Spontaneous inferior mesenteric arteriovenous fistula as a cause of severe portal hypertension and cardiomyopathy

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ABSTRACT

Inferior mesenteric artery (IMA) and inferior mesenteric vein (IMV) fistulas or malformations are extremely rare, with only 36 cases reported. Low incidence and nonspecific clinical signs and symptoms make mesenteric arteriovenous fistulas difficult to diagnose. We describe a case of a primary IMA-IMV fistula. Our patient presented with severe portal hypertension and cardiomyopathy along with robust arteriovenous connections between the IMA and IMV. Arterial embolization in this patient had to be followed by venous embolization for successful resolution of portal hypertension and cardiomyopathy. This case also highlights that close outpatient monitoring for treatment failure and recurrence is necessary for this disease process. (J Vasc Surg Cases and Innovative Techniques 2019;5:113-6.)

Keywords: Inferior mesenteric artery; Inferior mesenteric vein; Arteriovenous fistula and malformation; Amplatzer; Embolization; Portal hypertension

An arteriovenous fistula (AVF) is abnormal high-flow direct communication between an artery and vein without a capillary bed. The term is often used interchangeably with arteriovenous malformation (AVM), with malformation reserved mainly for presumed congenital lesions. AVMs or AVFs of the splanchnic circulation are exceedingly rare and occur more frequently in hepatic and splenic beds with rare occurrence in the mesentery.[1] Splanchnic AVF can result in a left to right shunt with development of visceral ischemia, persistent abdominal pain, and portal hypertension. Inferior mesenteric artery (IMA) and inferior mesenteric vein (IMV) AVFs can be of congenital or iatrogenic etiology. Congenital IMA-IMV fistulas often occur when multiple AVFs are formed and can originate from embryonic vessels that fail to regress or collagen defects. Iatrogenic fistulas between the IMA and IMV can be secondary to penetrating abdominal injuries, arterial catheterization, or surgery.[2-4] Our patient presented with spontaneous IMA-IMV AVF with severe portal hypertension and was treated with endovascular arterial and venous embolization. Informed consent was obtained from the patient to publish case details and images.

CASE REPORT

Our patient, a 46-year-old man, was referred to the vascular surgery service for a spontaneous IMA-IMV AVF with high output resulting in hepatic dysfunction secondary to severe portal hypertension and cardiomyopathy. The patient had no history of trauma or gastrointestinal surgery and presented with 3 months of abdominal pain. Abdominal computed tomography (CT) angiography with three-dimensional reconstruction revealed an AVF of the IMA near the region of the sigmoid colon (Fig 1). The patient wanted to pursue minimally invasive intervention before any open operations.

Selective inferior mesenteric arteriography was performed and confirmed the location of the AVF along the distal circulation of the IMA with a focal network of arteriovenous connections (Fig 2, *A*). We performed arterial embolization proximal and distal to the fistula bed and within the area with a plexus of arteriovenous connections. Coil embolization using the Retracta detachable coils (Cook Medical, Bloomington, Ind) of the distal sigmoidal branches of the IMA and the segment with AVF was performed first along with embolization of a higher origin of superior rectal artery with an Amplatzer 4 plug (Abbott, St. Paul, Minn). The main trunk of the IMA proximal to the fistula was embolized using Amplatzer 2 plugs. Completion arteriography revealed excellent coverage of the area of the fistula with a small amount of residual direct venous filling (Fig 2, *B*). Arteriography revealed patent bilateral internal iliac arteries and a mesenteric arterial arcade and intact perfusion of the colon through these collaterals before and after arterial embolizations.

Eight weeks after arterial embolization, the patient continued to have abdominal pain. CT angiography revealed persistent arteriovenous shunting in the region of the malformation. IMA arteriography showed that the area of previous embolization was thrombosed; however, there was significantly more neovascularization and shunting from the IMA proximal to previously placed plugs (Fig 2, *C*). Further embolization of the IMA was performed using 14-mm and 16-mm Amplatzer plugs proximally in the IMA (Fig 2, *D*). Arteriography then revealed better occlusion

