Congenital Extrahepatic Portosystemic Shunt with Multiple Visceral Aneurysms

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We herein report the case of a 48-year-old woman with a congenital extrahepatic portosystemic shunt (CEPS) who had multiple visceral aneurysms. She presented with dyspnea on exertion. On admission, her heart rate was 71 bpm and blood pressure 102/62 mmHg. Her arterial oxygen saturation was 95%. Cardiac auscultation revealed no loud S2 or murmurs. Chest X-ray and enhanced computed tomography (CT) revealed the dilation of the pulmonary arteries (PA) (Picture 1). Enhanced CT revealed direct flow into the inferior vena cava (IVC) from the portal vein (PV), with multiple artery aneurysms (Picture 2; HA: hepatic artery, RA: renal artery, SA: splenic artery). The hepatic vein was anatomically normal. Pulmonary hypertension was confirmed by a right heart catheterization. Treatment with oral ambrisentan was started after the patient refused liver transplantation.

Cases of CEPS complicated with multiple aneurysms are rarely reported in adults. The mechanism of aneurysm formation may be similar to the one responsible for visceral aneurysm formation in portal hypertension (1). It is unclear how or when the aneurysm formation occurred during her life. Therefore, close observation may be necessary, especially of adult CEPS patients who are not scheduled to undergo liver transplantation.

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Reference