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Giant Thrombotic Right Coronary Aneurysm in an Infant with Undiagnosed Incomplete Kawasaki Disease and Rapidly Progressive Cardiovascular Collapse

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INTRODUCTION

Kawasaki disease (KD) is an acute, self-limited medium-size-vessel vasculitis and is now the most common acquired heart disease in the pediatric age group.[1] The term *incomplete*, or alternatively *atypical*, is reserved for children with KD who fail to meet defined clinical criteria but otherwise have courses identical to those of patients with classical KD.[1,2] Although it is well known that coronary artery aneurysms (CAAs) are the most common cardiovascular sequelae of KD, this case describes acute, life-threatening coronary thromboses in an infant with incomplete and previously unrecognized KD and giant CAA. It illustrates the potential challenge of making the diagnosis when it is not suspected, particularly with thrombotic changes in the severely dilated distal coronary arteries that give the initial appearance of epicardial tumor.

CASE PRESENTATION

A 7-month-old male infant presented to our pediatric emergency department from a transferring outside hospital with 6 weeks of poor feeding, pallor, weight loss, and intermittent emesis. His emesis became bilious the day of presentation, with bloody stools, thus prompting his caregivers to seek emergent medical attention. His workup before transfer was notable for cardiomegaly on chest radiography (Figure 1) and abnormal laboratory test results, including C-reactive protein of 28.03 mg/dL, a white blood cell count of 27.8 $10^3/\mu$L, hemoglobin of 7.8 g/dL, a platelet count of 995 $10^3/\mu$L, bicarbonate of 20 mmol/L, and albumin of 2.7 g/dL. The infant had been evaluated during the preceding month by multiple providers for intermittent fevers and had been treated with multiple courses of antibiotics for presumed otitis media and pneumonia; by report he had developed a maculopapular rash during this time that was attributed to a drug reaction. In our emergency department he was noted to have tachycardia, lethargy, poor perfusion, abdominal distention, and hepatomegaly. He was admitted to the cardiac intensive care unit after a brief transthoracic echocardiographic examination revealed a large global pericardial effusion with preserved biventricular systolic function. A hyperechoic circular mass was also evident on this study in the anterior right atrioventricular (AV) groove, although it was not immediately clear whether the mass was intrinsic or extrinsic to the heart (Figure 2, Video 1). The proximal coronary arteries were not assessed. Shortly after arrival in the cardiac intensive care unit, the patient acutely decompensated with the development of ventricular tachycardia and ventricular fibrillation. He was successfully resuscitated with chest compressions and defibrillation, achieving return of spontaneous circulation with sinus rhythm. Given concern that the large pericardial effusion was causing hemodynamic compromise, an emergent pericardiocentesis was performed under echocardiographic guidance, resulting in the drainage of 100 mL of serous fluid.

Additional transthoracic imaging was performed after the pericardiocentesis was completed. The hyperechoic epicardial mass, measuring 18 mm in diameter, was again appreciated along the lateral AV groove of the right ventricle and tracked anteriorly toward the right coronary sinus (Figures 3-5, Videos 2-4). The right coronary artery (RCA) appeared to be compressed by the mass, with anterograde flow in the coronary documented by color Doppler (Figure 6, Video 5). The left main coronary artery appeared to be dilated proximally with anterograde filling by color Doppler, but the left coronary branches were not well seen (Figure 7, Video 6). There was good biventricular function with mild right atrial and right ventricular dilation. Computed tomography of the chest and abdomen was then performed because of the mass and the gastrointestinal symptoms of emesis and bloody stools. Computed tomography confirmed a round mass of hypoattenuation present within the lateral aspect of the right AV groove measuring 18 × 23 × 22 mm, with the RCA appearing to pass through the mass, but delineation of coronary anatomy was limited (Figure 8). No abdominal abnormalities or masses were appreciated.

