CASE REPORT

Unusual Cause of Massive Upper Gastrointestinal Bleeding: A Pancreatic Arteriovenous Malformation

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ABSTRACT

Context Upper gastrointestinal bleeding is one of the most common emergencies in gastroenterology. The common causes of the upper gastrointestinal bleeding include peptic ulcer disease, gastric erosive mucosal disease and portal hypertension. Gastrointestinal arteriovenous malformation is a less common cause of gastrointestinal bleeding and these arteriovenous malformation are most commonly located in the large and small intestine. Pancreatic arteriovenous malformation is a rare condition in which there is tumor-like formation or vascular anomaly built up via an aberrant bypass anastomosis of the arterial and venous systems in the pancreas. Splenic artery is most commonly involved (42%), followed by gastroduodenal artery (22%) and small pancreatic arteries (25%). Clinically it may present as gastrointestinal hemorrhage which is occasionally fatal. Other presentations are abdominal pain, pancreatitis, duodenal ulcer, jaundice, and portal hypertension. **Case report** We present a rare case of pancreatic arteriovenous malformation presenting as massive upper gastrointestinal bleeding. **Conclusion** Since early surgery is a life saving treatment for such cases, hence, a high index of suspicion should be maintained especially when massive bleeding is detected from the medial wall of second part of duodenum.

INTRODUCTION

Upper gastrointestinal bleeding is one of the most common emergencies in gastroenterology. The common causes of the upper gastrointestinal bleeding include peptic ulcer disease, gastric erosive mucosal disease and portal hypertension. Gastrointestinal arteriovenous malformation is a less common cause of gastrointestinal bleeding and these arteriovenous malformations are most commonly located in the large and small intestine. We present a rare case of pancreatic arteriovenous malformation presenting as massive upper gastrointestinal bleeding.

CASE REPORT

A 37-year-old man presented with history of abdominal pain for 3 months and melena for 2 days. He had similar episode of melena two weeks prior to presenting to our hospital when he was admitted to a local hospital and was transfused four units of packed red blood cells. The pain was of moderate to severe intensity, non-colicky, upper abdominal, not radiating to back, and had no relation to meals. There was no history of hypertension, diabetes mellitus, or ingestion of NSAIDs. On examination, he was pale and had mild tenderness in the upper abdomen. After admission, he was resuscitated with colloids and two unit of packed red blood cell transfusion.

Investigations revealed hemoglobin of 4.5 g/dL (reference range: 11.0-15.0 g/dL), total lymphocyte count $12x10^3$ mm^{-3} (reference range: 4-10 $x10^3$ mm^{-3}), platelets 180 $x10^3$ mm^{-3} (reference range: 150-450 $x10^3$ mm^{-3}), INR 1.2, serum creatinine 1.1 g/dL (reference range: 0-6-1.3 g/dL), serum bilirubin 0.8 mg/dL (reference range: 0.2-1.0 mg/dL), AST 42 IU/mL (reference range: 0-42 IU/mL), ALT 32 IU/mL (reference range: 0-60 IU/mL), serum alkaline phosphate 126 IU/L (reference range: 39-117 IU/mL). An ultrasound examination of the abdomen revealed mildly coarse echo-texture of liver, with portal vein diameter of 13 mm, and mild periportal collaterals. Intrahepatic bile ducts were normal; the common bile duct was of 8 mm and gallbladder and spleen were normal. Pancreatic head was bulky and heterogeneously hypoechoic while body and tail of the pancreas were normal. Upper gastrointestinal endoscopy revealed an ulcer in the medial wall of the second part of the duodenum with fresh oozing of

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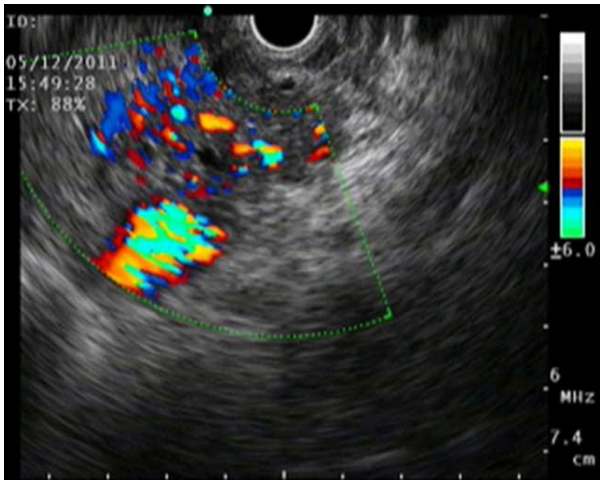

