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CASE REPORT

Arteriovenous malformation of the pancreatic head – difficulties in diagnosis and treatment

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Abstract A patient with pancreatic arteriovenous malformation who presented diagnostic and therapeutic difficulties is presented. The initial tests appeared to suggest inflammatory bowel disease, but the diagnosis was clinched by the finding of blood issuing from the ampulla of Vater. Repeated angiographic embolization did not obliterate the vascular malformation, and the symptoms eventually resolved after Whipple's pancreaticoduodenectomy.

Keywords Angiography · Pancreas · Vascular malformation · Whipple's procedure

Introduction

Pancreatic arteriovenous malformation (AVM) is a rare condition [1]. It was first described by Halpern et al. in 1968 [2]. It is usually asymptomatic. Gastrointestinal hemorrhage and abdominal pain are the most common symptoms. Advances in imaging and the use of angiography have resulted in precise diagnosis of this condition. We report a case of pancreatic AVM presenting with gastrointestinal bleeding and review the published literature. The case also illustrates the difficulties in diagnosis and management.

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Case report

A 26-year-old man presented to hospital in June 2006 with recurrent episodes of abdominal pain over the past 2 years, associated with vomiting. There were no symptoms of bleeding at that time. He had undergone surgery for intussusception at 6 months of age. The barium series was normal; diagnostic laparoscopy was performed as he continued to have pain. Laparoscopy revealed adhesions between small bowel loops and the scar. These were lysed and he remained asymptomatic thereafter.

In May 2007, he presented again to hospital with recurrent colicky abdominal pain and melena associated with hemodynamic symptoms. Hemoglobin was 7.3 g/dL; he received blood transfusions as required. Upper GI endoscopy and colonoscopy were normal. CT scan of abdomen revealed thickening of the distal small bowel and a normal-looking pancreas, and his laboratory findings (positive C-reactive protein, positive antisaccharomyces cervicisiae antibody) suggested a diagnosis of inflammatory bowel disease. Enteroscopy did not reveal any abnormality. Hence laparoscopy and laparoscopy-assisted pan enteroscopy (LAPE) was performed in June 2007. Enteroscopy was normal and biopsies taken from several sites in the jejunum and ileum showed within normal mucosa at histology.

One month later, he developed melena again. Side-viewing endoscopy revealed blood oozing from the ampulla of Vater. Celiac angiogram revealed a large arteriovenous malformation arising from the superior pancreaticoduodenal artery (Fig. 1). Embolization using steel coils was performed at the level of the gastroduodenal artery. Superior mesenteric artery injection did not reveal any filling of the AVM. However, melena recurred after 10 days. A second angiogram revealed filling of the AVM through a

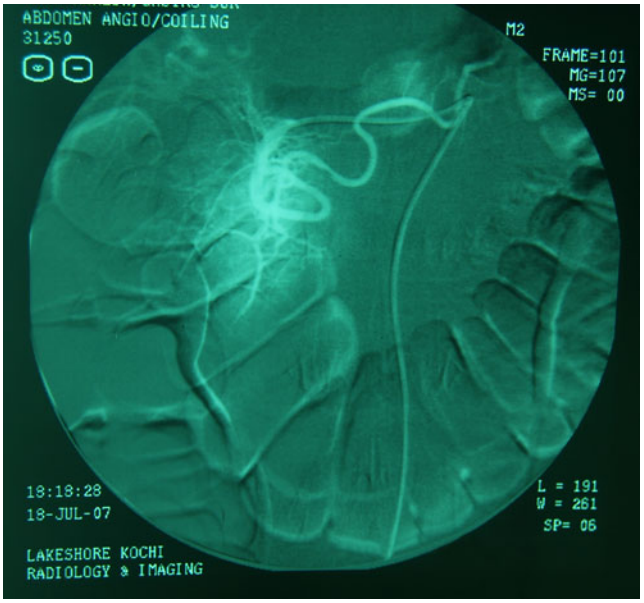

Fig. 1 Celiac angiogram showing arteriovenous malformation of the pancreas

separate vessel from the hepatic artery. Embolization of this vessel was also done (Fig. 2). A repeat CT scan revealed specks of calcification in the pancreas with focal pancreatitis but no clearcut features of AVM (Fig. 3). He continued to bleed, and a third angiogram showed an intense blush in the pancreaticoduodenal area with early filling of the portal vein. Embolization was considered not feasible as there was a risk of devascularization of the duodenum. Hence he was operated. During surgery, duodenoscopy confirmed the presence of bleeding from the ampulla of Vater. Palpation of the head of the pancreas revealed some nodularity of the