The infant stabilized briefly after pericardiocentesis but subsequently experienced a second cardiac arrest with rapid deterioration to pulseless electrical activity. Emergency cannulation for venoarterial extracorporeal membrane oxygenation (ECMO) was performed at the bedside using the right carotid artery and right internal jugular vein. Brief postevent transthoracic echocardiography on ECMO showed severely diminished biventricular function (Figure 9, Video 7). Because of inadequate left ventricular

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VIDEO HIGHLIGHTS

Video 1: Transthoracic echocardiography from a long-axis window shows a large pericardial effusion (PE) with good left ventricular and right ventricular systolic function. A hyperechoic circular mass (*arrow*) is seen in the right AV groove anterior to the aorta (Ao). *LV*, Left ventricle; *RV*, right ventricle.

Video 2: Long-axis imaging angled toward the right atrium (RA) and right ventricle (RV) after pericardiocentesis shows a hyperechoic circular mass (*arrow*) in the anterior right AV groove. Right ventricular function is good, with mild right atrial and right ventricular dilatation.

Video 3: Subcostal coronal imaging shows a hyperechoic circular mass (*arrow*) extending from the lateral right AV groove anteriorly. The right atrium (RA) and right ventricle (RV) appear mildly dilated, with good systolic function.

Video 4: Additional subcostal coronal imaging angled more anteriorly shows anterior extension of the hyperechoic circular mass (*arrows*) adjacent to the right ventricular outflow tract. There is a small global pericardial effusion. *RV*, Right ventricle.

Video 5: Short-axis imaging at the level of the aortic root demonstrates the RCA, which appears to be compressed by the large hyperechoic mass (*large arrows*) with anterograde flow in the coronary documented by color Doppler (*small arrows*). *Ao*, Aorta.

Video 6: Short-axis imaging at the level of the aortic root demonstrates anterograde filling by color Doppler in the dilated left main coronary artery (*arrows*). The pulmonary artery (PA) is seen anteriorly. *Ao*, Aorta.

Video 7: Apical imaging after ECMO support shows severely diminished left ventricular and right ventricular systolic function. *LV*, Left ventricle; *RV*, right ventricle.

Video 8: Aortic root angiography demonstrates a mildly dilated left main coronary artery (*small arrows*) with diffusely dilated left anterior descending and circumflex coronary arteries with luminal irregularities and no distal coronary flow (*large arrows*). There is no filling of the RCA.

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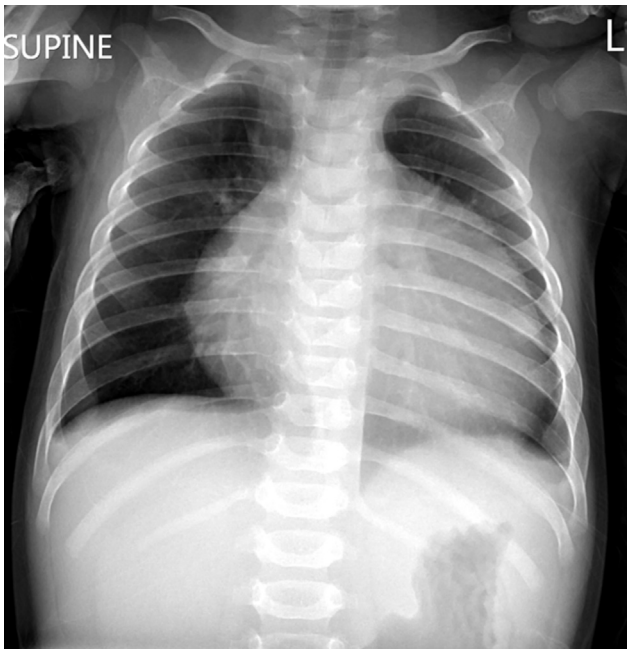

Figure 1 Chest x-ray reveals significant cardiomegaly.