Figure 1. Endoscopic ultrasonographic image of the pancreatic arteriovenous malformation.

blood (Figure 1). The oozing was controlled by injection of 10 mL of 1:10,000 adrenaline at the base of the ulcer. Serum amylase was found to be 462 IU/mL (reference range: 5-100 IU/mL), suggestive of pancreatitis.

CECT of the upper abdomen revealed bulky head and uncinate process of pancreas with altered attenuation and abnormal enhancement with multiple vascular channels within and around the head and uncinate process of pancreas with abnormal enhancement in adjacent wall of the duodenum and a cystic lesion in the head of the pancreas with haziness and stranding in the peripancreatic fat (Figure 2). Endoscopic ultrasound revealed multiple dilated vascular channels in the head of the pancreas (Figure 3) with portal vein thrombosis.

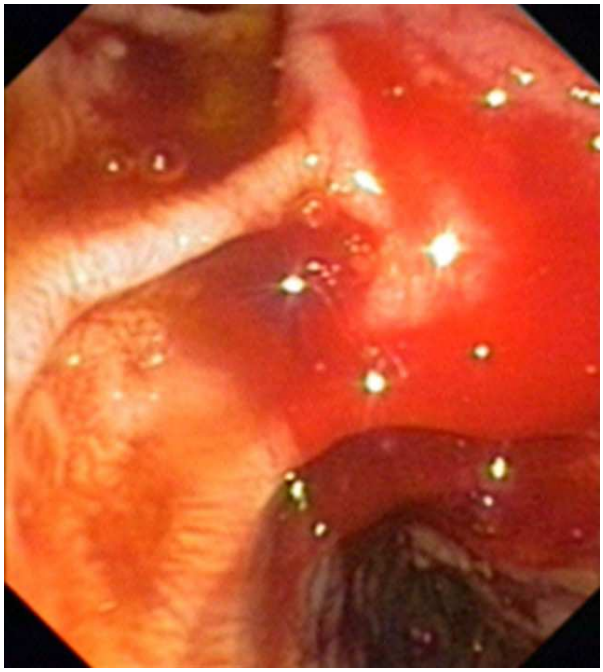

Figure 2. Endoscopic image of the bleed from the medial wall of the second part of duodenum.

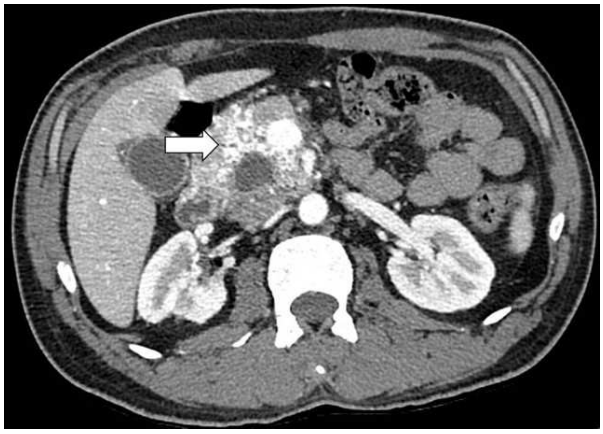

Figure 3. CT angiography of the pancreatic head showing the increased vascularity in arterial phase.

The patient had recurrence of upper gastrointestinal bleeding three days after the endoscopic intervention, requiring transfusion of two more units of packed red blood cells. He was taken up for emergency endoscopy which revealed bleeding ulcer in the proximal duodenum and a repeat session of injection sclerotherapy was done; however, the bleeding did not stop. In view of continuing bleeding, the patient underwent conventional angiography which revealed a diffuse vascular blush in the head of the pancreas (Figures 4 and 5) with early filling of the veins suggestive of tumor blush. Feeder from the gastroduodenal artery was embolized with gelfoam and coil. Check angiography showed no early filling of veins. Superior mesentery artery angiogram showed similar blush in the same region but could not be embolized.

