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A School-Goer's Nightmare: Case Report of CABG in an 8 Year-Old with Kawasaki Disease

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Abstract **Case Report**

Introduction: Kawasaki disease (KD) is an acute vasculitis primarily affecting medium-sized arteries, particularly the coronary arteries (CA). Its impact varies, with some patients developing transient dilation while others experience severe coronary artery aneurysms (CAA), leading to complications such as thrombosis, stenosis, and myocardial ischemia. Male gender, delayed diagnosis, incomplete presentation of KD, and resistance to Intravenous Immunoglobulin (IVIG) therapy are risk factors for CAA. The Japanese Ministry of Health classifies CAAs by size, with giant aneurysms (>8 mm) carrying significant long-term risks, including coronary thrombosis and acute coronary syndrome. ***Case Report:*** We report the case of an 8-year-old Malay boy with giant CAA secondary to KD. The patient presented with classical KD symptoms and inflammatory markers, with echocardiography revealing a giant aneurysm (8.7 mm) in the left anterior descending artery (LAD), alongside smaller aneurysms in the left circumflex and right coronary arteries (RCA). Initial management included IVIG, aspirin, and heparin, with warfarin and clopidogrel added subsequently. Coronary angiography indicated severe LAD stenosis (95%) with total occlusion and an aneurysmal segment distal to the occlusion. A multidisciplinary team determined that coronary artery bypass grafting (CABG) using bilateral internal mammary arteries was the optimal intervention. The patient had a favourable recovery and was stable at discharge. ***Discussion:*** CABG is recognized as a preferred treatment for severe or multivessel coronary involvement in KD, offering lower reintervention rates compared to percutaneous coronary intervention (PCI). Studies support CABG's efficacy in managing coronary complications in KD, especially for complex or multivessel cases. ***Conclusion:*** This case illustrates the complexities of managing giant CAAs in paediatric KD and underscores the necessity of a multidisciplinary approach and long-term monitoring to optimize patient outcomes and mitigate complications.

Keywords: Kawasaki Disease, Giant Aneurysm, Coronary Artery Aneurysm, Paediatric Coronary Artery Bypass Graft, Coronary Thrombosis, Bilateral Internal Mammary Artery.

INTRODUCTION

Kawasaki disease (KD) is an acute vasculitis which affects the medium-sized arteries, particularly the coronary arteries (CA). The extent of CA involvement varies from transient mild dilatation or ectasia, observed in approximately 40% of patients, to giant coronary artery aneurysms (CAA) [1]. Factors associated with a higher risk of CCAs include gender where males show greater propensity to develop CCAs, delayed diagnosis, incomplete presentation of KD, and KD that is resistant to Intravenous Immunoglobulin (IVIG) therapy [2].

The Japanese Ministry of Health defines CAAs as having an internal lumen diameter greater than 3mm in children under 5 years, or greater than 4 mm in children 5 years or older. Other diagnostic criteria include a segment which is 1.5 times greater than the adjacent segment, or when there is luminal irregularity. CAAs are further classified into small (< 5mm), medium (5-8mm), and giant (>8mm) aneurysms [2].

Despite the use of IVIG, the incidence of CAAs remains significantly high at 9% in Malaysia [2]. Patients with aneurysms are at lifelong risk of coronary thrombosis or developing stenosis, potentially leading to myocardial ischaemia, infarction or death [3].

Histopathological studies in patients with a history of KD show widespread arterial changes including inflammatory cell infiltration, intimal and medial disruption, intimal myofibroblastic proliferation,

and replacement of myocytes with fibroblasts and connective tissue [4].

Large or giant aneurysms (≥8 mm or Z score ≥10) are the least likely to resolve, and are associated with up to a 50% risk of thrombotic coronary occlusion, progressive stenosis requiring revascularization, or acute coronary syndrome within 30 years post illness [5, 6].

