Congenital Absence of the Left Circumflex Coronary Artery Associated With Acute Myocardial Infarction

A Case Report

Nobuyuki Hashimoto, MD; Junzo Nagashima, MD; Osamu Miyazu, MD; Yoshihiro Akashi, MD; Kensuke Kawasaki, MD; Yukiko Imai, MD; Katsuhiko Tsuchiya, MD; Atsushi Ozawa, MD; Takehiko So, MD; Haruki Musha, MD; Masayoshi Sakakibara, MD*; Fumihiko Miyake, MD*

Among the congenital anomalies of the coronary arteries, a left circumflex artery (LCX) defect is extremely rare. A 49-year-old man who developed an acute anterior infarction underwent coronary angiography, which revealed complete occlusion of the left main trunk, but the territory usually supplied by the LCX had been perfused by the superdominant right coronary artery. Treatment of the left main trunk by percutaneous coronary intervention produced a favorable result. Accurate evaluation of the principal vessels and the extent of compensatory perfusion is important when diagnosing ischemic heart disease accompanied by anomalous coronary arteries and for choosing the best treatment modality. (Circ J 2004; 68: 91–93)

Key Words: Acute myocardial infarction; Circumflex coronary artery; Congenital anomaly; Coronary artery; Left main trunk

Congenital absence of the left circumflex (LCX) coronary artery is a very rare vascular anomaly in which the artery fails to develop in the left atrioventricular groove. Yamanaka and Hobbs found it in only 4 of 126,595 patients who underwent coronary angiography and with a frequency of only 0.003% in all the patients and 0.24% in the patients with other coronary artery anomalies, they confirmed that this anomaly is very rare. In previous case reports of absent LCX, there has been an association with systolic click syndrome, ischemic changes in the zone of hypoperfusion, which causes chest pain, and a poor prognosis related to dilated cardiomyopathy. However, its association with acute myocardial infarction (AMI) has not been reported previously and we report our clinical experience of just such a case.

Case Report

The patient was a 49-year-old man who presented with chest pain. In 1993, he had been diagnosed as having hypertension and hyperlipidemia, but did not receive any treatment. Since January 1998, he had suffered from chest pain on exertion and on May 23, he developed chest pain around midday that forced him to rest, but by the next day he was dyspneic and presented to the Emergency Center. His risk factors included hypertension, hyperlipidemia, diabetes mellitus, and smoking. At the time of admission, he was 165 cm tall, weighed 73 kg, had a blood pressure of 214/126 mmHg, and a pulse rate of 110 beats/min (regular). He had clear consciousness and there was no cyanosis. No cardiac murmurs were heard, but there were bilateral moist rales. No edema of the legs was detected.

Admission laboratory tests revealed a white blood cell count of 8,900/μL, red blood cell count of 548 × 10⁴/μL, hemoglobin of 18.4 g/dl, hemafocrit of 55.0%, aspartate aminotransferase of 85 IU/L, alanine aminotransferase of 50 IU/L, lactate dehydrogenase of 554 IU/L, creatine kinase of 827 IU/L, and CK-MB of 74 IU/L, indicating an increase in the concentrations of the myocardial enzymes. The admission ECG revealed ST segment elevation in leads I,
aVL, and V1-5 (Fig 1A). The chest X-ray obtained at the time of admission showed mild pulmonary congestion and a cardiothoracic ratio of 55% (Fig 2). Echocardiography revealed severe hypokinesis of the left ventricular anteroseptal wall and thinning at the apex with dyskinesia.