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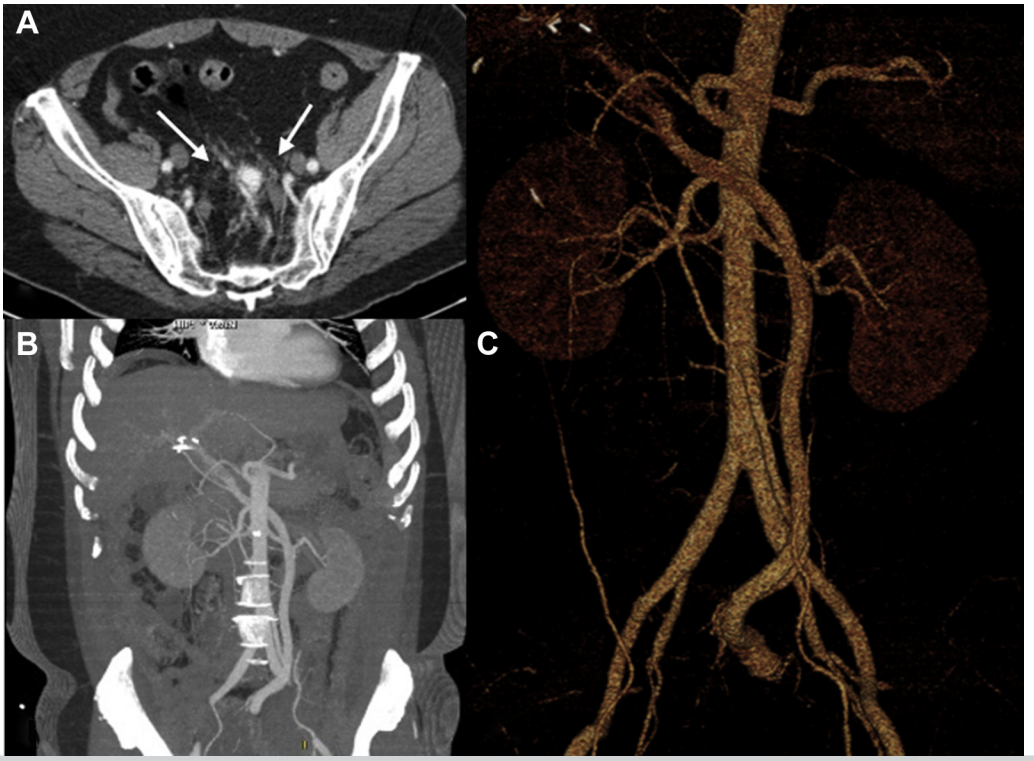

Fig 1. Computed tomography (CT) angiography demonstrates the inferior mesenteric artery (IMA) with irregular filling of the inferior mesenteric vein (IMV; *arrows*). **A,** Axial view. **B,** Coronal view. **C,** Three-dimensional reconstruction.

