—Case Reports—

Pancreatic Arteriovenous Malformation Involving Adjacent Duodenum with Gastrointestinal Bleeding: Report of a Case

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Abstract

A 54-year-old man was admitted to our hospital with the symptoms of palpitation, dyspnea, and tarry stool. Upper gastroduodenal endoscopy revealed submucosal lesions with vascular ectasia in the second part of the duodenum. Dynamic computed tomography (CT) detected a hypervascular lesion in the pancreatic head and the duodenum. Selective angiography showed proliferation of a vascular network and early filling of the portal vein at the early arterial phase. With a diagnosis of pancreatic arteriovenous malformation (AVM), we performed pylorus-preserving pancreaticoduodenectomy. At laparotomy, localized and meandering vessels were seen on the surface of the head of the pancreas. Histological examination showed dilated tortuous vessels accompanied by severed elastic fibers in the vessel media and blood clot formation. The incidence of pancreatic AVM remains extremely low, and recurrent gastrointestinal bleeding is a frequent complication. To prevent recurrent bleeding and progressive portal hypertension, surgery may be the definitive management of symptomatic AVM.
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Key words: pancreatic arteriovenous malformation, gastrointestinal bleeding

Introduction

The number of reported cases of arteriovenous malformation (AVM) in digestive organs has recently increased in recent years because of the widespread use of imaging techniques such as angiography and color Doppler ultrasonography; however, the incidence of pancreatic AVM remains extremely low. Since the first description by Halpern et al.[1] in 1968, about 80 cases of pancreatic AVM have been reported in the English and Japanese literatures[2–4]. We report a case of pancreatic AVM involving the adjacent duodenum with gastrointestinal bleeding, and review the literature concerning this disease.

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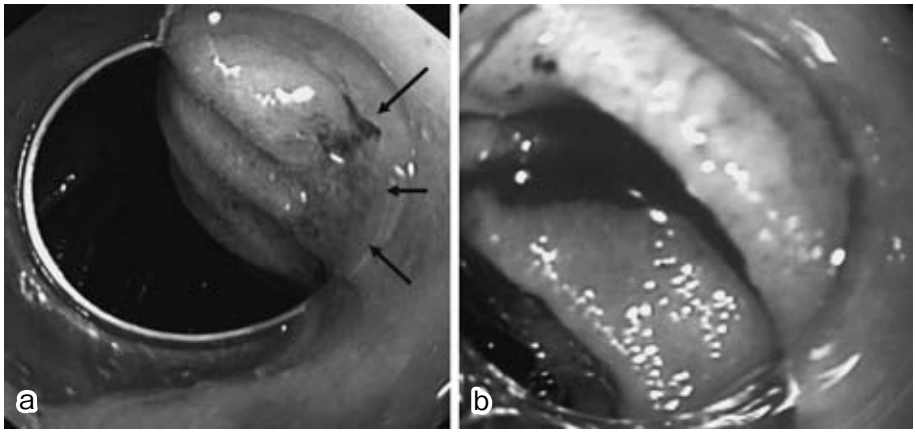

Fig. 1 **a**: Upper gastroduodenal endoscopy revealed submucosal lesions with vascular ectasia **(arrows)** in the second part of the duodenum. **b**: A duodenal lesion involving a pancreatic AVM was the site of gastrointestinal bleeding.

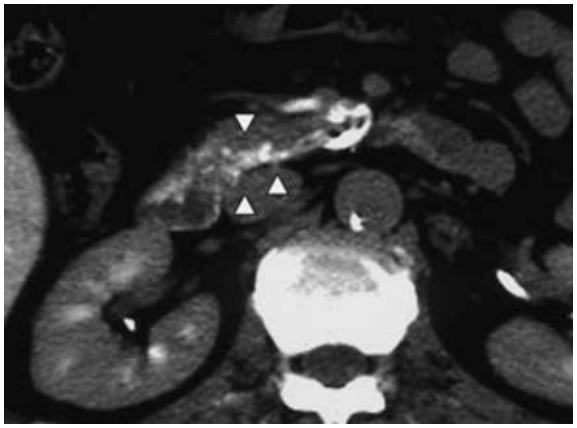

Fig. 2 Dynamic computed tomography revealed a hypervascular lesion in the head of the pancreas and the second part of the duodenum **(white arrowheads)**.

