Acute Myocardial Infarction in a Patient With Anomalous Left Coronary Artery Origin and Primary Antiphospholipid Syndrome

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Anomalous left main coronary artery (LMCA) originating from the right coronary sinus and running between the aorta and pulmonary trunk is a rare congenital condition. Although this disease is known to be associated with myocardial infarction and sudden death, the precise mechanism is uncertain. A 14-year-old male with this anomaly developed myocardial infarction during exercise complicated by primary antiphospholipid syndrome. He was admitted to hospital with persistent chest pain and sudden cardiac collapse that occurred while he was running. Cardiac catheterization demonstrated a narrowed segment in the LMCA and impaired blood flow, prompting a diagnosis of extensive anterior myocardial infarction. Emergency bypass surgery was performed using a single saphenous vein graft to the left anterior descending artery. Postoperative angiography showed the presence of an anomalous LMCA arising from the right sinus of Valsalva and running between the great vessels. The aortic samples were pathologically normal. He was discovered to have primary antiphospholipid syndrome and was discharged without symptoms after warfarin therapy. Complicated primary antiphospholipid syndrome may trigger myocardial infarction in asymptomatic patients with this type of coronary anomaly. (Jpn Circ J 2000; 64: 214–217)

Key Words: Antiphospholipid syndrome; Congenital heart disease; Coronary vessel anomalies; Myocardial infarction; Coronary artery bypass grafting

Anomalous origin of the left main coronary artery (LMCA) arising from the right sinus of Valsalva (RSV) and running between the aorta and pulmonary trunk is extremely rare. However, this condition is one of the most important congenital heart diseases because of all coronary anomalies it poses the greatest risk of sudden death! The diagnosis is often made at autopsy. In addition, it remains uncertain how this anomaly leads to symptomatic onset and why the life span of those affected is highly variable? We describe a young male with this coronary anomaly who developed myocardial infarction complicated by primary antiphospholipid syndrome (APS) during exercise.

Case Report

A 14-year-old healthy-looking male experienced anterior chest oppression while running for about 1 h, with no history of previous episodes. His daily physical activity level was very high and he had no history of Kawasaki disease, thrombotic disorder or illicit drug use. He was transported to hospital with persistent chest pain.

On admission, he was diaphoretic and cyanotic, systemically. His pulse was regular at 120 beats/min and blood pressure was 70/50mmHg. Physical examination showed no other remarkable findings, except for an S1 gallop. He did not have fever or any signs of trauma. Electrocardiography showed sinus tachycardia with diffuse ST segment elevation and wide QRS complex in leads I, II, III, aVR and V5-6 (Fig 1). Echocardiography showed severely depressed left ventricular systolic function with sparing of the inferior wall. The estimated ejection fraction was 40%. No structural abnormality was demonstrated. These findings suggested extensive myocardial ischemia, and the patient was taken to the catheterization laboratory.

It was difficult to engage the left coronary ostium, thus we selected aortography to evaluate the left coronary artery flow. Ascending aortography revealed a crescent-shaped ostium and impaired blood flow without peripheral contrast delay image (Fig 2). The coronary arteries were otherwise normal. Thus, the diagnosis of acute extensive anterior myocardial infarction was made. During the procedure, the boy developed dyspnea and bilateral basilar rules, which rapidly progressed and required mechanical ventilatory support and a femoral cardiopulmonary support system. Furthermore, he developed sustained ventricular tachycardia, which was eliminated by 2 cardioversions. Emergency coronary artery bypass grafting (CABG) was performed using a single saphenous vein graft to the left anterior descending artery.

The postoperative course was uneventful although the thallium-201 scintigraphy at rest demonstrated a perfusion...
Fig 1. Electrocardiography on admission showing widening of the QRS complex with significant ST changes in almost all 12 leads.

Fig 2. Ascending aortography on admission (left anterior oblique caudal view) showing a narrowed segment (arrow) in left main coronary trunk with impaired blood flow. LCC/ NCC/ RCC, left coronary/ noncoronary/ right coronary cusp; LAD, left anterior descending artery; LCX, left circumflex artery.
defect in the left ventricular apical and anterolateral walls consistent with myocardial infarction (Fig. 3). His second angiography revealed the presence of an anomalous LMCA originating from the RSV and following a course posteriorly between the great vessels (Fig. 4). It was also discovered that he tested positive for antiphospholipid IgG antibody (2.6 and 1.4 immediately and 5 months after the event, respectively; normal, <1.0). He was negative on rheumatic factor, antinuclear antibody and syphilis tests. Aortic tissue samples showed no evidence of invasion of inflammatory cells or atherosclerotic disease. The patient was discharged without symptoms after warfarin therapy and cardiac rehabilitation, about 1 month after the CABG.

**Discussion**

Anomalous origin of the LMCA from the RSV is extremely rare and reported to constitute 0.017% of coronary arteriography examinations. However, Taylor et al reported the incidence of sudden cardiac death to be 57% among 49 cases of anomalous left coronary artery in an autopsy study.

As far as we know, the present patient is the first to undergo emergency CABG and survive acute myocardial
infarction complicating this congenital disease. His survival might be explained by the relatively unimpaired LMCA flow, transport within 20 min of onset to the emergency room and the prompt diagnosis and surgical treatment with respiratory and circulatory support. In addition, the LMCA perfusion bed may have been small and/or the LMCA flow reserve may have been conserved.

Although acute myocardial infarction can be a presenting manifestation in cases of this type of coronary anomaly, the precise mechanism of myocardial ischemia is uncertain. No significant pathological or morphological differences have been found between patients with and without sudden cardiac death. The pathological features include a slit-like orifice and acute coronary takeoff, indicating the presence of an ostial valve-like ridge and phasic compression between the great vessels. Taylor et al recently proposed that the most likely mechanism of the ischemia in cases of anomalous LMCA arising from the RSV and coursing between the great vessels is impaired coronary flow reserve secondary to the multifactorial anatomical basis!

APS is a thrombotic disorder characterized by arterial and venous thrombosis associated with anticardiolipin antibody and lupus anticoagulant. The present patient was diagnosed with primary APS because of the episode of myocardial infarction, moderately high titers of anticardiolipin antibody and no evidence of underlying rheumatic disease. Antiphospholipid antibody levels are reported to increase in one-fifth of young (<45 years) survivors of myocardial infarction. Therefore, we cannot exclude the possibility that APS existed concomitantly with the coronary anomaly or that APS played a major role in the development of myocardial infarction. However, the patient had not shown previously either chest pain or syncope even under severe conditions of physical exercise and dehydration, and the intracoronary thromboses did not seem sufficient to give rise to coronary obstruction. Accordingly, it was suggested that, in this case, primary APS complicated the anomalous origin of the LMCA, leading to myocardial infarction.

References