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Gastrointestinal

Pancreatic arteriovenous malformation mimicking pancreatic neoplasm: a systematic multimodality diagnostic approach and treatment

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ABSTRACT

Pancreatic arteriovenous malformation (pAVM) is a very rare entity, as less than 100 cases are reported in the international literature. Patients with pAVM may be asymptomatic or may present a wide range of symptoms, such as vague pain, feeling of fullness, gastrointestinal bleeding, or even portal hypertension. We present the multimodality approach in the diagnosis of a patient with pAVM and treatment via transcatheter arterial embolization of the lesion using steel coils. The patient was free of symptoms 12 months later.

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Introduction

Pancreatic arteriovenous malformation (pAVM) is defined as an abnormal vascular net, developed from one or more feeding arteries and enlarged early draining veins, creating an arteriovenous shunting.

The incidence of pAVM is extremely rare, with less than 100 reported cases in the international literature. Arteriovenous malformation (AVM) of the pancreas causes a wide range of symptoms such as vague pain, feeling of fullness, or even portal hypertension, gastrointestinal (GI) tract bleeding, and pancreatitis. Nevertheless, the majority of the patients remain asymptomatic.

This case describes the use of multimodality imaging including ultrasound (US), computed tomography (CT) scan, magnetic resonance imaging (MRI), and digital subtraction angiography (DSA) in the precise diagnosis of pAVM. Additionally, therapy of this entity via transcatheter arterial embolization (TAE) using steel coils is presented.

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Case report

A 54-year-old man was referred to our hospital for evaluation of melena and vague pain at the right upper quadrant, after laparoscopic cholecystecomy 2 months ago and hospitalization for 2 weeks. Laboratory data revealed a normal hepatic and pancreatic function. The patient tested negative for hepatitis B and C viruses. Hemoglobin levels were 6.2 g/dL and the patient received blood transfusion.

Upper GI endoscopy revealed small red spots in the mucosa of the anterior wall of the second part of the duodenum but no active bleeding. Colonoscopy did not reveal any pathology. Transabdominal color Doppler ultrasound demonstrated an area of turbulent flow just lateral to the proximal superior mesenteric vein (Fig. 1), where an endoscopic ultrasound of the pancreas showed no pancreatic lesion. An abdominal CT revealed multiple vascular formations in the anatomic area between the uncinate process of the pancreas, the second part of the duodenum, and the superior mesenteric vein. The largest vascular formation drained into the superior mesenteric vein (Figs. 2 and 3). Additional abdomen MRI showed multiple small flow voids in the area of the aforementioned lesion with mixed signal after gadolinium contrast injection (Fig. 4).

Finally, TAE was indicated for diagnostic and therapeutic reasons. DSA angiography revealed the vascular nidus supplied by branches of the gastroduodenal artery and an instant outline of the splenoportal axis (Fig. 5A-C). Embolization of the main feeding arteriole was performed. Postembolization infusion showed a smaller shunt between the arteriole and the splenic vein, which was treated with a coil placement, and no further venous enhancement was seen (Fig. 5D).

After 6 and 12 months of follow-up, the patient was asymptomatic.

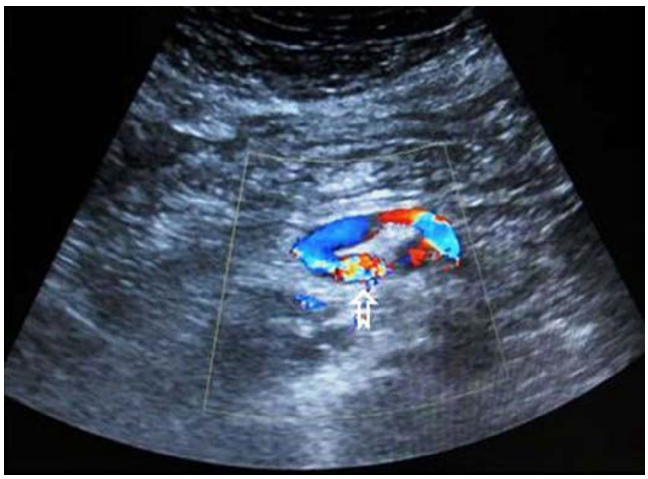

Fig. 1 – Color Doppler transabdominal ultrasound transverse image demonstrates a vessel with turbulent flow (open arrow) communicating with the proximal superior mesenteric vein.

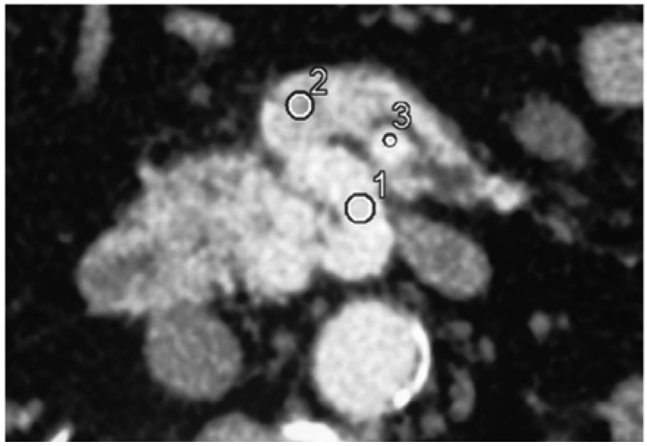

Fig. 2 – The pancreatic arteriovenous malformation is located in the anatomic area between the uncinate process of the pancreas and the second part of the duodenum. Note that the enlarged draining vein has a similar enhancement (1 = 129 HU) as the superior mesenteric artery (3 = 130 HU) has, and the superior mesenteric vein shows early opacification (2 = 79 HU), indicating the presence of an arteriovenous shunt.

Discussion

Although it can occur anywhere in the body, AVM of the GI tract and particularly in the pancreas is extremely rare. Only 0.9% of all GI vascular malformations are located in the pancreas [1] and 90% of them are classified as congenital. GI vascular malformations can be found sporadically, indicating a persistent remnant of the fetal pancreatic vascular network, or being part of the wide spectrum of hereditary hemorrhagic telangiectasia, also known as the Osler-Weber-Rendu syndrome [2]. Acquired pAVMs (10% of the cases), derive as a consequence of trauma, tumor, or inflammation of the pancreas [1]. Our patient did not reveal any other telangiectasia, and his personal and family histories were negative.

Although pAVM can occur everywhere in the pancreatic parenchyma, the commonest affected part is the head [3]. They tend to grow progressively in size. Large pAVMs usually lead to portal hypertension [4].

The splenic artery is most commonly involved in a pAVM (47%), followed by the gastroduodenal (22%) and small pancreatic arteries (25%) [5].

Small lesions do not cause symptoms. Larger lesions usually cause abdominal pain most possibly caused by steal syndrome as the pAVM shunts away the blood from the mesenteric circulation and, mainly, GI bleeding. In our case, the main cause of the patient's GI bleeding was erosion into the adjacent duodenum. Other causes of bleeding include the rupture of gastroesophageal varices caused by portal hypertension and the erosion of the AVM into the pancreatic duct or the bile ducts [6]. Symptoms usually occur in the fifth decade of life (mean age 49.8 years), and there is a clear male predilection [7].

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