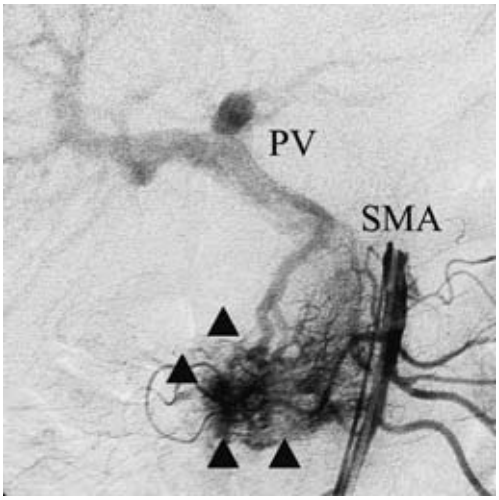

Fig. 3 Selective angiography showed proliferation of a vascular network and early filling of the portal vein in the early arterial phase **(black arrowheads)**.

Case Report

A 54-year-old man was admitted to our hospital in October 2004 with the symptoms of palpitation, dyspnea, and tarry stool. He had a 2-year history of gastric ulcer but no relevant family history. On physical examination, the patient was pale and in a preshock condition. The abdomen was soft and flat, and no masses were palpable. Laboratory data on admission showed an erythrocyte count of 1.29 million/mm^3, a hemoglobin concentration of 5.1 g/d*l*, and a serum iron level of 10 μg/d*l*. Levels of various tumor markers, such as carcinoembryonic antigen, CA19-9, and elastase-1, were normal. The patient received a transfusion of 8 units of packed red blood cells.

Upper gastroduodenal endoscopy after admission revealed submucosal lesions with vascular ectasia in the second part of the duodenum **(Fig. 1a)**, which was the site of active bleeding **(Fig. 1b)**. Endoscopic clipping was performed immediately to control bleeding. Dynamic computed tomography detected a hypervascular lesion in the head of the pancreas and the second part of the duodenum **(Fig. 2)**. Selective angiography showed the proliferation of a vascular network in the head of the pancreas and early

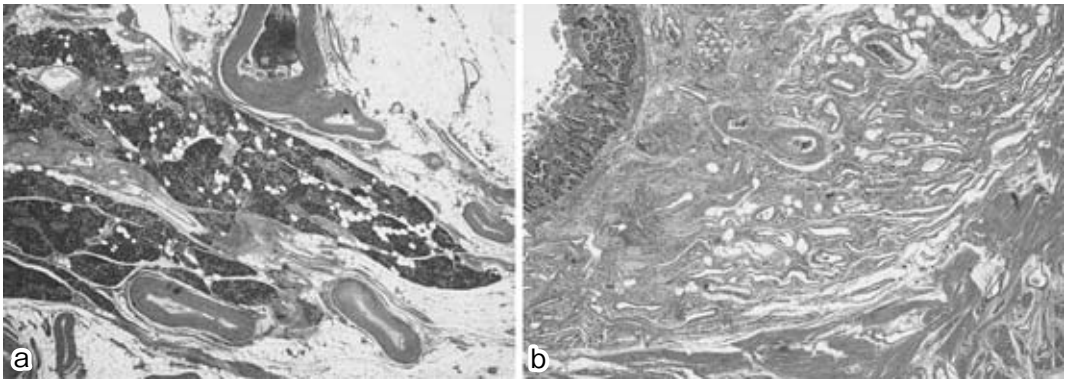

Fig. 4 Histologic examination showed dilated tortuous vessels accompanied by severed elastic fibers in the vessel media and blood clot formation.

visualization of the portal vein at the early arterial phase **(Fig. 3)**.