The patient continued to have abdominal pain with bleeding requiring multiple blood transfusions. The patient underwent Whipple's surgery in view of failure

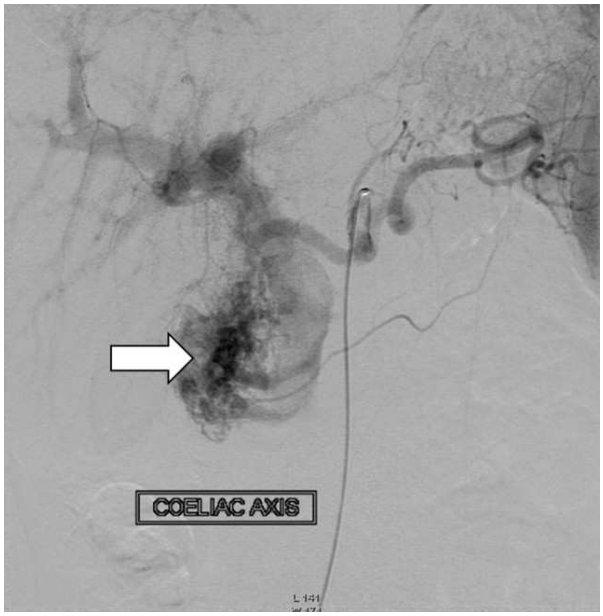

Figure 4. Selective celiac axis angiography showing vascular blush.

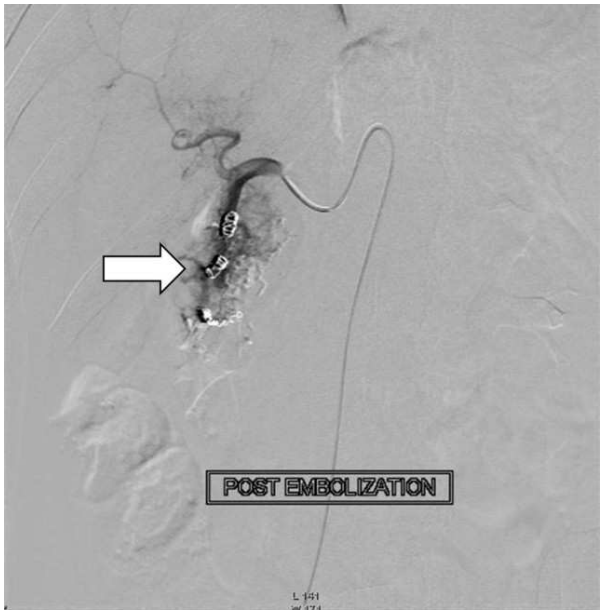

Figure 5. Selective celiac axis angiography post embolization.

of endoscopic and radiologic interventions and suspicion of bleeding lesion in the head of pancreas. The surgical findings revealed bulky pancreatic head with multiple collaterals around the head. There were multiple cystic spaces in pancreatic head and uncinate process. The pancreas distal to the neck was normal (Figure 6). Histopathology of the resected specimen (Figure 7) revealed the diagnosis of the pancreatic arteriovenous malformation. Patient had an uneventful post-operative course and was discharged in four weeks in a stable condition.

DISCUSSION

Pancreatic arteriovenous malformation, first reported by Halpern *et al*. [1], is a rare condition in which there is tumor formation or vascular anomaly built up via an aberrant bypass anastomosis of the arterial and venous systems in the pancreas. Clinically it may present as gastrointestinal hemorrhage which is occasionally fatal [2, 3]. Other presentations are abdominal pain, pancreatitis, duodenal ulcer, jaundice, and portal hypertension [4, 5]. In the series by Song *et al*. 92% patients had abdominal pain, 58% had gastrointestinal bleeding, 45% had pancreatitis, 27% had pseudocysts of the pancreas, 45% had duodenal or gastric ulcers and 9% had mild portal hypertension [6]. Pancreatic arteriovenous malformation occurs most frequently in fourth to sixth decades with a male predominance. Splenic artery is most commonly involved (42%), followed by gastroduodenal artery (22%) and small pancreatic arteries (25%) [7]. Many explanations have been proposed as the cause of gastrointestinal bleeding in pancreatic arteriovenous malformation which are: bleeding from pancreatic duct; bleeding from duodenal intestinal mucosa; and bleeding from esophagogastric varices which may occur due to associated portal hypertension [8]. Gastrointestinal bleeding may be fatal in 30-50% of patients with pancreatic arteriovenous malformation. Pancreatic arteriovenous malformation can be congenital as in Rendu-Osler-Weber syndrome or acquired which can be due to inflammation, trauma or tumor [9, 10].

