

Angiographic and intravascular ultrasonographic features of Kawasaki coronary artery disease

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ABSTRACT We report a 38-year-old man with previous Kawasaki disease who presented with exertional dyspnoea and was found to have multivessel coronary aneurysm and stenoses on coronary angiography. Coronary artery bypass surgery was subsequently performed. This case highlights the angiographic and intravascular ultrasonographic imaging features of this relatively uncommon condition presenting to an adult general cardiology service in Singapore.

Keywords: coronary aneurysm, coronary angiography, coronary stenosis, Kawasaki disease
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INTRODUCTION

Kawasaki disease is an acute, self-limiting vasculitis of unknown aetiology that occurs predominantly in infants and young children, with a predilection for those of Asian descent.⁽¹⁾ First described in Japan in 1967 by Tomisaku Kawasaki, the disease is now reported worldwide as an important cause of acquired heart disease.⁽²⁾ The sequelae incurred by the coronary arteries is a major determinant of mortality.⁽³⁾

CASE REPORT

A 38-year-old man with a history of Kawasaki disease diagnosed at age nine presented to our institution with recurrent exertional dyspnoea. His cardiovascular risk factors included hypertension, hyperlipidaemia and a family history of ischaemic heart disease. The patient had a previous history of angina pectoris two years ago, for which he underwent cardiac catheterisation at another hospital. Coronary angiography at that time revealed marked aneurysmal dilatations in the left main stem, proximal left anterior descending (LAD) artery and proximal circumflex arteries. There was also significant stenosis in the mid LAD artery, for which he underwent coronary stenting using a sirolimus-drug eluting stent. His right coronary artery was noted to be totally occluded at that time.

Cardiovascular examination at the time of presentation was unremarkable. The patient's resting 12-lead electrocardiogram showed normal sinus rhythm at 60 beats per minute and isolated Q wave in lead III. His two-dimensional echocardiogram revealed concentric left ventricular hypertrophy with preserved left ventricular (LV) systolic function (LV ejection fraction was visually estimated to be 65%). There was mitral valve prolapse involving the anterior leaflet, with posteriorly directed mitral regurgitation of moderate severity. The aortic root was dilated at 39 mm. A dipyridamole nuclear myocardial stress imaging study showed reversible moderate defect in the basal inferior segment,

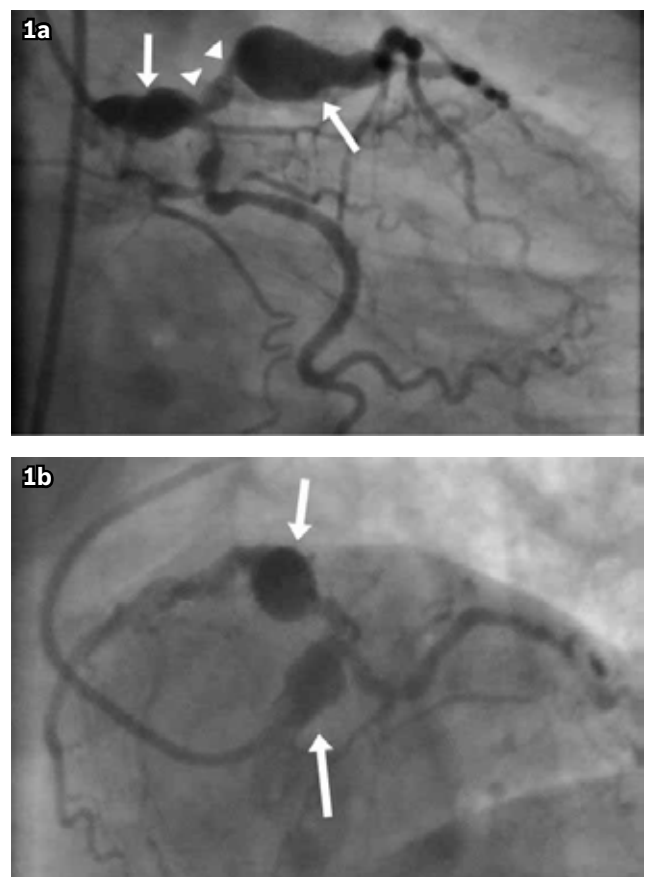


Fig. 1 (a) Right anterior oblique caudal and (b) left anterior oblique caudal projections of the left coronary arteries show aneurysms (arrows) and stenotic segments (arrowheads) of the left main and proximal left anterior descending arteries.

consistent with impaired coronary flow reserve in the right coronary artery territory.