These findings confirmed the preoperative diagnosis of pancreatic AVM in the head of the pancreas. To prevent recurrent bleeding and progressive portal hypertension, we performed pylorus-preserving pancreaticoduodenectomy. At laparotomy, localized and meandering vessels were seen on the surface of the head of the pancreas. Histological examination showed dilated tortuous vessels, which were often accompanied by severed elastic fibers in the vessel media and blood clot formation **(Fig. 4a)**. Moreover, these dilated capillaries were also found in the adjacent duodenal wall **(Fig. 4b)**. The pancreatic exocrine glands were normal. A pancreatic fistula developed postoperatively but healed with conservative treatment. The patient was discharged for further follow-up and remained well without recurrence of AVM.

Discussion

Pancreatic AVMs are either congenital or acquired[2-4]. Congenital AVMs originate from anomalous differentiation of the rudimentary plexus of primordial blood vessels, whereas acquired AVMs are usually caused by inflammation, tumor, or trauma[3,5]. Most cases of pancreatic AVM outside Japan are associated with Rendu-Osler-Weber syndrome and is known to be a part of the visceral angiodysplasia of hereditary hemorrhagic telangiectasis[3,6]. Because of the lack of pancreatitis, tumor, or history of trauma, the pancreatic AVM in the present case seemed to be congenital in origin.

Recurrent gastrointestinal bleeding is a frequent complication of AVM. Other symptoms include abdominal pain, abdominal discomfort, and abdominal hemorrhage[3,4]. The mechanism of gastrointestinal bleeding may be classified into three types as follows: 1) bleeding from AVM extending into the submucosal layer of the duodenum, 2) bleeding from the pancreatic duct through the orifice of the ampulla of Vater, 3) esophageal or gastric variceal rupture due to portal hypertension caused by pancreatic AVM[5,6]. According to previous reports, the presenting symptom in most cases is bleeding from varices due to portal hypertension[7]. Chuang et al.[6] have suggested that elevated portal venous pressure resulting from unrestricted overflow of the arterial blood into the portal system might cause variceal rapture. Mizutani et al.[8] have reported a case of pancreatic AVM with rupture into the pancreatic duct and also mentioned that bleeding from AVM through the pancreatic duct, in spite of massive bleeding, did not always cause sharp abdominal pain or hyperamylasemia. In the present case, there was no evidence of esophageal or gastric varices, duodenal ulcers, or hemosuccus pancreaticus. Intermittent gastrointestinal bleeding resulted from AVM involving the adjacent duodenum.

Angiography is important for the definitive diagnosis of pancreatic AVM and for the treatment

of gastrointestinal hemorrhage. The angiographic findings of pancreatic AVM include: 1) dilated tortuous feeding arteries, 2) a racemose intrapancreatic vascular network, and 3) early filling of veins, such as the portal vein[6]. Moreover, Chuang et al. have explained these characteristics in detail as follows. The racemose vascular network causes a transient, dense staining of the pancreas. Early filling of veins appears as early as the midarterial phase. An important feature is early clearance of the AVM complex. A unique feature of AVM is its hyperkinetic nature, which distinguishes it from all other pancreatic lesions[6]. In the present case, selective angiography demonstrated proliferation of a vascular network in the head of the pancreas and early visualization of the portal vein at the early arterial phase, which allowed a definitive diagnosis of pancreatic AVM.

Recently, Doppler ultrasonography and magnetic resonance imaging have proven useful in detecting pancreatic AVM. Kurosaki et al.[9] have reported that the subsequent detection of a pulsatile turbulent Doppler wave form led to the diagnosis of AVM, and that, on magnetic resonance, the lesion was demonstrated as a "signal-void" area, characteristic of rapid blood flow. Tano et al.[10] have described the characteristics of pancreatic AVM with color Doppler endoscopic ultrasonography and extracorporeal color Doppler ultrasonography as many pulsatile color flow signals in the duodenal wall, abnormal vessels in the pancreatic head, and a mosaic-like color signal in the portal trunk. These imaging studies are extremely useful for both diagnosis and the physiological assessment of AVMs.

