CASE REPORT

Idiopathic splenic arteriovenous fistula without splenic artery aneurysm

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SUMMARY

Splenic arteriovenous fistula (SAVF) without a splenic arterial aneurysm is a rare entity and may either be congenital or acquired. Acquired SAVFs are commonly due to adjacent splenic arterial aneurysm rupture, trauma or infection. We report a large idiopathic SAVF in a 50-year-old woman who had no symptoms. CT showed that the fistula was around the splenic hilum and no other specific findings. SAVF has a risk of sudden onset of portal hypertension and it should be treated as soon as diagnosed. Endovascular treatment for SAVF is now being performed successfully with increasing frequency, however, there is still no definitive evidence of treatment for SAVF. Treatment options should be determined appropriately depending on individual case characteristics and vascular anatomical variations. Our case was treated successfully with splenectomy, and long-term results are good without need of reintervention.

BACKGROUND

Splenic arteriovenous fistula (SAVF), an unusual lesion, may be either congenital or acquired. Congenital SAVFs occur most frequently in patients with Ehlers-Danlos or Rendu-Osler-Weber syndrome.¹ Acquired SAVFs are commonly due to blunt trauma, surgery or a penetrating injury that results in direct arteriovenous communication. Large SAVFs are known to cause portal hypertension, which may result in gastrointestinal bleeding. Surgical treatment and transarterial embolisation for SAVF have been reported to be equally successful. We describe a patient who had a large SAVF without a splenic artery aneurysm (SAA) or any of the symptoms or conditions commonly associated with a SAVF.

CASE PRESENTATION

A 50-year-old woman (weight, 45 kg; height, 153 cm) was referred to our institution for management of a SAA, the presence of which was suggested by an ultrasound examination. The patient had previously delivered two children, with both deliveries being normal. She had no symptoms of illness. There was no history of liver and pancreatic disease or alcohol abuse. She had never experienced abdominal trauma, and had never been submitted to abdominal surgery. A physical examination yielded the following: pulse, 92 bpm; blood pressure, 103/62 mm Hg; and body temperature, 36.8°C. The patient's abdomen was not distended, her liver was of normal size on percussion and her spleen was not palpable. Slight bruits were detected in her left

upper abdomen. The patient had no clinical or laboratory findings indicative of portosystemic encephalopathy or portal hypertension.

INVESTIGATIONS

Enhanced CT revealed enlargement of the splenic vein but no varices, ascites, changes in liver parenchyma, or splenomegaly. Transfemoral coeliac arteriography showed a tortuous splenic artery, without dilation (figure 1A), and premature filling

Figure 1 Selective coeliac axis angiography findings. (A) Tortuous splenic artery without dilatation. Splenic arteriovenous fistula is shown (arrow). (B) Early phase shows premature filling of the enlarged (4 cm) splenic vein (arrowheads).

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of an enlarged (>4 cm) splenic vein (figure 1B), findings highly suggestive of a SAVF.

TREATMENT

Because the fistula was located around the splenic hilum, the patient underwent laparotomy through a transverse incision. At operation, neither ascites nor collateral vessels were observed and the liver appeared normal. The lesser sac was opened by lifting the stomach, and proximal control of the splenic artery was obtained by encircling its origin. The splenic vein was dilated to 4 cm proximal to the splenic hilum, but there was no aneurysm in the splenic artery. When the splenic artery was ligated just proximal to the venous dilation, it shrunk immediately (figure 2).

The apparently idiopathic SAVF was treated with splenectomy. A macroscopic finding was shown in figure 3. A pathological examination of the resected specimen showed a fistula between splenic artery and dilated splenic vein. Although medial fibrosis was observed in the splenic artery and in the vein, there were no signs of aneurysm, haemangioma, or mycotic infection.

OUTCOME AND FOLLOW-UP

The patient's postoperative course was uneventful and she was discharged 7 days after surgery. She remains well and has been without need of additional treatment for more than 5 years.

DISCUSSION

The first SAVF reported was found during an autopsy in 1886. SAVFs are thought to arise from rupture of a pre-existing SAA into the adjacent splenic vein due to high intra-abdominal pressure during pregnancy or labour, or abdominal trauma, surgery or mycotic infection.^{2–5} Approximately 80% of SAVFs occur in women, many of whom are multiparous.⁶ Congenital cases are characterised by a tangle of tortuous vessels of small diameter, and usually have one or more large communications between the artery and the vein. Our patient with a SAVF had no history of trauma, abdominal surgery, aneurysmal changes in the splenic artery or signs or symptoms of infection. Therefore, the aetiology of her condition was unknown.