Clinical Course After Admission

Based on the results of the examinations, the patient was diagnosed as having an acute anteroseptal infarction, but he did not have any ischemic symptoms from 24 h after the onset of chest pain and the myocardial enzyme concentrations were below the peak values. Therefore, primary PCI was not indicated and conservative therapy and general management for heart failure were instituted. On June 9, 1998, the patient underwent coronary angiography. Left coronary arteriography showed complete occlusion of the origin of the left main trunk (LMT), and right coronary arteriography revealed no significant stenosis, but marked development of the posteriolateral branch (4PL). There was good collateral circulation from the right coronary artery (RCA) to the left anterior descending artery (LAD) (Fig 3). Left ventriculography also showed dyskinesia of the left ventricular anterior wall and the apex, with akinnesia of the septum and normal posterior wall motion (Fig 4). Complete obstruction of the LMT often leads to a fatal outcome, but in the present patient, the RCA was well developed and perfused a zone extending to the left ventricular posterior wall, thus preserving cardiac function. Although coronary artery bypass grafting was considered to improve the long-term prognosis, PCI of the LMT was selected because it would be relatively safe owing to the unusual development of the RCA, and early reperfusion was thought likely to improve the short-term prognosis. The guide catheter was an 8F Judkins Left 4.0 and a 0.014-inch guide wire was passed through the guide catheter to the LAD. Using a balloon (1.5 mm in diameter), the LMT lesion was dilated and re-perfusion was achieved. The distal LAD was subsequently visualized, but the LCX was not. Using a larger balloon (3.0 mm in diameter), dilatation was attempted at a maximum of 14 millibars for 60 s, but this resulted in coronary artery dissection. Therefore, coronary stents were inserted across the LMT into the LAD. Two GFX stents (3.0 mm in diameter) were placed using a pressure of 10 bars for 60 s, followed by post-dilatation using a 3.0 mm balloon at 18 bars for 30 s. After stent placement, angiography showed good dilatation of the LMT and LAD, but as we still could not visualize the LCX or its origin (Fig 5), it suggested that there was agenesis of the LCX. No other complications occurred and the course after PCI was uneventful. The patient was discharged on July 8, 1998.

At discharge, the ECG revealed a QS pattern in leads V1 and V2, as well as inverted T waves in leads I, aVL, and V1-5 (Fig 1B). After 3 months, TI myocardial scintigraphy showed an infarct focus in the anteroseptal area on the short-axis, horizontal view, but did not show any obvious defect in the left ventricular posterior wall or the lateral wall.

Discussion

The lack of a coronary artery in the left atrioventricular groove on left coronary arteriography confirms the diagnosis of absent LCX. However, it is necessary to differentiate it from other congenital coronary anomalies, such as single coronary artery, anomalous origin of the coronary arteries, and anomalous origin of the left coronary artery from pulmonary artery (Bland-White-Garland syndrome), as well as from acute myocardial infarction secondary to occlusion of the origin of the LCX.1 In the present case, initial RCA angiography revealed bifurcation of the left atrial artery and a high-position lateral wall artery branching from the peripheral RCA, and it showed that these branches were the source of collateral blood flow to the proximal LAD, although there was no collateral flow from the RCA to the LCX. The coronary sinus was detected near the LAD by enhancement in the venous phase and it extended along the RCA. Follow-up angiography was performed at 3 and 6 months after PCI and confirmed the absence of the LCX. Neither aortography nor pulmonary artery angiography showed other coronary artery anomalies and furthermore, TI myocardial scintigraphy did not show any obvious de-
fect in the territory of the circumflex artery. Based on these findings, congenital agenesis of the LCX and anteroseptal infarction were diagnosed. There was no significant restenosis of the LAD, in which a coronary stent was placed.

When the LCX is absent, RCA dominance is important. Normally, the LCX and RCA run around the atrioventricular groove and form a circle. If a patient has a small LCX, a well-developed RCA ascends the posterior atrioventricular groove beyond the crux and perfuses the left ventricular free wall. In fact, the present patient had a very well-developed, superdominant RCA and in cases with a dominant compensating artery, the absence of a coronary artery is not of clinical significance as long as coronary artery disease does not develop. Although absence of the LCX associated with a poorly-developed RCA needs to be mentioned as a subtype of coronary artery hypoplasia syndrome, there are currently no diagnostic criteria to clearly classify these anomalies. In patients with coronary artery hypoplasia, myocardial fibrosis occurs as a result of chronic myocardial ischemia in the area of hypoperfusion and features of dilated cardiomyopathy are seen, or else congestive heart failure arises from mitral valve incompetence secondary to papillary muscle dysfunction. Although there are currently no angiographic criteria to clearly classify these coronary anomalies, such anomalies have little clinical significance if the patient has a dominant coronary artery that maintains collateral circulation and there is no coronary artery disease, as in the present case. Conversely, if there is hypoplasia and collateral circulation is not formed, evaluation of myocardial damage in the hypoperfused region using echocardiography and nuclear medical studies is important for prognosis and selection of treatment.

In general, patients with congenital anomalies of the coronary artery have a favorable prognosis. In the case of single coronary artery or anomalous origin of coronary vessels, sudden death or ischemic events such as myocardial infarction can occur depending on the distribution of the major arteries. When a coronary artery anomaly is accompanied by impaired cardiac function, it is necessary to consider a diagnosis of coronary hypoplasia. Although the present patient had congenital absence of the LCX, its territory was perfused by a superdominant RCA, so that PCI of the LMT could be performed with relative safety. When diagnosing coronary artery anomalies and selecting treatment, it is important to accurately evaluate the distribution of the major arteries and the extent of compensatory perfusion.

References
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