CASE REPORT

Calcified occlusion of the right coronary artery in Kawasaki disease: evidence of myocardial ischaemia using cardiac technetium-99m-tetrofosmin perfusion single-photon emission computed tomography

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We report the case of a 14-year-old boy who developed Kawasaki disease at 5 months of age. The patient developed severe aneurysmal disease of both the left and right coronary arteries. He eventually developed total calcified occlusion of the right coronary artery despite long-term treatment with aspirin. Catheterisation showed no antegrade flow into the right coronary artery, with retrograde flow from the left coronary system into the right coronary. At the most recent follow-up he was asymptomatic, with normal exercise tolerance and a negative exercise stress test. Single-photon emission computed tomography (SPECT) myocardial perfusion imaging was carried out during stress and at rest using intravenous persantine (dipyridamole) and technetium-99m tetrofosmin. During stress, there were prominent left ventricular apical and anteroseptal defects, which normalised at rest. SPECT during stress and at rest may detect subclinical ischaemia and influence further management options in such patients.

Abbreviation: SPECT, single-photon emission computed tomography

A boy presented at the age of 5 months with a 2-week history of cough and poor feeding. He was being treated at the time with oral antibiotics for an upper respiratory tract infection. He had a maculopapular rash but no evidence of conjunctivitis. He was in mild respiratory distress and a cardiovascular examination disclosed muffled heart sounds, with no added sounds or murmurs. Normal brachial and femoral pulses were observed. His liver was palpable 4 cm below the costal margin. No documented fever or lymphadenopathy was detected at the time.

His full blood count showed a thrombocytosis (1093 x 10^6 platelets/mm^3). A chest radiograph showed cardiomegaly and increased pulmonary vascular markings. His electrocardiograph was normal, with no evidence of ST segment changes. An echocardiogram was obtained, showing a large pericardial effusion and a dilated left coronary artery. Cardiac catheterisation showed a large right coronary artery aneurysm and a smaller left coronary artery aneurysm. A diagnosis of Kawasaki disease was made, and the child was started on a high dose of oral aspirin (100 mg/kg/day divided QDS).

Serial echocardiograms and cardiac catheterisations were carried out to evaluate the coronary arterial ectasia in this child. At the age of 3 years, catheterisation showed calcification in the rim of the large proximal right coronary aneurysm (18 mm), with passage of blood in the thrombus and retrograde filling from the left. The proximal left coronary artery was aneurysmal (8 mm) but patent. At 7 years of age, the right coronary artery was occluded with a thrombus, but considerable collaterals had developed between the left and right coronary arteries. At 10 years of age, the diameter of the right coronary artery had increased to 20 mm and that of the left coronary artery to 15 mm. At 13 years of age, his arteriogram showed total occlusion of the proximal right coronary artery with retrograde filling from the left coronary artery from a collateral circulation (fig 1). He was started on oral clopidrogel at this stage.

The child remained asymptomatic throughout the follow-up, with normal exercise tolerance by history. His cardiovascular examination remained normal, and serial transthoracic echocardiography failed to show any regional wall-motion abnormalities. An exercise stress test (Bruce protocol) at his most recent review was negative with an exercise time of 15 min. Single-photon emission computed tomography (SPECT) was carried out 1 hour after intravenous persantine (dipyridamole) and at rest after injection of technetium-99m tetrofosmin. Myocardial perfusion was evaluated using the stress and rest images. During stress, there were prominent left ventricular apical and anteroseptal defects, which normalised at rest (fig 2). Assessment of left ventricular regional wall motion post stress also showed moderate hypokinesia of the left ventricular apex, which correlated with the region of abnormal perfusion (fig 2).

Figure 1  Coronary angiography showing aneurysmal left coronary artery. Note the calcification in the right coronary artery with retrograde flow from the left coronary artery supplying the left coronary artery.

Abbreviation: SPECT, single-photon emission computed tomography

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DISCUSSION

This case shows an atypical presentation of Kawasaki disease in the absence of any fever, lymphadenopathy or oral mucocutaneous changes, but the development of coronary arterial ectasia eventually unveiled the disease process. This report highlights the potential of developing total occlusion of a coronary artery and yet, in the presence of an adequate collateral circulation such patients remain asymptomatic, with a negative stress test and no evidence of regional wall-motion abnormality by transthoracic echocardiography. Although he received aspirin and clopidrogel, this patient went on to develop thrombosis of the right coronary artery, indicating that despite these treatments late occlusion may develop.