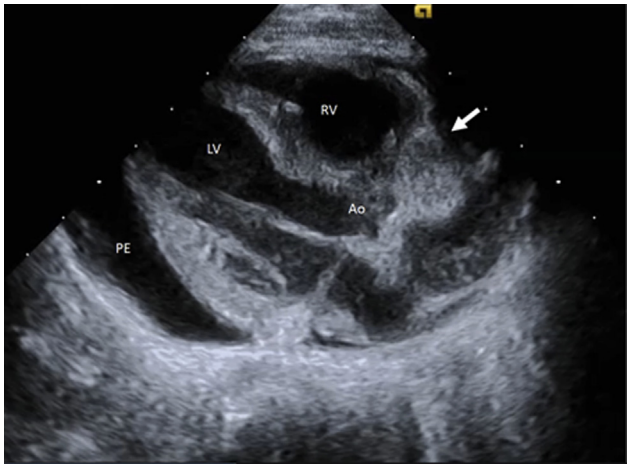

Figure 2 Transthoracic echocardiography from a long-axis window shows a large pericardial effusion (PE) with good left ventricular and right ventricular systolic function. A hyperechoic circular mass (*arrow*) is seen in the right AV groove anterior to the aorta (Ao). *LV*, Left ventricle; *RV*, right ventricle.

decompression on ECMO, the patient was taken to the hybrid cardiac catheterization laboratory for placement of a left ventricular vent. Direct inspection of the myocardial surface showed extensive dilatation and clot burden in the left anterior descending coronary artery and RCA, with severely diminished biventricular contractility. Both the right and left ventricles appeared ischemic, suggestive of massive infarction. Coronary angiography demonstrated total occlusion of the RCA and a mildly dilated left main coronary artery with diffusely dilated left anterior descending and circumflex coronary arteries with luminal irregularities and no distal coronary flow (Figure 10, Video 8). Biopsies obtained during the procedure revealed thrombus with fibrin in the coronary arteries and extensive myocardial necrosis, consistent with myocardial infarction. It was clear that the infant had experienced irreversible myocardial injury secondary to extensive coronary thromboses.

The infant was diagnosed with KD and giant CAA and rapidly progressive obstructive coronary thromboses, as it was now clear that the epicardial mass seen on echocardiography represented extensive thrombus in the massively dilated RCA. Supportive evi-

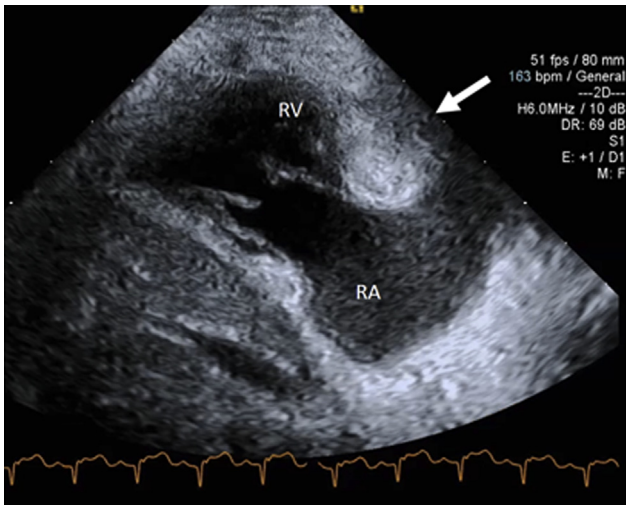

Figure 3 Long-axis imaging angled toward the right atrium (RA) and right ventricle (RV) after pericardiocentesis shows a hyperechoic circular mass (*arrow*) in the anterior right AV groove. Right ventricular function is good, with mild right atrium and right ventricular dilatation.