CASE REPORT

This case report details the presentation and management of an 8-year-old Malay boy diagnosed with KD and a giant CAA, treated at Hospital Serdang through a multidisciplinary approach. The patient, born at full term with a birth weight of 2.8 kg, presented at 6 years and 10 months old in December 2019 with classical symptoms of KD, including high-grade fever for 10 days, bilateral non-exudative painless conjunctival injection, polymorphous rash, unilateral cervical lymphadenopathy, strawberry tongue, and dry, red, and chapped lips. Laboratory findings revealed elevated C-reactive protein (CRP 124), elevated erythrocyte sedimentation rate (ESR >120), thrombocytosis (platelet 776), leukocytosis (leukocyte 17), and hypoalbuminemia (albumin 26). An abdominal ultrasound was normal, but an echocardiogram revealed a giant aneurysm (8.7 mm) in the left anterior descending artery (LAD), multiple smaller aneurysms in the left circumflex artery (4.1 mm), mid-right coronary artery (RCA) (4.97 mm), distal RCA (4.4 mm), and mild dilation of the left main coronary artery (3.6 mm). The patient was treated with IVIG (2 gm/kg) 6 hours after admission and remained afebrile thereafter. Additional treatment included intravenous heparin infusion (25 IU/kg/hour), oral warfarin (2.5 mg OD), and oral aspirin (75 mg OD). The patient was discharged after one week of hospitalisation, having achieved an INR target of 2–3.5. A second antiplatelet agent, clopidogrel (3.6 mg OD), was added during a follow-up in February 2020, after echocardiography showed enlarged aneurysms.

In June 2020, the patient experienced his first episode of central chest pain, characterised by a pricking sensation that resolved spontaneously within 10 minutes. Electrocardiography (ECG) showed no evidence of ischemia, and blood tests were normal. The echocardiogram revealed no coronary thrombus or increase in aneurysm size, and no active intervention was necessary. The patient experienced five more episodes of exertional chest pain in the same month, resolving at rest. A routine echocardiogram in September 2021 revealed a thrombus within the giant LAD aneurysm, with no significant changes in the size of other aneurysms. A CT angiography (CTA) in September 2021 confirmed a total calcium score of zero, a thrombosed giant proximal LAD aneurysm with distal LAD reconstitution likely from intramyocardial collateral vessels from the distal RCA, and three small to medium fusiform RCA aneurysms with a small mural thrombus in the proximal RCA aneurysm. There was no significant stenosis.

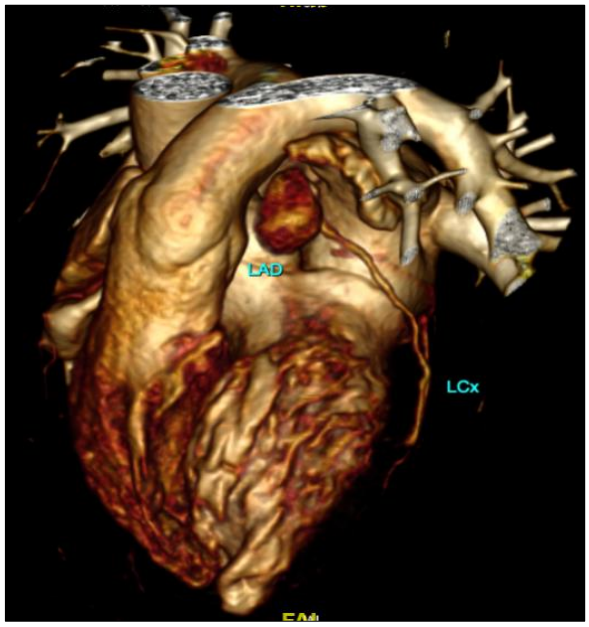

Figure 1a: Showing the CTA image of left anterior descending artery (LAD) giant aneurysm

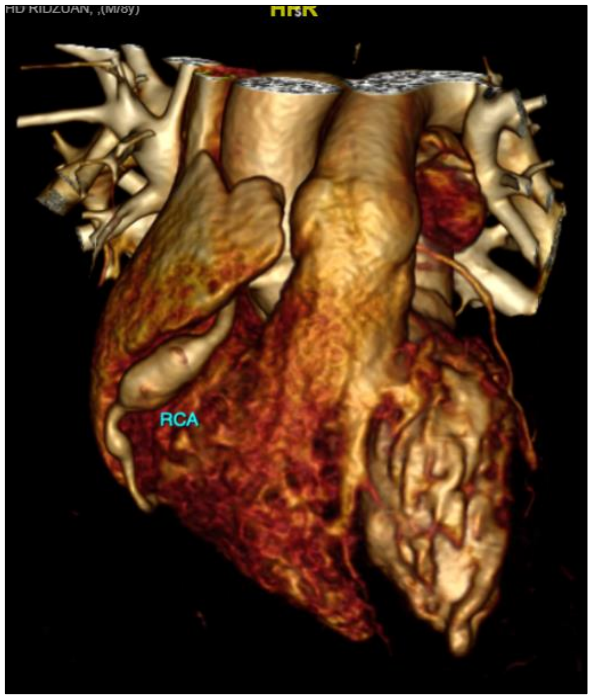

Figure 1b: Showing the CTA image of right coronary artery (RCA) aneurysms.

