Congenital portosystemic shunt with multiple splenic artery aneurysms: Reversing pulmonary hypertension and preventing aneurysm rupture

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ABSTRACT

We report an unusual occurrence of multiple splenic artery aneurysms and splenomegaly in a young woman with severe pulmonary hypertension, secondary to a congenital portosystemic shunt (CPS). The splenic artery was occluded using an Amplatzer Duct Occluder-II device, and closure of the large intrahepatic CPS was achieved using a muscular ventricular septal defect occluder. There was resolution of splenomegaly with normal pulmonary artery pressures, a few months after the procedure.

Keywords: Aneurysm, device, portosystemic shunt, pulmonary hypertension, splenic artery

INTRODUCTION

Congenital portosystemic shunts (CPSs), although well recognized, are rare causes of secondary pulmonary hypertension (PHT) and pulmonary arteriovenous malformations (PAVMs).[1] Computed tomography (CT) or magnetic resonance imaging to rule out portosystemic shunts (congenital or acquired) now forms part of essential workup in unexplained PHT.

CASE REPORT

A 23-year-old woman with a history of dyspnea and fatigue (WHO Class II) for 2 years and episodes of hemoptysis for 6 months was referred to our unit for PHT workup. Her resting saturation was 86% in room air. Clinical examination was conspicuous for digital clubbing, a loud second heart sound, and splenomegaly. Chest X-ray showed an enlarged main

pulmonary artery, mild right atrial enlargement, and multiple, tiny homogenous opacities in both lung fields. Two-dimensional echocardiogram showed a mildly dilated right heart, moderate PHT (assessed on pulmonary vasodilators), and no intracardiac shunt to explain the cyanosis. The inferior vena cava (IVC) and the visualized portion of the portal vein (PoV) were enlarged, with flow from the PoV to IVC. Agitated saline contrast echo showed opacification of the left atrium after three cardiac cycles, suggestive of PAVM [Video 1]. Contrast CT showed an enlarged spleen (14 cm) supplied by a dilated main splenic artery (6 mm) with multiple aneurysms (largest aneurysm measured 2.6 cm × 2.3 cm) in the retropancreatic, intrapancreatic, splenic hilar, and intrasplenic segments [Figure 1]. An accessory hepatic artery was also noted to arise from the celiac artery. There was a large communication (16 mm × 19 mm) between the IVC and the right PoV, immediately after the bifurcation of the

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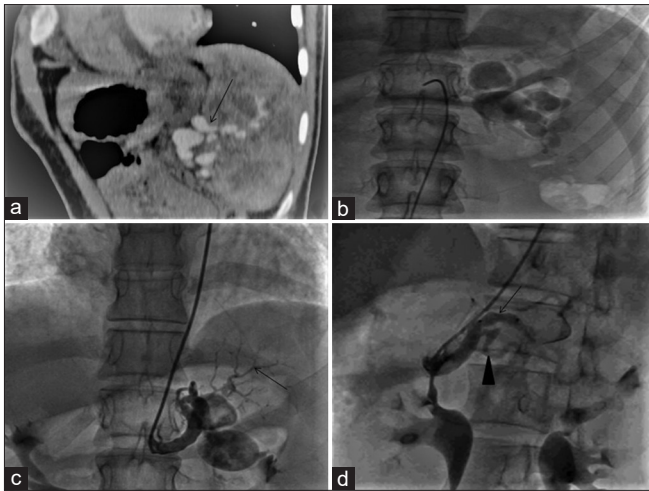
Figure 1: (a) CT sagittal oblique section showing splenic artery aneurysms and splenomegaly, (b) Splenic artery angiogram in AP view using Cobra catheter showing multiple splenic artery aneurysms, (c) Splenic artery angiogram in shallow LAO view; the artery cannulated from the right axillary artery. The arrow points to branches to the spleen from an accessory branch. Multiple aneurysms in the main splenic artery are seen, (d) Splenic artery angiogram post 5/6 ADO-II placement (arrow). Arrowhead points to pancreatic branches from the splenic artery, proximal to the device. CT: Computerized tomography, AP: Anteroposterior, LAO: Left anterior oblique, ADO-II: Amplatzer Duct Occluder-II

main PoV. While small radicles of the left PoV could be demonstrated, the right PoV ended blindly [Figure 2]. The liver parenchyma showed regenerative nodules and no features of cirrhosis. The lung parenchyma showed multiple, small PAVMs, no other pulmonary cause for hemoptysis. There were no features suggestive of hypersplenism (normal retic count and peripheral smear). Her serum ammonia level was 75 mmol/L. While the portosystemic shunt could explain PHT and PAVMs, the cause of splenomegaly could not be explained. Liver function tests and coagulation profile were within normal limits. An upper gastrointestinal endoscopy did not show esophageal or gastric varices to suggest noncirrhotic portal hypertension. A large splenic artery aneurysm (SAA), with its potential for rupture, needed urgent management.

