Right Coronary Artery Originating From Distal Left Circumflex Artery in a Patient With an Unusual Type of Isolated Single Coronary Artery

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SUMMARY

Single coronary artery is a rare congenital anomaly and is commonly associated with other congenital cardiac malformations. This report describes a 42-year-old man with an isolated single coronary artery, in whom the right coronary artery did not originate from the aorta but rather from the distal left circumflex artery. This patient did not have any other cardiovascular anomaly. However, he experienced angina pectoris and evidence of myocardial ischemia. Coronary angiography revealed insignificant coronary artery stenosis. He received medical treatment and responded well. An isolated single coronary artery is extremely rare, and this case may be only the 12th case reported in the literature. (Jpn Heart J 2004; 45: 337-342)

Key words: Angina pectoris, Congenital anomaly, Coronary artery disease

SINGLE coronary artery (SCA) is a rare congenital anomaly of the coronary arteries where only one coronary artery arises from the aortic trunk by a single coronary ostium, supplying the entire heart. SCA is commonly associated with other severe congenital cardiac malformations.1) The incidence of isolated SCA is only 0.024% to 0.066% in the general population2-5) undergoing coronary angiography. The isolated single coronary artery is rather rare, especially if the right coronary artery arises from the distal left circumflex artery. To our knowledge, only 11 cases of this particular congenital anomaly have been reported previously (Table). We report a case with the unusual feature of an isolated single coronary artery and present a review of the relevant literature.
A 42-year-old Taiwanese man had a history of occasional exertion-induced chest pain for 2 years. He denied a history of hypertension, diabetes mellitus, or hyperlipidemia, but had a history of smoking 1 package of cigarettes daily for twenty years. His family history included diabetes mellitus, hypertension, and myocardial infarction. Physical examination at admission revealed a blood pressure of 130/82 mmHg, heart rate of 86/min, respiration rate of 18/min, and no other remarkable abnormal findings. Electrocardiography showed left ventricular hypertrophy by voltage criteria but without ST depression or T-wave inversion. A lipid profile revealed total cholesterol of 209 mg/dL, high density lipoprotein-cholesterol of 41 mg/dL, low density lipoprotein-cholesterol of 138 mg/dL, and triglycerides of 120 mg/dL. Fasting blood glucose was 96 mg/dL. Other biochemistry, blood, and urine test values were within normal limits. Treadmill exercise testing with the Bruce protocol displayed horizontal depression of ST-segments for 1.5 mm over V4 to V6, II, III, and aVF from stage III that recovered 4 minutes after exercise (Figure 1). Thallium-201 heart scan demonstrated anteroapical myocardial ischemia. Subsequent coronary arteriography revealed normal hemodynamics and a left ventricular ejection fraction of 68% with a normal pattern of contractility. The left coronary artery angiography showed normal features of the left main coronary artery. The left anterior descending coronary artery and its branches were relatively small. In addition, the left circumflex artery was a dominant vessel with an aberrant branch from the distal left circumflex artery which crossed the crux and continued to the right atrioventricular groove. Finally, the vessel terminated near the right sinus of Valsalva. There was
a 40% diameter stenosis at the beginning of the aberrant right coronary artery. The attempt to engage the right coronary catheter to the right coronary artery was unsuccessful, and aortography revealed the absence of the right coronary ostium (Figure 2). The patient was advised to quit smoking. He was treated with aspirin, calcium channel blockers and simvastatin, and responded well. He has been followed up at the outpatient clinic uneventfully for more than 1.5 years.
Figure 2. **Upper panel:** Features of the left coronary artery in the right anterior oblique (RAO) view with a caudal projection. A small left anterior descending artery which terminated before the apex was observed. A dominant left circumflex (LC) artery giving off branches to the right atrioventricular groove and 40\% stenosis at take-off of the aberrant vessel (arrow). **Middle panel:** Left coronary angiogram, left anterior oblique (LAO) view. Note the small left anterior descending artery and the distribution of the predominant LC artery. A mild atherosclerotic lesion was observed (arrow). **Lower panel:** Aortography in LAO projection showing the absence of the right coronary artery (arrow). LAD = left anterior descending coronary artery; LC = left circumflex coronary artery.
DISCUSSION