Cardiac catheterisation was performed, which revealed aneurysmal left main and proximal LAD coronary segments (Fig. 1). There were also significant stenoses at the ostial and proximal LAD segments, but the previously stented mid LAD

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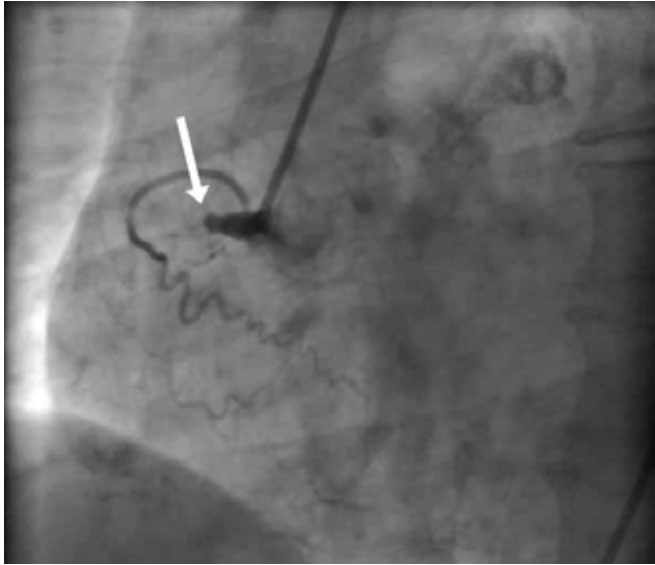


Fig. 2 Left anterior oblique projection of the right coronary artery shows a completely occluded right coronary artery (arrow).

segment was widely patent (Fig. 1). The right coronary artery was totally occluded (Fig. 2). Fractional flow reserve measurements across the ostial LAD and proximal LAD lesions with intracoronary papaverine (15 mg) challenge were 0.91 and 0.70, respectively, the latter being haemodynamically significant. Intravascular ultrasonography (IVUS) study using a Volcano Revolution 45 MHz catheter revealed that the minimum luminal area of the ostial LAD artery was 5.8 mm² and that of the proximal LAD was 4.1 mm². The largest diameter of the aneurysm in the LAD artery measured was 11 mm, with a thin layer of echo-dense intimal thickening visualised (Fig. 3).

Coronary artery bypass surgery was recommended in view of the large aneurysm with its attendant risk of rupture as well as the technical difficulty of percutaneous revascularisation. This was successfully performed with ligation of the LAD aneurysm, left internal mammary artery grafting to the mid-LAD artery, and three saphenous vein grafts to the first diagonal, circumflex and right posterior descending vessels. Mitral valve repair was also performed with Alfieri stitching and closure of cleft valve leaflet.

DISCUSSION

Our case study showed the angiographic findings of a young patient with Kawasaki disease, with its myriad morphological forms of coronary stenosis and occlusion, coronary calcification and coronary aneurysm occurring in the same coronary vasculature. The natural progression of the disease culminated in the patient undergoing successive percutaneous and surgical revascularisation over a period of two years.

Kawasaki disease is not commonly encountered in adult general cardiology clinics in Singapore. It is an example of high-risk paediatric conditions where cardiovascular adverse events occur in childhood and early adult life, and which warrants important risk reduction. The coronary artery lesions may change with time into a variety of forms. They may regress, remain unchanged, progress to stenotic or obstructive lesions

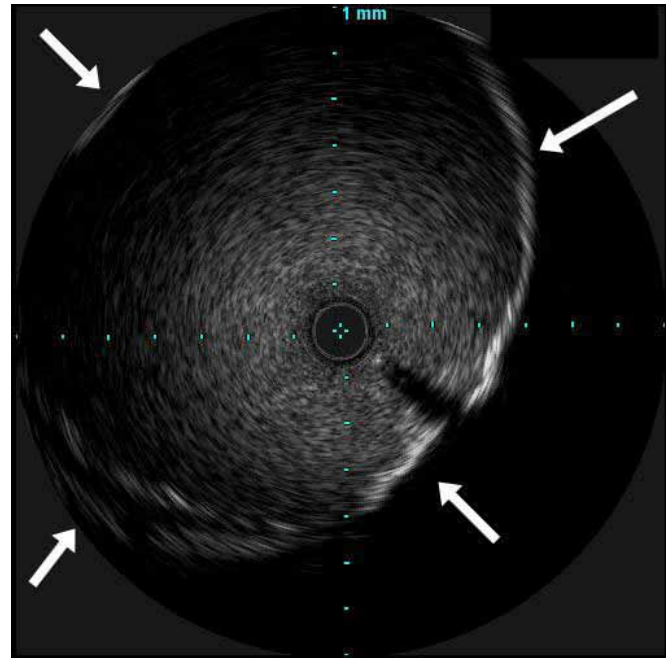


Fig. 3 IVUS image shows an aneurysmal proximal LAD lesion measuring > 11 mm in diameter. The wall of the aneurysm is marked out by arrows. This can be contrasted with Fig. 4, which shows the IVUS image of a non-aneurysmal LAD artery of normal diameter (3–4.5 mm).

with or without recanalisation and collateral formation, and rarely, coronary rupture. Smaller lesions (< 5 mm) have a higher likelihood of angiographic resolution. Calcification is a marker of chronicity and usually develops five or more years after disease onset. Coronary rupture occurs as a result of rapid dilatation of the aneurysm during the acute phase, but can also occur late (> 20 years after disease onset), albeit rarely.⁽⁴⁾

Coronary artery aneurysm occurs in about 25% of patients with untreated disease.⁽⁵⁾ Giant aneurysms (≥ 8 mm) are associated with a high risk for late complications, including thrombosis, stenosis and calcification, and can potentially lead to myocardial infarction and late mortality. Large aneurysms (6–8 mm) are similarly high risk and would require follow-up, testing and management. Fortunately, patients with giant and large aneurysms constitute the minority (< 1%) of patients with Kawasaki disease, but they would require aggressive lifelong anticoagulation, with stress tests and coronary angiogram where appropriate.