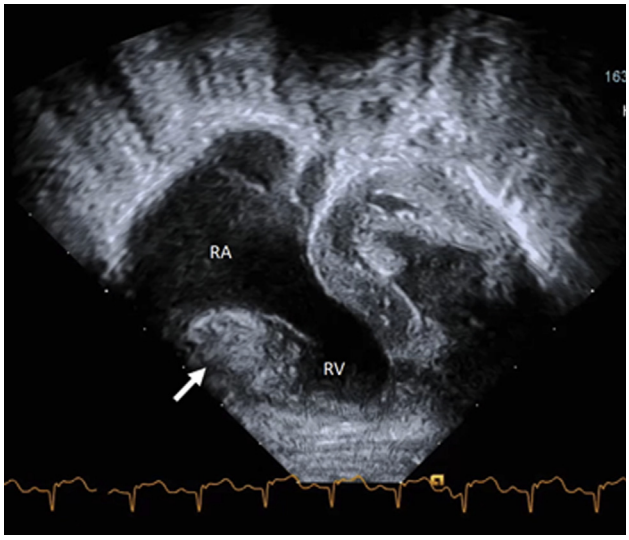

Figure 4 Subcostal coronal imaging shows a hyperechoic circular mass (*arrow*) extending from the lateral right AV groove anteriorly. The right atrium (RA) and right ventricle (RV) appear mildly dilated, with good systolic function.

dence of KD included his intermittent fevers before admission, biomarkers suggestive of systemic inflammation, and bilateral common iliac artery aneurysms found on subsequent computed tomographic imaging, consistent with the extracoronary midsize artery involvement seen in KD (Figure 11).[1-3] The patient was given infliximab to treat the fulminant systemic inflammatory response of active KD and systemic high-dose anticoagulation while he was maintained on continued ECMO support. Although there was no evidence of improvement in heart function, all other organs recovered on ECMO, so biventricular Berlin ventricular assist devices were implanted for long-term mechanical circulatory support. The patient is currently listed for orthotopic heart transplantation. The explanted heart had diffuse left and right CAAs with organized thrombus in the RCA (Figure 12).

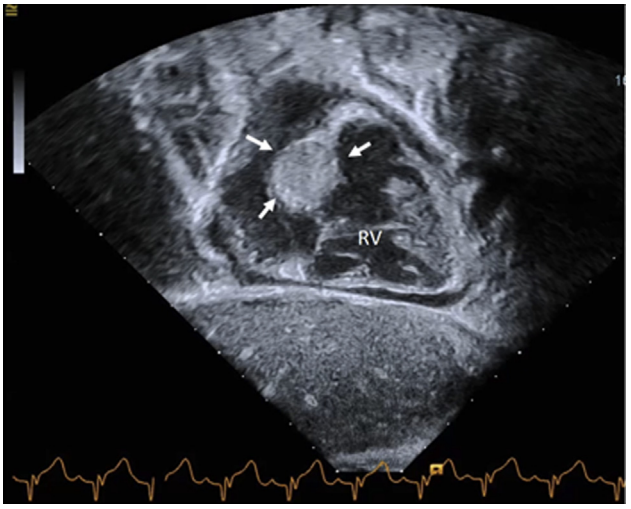

Figure 5 Additional subcostal coronal imaging angled more anteriorly shows anterior extension of the hyperechoic circular mass (*arrows*) adjacent to the right ventricular outflow tract. There is a small global pericardial effusion. *RV*, Right ventricle.

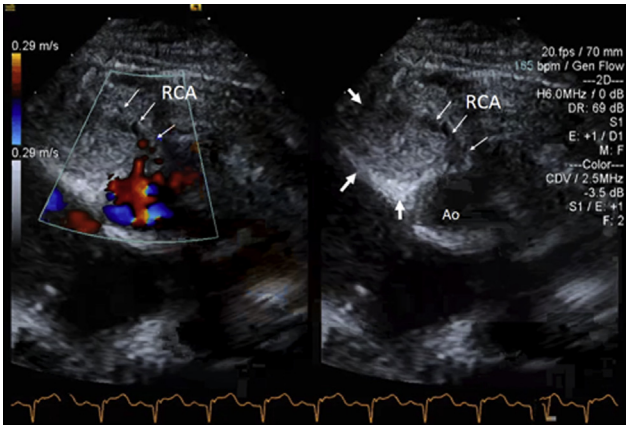

Figure 6 Short-axis imaging at the level of the aortic root demonstrates the RCA, which appears to be compressed by the large hyperechoic mass (*large arrows*) with anterograde flow in the coronary documented by color Doppler (*small arrows*). *Ao*, Aorta.

