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RESEARCH LETTER

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A child with Kawasaki disease who survived after rupture of a coronary artery aneurysm

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We report on a child with Kawasaki disease complicated by the development of a giant coronary artery aneurysm (CAA) progressing to rupture, successfully resuscitated and treated by emergency coronary artery bypass.

A 5-year-old boy was initially treated for lymphadenitis by a general practitioner; however, the fever persisted and he developed conjunctival injection, skin rash and cracked lips. He was then treated for Kawasaki disease with intravenous immunoglobulins (400 mg/kg per day) on the 9th day of illness for 3 consecutive days; fever subsided, but an echocardiography on day 13 revealed a grossly dilated right coronary artery and he was transferred to our hospital for further management. He was afebrile on admission and echocardiography revealed a giant right CAA measuring 18.6 mm in maximal diameter with a huge thrombus obliterating nearly the whole lumen (Fig. 1). The left coronary artery was also uniformly dilated; the platelet count was 1110×10^9/l and the ESR was 143 mm/h. In view of the high risk of complete coronary occlusion, he was heparinised together with administration of aspirin and warfarin. Abciximab was also given because of the severe thrombocytosis. The platelet count dropped to 786×10^9/l on the following day and the ESR dropped to 35 mm/h on the 4th day after admission. However, fever recurred on the 7th day with elevated ESR and persistently high platelet count. Therefore intravenous immunoglobulin (1 g/kg) was administrated again and he became afebrile afterwards. Nevertheless, the right CAA continued to enlarge, measuring 30 mm at its maximal diameter on MRI at 4 weeks (Fig. 2).

After discussion with cardiothoracic surgeons, coronary arterial bypass surgery was deemed necessary to prevent the imminent rupture. Unfortunately, the day before surgery, he suddenly collapsed in the ward during a temper tantrum. Immediate echocardiography and pericardiocentesis confirmed cardiac tamponade with blood. He was resuscitated and a pericardial window was created urgently at the bedside. He was successfully revived and rushed to the operating theatre, where a right CAA rupture was confirmed and emergency coronary artery bypass surgery was performed utilising the left saphenous vein as the graft. Post-operative inotropic support was weaned off within 36 h. He was extubated 2 days after surgery. Right ventricular function normalised within 1 month. He has remained well 6 months thereafter, receiving only aspirin for persistent left coronary artery dilatation.

Giant CAA occurs in 0.5%–1% of patients with Kawasaki disease with a fatality rate of 4% [3,6]. Risk factors for the development of a giant CAA during the acute phase of Kawasaki disease are not entirely clear. However, late start of intravenous immunoglobulin administration and the requirement of additional doses of immunoglobulin to control fever have been shown to be important risk factors, which are probably exemplified in this case. Abciximab, a platelet glycoprotein IIb/IIIa receptor inhibitor, which has been increasingly investigated for its thrombolytic effect and its potential role in regression in CAA [5] was used in our case. It might have helped in the reduction of the size of coronary artery thrombus, but failed to arrest the progression of enlargement of a giant CAA.

Rupture of a CAA inevitably results in death due to cardiac tamponade [4]. So far, only one patient with Kawasaki disease has survived after rupture of a giant CAA, this patient had an emergency coronary artery

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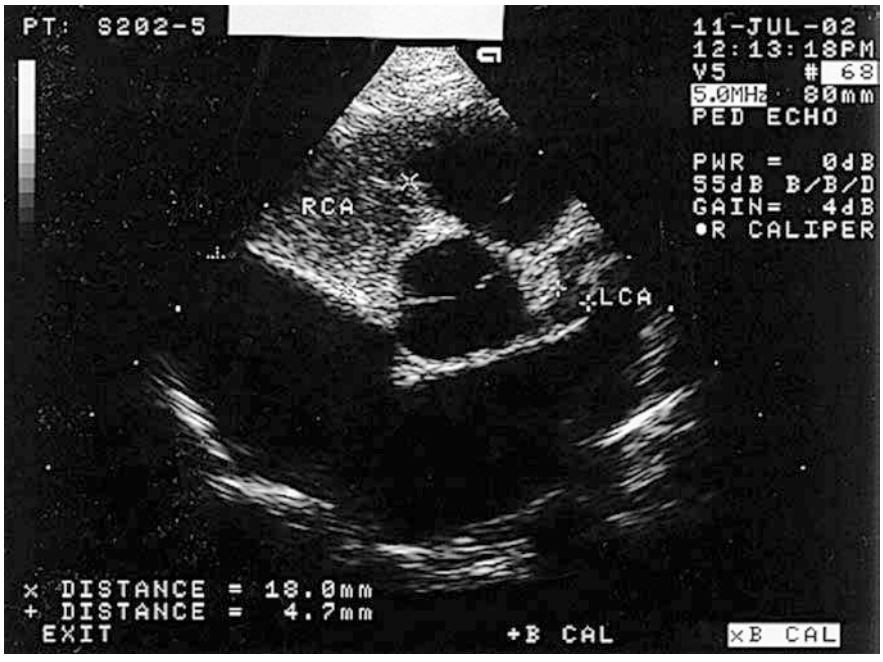