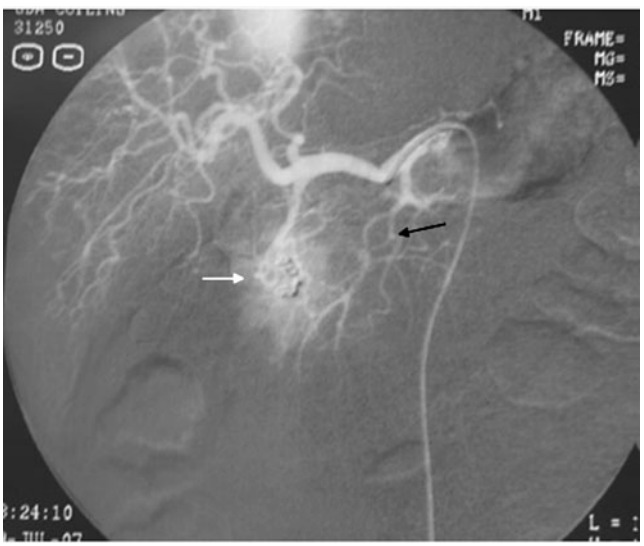

Fig. 2 Second angiogram with feeder from hepatic artery (*black arrow*). White arrow indicates previous embolization

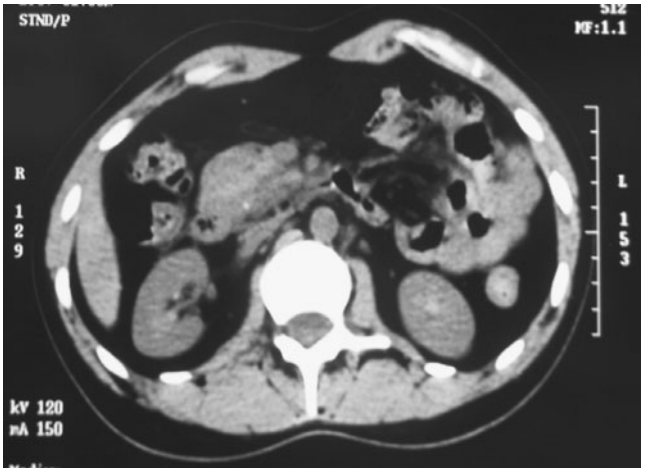

Fig. 3 CT scan showing calcification and "focal pancreatitis"

head, but no vascular malformation was seen. A Whipple's pancreaticoduodenectomy was performed. When the neck was divided, a dilated pancreatic duct containing blood was seen. Reconstruction was achieved by pancreaticojejunostomy. Cut section of the pancreatic head specimen revealed a vascular malformation in the head communicating with the pancreatic duct in the head (Fig. 4). Postoperative recovery was uneventful except for

