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CASE REPORT

ADVANCED

CLINICAL CASE: CORONARIES

A Ruptured Coronary Artery Aneurysm Secondary to Kawasaki Disease

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ABSTRACT

Coronary artery aneurysm occurs in 0.3%-0.8% of patients with Kawasaki disease, and cases of rupture are extremely rare. Only 2 cases have been reported in which the patients survived. We report a case of ruptured coronary artery aneurysm that was treated with coronary artery bypass grafting and extracorporeal membrane oxygenation. (**Level of Difficulty: Advanced.**) (J Am Coll Cardiol Case Rep 2022;4:790–793) © 2022 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

HISTORY OF PRESENTATION

A 3-year-old boy with a history of Kawasaki disease (KD) was hospitalized due to high-grade fever and cervical lymphadenopathy. On physical examination, bilateral conjunctival injection, erythematous lips and palms, swollen feet, and a polymorphous rash were observed. He was diagnosed with a recurrence of KD, and immunoglobulin (2 g/kg/d) and flurbiprofen were administered. However, the initial treatment was not effective, and the medications were shifted to aspirin (3 mg/kg/d) and prednisolone (2 g/kg/d). Except for a low-grade fever, all symptoms improved. Echocardiography revealed that the main trunk of the left coronary artery (LCA) was dilated (3.9 mm). Within a week, both the left and right coronary arteries developed aneurysms, which appeared to grow rapidly (**Figure 1,** Video 1). He was transferred to our hospital for multidisciplinary therapy. The day after the transfer, he lost consciousness. Assessment of his vital signs revealed he was bradycardic and hypotensive, but a specific cause could not be determined. Echocardiography showed pericardial effusion; thus, cardiac tamponade due to rupture of the aneurysms was suspected, and he was immediately transferred to the intensive care unit while performing cardiopulmonary resuscitation. Extracorporeal membrane oxygenation (ECMO) was introduced through his neck. Pericardial drainage was performed via a small incision, and cardiac tamponade resolved. The pericardial effusion was not as viscous as blood; hence, we thought the bleeding was already controlled, and surgery was planned for later. Although there was no bleeding

LEARNING OBJECTIVES

- To learn the urgent management of a patient with CAA rupture secondary to KD.
- To learn the effectiveness of LA/LV venting in patients with low LV function with ECMO.
- To be able to make treatment decisions by CAG in patients with prolonged cardiac decompensation, even with ECMO and after cardiopulmonary resuscitation.

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The authors attest they are in compliance with human studies committees and animal welfare regulations of the authors' institutions and Food and Drug Administration guidelines, including patient consent where appropriate. For more information, visit the Author Center.

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for a few hours, the amount of drainage increased, and the ECMO flow became unstable. Rebleeding was suspected, and he was immediately transferred to the operating room. Full sternotomy was performed, which revealed a large amount of coagulum in the pericardial space. The source of bleeding was located at the anterior aspect of the left coronary artery aneurysm (CAA). Cardiopulmonary bypass was established with an ascending aorta perfusion and right atrium cannulation for drainage. Because the aneurysm wall was too fragile to be repaired while the heart was beating, his body was cooled to 32°C, and cardiac arrest was induced. We sutured the ruptured portion by using pericardium pledgets and used fibrin sealant patches for hemostasis (Video 2). The bleeding was controlled, and it was confirmed that cardiac activity had restarted. Because the cardiac function was well preserved compared with the preoperative status and there was no evidence of stenosis or occlusion of the coronary arteries throughout the course, we ascertained that an additional invasive procedure, including coronary artery bypass grafting (CABG) during the emergency operation, was not required. He was transferred to the intensive care unit while still under ECMO. On the first postoperative day, the cardiac function decreased (ejection fraction <20%) (Video 3), although electrocardiography and blood test results did not indicate ischemic disease. A cannula was directly inserted into the left atrium (LA) for venting on the first postoperative day to decompress and minimize barotrauma of the dilatation of the left ventricle (LV) and the LA.

MEDICAL HISTORY

The patient was diagnosed with KD at 2 years of age.

DIFFERENTIAL DIAGNOSIS

The diagnosis was acute myocardial infarction, pericardial effusion, and cardiomyopathy.

INVESTIGATIONS

There were no signs of improvement in cardiac function after adding LA venting. Hence, on the third postoperative day, coronary angiography (CAG) was performed to rule out ischemic disease, which showed obstruction of the left anterior descending artery (LAD) (**Figure 2,** Video 4).

MANAGEMENT (MEDICAL/INTERVENTIONS)

We performed CABG from the ascending aorta to the LAD using a saphenous vein graft on the same day as the CAG (Video 5). During the procedure, antegrade flow of the LCA was detected despite the preoperative evaluation. We concluded that the low cardiac function could have been caused by a temporary clot in the LCA. At that time, we changed the LA venting to LV venting to more effectively prevent LV distention. On the 13th postoperative day, ECMO was shifted to a left ventricular assist device. After 3 weeks, the cardiac function improved significantly (ejection fraction = 56%), and he was weaned from the left ventricular assist device (Video 6).

