Infective endocarditis of the aortic valve in a patient with a coronary artery fistula

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A 38-year-old man was admitted to our institution because of acute biventricular heart failure. He reported dry cough, reduced exercise tolerance, exertional dyspnea, and a single episode of fever 4 months earlier, with a sudden deterioration 2 weeks before admission. Laboratory tests showed an increased leukocyte count, elevated C-reactive protein level, and evidence of renal and liver failure. Blood cultures were negative. Trans-thoracic echocardiography revealed tricuspid aortic valve, ruptured left coronary leaflet with a mobile vegetation protruding into the left ventricular outflow tract (15 × 3 mm), and significant aortic regurgitation. The left ventricle was dilated (71 mm) with reduced ejection fraction (EF, 35%). Transesophageal echocardiography demonstrated marked dilation of the left main coronary artery (LMCA), up to 1 cm with turbulent flow. Invasive coronary angiography showed ecstasia of both LMCA and the left circumflex artery (Cx), with a communication between the Cx and right atrium (RA) (FIGURE 1A). Computed tomography (CT) revealed a fistula between the Cx and coronary sinus (FIGURE 1B, C). A surgical evaluation confirmed inflammatory destruction of the left coronary cusp and a coronary artery fistula (CAF) between the Cx and right atrium (FIGURE 1D). The patient underwent uneventful mechanical aortic valve implantation (Sorin 27) and occlusion of the CAF.
At 5 months, the patient was free from symptoms, and EF significantly improved (51%). A follow-up CT scan confirmed the epicardial closure of the fistula with normal flow in the Cx (FIGURE 1E).

This case illustrates an uncommon association between a CAF and infective endocarditis of the aortic valve. The CAF is a rare congenital vascular anomaly that constitutes from 0.1% to 0.4% of cases with congenital heart disease and is an incidental finding in 0.1% of coronary angiograms. CAFs are mostly asymptomatic; however, some of them may cause myocardial ischemia, arrhythmias, or congestive heart failure and are associated with risk of complications such as infective endocarditis, coronary rupture, or sudden death. Infective endocarditis has been reported to occur in 4% to 10% of patients with a CAF. In the literature (publications between 2000 and 2015), we identified 18 reports of patients with a CAF complicated by infective endocarditis. In our patient, in the absence of positive blood culture results, we were unable to determine the cause of infective endocarditis. Therefore, the CAF could either cause or contribute to the rapid progression of the disease with a deterioration of the patient’s clinical status.

REFERENCES