In the same month, a coronary angiogram was performed, which revealed severe stenosis (95%) in the proximal LAD, with total occlusion of the LAD. An aneurysmal segment was identified distal to the occluded portion. Following a multidisciplinary team discussion, the decision was made to proceed with coronary artery bypass grafting (CABG).

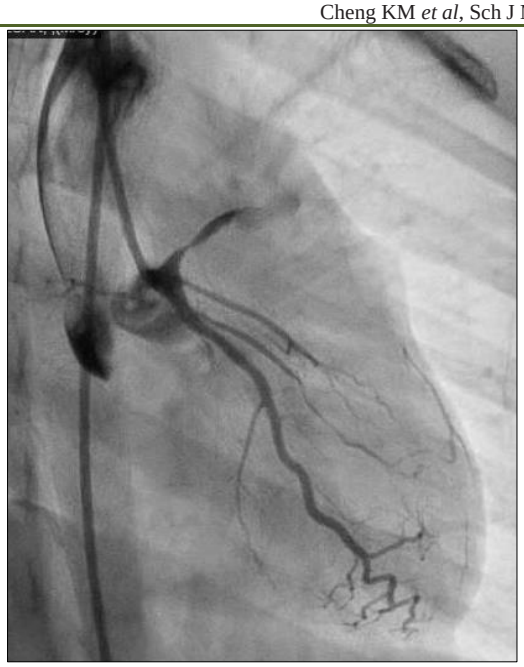

Figure 2a: Showing the coronary angiogram image of giant aneurysm in the LAD with total occlusion of the LAD

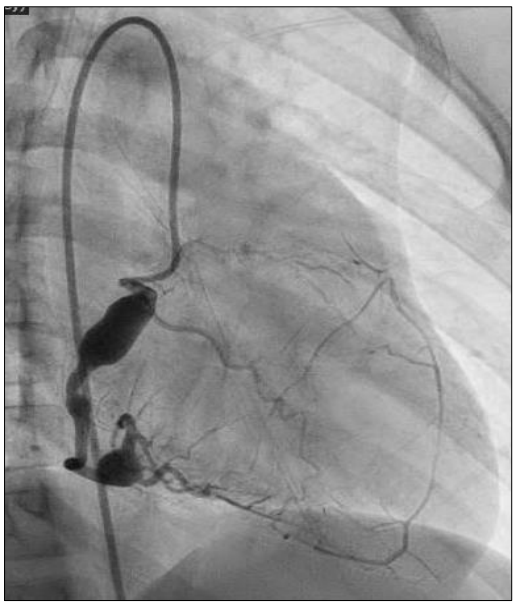

Figure 2b: Showing the coronary angiogram image of RCA aneurysms and retrograde supply to the LAD

The patient underwent CABG on 2nd December 2021. Intraoperatively, an aneurysm was visualised over the proximal LAD, measuring 1.5cm x 3cm. Another two aneurysms were located over proximal and distal RCA, each measuring 1cm x 2cm. Bilateral internal mammary arteries were used as conduits: the left internal mammary artery was anastomosed at the mid LAD, which was distal to the aneurysm, while the right internal mammary artery was anastomosed at the mid RCA, between the two aneurysms. The diameters of the mid LAD was 1.5mm, and the mid RCA was 2mm, respectively. The distal end of the LAD aneurysm was ligated with Prolene 5/0 suture.

Postoperatively, the patient was transferred to the paediatrics cardiac intensive care unit with stable haemodynamics. An intravenous infusion of glyceryl trinitrate was initiated to maintain patency of the bilateral internal mammary arteries. A stat dose of Aspirin was administered six hours postoperatively to improve graft patency. The patient was successfully extubated on postoperative day 1. Throughout the admission, the patient remained stable and was discharged on postoperative day 6 with dual antiplatelet therapy (tablet cardiprin and syrup clopidogrel), syrup carvedilol, syrup atorvastatin, syrup furosemide, syrup pantoprazole, and syrup paracetamol. The patient has since been under regular follow-up at the paediatric cardiology clinic and remains in good health.