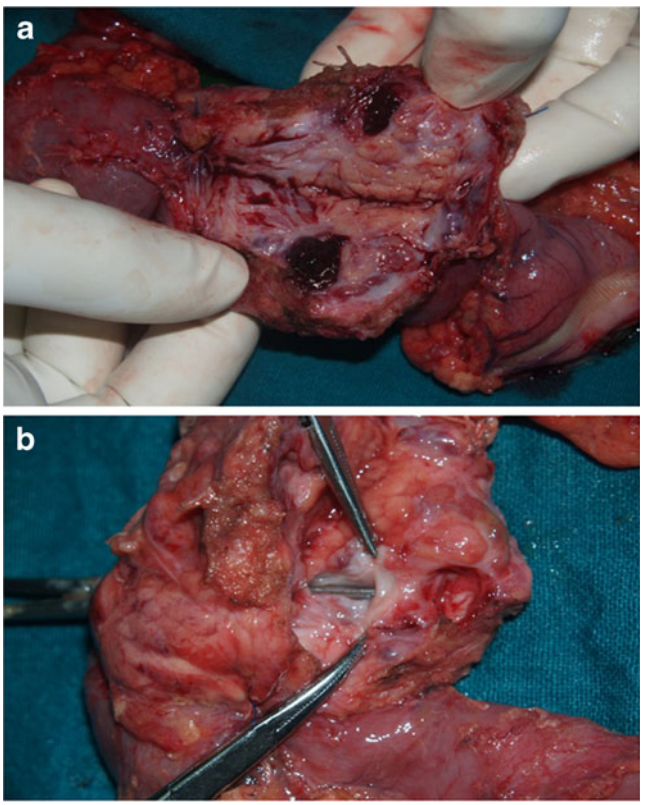

Fig. 4 (**a**) Cut specimen of pancreas showing blood clot within the arteriovenous malformation. (**b**) Probe demonstrating communication of AVM with the pancreatic duct

delayed gastric emptying which lasted 14 days. Histopathology confirmed the presence of arteriovenous malformation. The patient is asymptomatic on follow up 4 months after surgery.

Discussion

Pancreatic AVMs may be congenital (90%) or acquired in origin. It can be an isolated finding or may be associated with Osler-Weber-Rendu disease, which is an autosomal dominant syndrome characterized by recurrent epistaxis, telangiectasia of skin, intraabdominal arteriovenous malformations and a positive family history [3, 4]. Acquired AVM may also be secondary to pancreatitis, tumor or trauma [5]. The most common site of pancreatic AVM is the head of the pancreas and the mechanism of hemorrhage is believed to be either rupture of the malformation into the pancreatic duct or intestinal mucosa or rupture of esophageal or gastric varices because of secondary portal hypertension produced by the AVM [4–6]. Our patient also bled due to rupture into the pancreatic duct. Bleeding may also occur rarely into the bile duct and present as hemobilia [7]. Angiography provides a definitive diagnosis and the characteristic features are: (a) dilated and tortuous feeding arteries, (b) complex intrapancreatic vascular network, and (c) early filling of draining veins such as portal vein. The vessels most commonly affected are the splenic artery (42%), gastroduodenal artery (22%) and small pancreatic arteries (25%) [8]. CT scan appearances may clinch the diagnosis with a peculiar vessel structure suggestive of AVM [9], but it failed to identify the malformation in our case. The presence of hypoechoic nodules in the pancreas on colour Doppler may also suggest an arteriovenous malformation. Doppler ultrasonography also helps to determine the relationship of the AVM to the portal vein, the direction of blood flow and for detecting AV shunt [7, 10]. Magnetic Resonance Imaging may also be useful in identifying a vascular lesion on T1 and T2 weighted images [11].

The presentation at a young age and the absence of features of Osler-Rendu-Weber syndrome suggest a congenital etiology in our patient. However, the AVM was not large and was not even apparent on the surface of the pancreas on laparotomy. CT scan revealed a speck of calcification in the region of the AV malformation and features of pancreatic inflammation. When the neck of the pancreas was transected, the duct was clearly seen to be dilated. It is not clear whether an associated chronic pancreatitis coexisted or if the dilatation of the duct was merely a result of distension due to the blood collecting within it. There were no gross histological changes of chronic pancreatitis and the calcific focus was in the AV malformation itself.

Angiographic embolization is useful in reducing the vascularity of the malformation as an aid to surgery [12]. However, recurrent bleed has been reported in up to 37% of patients embolized successfully and who were waiting for surgery [13]. Could this case be just a hypervascularity of the head of the pancreas due to inflammation? This is unlikely as (a) hypervascularity is unlikely to cause significant large volume episodic bleeding, (b) A clear cut vascular cavity was found (Fig. 4) which communicated with the main pancreatic duct, and (c) histologic appearance. The case illustrates the following: (a) a diagnosis of pancreatic arteriovenous malformations though rare, must be considered in all patients with hemosuccus pancreaticus; (b) angiography is the ideal investigation, and CT scan may fail to identify the lesion; (c) although embolization may be tried, surgical resection may become necessary.

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