A SAVF may exist for a substantial period of time without causing symptoms, but the haemodynamic changes that result from the arteriovenous shunt can cause a sudden increase in portal vein pressure.⁴ On the other hand, portal hypertension due to chronic liver disease develops gradually.⁷ Because of the risk of rapid onset of portal hypertension in patients with a

Figure 2 At operation, the splenic artery was ligated and divided (arrows) just proximal to the venous dilation (arrowhead).

Figure 3 A fistula (arrowhead) between the splenic artery (black arrow) and splenic vein (white arrow) is visible in the surgical specimen.

SAVF, all detected SAVFs should be excised or embolised, even if the patient is asymptomatic.⁸ Signs and symptoms of SAVF-related portal hypertension include ascites (35% of patients), oesophageal or gastric varices (52%) and splenomegaly (55%).^{2 5 6 9} Variceal bleeding, which occurs in about 45% of patients,⁶ is the most life-threatening manifestation of a SAVF. Diarrhoea, perhaps caused by congestion of the gastrointestinal vasculature, is uncommon. An abdominal bruit, usually of the machinery type, is heard over the left flank in only about 30% of cases,¹⁰ whereas a continuous machinery bruit is often present in the left flank (61% of patients).^{8 11} SAVF-associated congestive heart failure has also been reported.⁸

Prevention of SAVF-related portal hypertension, hepatosclerosis and cirrhosis, depends on early diagnosis and treatment of the lesion. Until recently, the standard technique for detecting a SAVF was coeliac or splenic arteriography. Now, however, ultrasonography or CT scanning can be used to establish the diagnosis.^{1 3–5 12} When a bruit is heard over the left flank, colour Doppler ultrasonography should first be performed to characterise the direction and velocity of blood flow. Dynamic dual-phase multidetector CT angiography should then be employed to delineate the morphological characteristics, number, location and extent of the arteriovenous connections and thereby facilitate treatment planning.

The traditional treatment for a SAVF is open or laparoscopic splenectomy.^{4 13–15} Endovascular treatment (EVT) is now being performed successfully with increasing frequency and is the first choice of treatment.^{4–6} Selective coil embolisation of SAVFs has been reported,^{4 13 16} as has exclusion of the fistula by using a covered graft or Amplatz occlusion device.⁶ Nevertheless, the treatment option is still based on individual decisions depending on patient characteristics and vascular anatomical variation. As of now, there is no definitive evidence of treatment for SAVF, because it is rare. Recently, Hogendoorn *et al*¹⁷ published a

review article of treatment for SAA. They demonstrated that surgical treatment revealed good long-term results but high pre-operative mortality. In contrast, EVT has shown low short-term morbidity but a high reintervention rate. A postembolisation syndrome, such as portal vein thrombosis, is one of the complications after EVT. Ding *et al*¹⁶ reported a portal thrombosis case after EVT for SAVF though low-molecular-weight heparin was used. Preservation of the spleen has been thought to be important, but there is no large scale of study assessing the effect of splenectomy on the immune system. The effect of splenic artery embolisation on splenic immune function was also not well understood.¹⁸ In addition, Nakae *et al*¹⁹ have shown no difference in antipneumococcal antibodies after the treatment between splenectomy and splenic artery embolisation.

In our patient, insertion of a covered graft was not considered because of the location of the fistula and tortuosity of the splenic artery. We also did not perform transcatheter arterial embolisation because it carries a risk of abscess formation caused by splenic segmental infarction and the risk is increased in patients who have a splenic hilar fistula.^{13 16 17} In addition, our patient was not at high risk of surgery, therefore we deemed complete resection of the SAVF, and we thought that it was important to obtain a pathological evaluation of the surgical specimen. Our patient's postoperative short-term and long-term course is good, and she remains well without need of reintervention.

Learning points

- Splenic arteriovenous fistula (SAVF) should be treated as soon as diagnosed.
- Treatment options for SAVF should be determined appropriately depending on individual case characteristics and vascular variations.
- Splenectomy still remains one of the treatment options for good risk patients whose SAVF is located around the splenic hilum.

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