Indian Journal of Thoracic and Cardiovascular Surgery (May-June 2021) 37(3):316–319
https://doi.org/10.1007/s12055-020-01037-5

CASE REPORT

A rare case of Kawasaki disease with giant coronary artery aneurysm

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Received: 26 May 2020 / Revised: 29 July 2020 / Accepted: 12 August 2020 / Published online: 21 September 2020

Abstract

Kawasaki disease (KD) is an acute, self-limiting vasculitis that occurs in children of all ages. This was first described by Kawasaki in 1967. Spontaneous regression is observed; however, 25% of patients develop coronary artery aneurysm (CAA). These may result in ischaemic heart disease causing myocardial infarction, rupture leading to pericardial tamponade and distal embolization which culminate in sudden cardiac death. Diagnosis of KD relies on clinical suspicion with no gold standard diagnostic test. A case of KD with giant CAA in a 14-year-old female is described with emphasis on challenges pre- and peri-operatively. The review provided post description of the case emphasizes on pathophysiology with clinical course of CAA in association with KD and justification of our approach with an insight into newer treatment modalities.

Keywords Kawasaki disease · Coronary artery · Aneurysm · Internal thoracic artery · Coronary artery bypass grafting

Introduction

Kawasaki disease (KD) is a systemic vasculitis syndrome of unknown aetiology wherein vasculitis is responsible for all manifestations. Although rare, coronary artery aneurysms (CAA) are seen in 25% cases of KD of which 50% either persist or progress with stenosis causing myocardial ischaemia accounting for most of the mortality in KD [1]. The aneurysms which persist beyond the first year are unlikely to regress [1]. Giant CAA can thrombose, calcify and rarely rupture. A case of a female child with giant CAA due to KD is described wherein coronary revascularization with bilateral Internal thoracic artery (ITA) grafting was done. This is followed by literature review focussing on pathophysiology of CAA with KD and treatment with an insight into newer modalities.

Case report

A 14-year-old female child presented with complains of progressive dyspnoea and angina on exertion since 5 years of age. Cardiology evaluation confirmed the diagnosis of KD with thrombosed CAA. She was kept on medical follow-up with oral anticoagulants, antiplatelet, beta blocker and nitrates. There was an episode of fungal pneumonia 7 years back for which she required hospitalization, prolonged mechanical ventilation and tracheostomy. She was discharged after a month and subsequently decannulated. After the discontinuation of antifungals, she was tested for fungal infection by two fungal cultures serum galactomannan which were negative. Cardiology evaluation by echocardiography revealed no evidence of fungal endocarditis in subsequent follow-up. Steroids were discontinued post this episode. She developed subglottic stenosis and left vocal palsy for which she presented with stridor 2 years back and underwent treatment under otolaryngology care. Preoperative electrocardiography was unremarkable. Chest X-ray revealed cardiomegaly with left ventricular type apex. Echocardiographic evaluation revealed no areas of wall motion abnormalities with normal valves and ventricular function. Computerized tomographic (CT) angiography was done which demonstrated a thrombosed aneurysmal right coronary artery (RCA) in proximal and mid part measuring 6.8 mm. Distal part of left main with proximal-mid left anterior descending (LAD) was also aneurysmal measuring 12.8 mm with peripheral thrombosis and calcification (Fig. 1). Distal RCA and LAD were normal. Blood tests were unremarkable. Blood investigations to assess the disease process and inflammation as described in literature were performed [2]. Haemoglobin with differential leucocyte count, platelet count, albumin, erythrocyte sedimentation rate and C-reactive protein were unremarkable suggesting no active