SPECT myocardial perfusion scanning highlighted a severe apical and anteroseptal defect using intravenous dipyridamole in this patient. The utility of SPECT in patients with Kawasaki disease and coronary anomalies has been previously reported. The finding of a reversible apical defect in this patient, corroborated by apical hypokinesis during stress, showed that this patient had compromised myocardial perfusion and function despite a normal transthoracic echocardiogram and exercise stress test. This highlights the importance of carrying out additional stress imaging studies in patients after Kawasaki disease rather than relying solely on conventional echocardiography or cardiac catheterisation. Intravenous dipyridamole pharmacologic stress produces a marked increase in myocardial blood flow exacerbating resting perfusion defects. Technetium-99m-tetrofosmin provides superior image quality and requires lower radiation dose than thallium myocardial perfusion imaging. Quantitative electrocardiographically gated SPECT myocardial perfusion imaging during low-dose dobutamine infusion has also been shown to be a safe and efficacious method for the combined evaluation of myocardial contractile reserve and myocardial perfusion. Furthermore, in one study of 90 patients with coronary aneurysms, using multivariate regression, reversibility by cardiac thallium SPECT was the most predictive factor of patients developing late cardiac events, including unstable angina, myocardial infarction or death.

The gradual development of thrombosis may have aided in the development of collateral supply to the right coronary territory. An acute thrombotic episode may not have been tolerated in the same setting. The earlier use of catheter-delivered thrombolytic agents may have allowed re-establishment of vessel patency in this patient, and more recent guidelines have highlighted the potential of abciximab to reduce the potential of coronary thrombosis in patients with Kawasaki disease. Several reports have reported on revascularisation procedures in patients with demonstrable myocardial ischaemia, and this patient may warrant revascularisation to the left coronary arterial system.

This report highlights the clinical utility of cardiac SPECT in children with coronary arterial disease as sequelae of Kawasaki disease. Such investigations may alter strategic intervention or medical therapy in such children. Care should be taken to minimise the risks of radiation exposure in such patients by adjusting isotope dosage according to patient weight.

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REFERENCES
Idiopathic infantile arterial calcification presenting as fatal hypertensive cardiomyopathy

A 5-month-old girl was admitted to our paediatric intensive care unit with cardiorespiratory failure and hypertension. Chest radiography showed cardiomegaly and pulmonary plethora. Electrocardiography showed left ventricular hypertrophy and strain pattern. Echocardiography showed a dilated and hypertrophied left ventricle, with bright and hyper-reflective myocardium (fig 1). Fractional shortening was 12.5%. The concentration of creatine kinase was 493 U/l (normal range 20–190) and that of troponin I was 3.1 µg/l (normal range <0.03).

The patient initially responded to treatment but collapsed and died 18 h later.

Postmortem examination showed left ventricular hypertrophy with no outflow obstruction or coarctation. The coronary arteries had a normal origin and distribution but were thick walled and diffusely narrowed (fig 2). Cutting into the vessels was difficult. The thoracic and abdominal aorta and great vessels were also diffusely thickened. Both renal arteries were thickened and completely occluded. There were no thrombi.

Histological examination showed a deposition of calcium in the internal elastic lamina of the coronary and renal vessels, with fibroproliferative changes in the intima, resulting in occlusion (fig 3). At a magnification of × 100, appearances were typical of those seen in idiopathic calcification of infancy, a genetic disorder resulting from heterozygotic nonsense mutations in ectonucleotide pyrophosphatase phosphodiesterase-1. This causes calcification of the arterial internal elastic lamina, resulting in occlusive coronary artery disease. Calcification in a peripheral artery with electrocardiographic changes of occlusive coronary artery disease suggests the diagnosis. Many patients die from myocardial infarction in the first 6 months of life. Hypertension with cardiac failure is a common mode of presentation.

References

Images in Paediatrics

Figure 1: Parasternal long-axis echocardiogram showing hypertrophy and dilated left ventricle. Ao, aorta; LA, left atrium; LV, left ventricle.

Figure 2: Gross pathology of the left ventricle at necropsy showing hypertrophied left ventricle (LV), normal aortic outlet (Ao) and thick-walled coronary artery with narrow lumen (arrow).

Figure 3: Microscopic pathology of the left coronary artery showing deposition of calcium in the internal elastic lamina and fibroproliferative changes in the intima.