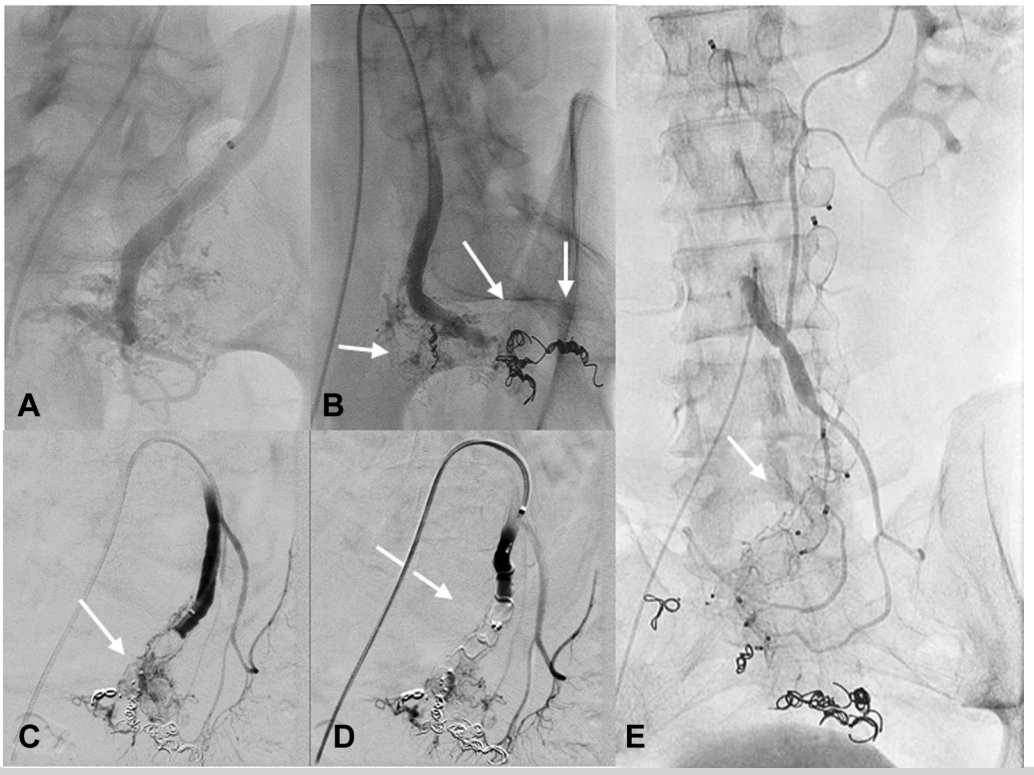

Fig 2. A, Selective arteriogram of the arteriovenous fistula (AVF) originating off the distal inferior mesenteric artery (IMA). **B,** Completion coil embolization arteriogram of the distal sigmoidal branches of the IMA, higher origin of superior rectal artery, and main trunk of IMA proximal to the fistula (*arrows*). **C,** Arteriogram shows previous embolization sites of the IMA with increased proximal neovascularization (*arrow*). **D,** Arteriogram after placement of proximal Amplatzer plugs (*arrow*). **E,** Post-venous embolization (*arrow*) IMA arteriogram showed complete cessation of flow across the arteriovenous malformation (AVM) into the inferior mesenteric vein (IMV).

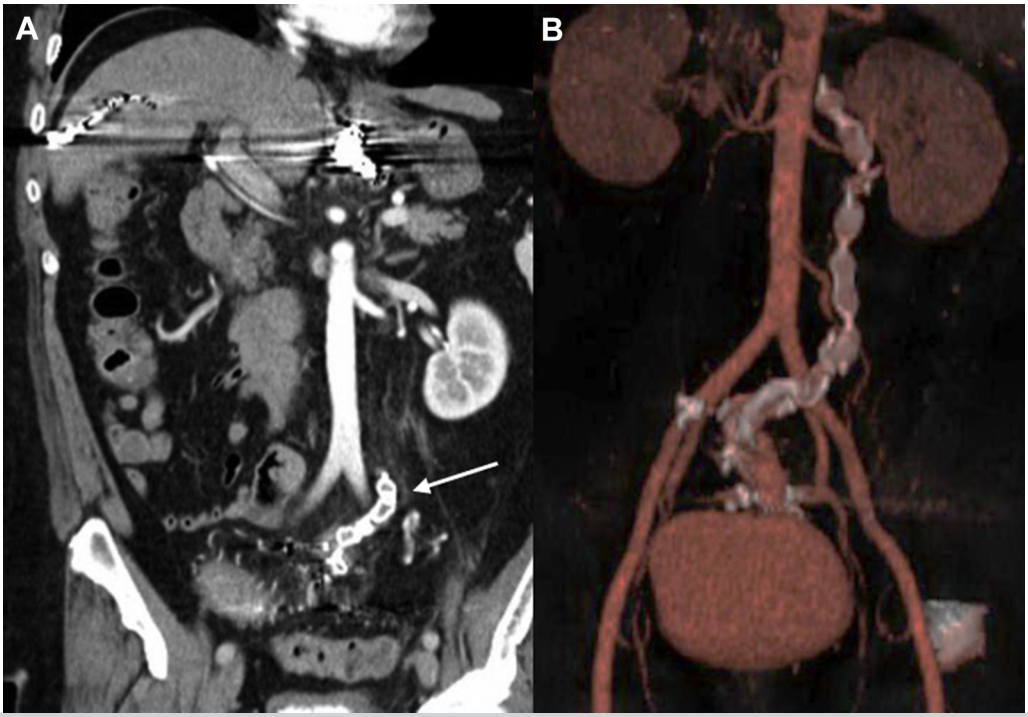

Fig 3. A, Computed tomography (CT) angiography shows no contrast material within the embolized inferior mesenteric vein (IMV) or downstream from the embolized inferior mesenteric artery (IMA). **B,** Three-dimensional reconstruction of segmental embolization of IMA and IMV.