DISCUSSION

The diagnosis of KD may be quite challenging in children <1 year of age, as infants are likely to present with fewer classical clinical features and are thus less likely to fulfill diagnostic criteria for typical KD.[1] A small prospective study by Heuclin *et al.*[4] found that following the American Heart Association guidelines, the early

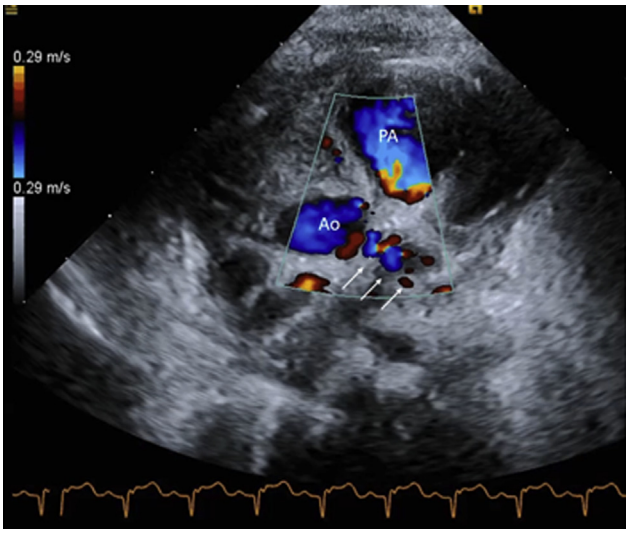

Figure 7 Short-axis imaging at the level of the aortic root demonstrates anterograde filling by color Doppler in the dilated left main coronary artery (*arrows*). The pulmonary artery (PA) is seen anteriorly. *Ao*, Aorta.

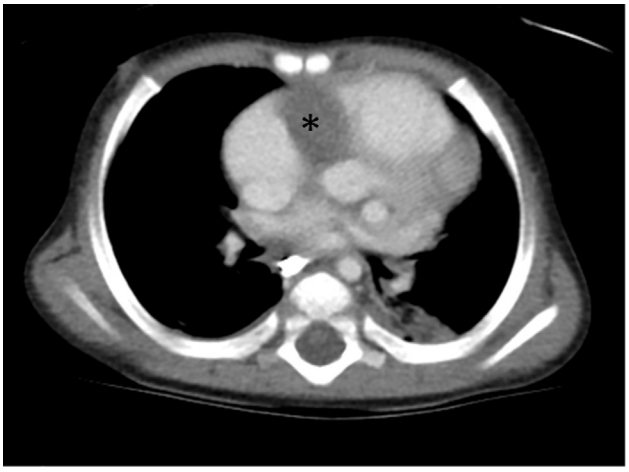

Figure 8 Computed tomography of the chest demonstrates a round focus of hypoattenuation (*asterisk*) present within the lateral aspect of the right AV groove extending superiorly and measuring 18 × 23 × 22 mm.

use of echocardiography increases both the diagnostic sensitivity and treatment rate for KD. Because incomplete KD can mimic many infantile intercurrent illnesses, echocardiography is not requested as part of the diagnostic workup during the initial phases of the disease in most infants. Additionally, KD is associated with increasing complications in infants.[1-3,5,6] An increased risk for coronary artery events with KD has been related to delayed administration of intravenous immunoglobulin, and infants are at particular risk for late treatment because KD masquerades as common infectious illnesses.[3,5,7] This case underscores the need for a high index of suspicion for incomplete KD in infants with persistent fevers and emphasizes the value of careful echocardiographic coronary interrogation when cardiovascular compromise is identified.

