

Kawasaki disease with giant aneurysm of left coronary artery and complete occlusion of right coronary artery

Brief Report

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Abstract

Kawasaki disease is an acute febrile medium-vessel vasculitis affecting children. Though coronary artery aneurysm is a common complication, stenosis and occlusion are rare. Here, we report a case of Kawasaki disease with concomitant aneurysm and complete occlusion of different coronary arteries in the same patient.

Case report

A 1-year-old boy presented to our department with history of fever and maculopapular rash of 1-week duration. On examination, he had erythema of hands and feet, cervical lymph node enlargement, and conjunctival congestion on both sides. Systemic examination was within normal limits. Haematological investigations revealed leucocytosis with elevated erythrocyte sedimentation rate. Electrocardiogram showed normal sinus rhythm without any abnormalities. A clinical diagnosis of Kawasaki disease was made.

He was treated with intravenous immunoglobulin 2 g/kg as single infusion. Aspirin was administered orally at a dose of 1000 mg/day (80 mg/kg/day in four divided doses) for 3 days when defervescence occurred. Aspirin was continued at a dose of 75 mg/day. He improved symptomatically, fever and rash subsided. His echocardiography at the time of admission revealed a giant aneurysm of left main coronary artery measuring around 12 mm. Right coronary artery was also enlarged measuring 8 mm (Fig 1). He was initiated on oral anticoagulation (warfarin) and maintained at an international normalised ratio between 2 and 3.

A follow-up echocardiogram after 3 months showed increase in the size of left coronary artery aneurysm (16 mm). Right coronary artery was not visualised. He was asymptomatic. As there was progressive increase in the size of the aneurysm of left coronary artery and as right coronary was not visualised, a coronary angiography was performed to delineate the coronary anatomy. Electrocardiogram at this point showed Q waves in inferior leads. Coronary angiography showed a giant aneurysm (18 mm) of left main coronary artery with swirling of contrast within the aneurysm (Video 1 in Supplementary material). Right coronary artery was occluded proximally and there were numerous collaterals supplying the distal part of right coronary artery (Figure 2 and Video 2 in Supplementary material). Oral anticoagulation (warfarin) was continued along with aspirin 75 mg/day. Beta blocker (metoprolol) was also initiated. He remains asymptomatic for the past 1 year. There is no left ventricular dysfunction and is being closely monitored.

Discussion

Complications of Kawasaki disease range from asymptomatic coronary artery dilatation to large coronary artery aneurysms. These are prone for thrombosis with resultant myocardial infarction and sudden death.¹ Treatment with intravenous immunoglobulins has resulted in significant decline in its incidence. Myocarditis, pericarditis, and valvular regurgitations culminating in heart failure are also reported sequelae.

Aneurysms occur mostly during the initial phase and predominantly involve the proximal segments of the coronary arteries. Approximately 50% of the aneurysms regress within 5 years of disease onset.² These aneurysms also develop stenosis over the course. Chronic endothelial shear stress at the site of aneurysm will trigger the transformation of smooth muscle cells to myofibroblasts.^{3,4} This causes arterial narrowing.

As per guidelines, all patients diagnosed with Kawasaki disease should undergo serial echocardiography on diagnosis, at 2 and 6 weeks after the onset of the disease. Those with evolving coronary artery abnormalities should undergo echocardiography twice/week until the lesions have stopped progressing.⁵ Children may not exhibit typical features of myocardial ischemia. Hence, they should be carefully monitored and thromboprophylaxis should be judiciously administered. Those with giant aneurysms (>8 mm) should be subjected to coronary

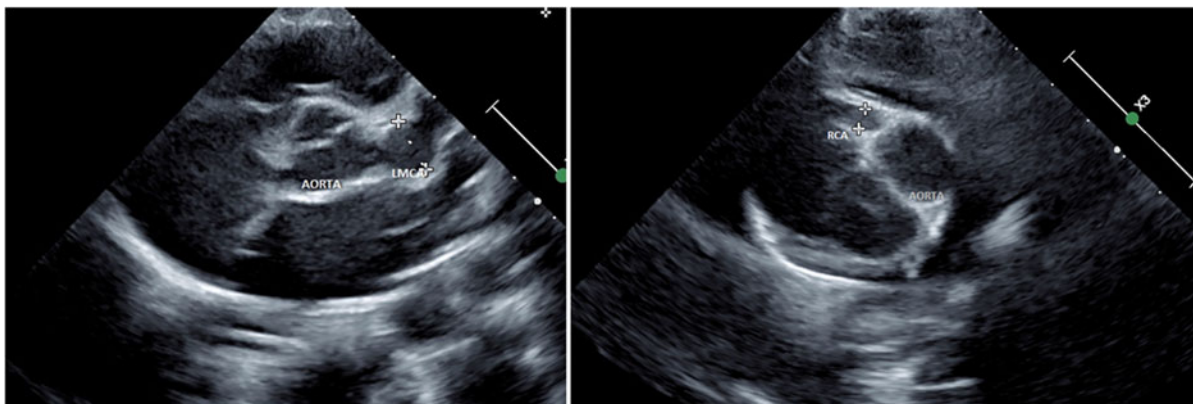


Figure 1. Transthoracic echocardiography parasternal short axis view showing dilated left main coronary artery (LMCA) and dilated right coronary artery (RCA).



Figure 2. Coronary angiogram showing complete occlusion of proximal right coronary artery with collaterals supplying the distal part of the vessel.

angiography to identify stenotic lesions. An extensive collateral supply to distal part of right coronary artery in our patient was probably due to chronic stenosis and occlusion.

Flow stasis in large aneurysms forms the pathophysiological basis of thrombus formation, and hence oral anticoagulation with warfarin should be given to patients with giant aneurysms. Revascularisation is not indicated in an acute phase of the illness. Aneurysms usually do not rupture and surgical plication of the aneurysm has not been fruitful and has even caused catastrophic complications and death. Revascularisation is indicated in the chronic phase if there are symptoms of ischemia or if more than 10% of left ventricular myocardium is ischemic. Coronary artery bypass grafting appears to be superior to percutaneous coronary intervention.⁶ Long-term outcome of coronary artery bypass grafting in Kawasaki disease is not known.⁷ As our patient was asymptomatic without left ventricular dysfunction, we decided to treat him conservatively with warfarin and aspirin.

This report shows different morphological spectrums of Kawasaki disease in different coronary arteries in the same patient.

This report emphasises the fact that all patients of Kawasaki disease with large aneurysms should be meticulously maintained on aspirin and anticoagulation to prevent myocardial ischemia. Aneurysms are prone for thrombosis due to shear endothelial stress. Acute thrombotic occlusion results in myocardial infarction and rarely sudden cardiac death.

Supplementary Material. To view supplementary material for this article, please visit <https://doi.org/10.1017/S1047951120000426>

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Conflicts of Interest. None.

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