Therapeutic endoscopy is the first-line treatment to control gastrointestinal bleeding from a mucosal vascular lesion. However, for a large submucosal lesion, as seen in the present case, endoscopy is less likely to be successful. Therefore, endoscopic treatment is considered a temporary measure that should be followed by a more definitive therapy. Angiographic management by embolization can be used as a definitive treatment, especially in high-risk patients[11,12]; however, complete embolization of multiple vessels is difficult to achieve. The proliferation of new collateral veins after angiographic treatment may lead to recurrent bleeding and the progression of portal hypertension[5,12]. Therefore, complete surgical resection was preferable and was most previously described cases. Outcomes after partial resection and multiple ligations seem unsatisfactory. Although preoperative assessment of the size, location, and number of lesions is needed, surgery may be the only definitive treatment for symptomatic AVM[5,12,13]. In the present case, because the pancreatic AVM involved the adjacent duodenum, pylorus-preserving pancreaticoduodenectomy was performed and a favorable outcome was obtained.

References

1. Halpern M, Turner AF, Citron BP: Hereditary hemorrhagic telangiectasia. Radiology 1968; 90: 1143–1149.
2. Grannis FW, Foulk WT, Miller WE, Payne WS: Diagnosis and management of an arteriovenous fistula of pancreas and duodenum. Mayo Clin Proc 1973; 48: 780–782.
3. Uchino A, Ishino Y, Ohno M: Arteriovenous malformation of the pancreas associated mesenteric varices: Case report and review of the literature. Radiat Med 1989; 7: 6–9.
4. Kato T, Takahashi M, Okawada T, Miyazaki Y, Kaneko M: Pancreatic arteriovenous malformation treated by transcatheter embolization: Report of a case with hepatocellular carcinoma. Radiat Med 1991; 9: 19–21.
5. Nishiyama R, Kawanishi Y, Mitsuhashi H, et al.: Management of pancreatic arteriovenous malformation. J Hepatobiliary Pancreat Surg 2000; 7: 438–442.
6. Chuang VP, Pulmano CM, Walter JF, Cho KJ: Angiography of pancreatic arteriovenous malformation. Am J Roentgenol 1977; 129: 1015–1018.
7. Uda O, Aoki T, Tsuchida A, et al.: Pancreatic arteriovenous malformation observed to bleed from the bile duct and a duodenal ulcer: Report of a case. Jpn J Surg 1999; 29: 462–466.
8. Mizutani N, Masuda Y, Naito N, Toda T, Yao E, Fukumoto M: Pancreatic arteriovenous malformation in a patient with gastrointestinal hemorrhage. Am J Gastroenterol 1981; 76: 141–144.
9. Kurosaki M, Hattori K, Minato Y, et al.: Asymptomatic arteriovenous malformation of the pancreas. Demonstration by doppler ultrasonography and magnetic resonance imaging. Dig Dis Sci 1993; 38: 1342–1346.
10. Tano S, Ueno N, Ueno T, Wada S, Aizawa T, Kimura K: Pancreatic arteriovenous malformation with duodenal ulcer. Demonstration by color doppler ultrasonography. Dig Dis Sci 1996; 41: 1232–1237.
11. Takahashi S, Tajima H, Kawamata H, et al.: A case of pancreatic arteriovenous malformation causing

portal hypertension. Jpn J Clin Radiol 1998; 43: 1065–1068 (in Japanese).
12. Poon RTP, Poon J: Massive GI bleeding due to a duodenal arteriovenous malformation. Gastrointest Endosc 2000; 52: 101–104.
13. Takiguchi N, Ichiki N, Ishige H, et al.: Pancreatic arteriovenous malformation involving adjacent duodenum in a patient with gastrointestinal bleeding. Am J Gastroenterol 1995; 90: 1151–1154.

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