The Coincidence of 3 Different Rare Coronary Artery Anomalies in an Adult Patient With Untreated Kawasaki Disease

Case Report

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Abstract: The coincidence of 3 different rare coronary artery anomalies is extremely rare, and has not been reported so far.

We report multiple imaging findings of a giant coronary artery aneurysm, which has a fistulous connection to the right ventricle associated with anomalous origin of the anterior descending coronary artery from the right coronary artery in a 67-year-old woman who suffered with a 20-year history of progressively chest distress on exertion and a history of untreated Kawasaki disease in her childhood.

The patient received surgical treatment. The aneurysm was resected and openings at both ends being oversewn. And the fistula was also closed directly. She recovered and discharged uneventfully.

The coincidence of 3 different rare coronary artery anomalies in an adult patient with untreated Kawasaki disease is a rare and complicated condition, in which surgical treatment is recommended.

CASE REPORT

A 36-year-old woman was admitted to our hospital with a 20-year history of progressively chest distress on exertion. This symptom relieved after oxygen inhalation. She did not have any cardiovascular risk factors, but had a history of acute febrile illness at 5 years of age who was diagnosed as having Kawasaki disease but without timely treatment. Physical examination revealed a grade 3/6 systolic murmur heard in the second and third intercostal space at left sternum. Blood pressure was 109/60 mmHg in the left arm, and 125/67 mmHg in the right arm. Laboratory tests revealed elevated levels of prothrombin time, fibrinogen, D-dimer, troponin, myoglobin, and creatine kinase. Electrocardiogram and chest X-ray were within normal limits.

Transsthoracic echocardiography showed dilation of the left coronary artery measuring 1.5 times the diameter of a normal adjacent segment and “Giant” CAA has a diameter >2 cm. The causative factors for giant CAA include atherosclerosis, Takayasu arteritis, congenital disorders, and Kawasaki disease. Coronary artery fistula (CAF) is abnormal communication between epicardial coronary arteries and cardiac chambers or vessels around the heart. CAA formation occurs in about 15% to 19% of patients with CAF, and giant CAA can be found in 5.9% of patients with CAF. Anomalies of the anterior descending coronary artery arising from the right coronary artery (RCA) is rare in a heart without a congenital defect. It occurs frequently in patients with Tetralogy of Fallot and a similar appearance is present in L-loop (corrected) transposition of the great arteries, owing to inversion of the coronary arteries. We present a patient with giant CAA that has a fistulous connection to the right ventricle (RV) in whom the anterior descending coronary artery arose from a dominant RCA, a complicated case with untreated Kawasaki disease.

INTRODUCTION

Coronary artery aneurysm (CAA) is defined as the dilation of the coronary artery measuring 1.5 times the diameter of a normal adjacent segment and “Giant” CAA has a diameter >2 cm. The causative factors for giant CAA include atherosclerosis, Takayasu arteritis, congenital disorders, and Kawasaki disease. Coronary artery fistula (CAF) is abnormal communication between epicardial coronary arteries and cardiac chambers or vessels around the heart. CAA formation occurs in about 15% to 19% of patients with CAF, and giant CAA can be found in 5.9% of patients with CAF. Anomalies of the anterior descending coronary artery arising from the right coronary artery (RCA) is rare in a heart without a congenital defect. It occurs frequently in patients with Tetralogy of Fallot and a similar appearance is present in L-loop (corrected) transposition of the great arteries, owing to inversion of the coronary arteries. We present a patient with giant CAA that has a fistulous connection to the right ventricle (RV) in whom the anterior descending coronary artery arose from a dominant RCA, a complicated case with untreated Kawasaki disease.

Abbreviations: CAA = coronary artery aneurysms, CAF = coronary artery fistula, LA = left atrium, LV = left ventricle, RA = right atrium, RCA = right coronary artery, RV = right ventricle.
Coronary angiography which showed the saccular aneurysm with feeding artery from the dilated left main coronary artery connected to the RV through the fistula by the contrast dye (Figure 4A), the anomalous origin of the anterior descending coronary artery from the dominant RCA and the course of the anterior descending coronary artery showed in MSCT (Figure 4B, C).

At operation, a small hole measuring only 0.3 cm width was detected in the aneurysm whose diameter was about 3.0 × 3.5 cm, through which the aneurysm drained to the RV. Under cardiopulmonary bypass via median sternotomy, both the proximal and distal openings were oversewn with pledgets after incision of the aneurysm and the secretion of the aneurismal contents, which was white thrombus and mixed thrombus. The fistula was closed directly. After the operation, the patient was released and discharged soon. Postoperative MDCT revealed that the aneurysm had disappeared (Figure 5).

DISCUSSION

Kawasaki disease is an acute systemic inflammatory disorder with coronary artery aneurysm or ectasia as its main complication. The most commonly affected part is in the proximal end of the main blood vessel and the left anterior descending part. The aneurismal sac may be saccular or fusiform, and the former is more easily to rupture, thrombosis or fistula formation. CAF can be associated with giant CAA and the RCA is more commonly affected. The drainage is carried out for low-pressure chambers, most commonly the RV. The clinical manifestations of CAF depend on the magnitude of the blood flow through the fistula and its location. Most CAFs are asymptomatic, but some patients present with palpitations, exertional dyspnea, angina, or myocardial infarction, all of which may be attributable to a coronary “steal phenomenon” and myocardial ischemia.

Anomaly of the anterior descending coronary artery arising from the RCA in patients without congenital heart defect has been rarely reported. Its incidence was reported to be 0.03% among 126,595 cases. In case reports of patients in whom the anterior descending coronary artery takes origin from the RCA, the RCA usually is the “dominant” vessel. And a report described a patient in whom the anterior descending coronary artery arose from a small, nondominant RCA.

Without doubt, it is reasonable to speculate that the patient developed a giant CAA in left main coronary artery owing to the delayed treatment of Kawasaki disease in the childhood. The CAF was secondary to the giant CAA, which leads to the occurrence of symptoms for the transient myocardial ischemia from the “coronary steal” effect in adulthood. In this case, Kawasaki disease mainly affected the left main coronary artery.

FIGURE 1. Echocardiography. (A) Four-chamber view. A cystic lesion (arrow) was showed in the apex of interventricular septum. (B) Short-axis view. A hyperechoic mass (arrow) was detected in the cystic lesion. (C) Doppler echocardiography. A diastolic flow in the dilated vessel was showed with peak velocity achieved 2.85 m/s.
and the RCA was relatively normal. We speculate that the occurrence of the anomaly of the anterior descending coronary artery arising from the RCA is good for the improvement of myocardial blood supply.

Presently, no treatment guidelines or follow-up recommendations exist for this rarity because the patient had missed the best diagnosis and therapy period. It is recommended that hemodynamically significant fistula and large, saccular, rapidly...
growing or symptom-causing aneurysms and very large or need surgical treatment.\textsuperscript{10,12} Surgical treatment was selected to release her symptom. And she was in a healthy status during the 12-month follow-up.

We report a giant CAA, which has a fistulous connection to RV associated with anomalous origin of the anterior descending coronary artery from the RCA, in a 67-year-old woman who suffered with a 20-year history of progressive chest distress on exertion and a history of untreated Kawasaki disease in the childhood. To the best of our knowledge with literature review, this is the first reported complicated case diagnosed with multiple imaging approaches. Surgical removal of the CAA and closure of the CAF were successfully performed, and the patient had an uneventful recovery and no serious complications.

ACKNOWLEDGMENTS

The authors specially thank the staff of the department of echocardiograph and radiology for the diagnosis of the disease.

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