A 63-year-old woman presented with a 12 months history of progressive retro-sternal chest pain. Symptoms worsened on exertion but were ultimately relieved after administration on nitrates, suggesting typical cardiac-related angina. Physical examination was unremarkable. Chest radiography and electrocardiography revealed no abnormalities. Transthoracic echocardiography showed a normal-sized heart and regular ventricular function without wall motion abnormalities. Serum cardiac enzymes were in the normal range. Conventional coronary angiography found no atherosclerosis but revealed a large fistula arising from the medial segment of right coronary artery (RCA). However, the site of drainage was not definitely identified. Therefore, contrast-enhanced electrocardiogram-gated 320-detector row computed tomography (CT) was performed and clearly demonstrated a tortuous and markedly dilated proximal RCA giving rise to a large fistula draining into the superior vena cava (SVC). Electrocardiogram-gated magnetic resonance imaging exhibited vigorous flow of 0.84 L/min and a left-to-right shunt volume of 1.2:1. Additional intra-operative multiplane transesophageal echocardiography also demonstrated a large fistula of the RCA to the SVC, confirming the CT and magnetic resonance imaging findings. On the basis of patient’s symptoms, surgical ligation of the fistula was recommended. Intra-operatively, a large fistula was seen and a palpable thrill noted. The proximal end of the fistula was dissected near the origin from the RCA, and both the proximal and the distal part of the fistula were ligated and carefully sutured. The patient returned to the intensive-care unit, recovered smoothly from surgery, and was doing well 6 months after discharge.