The hallmark histopathological feature of coronary artery lesions in Kawasaki disease is intimal thickening, which consists of extracellular matrix and smooth muscle cells that have migrated through the disrupted internal elastic lamina.⁽⁶⁾ The degree of intimal thickening varies from lesion to lesion, and can be found even in regressed coronary arteries. In extreme cases, the intimal thickening may cause progressive localised stenosis. Therefore, the term ‘atherosclerotic narrowing’ in patients with Kawasaki disease is a misnomer, for the underlying pathophysiological mechanism is completely different.

IVUS provides *in vivo* visualisation of the coronary wall histopathology and is especially useful in defining the morphology and dimensions of the coronary lesions, as well as for assessing

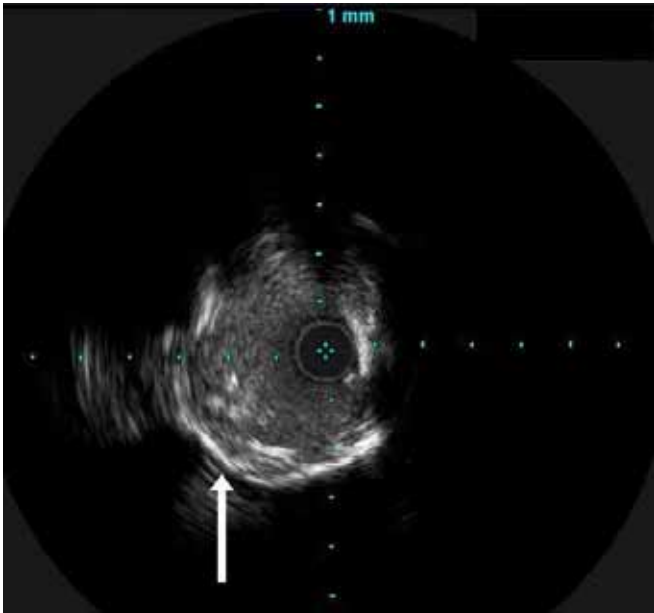


Fig. 4 IVUS image of the LAD artery shows calcification, as evident by the hyperechoic lesions with acoustic shadowing behind it (arrow).

the results of any percutaneous coronary intervention. The typical IVUS findings in patients with Kawasaki disease include the presence of intimal proliferation, demonstrated as a dense echo with a measurable symmetrical or asymmetrical thickening of the layer, and calcification (Fig. 4), as seen in our patient. This change can also be observed in angiographically regressed aneurysms with normal lumen diameters or even in angiographically normal coronary arteries.⁽⁷⁾

Besides coronary involvement, Kawasaki disease can also cause valvular insufficiencies, particularly involving the mitral and tricuspid valves, as a result of valvulitis or myocarditis-induced myocardial dysfunction. These may be transient and disappear with the acute illness, but some may persist as chronic valvular regurgitation as a result of the thickening or deformation of fibrosed valves.⁽⁸⁾ In our patient, there was intra-operative evidence of cleft anterior mitral valve leaflets with moderate mitral regurgitation. However, no sign of thickening or chronic inflammation was observed. It is uncertain if the mitral valve pathology was related to his Kawasaki disease or was an incidental finding.

As no strong prospective data exists to guide in the selection of an optimal treatment regimen, the management of coronary disease in patients with Kawasaki disease depends on the severity and extent of coronary involvement, as well as the physician's preference. Therapeutic regimens used include antiplatelet

therapy with aspirin, with or without dipyridamole or clopidogrel, anticoagulant therapy with warfarin or low-molecular-weight heparin, or a combination of antiplatelet and anticoagulant, usually aspirin and warfarin. Most experts believe that a predominantly platelet-directed strategy is sufficient in the setting of mild to moderate level disease.⁽⁹⁾

Recommendations for catheter intervention in patients with Kawasaki disease were recently formulated by Japan's Research Committee of the Ministry of Health, and include patients with ischaemic symptoms, reversible ischaemia on stress test and more than 75% stenosis in LAD (level C).⁽¹⁰⁾ Plain old balloon angioplasty is generally not successful due to dense fibrosis and calcification in the arterial wall. However, studies have shown that the use of stents and rotational ablation is able to achieve a success rate of more than 80%.⁽⁹⁾ If contraindications to percutaneous coronary intervention such as multiple, ostial or long segment stenosis, as well as severe left ventricular systolic dysfunction, are present, coronary artery bypass graft is the alternative option.

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