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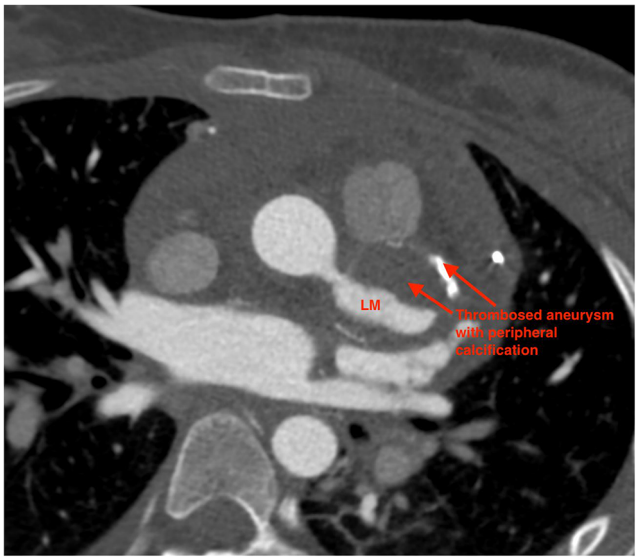

Fig. 1 Preoperative CT angiography showing giant CAA of distal left main with thrombosis and peripheral calcification

inflammatory process. The blood investigations were unremarkable with normal levels of serum galactomannan done before surgery to screen for persistence of fungal infection.

The presence of giant CAA in distal Left main and proximal LAD with moderate-sized aneurysm in RCA with the presence of thrombus and the ongoing angina prompted us for surgical revascularization. Lateral X-ray of neck was done to assess airway which seemed adequate. Otolaryngologists (ENT—ear, nose and throat specialists) were consulted before surgery for any residual stenosis of subglottic area requiring intervention. No such stenosis was found on examination by ENT team which could have warranted an intervention. Warfarin and aspirin were stopped 5 days before surgery and patient was kept on intravenous heparin until 6 h before surgery.

In supine position, median sternotomy was performed, and pedicled bilateral ITA grafts were harvested. After administering heparin at a dose of 400 units/kg, both ITAs were divided distally and adequate flow was observed in both the conduits. Cardiopulmonary bypass was instituted with aorta-right atrial cannulation. Myocardial protection was provided with moderate hypothermia at 28 °C and modified cold blood cardioplegia. There was no evidence of active vasculitis in form of erythematous vessels or friable tissues on table. The aneurysmal areas were hard on palpation (thrombosed and peripherally calcified) hence, left as such. Left internal thoracic artery (LITA) to LAD and right ITA (RITA) to RCA were anastomosed distal to aneurysmal site. Distal coronaries were normal with adequate lumen of 1.5 mm with no thrombus. The patient was rewarmed, aortic cross clamp was released and was slowly weaned off cardiopulmonary bypass. Patient was shifted to intensive care unit with stable haemodynamics on 5 microgram/kg/min of dobutamine and 0.5 microgram/kg/min of nitroglycerin. Postoperative course was uneventful. She was electively ventilated for 24 h due to past history of subglottic stenosis. Low molecular weight heparin was started 8 h after surgery once chest drain outputs were minimal and was continued for 48 h. Tablet aspirin was restarted along with warfarin from 1st postoperative day. Inhaled bronchodilator and intravenous hydrocortisone were administered before extubation. The rest of the postoperative course was uneventful and she was discharged on day 8. Computerized tomographic angiography done at 6 months of follow-up revealed patent grafts with no change in coronary aneurysms size (Figs. 2 and 3). She was discharged on antiplatelet (Ecosprin 75 mg once daily (OD)), oral anticoagulant (warfarin, started with 2 mg OD with International normalized ratio monitoring maintained around 2 with out patient department (OPD)-based testing), beta blocker (metoprolol 12.5 mg twice a day) and statin (rosuvastatin 20 mg OD at night). She is doing fine at 9 months follow-up and is asymptomatic. We planned for echocardiographic evaluation at 3, 6, and 12 months and yearly thereafter. The CT angiography will be repeated at 1 year and then further planning will be done in accordance with our multi-disciplinary team.

