Esophageal Varix in an Adult Fontan Survivor

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A 27-year-old woman was admitted to our hospital for the investigation of the causes of melena and anemia. At seven years of age, she had undergone a Fontan operation of total cavo-pulmonary connection (TCPC) for right isomerism, dextrocardia, single atrium, single ventricle, and common atrioventricular canal (Picture A). She had been treated with warfarin after the operation and had been in good condition. She did not drink alcohol and had no history of viral hepatitis. The serum aspartate aminotransferase (40 IU/L), alkaline phosphatase (424/IU/L), and γ-glutamyltransferase (92 IU/L) levels were slightly increased, although other hepatic-biliary enzymes and N-terminal pro-brain natriuretic peptide levels were normal. Transthoracic echocardiography showed an accelerated blood flow from the pulmonary vein into the atrium with a maximum velocity of 2.0 m/s with a normal ventricular systolic function and mild regurgitation of the atrioventricular valve. Gastroscopy revealed varix from the mid-portion of the esophagus to the cardiac part of the stomach, although active bleeding was not detected (Picture B, arrows). Contrast-enhanced chest and abdominal computed tomography showed hepatomegaly with multiple hyperplastic nodules, which suggested congestive liver (Picture C) and no thrombus in TCPC or the bilateral pulmonary arteries. We must consider chronic congestive hepatopathy, including portal hypertension, as potential causes of gastro-esophageal varices and regenerative liver nodules in Fontan survivors (1, 2).

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