Pulmonary vasodilators were discontinued a month before cardiac catheterization and splenic artery occlusion. Pulmonary artery pressures (systolic – 56/diastolic – 25/mean – 35 mmHg) at catheterization were less than half systemic (systolic – 145/diastolic – 80/mean – 102 mmHg). A right axillary artery access offered coaxial alignment and better engagement of the splenic artery over a femoral arterial approach. The splenic
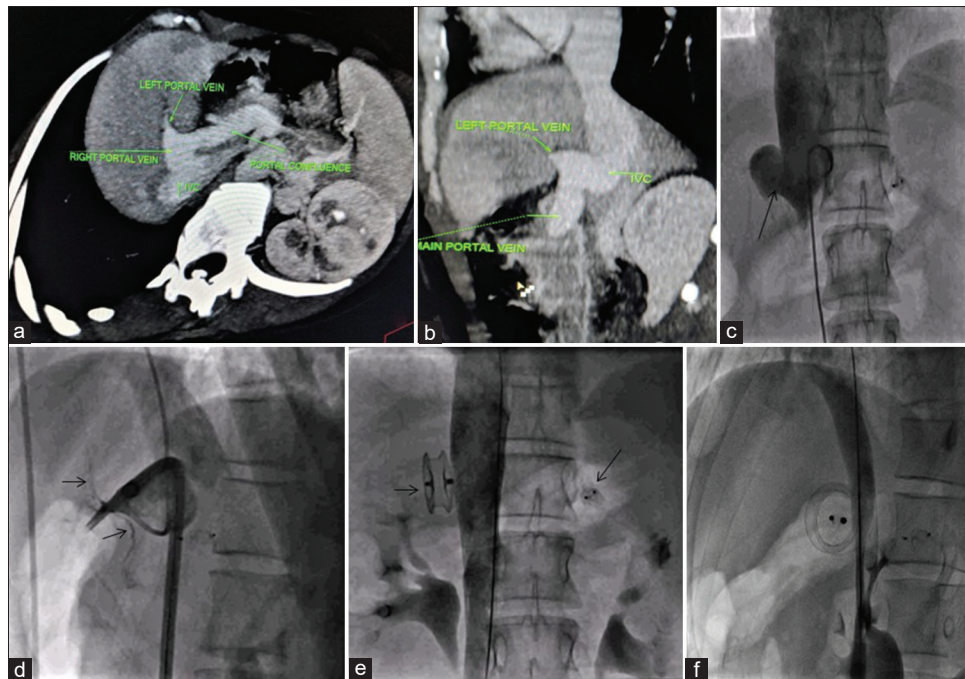

Figure 2: (a) CT axial oblique view depicting the point of communication between the right branch of PoV and IVC immediately after portal vein bifurcation. The labels on the figure from top right and clockwise indicate the left portal vein, portal confluence, IVC and right portal vein respectively, (b) CT coronal oblique view depicting the CPS. The labels on the figure from top right and clockwise indicate the left portal vein, IVC and the main portal vein, (c) Pigtail angiogram of IVC showing the CPS (arrow) in AP view, (d) Cook sheath with end-hole catheter introduced deep into the left portal vein showing hypoplastic portal vein radicles in steep LAO view. Note the JR diagnostic catheter introduced into the CPS from the IJV, (e) IVC angiogram in AP view showing no residual shunt after placement of 20-mm muscular VSD occluder (arrow) in the CPS. Note the ADO-II device in the splenic artery in the background (arrow), (f) IVC angiogram in lateral view showing a well-positioned muscular VSD occluder (en face view) and no residual shunt. CT: computerized tomography, PoV: portal vein, IVC: inferior vena cava, CPS: congenital portosystemic shunt, AP: Anteroposterior, LAO: Left anterior oblique, JR: Judkins right, IJV: Internal jugular venous, IVC: Inferior vena cava, VSD: Ventricular septal defect, ADO-II: Amplatzer Duct Occluder-II

artery was cannulated using a 5F diagnostic Cobra catheter (Cook Medical, Bloomington, USA) and 0.014" Whisper Extra Support Wire. The diagnostic catheter was exchanged for a 5F Judkins right (JR) guide catheter. Contrast injections were given by hand to delineate the aneurysms in anteroposterior (AP) and shallow left anterior oblique (LAO) views [Video 2]. A 5/6 Amplatzer Duct Occluder-II device (retention skirt diameter of 11 mm; twice the splenic artery dimension) was deployed in the main splenic artery just proximal to the first aneurysm and distal to the origin of a large pancreatic branch and a separate superior branch traversing to the spleen [Video 3]. The patient was monitored postprocedurally with serial serum amylase levels to rule out pancreatic ischemia.

