Arteriovenous malformation of the pancreas: A case report and review of literature

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Extrahepatic arteriovenous malformations (AVMs) of the gastrointestinal (GI) tract are rare and mostly asymptomatic congenital anomalies. The present case describes a 45-year-old woman with an AVM in the head of the pancreas, which caused massive GI bleeding that recurred after embolization, and which was subsequently treated with a pylorus-preserving Whipple pancreaticoduodenectomy. The authors then review the available literature pertaining to AVMs of the GI tract, the diagnostic modalities that have been used to identify them and the treatment approaches that have been described to date, which range from coil embolization of the feeding artery to radical resection of the affected organ. It is important to remember that these lesions shunt blood between the high-pressure arterial system and the low-pressure portal system, which leads to the much-dreaded consequence of portal hypertension.

Key Words: *Arteriovenous malformation; Osler-Weber-Rendu disease; Portal hypertension; Pylorus-preserving Whipple pancreaticoduodenectomy*

Extrahepatic arteriovenous malformations (AVMs) of the gastrointestinal (GI) tract are rare and mostly congenital anomalies. They are usually asymptomatic but may present with portal hypertension in the absence of primary liver pathology. They are also rare causes of massive GI bleeding. Treatment modalities may include ligation of the afferent artery, embolization of feeding vessels, portocaval shunting, or surgical resection of part or all of the affected organ (1).

We report a case of a patient with an AVM in the head of the pancreas, which caused massive GI bleeding that recurred after embolization and which was subsequently treated with a pylorus-preserving Whipple pancreaticoduodenectomy. The present case report also highlights the recent developments in the diagnosis and treatment of arterioportal malformations.

CASE PRESENTATION

A 45-year-old woman was admitted to our hospital with significant upper GI bleeding. She had no history of liver disease, chronic pancreatitis or alcohol abuse. Upper GI endoscopy showed grade II esophageal and gastric varices. A transjugular liver biopsy was negative for cirrhosis. Celiac arteriography showed a tangle of blood vessels in the region of the head of the pancreas, supplied from branches of the gastroduodenal artery, with early shunting of arterial blood into the portal vein (Figures 1A and 1B).

Subsequent venography demonstrated elevated pressure in the hepatic venous system, with a wedge hepatic vein pressure of 23 mmHg and free hepatic vein pressure of 17 mmHg. This was presumed to be secondary to arteriovenous shunting between the pancreatic branches of the gastroduodenal artery and the portal vein. The patient underwent transcatheter coil embolization of the feeding vessel with relief of symptoms for approximately six months, followed by another episode of GI bleeding. Upper GI endoscopy was repeated and showed grade I esophageal varices. A celiac angiogram showed persistence of the AVM and worsening of portal hypertension, with enlargement of venous collaterals. Re-embolization was not an option at that stage because the gastroduodenal artery had been coil embolized (Figure 2). The patient subsequently underwent a pylorus-preserving Whipple pancreaticoduodenectomy. She recovered uneventfully and has not reported any episodes of GI bleeding since her surgery six years ago.

DISCUSSION

AVMs of the GI tract are uncommon and, when present, may be found in the liver or in extrahepatic locations (2). Meyer et al (3) reviewed a series of GI AVMs and found that 78% of these were located in the cecum and the right colon, followed by 10.5% in the jejunum, 5.5% in the ileum, 2.3% in the duodenum, 1.4% in the stomach and 0.9% in the rectum. Only 0.9% were found in the pancreas.

Extrahepatic AVMs have been described, including arterioportal malformations between the accessory right hepatic artery, gastroduodenal artery, or superior mesenteric artery and the portal vein. Ninety per cent of AVMs of the pancreas are congenital in origin and 10% to 30% of them are associated with Osler-Weber-Rendu disease (1), 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 (4). Secondary AVMs have been described, and some of the postulated causes include pancreatitis, trauma and tumours (5). Iatrogenic development of fistulas between the superior mesenteric vein and the superior mesenteric artery has been described following pancreas transplantation surgery (6).

Pancreatic AVM is a rare entity first reported in 1968 by Halpern et al (7,8) in a patient with Osler-Weber-Rendu disease. In total, fewer than 80 cases have been reported in the literature. The true incidence of this pathology is difficult to estimate because many cases do not progress into clinical

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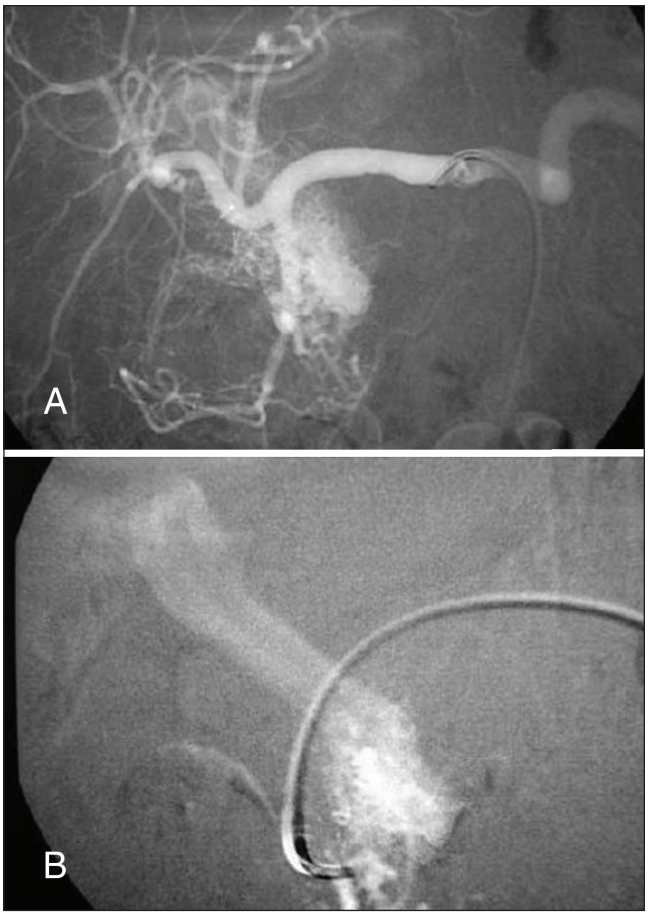

