Bilateral Coronary Artery Fistulas
With Multiple Sites of Drainage

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A 49-year-old woman presented with bilateral coronary fistulas with multiple sites of drainage. She had been referred to hospital for evaluation of a cardiac murmur, and a coronary arteriogram revealed multiple coronary fistulas. One fistula originated from the proximal part of the left anterior descending coronary artery and connected to the main pulmonary artery. Three coronary artery fistulas arose from a conal branch of the right coronary artery and drained into the main pulmonary artery, the right atrium and the great cardiac vein. This is the first reported case of bilateral coronary fistulas with multiple sites of drainage. (Jpn Circ J 1998; 62: 783–784)

Key Words: Coronary artery fistula; Multiple drainage

A coronary artery fistula is an uncommon congenital malformation of the heart. The majority of these anomalies involve only 1 coronary artery, and only rarely are 2 coronary arteries involved. It has been reported that approximately 5% of all coronary artery fistulas occur bilaterally. Bilateral coronary artery fistulas usually communicate with a single site in a cardiac chamber or vascular structures, and fistulas with multiple sites of drainage are extremely rare. This report describes a very rare case of bilateral coronary artery fistulas draining into the main pulmonary artery, the great cardiac vein and the right atrium.

Case Report

A 49-year-old woman was referred to hospital for evaluation of a cardiac murmur. On admission, her blood pressure was 148/82 mmHg. The pulse rate was 68 beats/min and regular. A high-pitched continuous murmur of grade 3/6 was heard best in the third intercostal space at the left sternal border. The laboratory data were all normal. The electrocardiogram was normal as was the chest X-ray. No significant ST–T wave changes were found at a maximal heart rate of 181 beats/min during treadmill exercise testing. Stress thallium-201 myocardial scintigraphy showed no perfusion defects.

At cardiac catheterization, the pressures, cardiac output and left ventricular function were all normal. Selective coronary arteriography revealed no fixed narrowing of any coronary artery, but showed 4 coronary artery fistulas. One fistula originated from the proximal part of the left coronary anterior descending coronary artery and connected to the main pulmonary artery (Fig 1). Three coronary artery fistulas arising from a conal branch of the right coronary artery also were noted (Fig 2). These fistulas drained into the main pulmonary artery, the right atrium and the great cardiac vein. The calculated pulmonary-to-systemic flow ratio (Qp/Qs) was 1.3:1.0. She received no medication and advice for endocarditis prophylaxis was given.

Discussion

The actual incidence of bilateral coronary fistulas is not known. Levin et al reviewed 363 cases of coronary fistulas and noted that such fistulas were single in the majority of cases, and only 19 cases (5%) were multiple, originating from both coronary arteries. The coronary artery fistulas drained into the right ventricle in 41%, the right atrium in 26%, and the pulmonary artery in 17% of their cases. Coronary artery fistulas to the cardiac chambers arise due
to failure of the intramyocardial sinusoids to obliterate. In contrast, coronary artery fistulas to the pulmonary artery arise due to abnormal communication between the coronary bud of the pulmonary trunk and the coronary artery. However, Levin et al did not find a case in which the fistulas drained into multiple sites of a cardiac chamber and vascular structures. In our case, there were multiple fistulas arising from the right coronary artery and left anterior descending coronary artery, which drained into the main pulmonary artery, the right atrium and the great cardiac vein. To our knowledge, this is the first reported case of bilateral coronary fistulas with multiple sites of drainage.

The pathophysiologic consequences of coronary artery fistulas are related to the amount of oxygenated blood shunted away from the myocardium. In our patient, myocardial ischemia could not be detected by treadmill exercise testing or by stress thallium-201 myocardial scintigraphy. Therefore, she received no medication, and advice for endocarditis prophylaxis was given.

Fig 2. Selective right coronary arteriogram in the right anterior oblique projection (A) and the left anterior oblique projection (B) showing 3 coronary artery fistulas arising from the conal branch of the right coronary artery and draining into the main pulmonary artery, right atrium and great cardiac vein.

References