Fig. 1 Echocardiography with short-axis view at 2 weeks after the onset of illness showing a huge right CAA with suggestion of thrombus formation. The left coronary artery was also dilated. (*RCA* right coronary artery, *LCA* left coronary artery)

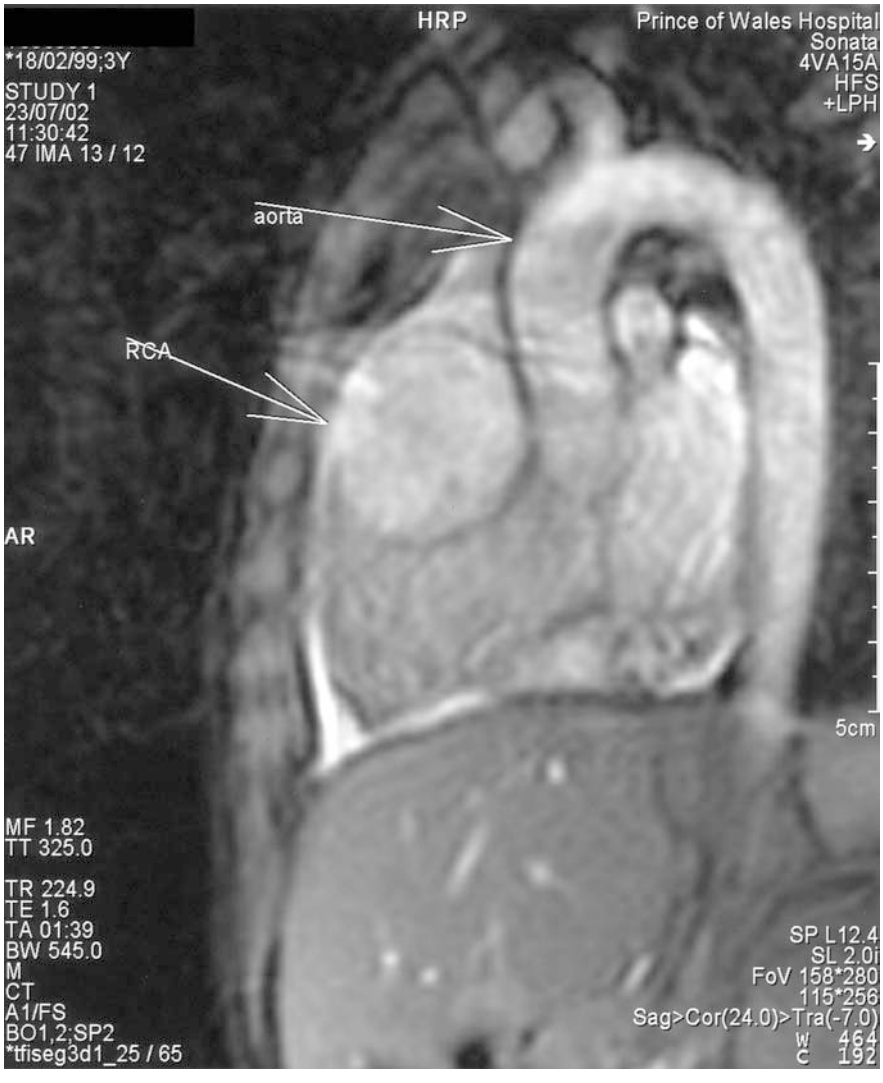

Fig. 2 Cardiac MRI at 4 weeks showing the huge right CAA. (*RCA* right coronary artery)

bypass surgery followed shortly by orthotopic heart transplantation [2]. Our patient is the first who survived after rupture of a giant CAA and successfully went through the emergency coronary artery bypass surgery. However, we were only able to perform saphenous venous bypass grafting due to the emergency life-saving nature of the operation, as opposed to the originally planned internal thoracic arterial graft which was reported to have a higher long-term patency rate [1]. Thus, we advocate consideration for elective coronary artery bypass surgery early in the course of disease in patients with Kawasaki disease and a giant CAA with relentless progression.

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