A 44-year-old man with the chief complaint of atypical chest pain, was referred to the echocardiography laboratory for further evaluation of an interventricular septal (IVS) defect. Physical examinations yielded no significant findings. Transthoracic echocardiography showed mild left ventricular (LV) enlargement with global hypokinesia and mild systolic (ejection fraction: 45%) and diastolic dysfunction. The right ventricle (RV) was also mildly enlarged and demonstrated mild systolic dysfunction. In the short-axis view at the level of the mitral valve, there was continuous, turbulent, high-gradient flow (peak pressure gradient: 64 mmHg) originating from the attachment point of the IVS and the inferior wall of the RV. In addition, the left main origin was dilated (16 mm) (Fig. A) and 1 branch had a tortuous course. The branch traveled from the antero-lateral LV wall to the posteroinferior wall, reached the inferior wall of the RV, and then entered the RV cavity below the posterior tricuspid valve leaflet. These findings were suggestive of a left coronary fistula to the RV. Further evaluation via transesophageal echocardiography substantiated the tortuosity in the left circumflex artery and connection to the RV. No other congenital cardiac defects were found (Figs. B and C). Selective coronary angiography demonstrated the tortuous course of the left circumflex artery, which connected to the right heart; however, an accurate connection site was not clearly confirmed. Ventriculography did not show the IVS defect (Fig. D). The patient was referred for corrective surgery. A coronary artery fistula to the cardiac chambers has a continuous flow with a high gradient; however, when the entrance site is near the IVS, it can be mistaken for a small IVS defect with a high-gradient systolic flow. Accordingly, the flow pattern (continuous vs. systolic) can help to differentiate between these 2 conditions.

CASE IMAGE

Figures—(A) Connection between the left circumflex artery and the right ventricle resembling an interventricular septal defect as seen on transthoracic echocardiography. RA: Right atrium; RV: Right ventricle. (B) Connection between the left circumflex artery and the right ventricle as seen in the right ventricular inflow-outflow view of 2-dimensional transesophageal echocardiography. RA: Right atrium; RVOT: Right ventricular outflow tract. (C) Connection between the left circumflex artery and the right ventricle as seen in the right ventricular inflow-outflow view of color Doppler transesophageal echocardiography. RA: Right atrium; RVOT: Right ventricular outflow tract. (D) Tortuous course of the left circumflex artery viewed with selective coronary angiography (arrow).