This report deals with a very unusual condition of an isolated single coronary artery with the right coronary artery arising from the distal left circumflex artery. Lipton, et al classified single coronary arteries into nine patterns according to the site of origin and anatomical distribution of the branch of the coronary artery.2) From their classification, a first division is made between the R-type (right type) and the L-type (left type) according to the site of origin of the artery, ie, in the right or left sinus of Valsalva. Next, the artery was designated as group I, II, or III depending on its anatomical course. The final designation refers to the relationship between the anomalous coronary artery and the aorta or pulmonary artery. The letters “A”, “B”, and “P” refer to “anterior”, “between”, and “posterior” patterns. Yamanaka and Hobbs modified the Lipton classification by adding “septal” and “combined” types, designated as “S” and “C”.6) The present case was classified as an “L-I” pattern, which is a markedly dominant left circumflex artery but absence of a right coronary artery. To our knowledge, only 11 cases of this peculiar anomaly have been previously reported. This case therefore may be only the 12th patient with an isolated single coronary artery having an L-1 pattern (Table I).

A single coronary artery can result in the development of cardiac ischemia, cardiomyopathy, and congestive heart failure.7) Myocardial ischemia may occur if there is only insignificant or absent coronary artery narrowing. Shirani and Roberts reported that 15% of patients with a single coronary artery had myocardial ischemia due to the direct consequence of the coronary anomaly.8) Vrolix and colleagues reported a patient with a single coronary artery who had 80% stenosis of the left circumflex artery in whom coronary artery bypass surgery but not coronary angioplasty was performed.4) The surgical treatment was preferred because coronary angioplasty could compromise the other important branches. Nevertheless, new advancements in coronary interventional devices and techniques have made angioplasty another feasible therapeutic choice. Takano, et al reported a 35-year-old man with an isolated single coronary artery who suffered an acute myocardial infarction. They found a culprit lesion at the middle segment of the left anterior descending artery using intravascular ultrasound and coronary angiography. This patient underwent successful coronary stent deployment.9) Yamanaka, et al found that the L-I group of a single coronary artery usually has a benign clinical course.6) However, according to data in the Table, 4 out of 12 patients who had a right coronary artery originating from the distal left circumflex artery required CABG. The incidence of CABG is about 33%. Due to limited information, it is difficult to make a conclusion regarding whether patients with the L-1 type of an isolated single coronary artery will be high risk or have a benign
course. We postulate that the prognosis of an isolated single coronary artery depends upon the extent of the underlying coronary artery stenosis. Patients with an insignificant atherosclerotic lesion and without other associated abnormalities most likely will have a benign course. In the present case, the exertional chest distress suggested myocardial ischemia, as evidenced by the positive treadmill and thallium exercise test findings. The anteroapical ischemia may be due to the small left anterior descending artery which terminated before the apex. Hypoperfusion of the right atrium and right ventricle by this special anatomy could compromise the myocardium. The borderline lesion of the aberrant right coronary artery (407. stenosis) is not a big problem and the interventional therapy is therefore not considered. Due to mild symptoms and mild atherosclerosis, our therapeutic strategy is aggressive medical therapy and close follow-up. This patient remains free of symptoms to date.

In summary, we report a patient with an unusual type of isolated single coronary artery in whom the right coronary artery arose from the distal circumflex artery. This patient did not have a significant coronary artery stenosis but had experienced angina pectoris and myocardial ischemia. The anatomic variations in the coronary arteries, including the small left anterior descending artery and inadequate branches from the right coronary artery, may have contributed to his myocardial ischemia. He responded well to the medical treatment provided.

REFERENCES