She presented 3 months later to the hospital with features of severe PHT (TR jet velocity of 4.2 m/s), right ventricular dysfunction, and cardiogenic shock. Following initial stabilization with intravenous fluids, inotropes, and oral sildenafil, she underwent closure of the CPS [Video 4].

Right femoral arterial, right femoral venous (RFV), and right internal jugular venous (RIJV) access was obtained. The portal venous pressure (7 mmHg) was monitored from the RFV using a 5F JR catheter. The PoV was entered from the RIJV using another 5F JR diagnostic catheter and 0.025" exchange length J-tipped Terumo wire. This was subsequently exchanged for a 0.035" Amplatz extra stiff wire to facilitate the delivery of an 11F Cook sheath [Video 5]. Contrast injections were given through the sheath in LAO 60°, LAO 80°, and AP views to confirm the position of the sheath tip in the CPS. A 20-mm muscular ventricular septal defect occluder (4 mm larger than the shunt dimension) (Lifetech Scientific Inc., Shenzhen, China) was deployed after angiographic confirmation. The hepatic venous wedge pressure (surrogate of portal venous pressure) stayed 7 mmHg, before and after device release. Since the device was deployed entirely within the malformation, a 22 mm × 70 mm self-expanding, uncovered Wallstent (Boston Scientific, USA) was positioned in the involved segment of the IVC (20-mm diameter) to prevent any remote chance of device embolization to the right heart [Video 6]. The placement of a self-expanding stent was not part of the initial procedural plan. As the left renal vein entered the IVC 1.3 cm distal to the CPS, a covered stent was not considered for closure due to insufficient landing zone. She was discharged on oral sildenafil. Her saturation improved to 98% over the next 6 months, and there was no evidence of PHT on follow-up echocardiograms [Video 7]. She was taken off PHT treatment and has remained asymptomatic for over 2 years now. Follow-up ultrasound abdomen showed a well-positioned device and stent in the IVC and no splenomegaly (11 cm).

DISCUSSION

The report serves to highlight a rare association and the importance of identifying portosystemic shunts as a potential cause of treatable PHT. Our patient had an intrahepatic CPS Type 1.[2] There are only few reports of SAA with CPS.[3-5] Indications for splenic artery occlusion in the setting of aneurysms include a large aneurysm (>2.5 cm in diameter), pseudoaneurysm of the artery, persistent abdominal pain, female sex, and before liver transplant.[6] Current guidelines recommend closure of all CPSs persisting beyond infancy to prevent complications such as hepatopulmonary syndrome, porto-PHT, hyperammonemia, and hepatoblastoma.[7,8] Treatment of SAA and CPS at the same time would have significantly increased procedural duration. We chose to initially occlude the splenic artery to prevent a potentially fatal aneurysm rupture. A reduction in splenic artery flow would also reduce the portosystemic shunt fraction. While the presence of hypoplastic portal radicles has permitted safe closure of Abernethy malformations without significant elevation of portal pressure (as the radicles tend to grow over time), the time taken for PoV regeneration is difficult to determine. Despite the presence of hypoplastic left PoV radicles, initial closure of the CPS could have resulted in transient increases in PoV pressure, congestive splenomegaly, and aneurysm rupture. Due to the presence of an additional branch to the spleen proximal to the device, splenic infarction was avoided. As splenomegaly resolved following arterial occlusion and shunt closure, we postulate that splenomegaly was due to excessive flow into an enlarged, tortuous splenic artery with multiple aneurysms. The portosystemic shunt prevented any efferent resistance to increased portal venous return, and increased splenic arterial flow over the years could have led to organ enlargement. Identification of a genetic cause for SAAs was not pursued in our patient. Previous reports of CPS and PHT have not found a significant long-term benefit of pulmonary vasodilators.[9,10] This was expected, as drugs used for PHT do not alter underlying pathophysiologic mechanisms. Although follow-up is limited to 2 years, the report highlights the role of closure of CPS in reversing the pathophysiologic process.[11] While long-term follow-up is imperative, such an interventional strategy might permit discontinuation of pulmonary vasodilators in selected patients.

CONCLUSION

Transcatheter treatment of two different pathologies helped in the resolution of cyanosis, PHT, and splenomegaly and aborted the risk of SAA rupture.

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