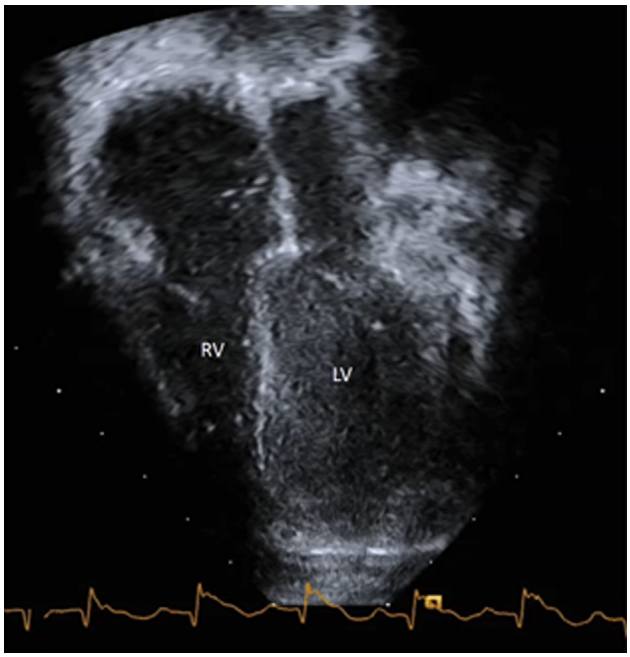

Figure 9 Apical imaging after ECMO support shows severely diminished left ventricular and right ventricular systolic function. *LV*, Left ventricle; *RV*, right ventricle.

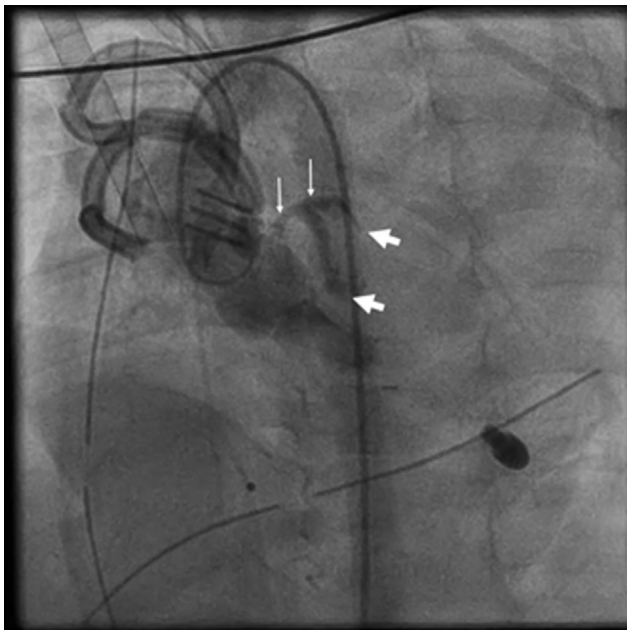

Figure 10 Aortic root angiography demonstrates a mildly dilated left main coronary artery (*small arrows*) with diffusely dilated left anterior descending and circumflex coronary arteries with luminal irregularities and no distal coronary flow (*large arrows*). There is no filling of the RCA.

