과 위 장간막 동맥혈관조영술에서 췌장 동정맥기형이 발견되었고 n-butyl-2-cyanoacrylate를 사용하여 경동맥 색전술을 시행하였다. 2개월 후 추적 전산화단층촬영에서 췌장 동정맥기형의 완전한 소실을 확인하였다.

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