of the mid IMA, with the left colic artery and proximal sigmoid patent to perfuse the colon; however, backfilling of the AVM was noted with some persistent shunting. Selective left iliac internal arteriography revealed ascending lumbar branches into the AVF, and selective right iliac internal arteriography revealed cross-filling of the fistula from the right sacral branches. We decided to proceed to venous outflow embolization as opposed to embolization of the internal iliac arteries. Two days later, the patient was brought back for embolization of the venous side. Venous embolization of the AVF outflow tract was then performed using two 22-mm Amplatzer plugs and one 20-mm Amplatzer plug through percutaneous transhepatic portal access. Post-venous embolization inferior mesenteric arteriography showed complete cessation of flow across the AVF into the IMV (Fig 2, *E*). Portal venous pressure decreased from 30 to 35 mm Hg to 8 to 12 mm Hg. The 1-month postprocedural CT angiography (Fig 3, *A*) and three-dimensional reconstruction (Fig 3, *B*) demonstrated complete thrombosis of the AVM.

One month postoperatively, the patient continued to have left lower quadrant abdominal pain. CT imaging showed venous congestion and colonic wall thickening of the distal sigmoid colon, probably secondary to venous embolization. Colonic resection was recommended for symptomatic relief. On surgical exploration, a segment of distal sigmoid colon noted to be significantly congested was resected, and an end-colostomy was created. The choice of colostomy was made because of concern of relative venous congestion of the resected ends and the risk of anastomotic ischemia. The embolized vascular malformation was completely thrombosed but scarred down and unable to be resected. The mesenteric embolization was intact with no thrill or bruit, and the liver appeared normal without any signs of cirrhosis or portal venous congestion. The patient underwent a reversal of the colostomy with coloproctostomy 4 months later. He was seen in follow-up 6 months later and continues to do well without any recurrent symptoms.

DISCUSSION

Diagnosis of splanchnic AVF or AVM can be established by CT angiography, mesenteric angiography, or direct examination findings of congested viscera with palpable thrill within the mesenteric bed. Treatment is case specific and debatable through surgical resection or percutaneous embolization.[2,5,6] Surgical resection of the affected bowel and entire AVM is curative yet carries a risk of hemorrhage and incomplete exclusion of the fistula. Percutaneous embolization is less invasive; however, it can lead to organ ischemia or recurrence if more than one feeding vessel is involved.[3,5,7,8]

IMA-IMV AVF or AVM is extremely rare, with only 36 cases reported.[1-10] For our case, the initial operative management was by arterial embolization of the AVF. Results of the management for IMA AVF demonstrate that endovascular arterial embolization of the feeder artery at the AVF junction is an

extremely effective approach. However, it does carry a risk of visceral ischemia, particularly for large (>8 mm) and high-flow feeding vessels.[1,2] Therefore, surgery is more suitable for these patients. In total, 18 cases received surgical treatment, 12 embolization, and 5 combination therapy, and 1 patient refused intervention.[1-10]

Our patient continued to have intermittent abdominal pain after the first arterial embolization. The CT scan showed multiple AVM shunts and severe portal hypertension. In our patient, arterial embolization was not successful, and a dual arterial and venous approach led to successful exclusion of the AVF. Combined inflow and outflow embolization resulted in the patient's portal venous pressure dropping from severe portal hypertension to normal range. Follow-up imaging showed no evidence of arteriovenous shunting or portal hypertension. In retrospect, we should have considered combined arterial and venous intervention at the index operation. Venous embolization, however, led to anticipated venous congestion of the sigmoid colon, eventually necessitating segmental colonic resection.

CONCLUSIONS

Splanchnic AVFs and AVMs can be a rare cause of unexplained portal hypertension and high-output cardiac failure. This case highlights that treatment with dual arterial and venous embolization may be needed to manage severe portal hypertension secondary to this rare pathologic process. Close outpatient monitoring for treatment failure and recurrence is necessary for these patients.

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