Echocardiography is a very effective tool to assess for the known cardiac complications of KD, such as the presence of coronary artery dilation, CAA formation, and pericardial effusion, in addition to evaluation of ventricular function.[8] The development of giant

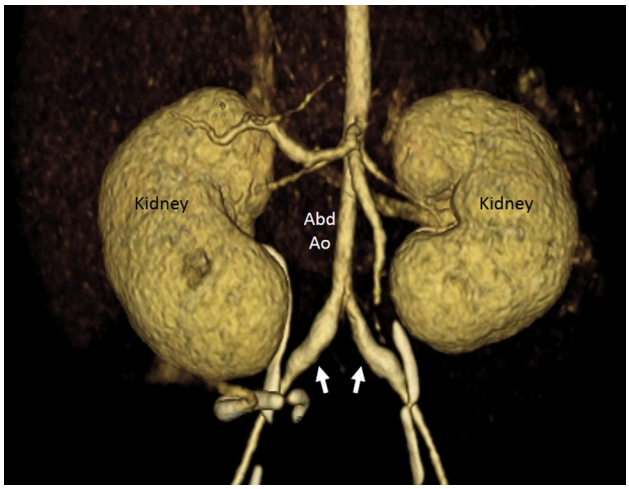

Figure 11 Computed tomographic angiography of the abdominal aorta (Abd Ao) demonstrates fusiform aneurysms of bilateral common iliac arteries (*arrows*).

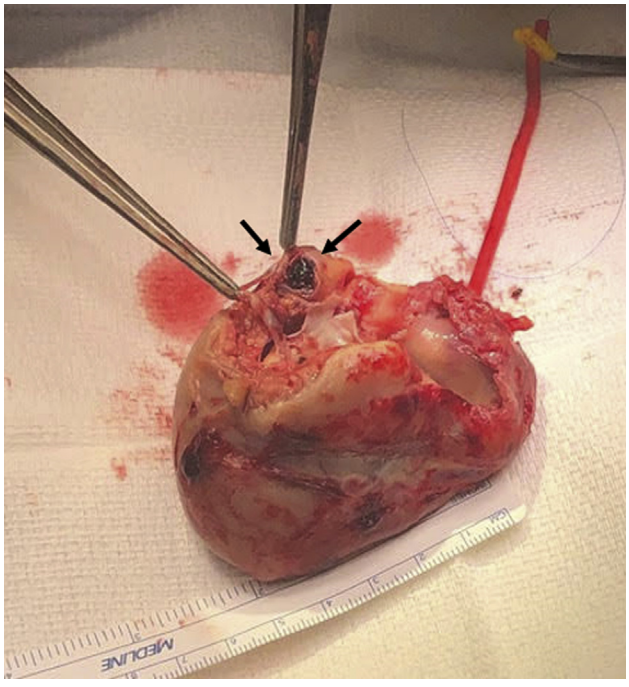

Figure 12 Operative photograph of the explanted heart after ventricular assist device placement with the aortic root removed and the proximal RCA sectioned opened (*arrows*). There is organized thrombus that can be seen completely filling the lumen of the dilated coronary.

thrombotic coronary aneurysms in an infant without clear clinical signs of KD, however, is exceedingly rare and unexpected. Because of the unusual echo-dense appearance of the thrombosed distal RCA in the AV groove and the unclear and rapidly evolving clinical picture, the diagnosis of KD was not immediately apparent in this case, leading to additional nondiagnostic imaging in which the focus was on epicardial mass assessment and not coronary artery disease. In retrospect, the coronary changes were apparent by echocardiography, especially after direct inspection of the coronary aneurysms on ECMO support. It is unlikely that any acute intervention at our hospital would have changed the unfortunate rapidly progressive disease in this infant, but the case emphasizes the need for complete interrogation of the coronary arteries (and suspicion of incomplete KD) when an infant presents with acute cardiovascular collapse.

CONCLUSION

Echocardiography is a mainstay in the diagnostic workup for KD. A high index of suspicion of KD is needed in those who present with an incomplete clinical picture. This is especially important in infants, as they frequently have atypical clinical presentations and are most at risk for delayed diagnosis and the development of CAA or life-threatening cardiac complications associated with this disease.[7]

SUPPLEMENTARY DATA

Supplementary data related to this article can be found at https://doi.org/10.1016/j.case.2019.08.006.

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