DISCUSSION: ASSOCIATION WITH CURRENT GUIDELINES/POSITION PAPERS

In Japan, CAAs, which rarely rupture, occur in approximately 0.3%-0.8% of patients with KD.[1] The treatment for fatal complications has not yet been

ABBREVIATIONS AND ACRONYMS

CAA = coronary artery aneurysm

CABG = coronary artery bypass grafting

CAG = coronary angiography

ECMO = extracorporeal membrane oxygenation

KD = Kawasaki disease

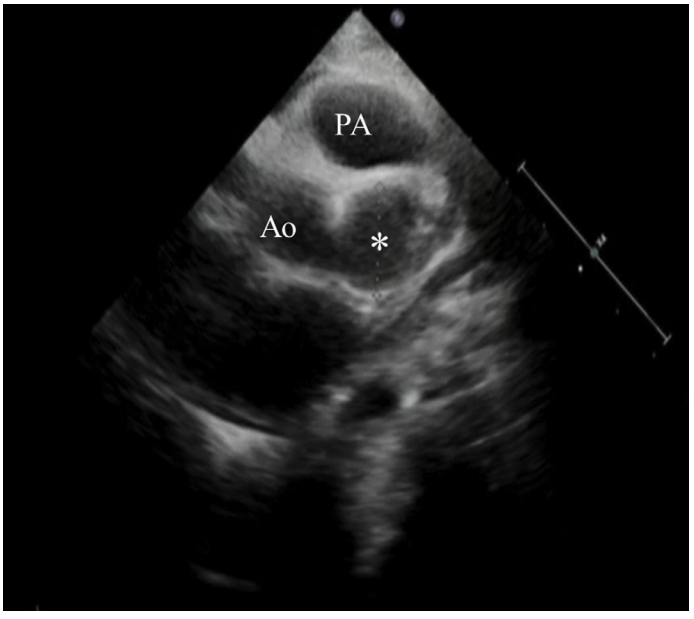
LA = left atrium

LAD = left anterior descending artery

LCA = left coronary artery

LV = left ventricle

FIGURE 1 Short-Axis View at the Level of the Coronary Artery



An aneurysm **(*)** developed at the left coronary artery and measured 17 mm in diameter. Ao = aorta; PA = pulmonary artery.

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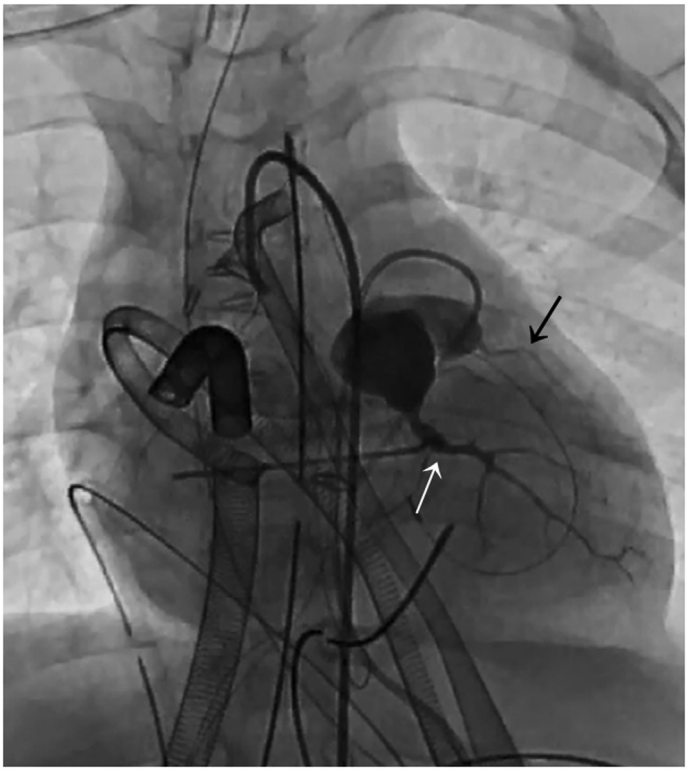
FIGURE 2 Aneurysm at the Left Coronary Artery With Contrast



It only showed the circumflex of the left coronary artery **(white arrow)** and the first branch of the left anterior descending artery **(black arrow)**; hence, we suspected left anterior descending artery obstruction.

established. Hence, when an aneurysm ruptures, the prognosis is poor.[2] Currently, only 2 cases of ruptured CAAs secondary to KD in which the patients survived have been reported. One case reported by Koutlas et al[3] in 1997 underwent emergency CABG (left internal thoracic artery-LAD, saphenous vein graft-left circumflex) but had difficulty being weaned off ECMO due to a low cardiac function and uncontrolled ventricular tachycardia; the patient subsequently underwent heart transplantation. Another case reported by Mok et al[4] in 2003 was treated with emergency CABG using a saphenous vein graft without heart transplantation.

