Coronary Artery Aneurysm After Kawasaki Disease in a Single Coronary Artery

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Figure 1. Transthoracic echocardiographic scans showing a giant coronary artery aneurysm (10 mm in diameter) and anomalous origin of the right coronary artery. (A) Parasternal short-axis view showing a giant aneurysm (maximal diameter, 10 mm) in the proximal left anterior descending coronary artery. (B) Parasternal short-axis view showing that the right coronary artery did not arise from the right sinus of Valsalva and coursed the anterior side of the aorta. Ao, aorta; CAA, coronary artery aneurysm; LMCA, left main coronary artery; PA, pulmonary artery; RCA, right coronary artery.
The subject of this report is a 21-month-old girl who had Kawasaki disease. Her symptoms included fever of 15 days’ duration, indurative edema in the fingers, polymorphous exanthema, bilateral non-exudative conjunctival injection, cervical lymphadenopathy, and indurations at the Bacille Calmette-Guérin inoculation site. She was treated with high-dose aspirin (30 mg · kg\(^{-1}\) · day\(^{-1}\)) and high-dose intravenous immunoglobulin (2 g/kg) twice. Because of refractoriness to immunoglobulin, she additionally received intravenous methylprednisolone, urinastatin, and cyclosporin A. Echocardiograms performed on the 14th day of the illness revealed a giant coronary artery aneurysm at the proximal segment of the left anterior descending coronary artery (Figure 1A). It also demonstrated that the right coronary artery did not arise from the right sinus of Valsalva and coursed the anterior side of the aorta. The origin of the right coronary artery could not be detected (Figure 1B). No other associated cardiac malformations were found.

Four months after the onset of the illness, she remained asymptomatic and echocardiograms showed a decrease in the diameter of the coronary artery aneurysm to 6 mm. Nuclear perfusion scans, using technetium sestamibi with intravenous adenosine, did not show myocardial ischemia. Selective left coronary angiograms showed an aneurysm in the proximal left anterior descending coronary artery with a maximum diameter of 6 mm, as well as normal courses of the left main and circumflex coronary arteries and the left anterior descending coronary artery. Notably, the right coronary artery abnormally arose as a separate branch from the left anterior descending coronary artery just distal to the aneurysm. It coursed anteriorly to the pulmonary artery and down to the right atrioventricular groove as the normal right coronary artery course (Figures 2A, 2B; Movies S1, S2). An aortic root injection confirmed the absence of the right coronary artery ostium in the right sinus of Valsalva. Although the size of the coronary artery aneurysm decreased from giant to moderate, both anticoagulation and antiplatelet therapies (0.16 mg · kg\(^{-1}\) · day\(^{-1}\) of warfarin and 6 mg · kg\(^{-1}\) · day\(^{-1}\) of aspirin, respectively) have been continued in view that the aneurysm is located proximal to the distal left anterior descending and entire right coronary arteries.

Isolated single coronary artery is a rare disorder; its incidence ranges from 0.008% to 0.067% of patients referred for coronary angiography.\(^1\) Anomalous origin of the right coronary artery from the left anterior descending coronary artery is one of the rarest anomalies of this disorder, which has been classified as type “IB1” by Shirani and Roberts.\(^2\) This variant is not related to the mechanical compression of the coronary artery between the aorta and the pulmonary artery; however, more than approximately 40% of cases required revascularization.\(^3\) The possible causes of the increased incidence of coronary artery disease in this variant are an acute angle of takeoff, kinking, and torsion of the anomalous vessel.

Coronary artery aneurysms are the predominant cause of morbidity and mortality in patients with Kawasaki disease.\(^3-6\) A case of a coronary artery aneurysm after Kawasaki disease accompanied with the anomalous origin of the right coronary artery from the left main coronary artery was reported by Muta et al.\(^7\) To the best of our knowledge, this is the first case where a development of a coronary artery aneurysm after Kawasaki disease was accompanied with the anomalous origin of the right coronary artery from the left anterior descending coronary artery. Generally, stenotic lesions occur at the proximal or distal end of significant coronary artery aneurysms.\(^8\) In this particular case, the risk of myocardial ischemia might be increased because the presence of significant stenosis adjacent to coronary artery aneurysms or thrombosis jeopardizes not only the myocardium supplied by the left anterior descending coronary artery but also that supplied by the anomalous right coronary artery.

Figure 2. Coronary angiograms showing a moderate-sized (6.9 mm) coronary artery aneurysm (CAA) in the left anterior descending coronary artery (LAD). They also show an anomalous origin of the right coronary artery (RCA) from the LAD distal to the CAA. (A) Lateral projection. (B) Right anterior oblique and caudal projections.
Disclosures
None.

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Supplementary Files
Supplementary File 1
Movie S1. Left coronary angiogram in the lateral projection.

Supplementary File 2
Movie S2. Left coronary angiogram in the right anterior oblique and caudal projection.
Please find supplementary file(s):
http://dx.doi.org/10.1253/circj.CJ-13-0128