Splenic vein aneurysm in association with extrahepatic portal hypertension

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Splenic vein aneurysm (SVA) is an extremely uncommon condition with only 50 cases having been reported in the literature.¹⁻⁵ We report a unique case of SVA in a child with extrahepatic portal hypertension. Since the condition was correctly diagnosed on preoperative computed tomography-assisted splenoportovenography (CTSPV), a planned surgical intervention was undertaken.

Case

A 12-year-old male child with extrahepatic portal hypertension was referred to our institution for a lienorenal shunt procedure. He had received sclerotherapy for esophageal varices, but his symptoms had continued unabated. On examination, there was massive splenomegaly; liver function tests and a coagulogram were within normal limits. Upper gastrointestinal endoscopic examination showed Grade III varices at the gastroesophageal junction.

Color Doppler examination revealed a portal cavernoma and a dilated distal splenic vein; visualization of the proximal part of the splenic vein was sub-optimal due to overlying bowel gas. On helical CTSPV, the spleen was enlarged; a small splenunculus was seen anterior to the splenic hilum. The splenic vein was patent and measured 12 mm in diameter. The main portal vein was blocked and showed evidence of cavernomatous transformation. Prominent collaterals were seen at the lower end of the esophagus, suggestive of esophageal varices. Aneurysmal dilatation of the proximal part of the splenic vein was seen at a distance of about 15 mm from the origin of the splenic vein; this venous aneurysm measured about 22 mm in diameter (Figure 1).

Using a left subcostal incision the abdomen was opened and a splenectomy performed. An SVA of more than 2 cm in diameter was detected behind the pancreatic tail. Distal pancreatectomy along with complete excision of the SVA was performed (Figure 2). The rest of the splenic vein was dissected out from the dorsal side of the pancreas and a central lienorenal shunt was performed. The histopathological examination of the resected specimen was performed and the diagnosis of SVA was confirmed.

Discussion

Abdominal venous aneurysms are rare. In the portal venous system, they could arise from the extra- or intrahepatic portion of the portal vein. While extrahepatic portal venous aneurysm is an uncommon condition, SVA is still a rare entity.² The etiology of splenoportal venous aneurysms is thought to be congenital, secondary to portal hypertension or associated with abnormal weakness of the venous wall due to trauma, inflammation or localized degenerative changes.¹ It has been postulated that a vessel in a congenitally or otherwise weakened area may dilate later in life, either under the influence of normal portal

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Figure 1. (top) CTSPV showing aneurysmal dilatation of the splenic vein (single arrow) and massive splenomegaly. A small splenunculus (double arrow) is seen anterior to the aneurysm.

Figure 2. (bottom) Resected specimen showing excised and opened splenic vein aneurysm (SVA) along with distal pancreatectomy (PT = pancreatic tail).

venous pressure or as a result of portal hypertension, with mechanical forces like blood turbulence playing a major role.⁶

It is important that SVA should be correctly diagnosed and appropriately managed to avoid life-threatening complications that could result from spontaneous rupture of the aneurysm.⁴ Awareness of the high possibility of encountering an aneurysm of the splenoportal axis is necessary when surgical intervention is planned for patients with portal hypertension. Recognizing the condition could also prevent such catastrophic events like obtaining an image

guided biopsy of the lesion. Ultrasound may reveal an echo-free lesion behind the pancreas, but color Doppler examination remains essential to differentiate other cystic lesions from a vascular aneurysm. However, in our patient color Doppler examination failed to reveal SVA, since the region was obscured by overlying bowel gas. With the introduction of helical CT, volumetric acquisition has become possible over a single breath hold, which has drastically reduced examination time. Such rapid developments in CT technology have made CTSPV a reality and now the mainstay in the evaluation of the splenoportal venous axis.⁶ An added advantage of CTSPV over color Doppler examination is in those situations where overlying bowel gas and obesity hinder adequate penetration of the ultrasound beams.⁷ Apart from demonstrating the morphological details of the aneurysm, CTSPV can also depict any intra-aneurysmal thrombus formation.² In our patient, CTSPV clearly demonstrated the SVA, including details of its site, size and relationship with local structures.

Management of patients with SVA varies from non-invasive follow-up to surgical intervention. Patients with incidental and asymptomatic SVA without associated portal hypertension may be spared surgical intervention and subjected to a close follow-up by imaging; patients with SVA and having risk factors for aneurysm rupture such as severe portal hypertension and bleeding tendencies due to liver failure need surgical exploration; prophylactic surgical intervention may also be considered for patients with abdominal pain or with evidence of expanding aneurysms.²⁻⁴ When excision of SVA is difficult, a decompression procedure may be the best option to drain the aneurysm associated with portal hypertension. In our patient, the location of aneurysm did not permit the making of a lienorenal shunt without first performing complete excision of the aneurysm along with distal pancreatectomy. It was then possible to mobilize the splenic vein for designing a central lienorenal shunt.

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