This case is the third report of its kind. If a CCA ruptures, it is critical to stabilize the circulation with rapid cardiopulmonary bypass induction and to perform pericardial drainage for cardiac tamponade. Meanwhile, procedures to manage the aneurysm and bleeding should be considered. As Miyamoto et al[2] reported, repairing the fragile wall of a ruptured CAA in the acute stage of KD was challenging. Without CAA plasty, minimal hemostasis should be performed for the aneurysm. In this case, the priority was to stabilize the circulation, and the treatment of the aneurysm was delayed because it was a high-risk and challenging procedure, which we believe contributed to the patient's survival.

Moreover, because there was no improvement in cardiac function, we performed LA venting, which was subsequently changed to LV venting, to prevent LA/LV distention on the first postoperative day. This may have contributed to the recovery of the LV function. Kotani et al[5] reported earlier timing of LA decompression appeared to be associated with a high probability of being weaned from ECMO and reasonable LV functional recovery.

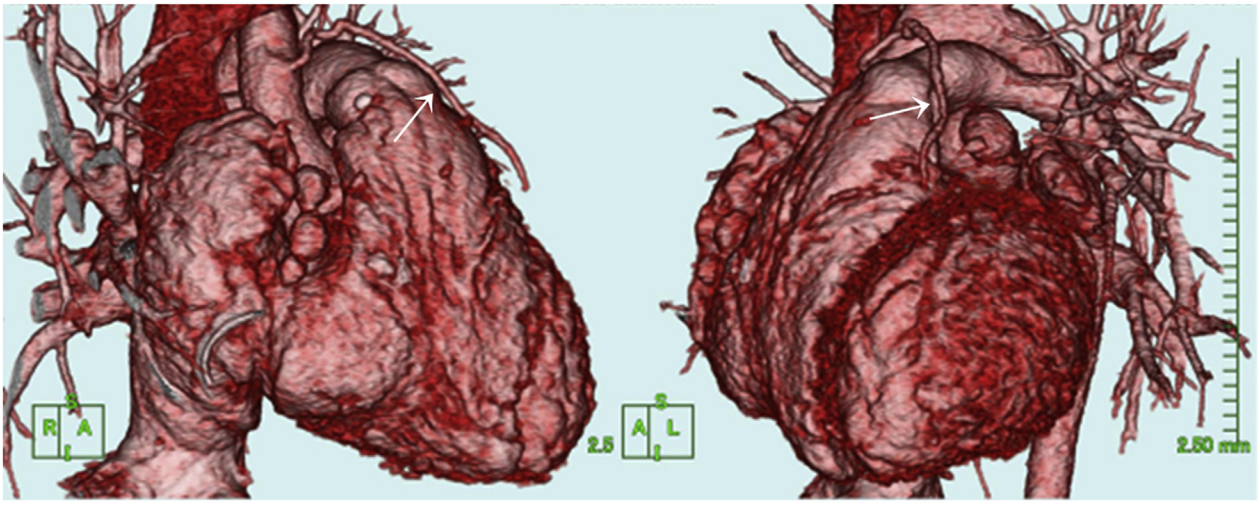
Last, although the patient was on ECMO, CAG was performed to determine the appropriate treatment plan. CAG revealed coronary artery occlusion, which necessitated CABG. CAG is recommended for patients with prolonged cardiac decompensation, even if they are on ECMO and after cardiopulmonary resuscitation.

Overall, heart surgery was performed thrice: for the release of tamponade, for aneurysm repair, and CABG. Complications of CAA include rupture and myocardial infarction. CABG should be performed simultaneously when repairing a ruptured aneurysm.

FOLLOW-UP

Postoperative computed tomography showed the graft was patent (**Figure 3**). There was no improvement in brain activity due to hypoxic-ischemic encephalopathy. Although the auditory brainstem response ruled out brain stem death, the electroencephalogram showed an almost flat pattern. After

FIGURE 3 **Postoperative Computed Tomography**



Postoperative computed tomography showed the graft **(arrow)** was patent.

undergoing a tracheostomy, he was discharged from the hospital.

CONCLUSIONS

This is a case of a ruptured CAA secondary to KD that was managed and treated by maintaining the circulation with ECMO and improving cardiac function with CABG.

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The authors have reported that they have no relationships relevant to the contents of this paper to disclose.

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KEY WORDS coronary artery aneurysm, coronary artery bypass grafting, extracorporeal membrane oxygenation, Kawasaki disease, rupture

APPENDIX For supplemental videos, please see the online version of this paper.