DISCUSSION

Despite extensive research, the exact aetiology of KD remains elusive. A systematic review done by Yuan based on 71 articles related to KD, 637 children with KD who have undergone cardiac procedures were analysed [7], and the incidence of cardiac sequelae in KD in males were higher as opposed to females with a ratio of 3.1:1. Recent studies show that, siblings are more than 10 times likelier to develop KD compared to the general population [8]. However, there is insufficient evidence to suggest that the KD is transmitted from person to person.

Multiple modes of management have been taken into account for effective management of CA thrombosis following KD.

Percutaneous Coronary Intervention (PCI) versus CABG

CABG is the preferred mode of treatment in KD with CA involvement. Based on a study by Dionne *et al*., amongst 22 children across Canada, where 11 of them underwent CABG, it was reported that none of the children required re-intervention post-CABG. However, 7 of those who had undergone PCI required reintervention [9]. A separate study done by Muta *et al*., in Japan, involving 148 children, found that there was no significant difference in immediate mortality rate amongst the children who have undergone PCI or CABG. However, there was significantly higher repeat revascularisation amongst the PCI group than that in the CABG group [10]. This shows that the results of CABG are superior particularly in multivessel intervention post KD as opposed to PCI.

Indications for CABG in KD

The American Heart Association guidelines states that (1) severe occlusion of the left main coronary artery, (2) severe occlusion of the proximal segment of the LAD, (3) severe occlusion of more than one major coronary artery, and (4) recurrent myocardial infarction are indications for CABG [11]. The indications for CABG in KD also include presence of clinical signs of myocardial ischaemia or positive signs of ischaemia by ECG or scintigraphic examination [11]. Surgical treatment is strongly recommended for children with a previous myocardial infarction (MI) because prognosis after MI was reported to be unfavourable [13].

The Conduit of Choice

The choice of vessels for coronary artery grafting has been subject to much discussion. In 1976, the Nara Medical University in Japan reported its first paediatric CABG procedure utilising a saphenous vein graft (SVG) for this condition [12], however the graft eventually occluded. This was then followed by the first use of the internal thoracic artery (ITA) in 1985 [13], wherein cases of ITA-LAD was done and documented to have a 100% patency, three years after the CABG. The authors of that study also found that the ITA grafts' growth corresponded to the somatic growth of the children [14], thus becoming the conduit of choice for future KD patients. The patency of arterial grafts based on a study by Yoshikawa *et al*., done on 100 patients with KD, was 94% in the first postoperative year, 82% in the fifth year, and 78% in the tenth, which was higher than the patency of venous grafts which was reportedly 82%, 63% and 36%, respectively. Approximately 70% of these children did not have any cardiac events in 10 years postoperatively [14]. To date, Nara Medical University has operated on 114 paediatric patients under the age of 20, demonstrating favourable long-term outcomes with ITA grafts [15].

In our case report, early CABG was done as the child was experiencing recurrent chest pain. Preoperatively, an extensive multidisciplinary team (MDT) discussion involving paediatric cardiothoracic surgeons, adult cardiologist, paediatric cardiologist, paediatric intensivist, and cardiothoracic anesthesiologist was held to discuss on the best treatment option and postoperative care plan for this child. The final consensus was for CABG. A median sternotomy approach with harvesting of bilateral IMA as conduit was planned. Central cannulation of ascending aorta and 2-stage venous cannulation via right atrium was decided upon. The operative findings were uneventful as described above. Post-operative recovery was uneventful and the child was eventually discharged home with recurrent follow-up with the paediatric cardiology team.

CONCLUSION

This case highlights the complexity and risks associated with managing giant CAAs in KD, emphasising the need for multidisciplinary approach and long term follow up to monitor and manage potential complications. CABG with bilateral IMA as arterial conduit is a feasible surgical option for Kawasaki disease in paediatric age groups with good outcome.

Ethical Approval: This case series has been registered under National Medical Research Register (NMRR).

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AUTHOR CONTRIBUTION

Cheng KM is the primary author who did the literature research and case write up. Bhavani K assisted in writing and proofreading the case report. Han JC prepared the case summary and images. Norliza A is the paediatric cardiologist in-charge managing this patient. Hamdan L and Mohamad Arif MN are the supervisors for this case report and were the consultant surgeons who had performed this surgery

Conflict of Interest Disclosure: The authors declare no conflict of interest with regards to the content of this report.

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