Case Reports

Congenital Absence of the Left Circumflex Coronary Artery

Tung-Chun LIN,1 MD, Wen-Shin LEE,1 MD, Chi-Woon KONG,1 MD, and Wan-Leong CHAN,1 MD

SUMMARY

We report a rare case of congenital coronary artery anomaly with recurrent chest pain. A 44-year-old Taiwanese female patient presented with exertional chest pain that had lasted for 2 years. An electrocardiogram showed right axis deviation and an rS pattern in leads I and aVL, and an exercise stress test was inconclusive. A thallium-201 myocardial perfusion study revealed perfusion defects in the septal and inferior walls which normalized in the delayed imaging. Coronary angiography revealed the absence of a left circumflex coronary artery and a superdominant right coronary artery with terminal branches supplying the left ventricular inferior and posterolateral walls. An aortogram revealed no evidence of the existence of a left circumflex coronary artery. Administration of nitrates and calcium antagonists could not ameliorate her symptoms, but she remained stable during ordinary activity. In young premenopausal female patients who present with symptoms suggestive of angina pectoris and a positive stress test, congenital anomaly of the coronary artery should be considered and an angiographic study should be performed. (Jpn Heart J 2003; 44: 1015-1020)

Key words: Coronary artery, Left circumflex, Congenital anomaly

CONGENITAL coronary artery anomalies are infrequently seen during coronary angiographic study, and have been reported to occur in 0.64 to 1.3 percent of patients.1 Most patients with a congenital coronary artery anomaly are asymptomatic; rarely, they may present with chest pain and may have myocardial ischemia or other life-threatening conditions. In premenopausal women with chest pain, it is important to differentiate coronary artery disease from other causes, since this symptom may provoke anxiety and fear. We describe an interesting case with chest pain and abnormal thallium-201 myocardial perfusion scan who had congenital absence of the left circumflex coronary artery.
CASE REPORT

A 44-year-old Taiwanese female, a victim of thalassemia minor, was admitted on account of intermittent exertional precordial chest pain for two years and received antiangina medication in a local clinic under the assumption of coronary artery disease. She had no history of systemic disease such as diabetes mellitus or hypertension; no history of cigarette smoking or alcohol drinking; and had regular menses. She visited our outpatient department because of persistent chest pain and exacerbation of symptoms in spite of medication. Physical examination revealed a well-nourished female with a blood pressure of 132/88 mmHg and a pulse rate of 78 beats per minute. Cardiac auscultation revealed regular heart sounds without cardiac murmur or S3. The lungs were clear. Peripheral pulsation of the radial, popliteal, posterior tibial, and dorsalis pedis arteries were normal. Chest X-rays showed normal findings. A 12-lead electrocardiogram showed right axis deviation and an rS pattern in leads I and aVL (Figure 1). An echocardiogram revealed normal left ventricular morphology and no abnormal regional wall motion. A treadmill exercise stress test (Bruce's protocol) was performed and the result was inconclusive. An exercise stress thallium-201 myocardial perfusion study revealed perfusion defects in the septal and inferior walls (Figure 2A, arrow-head) which normalized in the redistribution (delayed) imaging (Figure 2B, arrow-head). Therefore, she was admitted for coronary angiographic study.

Figure 1. Resting 12-lead electrocardiogram showing right axis deviation and rS pattern over leads I & aVL (arrow heads).
Selective coronary angiography was performed via the right femoral approach. Left coronary injection revealed one artery arose from the left sinus of Valsalva and continued as a single left anterior descending artery which gave off a large diagonal branch (Figure 3) to supply the left ventricular lateral wall. No circumflex artery was demonstrated after administration of nitroglycerin at repeated coronary angiograms. The right coronary artery, which arose normally from the right sinus of Valsalva, was a large (super-dominant) artery, crossed along the left atrioventricular groove, and had large branches supplying the left ventricular inferior and posterolateral walls (Figure 4). No discrete stenosis was detected in either artery. An aortic root angiogram (Figure 5) revealed no evidence of the left circumflex artery from the left coronary artery or from another separate origin. The patient was given some nitrates and diltiazem to treat her symptoms, but they appeared to be not very helpful. She still had some exertional chest discomfort, but usually during strenuous physical activity. She stopped taking these medications for one year and her condition remained unchanged.

Figure 2. Thallium-201 myocardial perfusion scan revealed perfusion defects (arrow heads) in the septal and inferior walls during stress phase (A) and these perfusion defects normalized (arrow head) in delayed imaging during the redistribution phase (B).
Figure 3. Left coronary angiogram in right anterior cranial oblique view. No circumflex artery is demonstrated.

Figure 4. Right coronary angiogram in right anterior oblique (A) and left anterior oblique (B) views. The right coronary artery was a dominant vessel which gave off large branches (arrowheads) supplying the left ventricular posterior-lateral wall.
Coronary artery anomaly is a group of diseases with various severities, which ranged from mild variations to marked deviations in anatomical basis and from asymptomatic behavior to life threatening condition in clinical basis. They are present at birth, and relatively few are symptomatic during childhood. Most anomalies are discovered as incidental findings during coronary angiographic study or at autopsy, and 0.64% to 1.3% incidence rates were reported in the literature.\textsuperscript{1,2) Yamanaka and Hobbs\textsuperscript{1)} described 126,595 patients undergoing cardiac catheterization between 1960 and 1988, the largest study in the literature. Separate origins of the left anterior descending and left circumflex arteries from the left sinus of Valsalva are the most common anomalies, occurring in about 0.41% of the patients studied. Absence of the left circumflex artery is very rare, being reported in only a few cases.\textsuperscript{2-4) Absence of the left circumflex artery is usually associated with a large “super-dominant” right coronary artery crossing the crux of the heart that ascends to the left AV groove and perfuses the inferior and posterolateral walls of the left ventricle.\textsuperscript{1) Anomalous origin of the left circumflex coronary artery from the right sinus of Valsalva or from the right coronary artery was reported by Ueyama, \textit{et al}\textsuperscript{21 and Page, \textit{et al},\textsuperscript{5) and the incidences were 0.39%}}
and 0.67%, respectively. Ott reported the rare case of a left circumflex coronary artery originating from the right pulmonary artery.\textsuperscript{6)}

In our case, a premenopausal female patient without any risk factors for coronary artery disease presented with symptoms suggestive of angina pectoris. The electrocardiogram showed right axis deviation and an rS pattern over leads I and aVL, while a thallium-201 myocardial perfusion study revealed perfusion defects in the septal and inferior walls which normalized in the late phase. Finally, a coronary angiogram proved that this was a case of congenital absence of the left circumflex artery. The symptoms of exertional chest pain were not well understood. We hypothesize that the chest pain may have resulted from transient ischemia of the left ventricular inferior and septal walls in conditions in which an increased oxygen demand is required. In the absence of the left circumflex artery, the oxygen demand of these areas may not be met by the blood supplied from the right coronary artery during physical exertion. The blood supply to the inferior wall was partially stolen from the posterior descending artery to supply the left ventricular posterior and lateral walls. The stress thallium-201 myocardial perfusion study supported this point since there were perfusion defects in the inferior and septal walls in our patient. The posterior descending artery supplies the inferior wall and the lower third of the interventricular septal wall.

Although the absence of a left circumflex artery is regarded as a benign condition,\textsuperscript{1)} we emphasize that the recognition and identification of this anomaly are of clinical importance because the symptoms cannot be differentiated from atherosclerotic coronary artery disease. Among low-risk female patients with chest pain and a positive stress test, coronary artery anomaly should be considered and an angiographic study should be performed.

REFERENCES