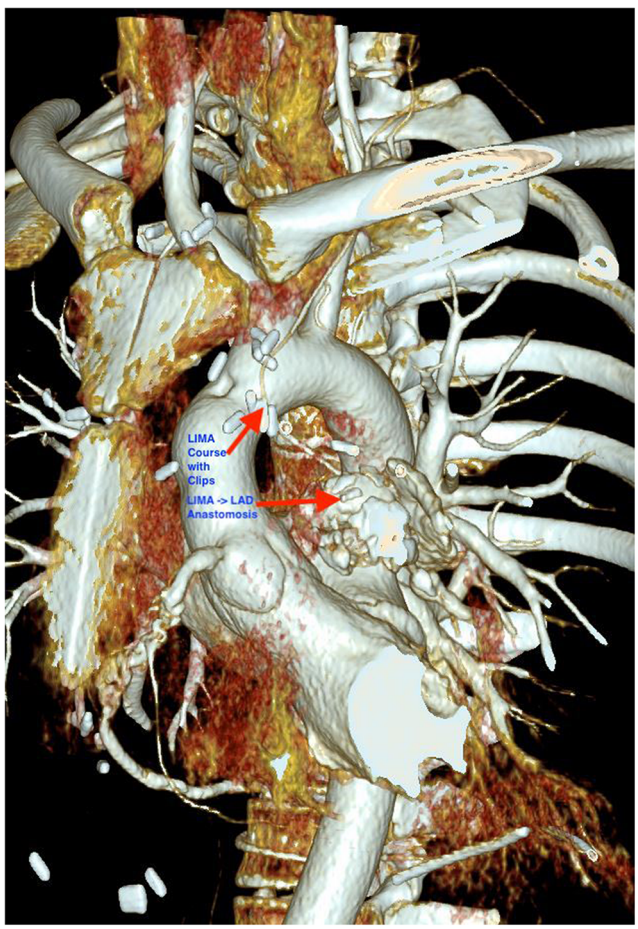

Fig. 2 3D reconstructed postoperative CT angiogram showing patent LIMA->LAD anastomosis

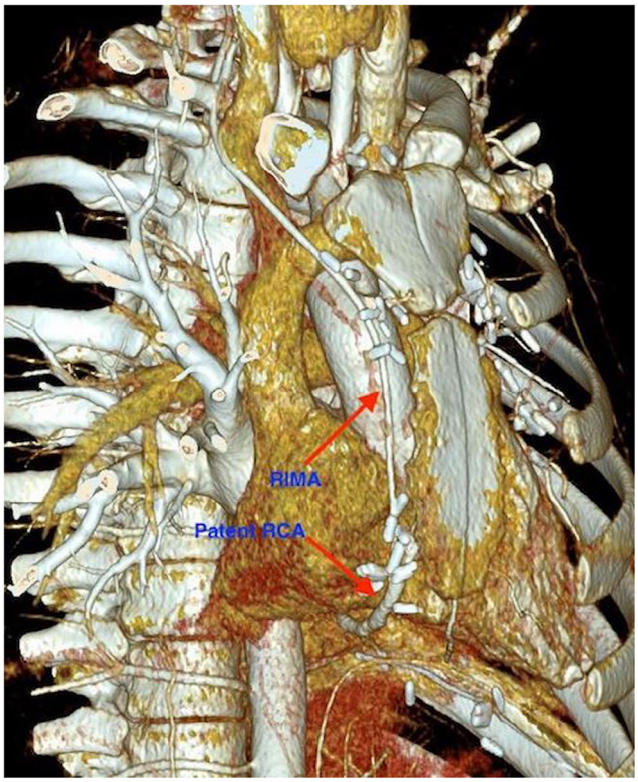

Fig. 3 3D reconstructed postoperative CT angiogram showing patent RIMA->RCA anastomosis

Discussion

KD is a systemic vasculitis having genetic predisposition being triggered by an unknown infection usually affecting children less than 5 years of age with female predominance. It is observed in Asian countries especially Japan [1].