Surgical resection is the treatment of choice with the intention of curative procedure [4] but it is associated with the risk of massive intraoperative bleeding. Thus other modalities like transarterial embolization [11], embolization performed with use of embolic liquid agent, like ethylene-vinyl alcohol copolymer (onyx) [12], radiation therapy [13] and placement of a transjugular intrahepatic portosystemic shunt [14] have been described. Most common surgical intervention for the pancreatic arteriovenous malformation is pancreatic resection followed by distal pancreatectomy with splenectomy and last is total pancreatectomy. In the series by Song BK *et al*. [6], of 11 patients undergoing surgery, pylorus preserving pancreaticoduodenectomy or pancreaticoduodenectomy was done in 3 patients, distal pancreatectomy in 4 patients, total pancreatectomy in 3 patients, and central pancreatectomy in 1 patient. Surgical resection of the affected pancreas is the most effective treatment of

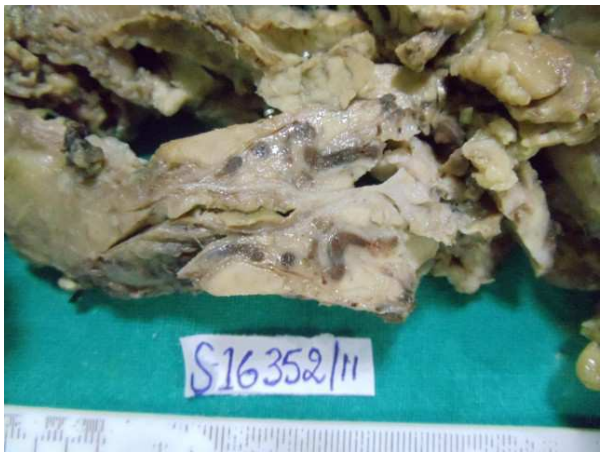

Figure 6. Gross specimen of the pancreatic arteriovenous malformation.

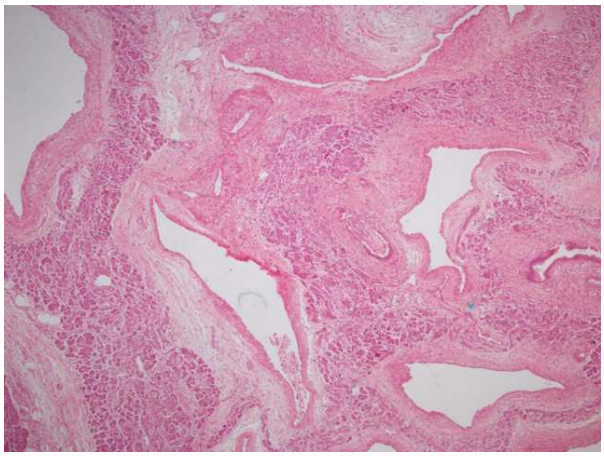

Figure 7. Photomicrograph showing variable sized thick and thin walled blood vessels arranged haphazardly within the pancreatic parenchyma (H&E, x40).

patients with symptomatic pancreatic arteriovenous malformation. Transjugular intrahepatic portosystemic shunt and radiation therapy might be other treatment options when a patient is at a high risk for surgical treatment [4].

In conclusion, pancreatic arteriovenous malformation is a rare cause of upper gastrointestinal bleeding and needs a high index of suspicion when bleeding is detected from the medial wall of second part of duodenum. Once diagnosed it can be successfully managed surgically; however, in high risk cases other modalities of treatment like transarterial embolization, transjugular intrahepatic portosystemic shunt, or radiation therapy may be used.

Conflict of interests The authors have no potential conflict of interests

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