Figure 1) *Angiographic images of the head of the pancreas showing the arteriovenous malformation* (**A**) *and early filling of the portal vein* (**B**)

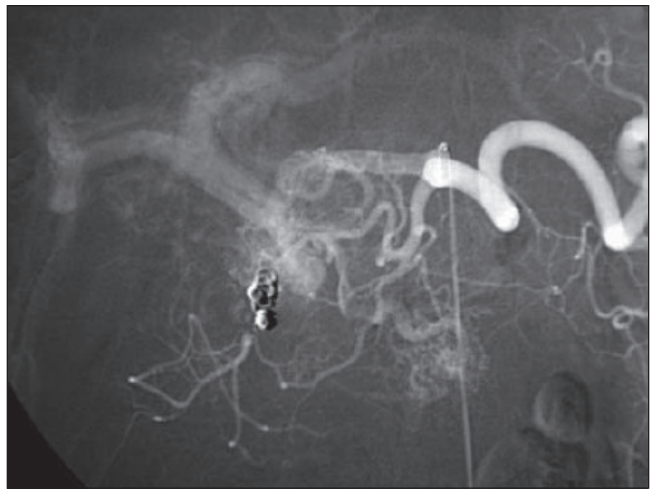

Figure 2) *Angiographic image of the head of the pancreas showing recurrent filling of the arteriovenous malformation despite previous coil embolization of the gastroduodenal artery*

disease. Nishiyama et al (1) reviewed 42 patients with AVMs of the pancreas. They reported that the mean age at diagnosis was 48.8 years (ranging from seven months to 67 years); 78.6% of the patients were men, and the cause was congenital in 91% of cases. Moreover, 50% had life-threatening GI bleeding. The AVM was located in the head of the pancreas in 56% of the patients, 31% of whom also had an extrapancreatic AVM. Treatment modalities included resection in 24 patients, pancreaticoduodenectomy in 10 patients, distal pancreatectomy in five patients, devascularization in four patients, resection of adjacent organs in five patients and total pancreatectomy in one patient for an AVM of the whole pancreas.

AVMs of the GI tract may be asymptomatic, or may present with pain or GI bleeding. Bleeding usually originates from a ruptured esophageal or gastric varix secondary to portal hypertension (9). Another potential etiology of the hemorrhage may be direct erosion of the AVM into the pancreatic duct or through the adjacent intestinal mucosa as a duodenal ulcer (10). Rarely, patients may present with jaundice as the initial symptom secondary to hemobilia, usually caused by erosion of the AVM into the bile ducts (11). Abdominal pain may be caused by the steal syndrome, with shunting of blood away from the mesenteric circulation through the AVM.

Diagnosis is usually made by ultrasonography, computed tomography (CT), magnetic resonance imaging (MRI) and/or angiography. The characteristic finding of an AVM on ultrasound imaging is that of a hypoechoic lesion with a mosaic appearance and a pulsatile waveform in the portal vein on colour Doppler ultrasonography. CT scanning typically shows nodular enhancement and early appearance of the contrast in the portal vein. On MRI, the lesion is seen as a 'signal-void' area, characteristic of rapid blood flow. Angiographic characteristics include dilated and tortuous feeding arteries, followed by a racemose network of vessels in the pancreatic parenchyma, a transient dense pancreatic stain, early appearance of the portal vein, and then early disappearance of the pancreatic stain (12).

Doppler ultrasonography is a noninvasive test useful for early detection of AVMs of the pancreas. CT scanning and MRI have improved the characterization of these lesions (5) and the delineation of their anatomical relationship to other structures. Although angiography is very important as a confirmatory test and in planning treatment, the angiographic characteristics of AVMs of the pancreas can also be seen in pancreatitis, hypervascular neoplasms and some metastases (13,14).

Angiography is necessary to guide treatment even if the diagnosis has been established by noninvasive diagnostic modalities. Transcatheter arterial embolization is a helpful method for controlling hemorrhage, which may be the definitive management for some lesions or a temporizing preoperative measure to control the signs and symptoms for others (15). Recurrent bleeding has been reported in 37% of patients after successful embolization (14) mainly due to the propensity of those lesions to grow new collateral vessels; however, this treatment modality remains valuable, especially in patients who are not good surgical candidates.

Pancreatic AVMs grow progressively in size, leading to the development of portal hypertension which, once established, becomes resistant to treatment even if the AVM is completely resected and all arterioportal communications are interrupted. Hence, it is important to adequately control the lesion at an early stage before portal venous pressure starts to rise. This is why many authors recommend early surgical resection of these lesions (1).

In the present case, the portal pressure was initially controlled with embolization of the AVM afferent vessel as evidenced by

the regression of the esophageal varices; however, the AVM recurred, leaving no other embolization access because the major feeder vessel had already been embolized and the AVM seemed to be feeding from both the celiac and superior mesenteric routes.

SUMMARY

We report a case of AVM of the head of the pancreas, presenting with portal hypertension and GI bleeding in the absence of primary liver pathology, which was successfully treated with a pylorus-preserving Whipple pancreaticoduodenectomy.

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