The inflammatory process with abundant monocytes and macrophage proceeds via acute, healing and cicatricial phase. The pan vasculitis damages the elastic laminae and disrupts the intima leading to aneurysmal dilatation which is usually observed around the 12th day [2, 3]. If diagnosis of KD is confirmed, administration of intravenous immunoglobulin within 10 days at a single dose of 2 g/kg significantly reduces the risk of CAA from 25 to 5%; however, early fever and constitutional symptoms are often missed and the disease progresses [2]. The delayed healing progresses to giant aneurysm which are ≥ 8 mm size in children < 5 years of age or the internal diameter of the area being 4 times the adjacent segment. CAA are described as 1.5 times that of the native coronary artery [3, 4].

Altered haemodynamics in the aneurysm lead to thrombosis and reduced flow reserve of myocardium and therefore, these patients are kept on anticoagulant and antiplatelet therapy.

The CAAs may have adverse sequelae like coronary dissection, rupture resulting in tamponade, thrombus with ischaemia to the myocardium, compression of structures in vicinity or formation of a fistulous tract. The risk of thrombus formation exceeds with size with preponderance in those above 5 mm in size [3]. There is also a risk of distal embolism in rare scenarios; however, the low prevalence of CAAs, studies having small size cohorts with varied outcomes, and exact incidence cannot be predicted emphasizing the need for long-term follow-up establishing the course. A meta-analysis of various studies revealed that these adverse cardiac events reduced significantly when patients were maintained on antiplatelet and anticoagulant together versus in isolation [5]. The risk of rupture is rare even in giant CAAs. Small (< 4 mm) to moderate-sized CAA (4–8 mm) may produce post aneurysmal stenosis and a risk of distal thrombo-embolism prevails; hence, these lesions should also be re-vascularised while treating the giant CAAs [3, 5]. This was followed in this case too by revascularization of right-side moderate CAA along with the left-sided giant CAA.

Although, coronary angiography is the gold standard; however, being invasive, lack of anatomical delineation, underestimation of size in presence of thrombus and the relative absence of calcium in coronary vasculature of smaller children, CT angiography is considered better preoperative imaging modality [3].

Regarding the choice of intervention, the smaller children with KD with aneurysmal thrombosis and calcification of coronary arteries with access problems and because of absence of growth potential, percutaneous coronary intervention (PCI) is usually not preferred [3].

Since its first description by Kitamura et al., surgical revascularization has become the mainstay treatment strategy. Pedicled ITA is a live conduit due to its growth potential with no tenting phenomena at anastomotic site (compared with autologous saphenous vein), adequacy of length (as compared with the gastro-epiploic artery [6] when used in paediatric population) and seldom involvement in KD inflammation, it has emerged as the most suited graft for revascularization [3]. But it also is a surgical challenge to harvest bilateral ITAs in young children; however, it was successfully done in our case. The coronary artery aneurysm registry which is by far the largest multicentric database has found the mortality and major adverse cardiac event rate to be 15.3% and 31% respectively without intervention [7]. Postsurgical revascularization there has been observed excellent graft patency of ITA (87%) with overall survival rate of 95% as quoted by Kitamura et al. a over 25-year period of observation in 114 patients [8].

Patch angioplasty can be done for giant CAA; however, non-treated patches may undergo aneurysmal changes and glutaraldehyde treated ones undergo calcification increasing shear stress on opposite coronary wall. Presence of thrombus also precludes use of this technique [9].

Reconstructive down-sizing has been described to ensure laminar flow and reduce the size to avoid use of anticoagulants

postoperatively, but it was complicated with RCA occlusion, narrowing at the aneurysmal site and difficulty in the presence of calcification; therefore, it has not gained popularity [3, 9]. Newer techniques of rotational atherectomy and drug-coated balloon have been reported which need further development [10]. A follow-up strategy needs to be individualized due to rarity of incidence.

Conclusion

CAA should be kept in mind as differential for angina in paediatric population. Coronary artery bypass grafting using arterial conduits should be the preferred treatment strategy for CAA. The follow-up needs to be individualized with a multidisciplinary team for both cardiac and other systemic complications.

Funding Nil.

Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest.

Ethical statement The manuscript is original and has not been submitted to any other journal for simultaneous consideration, and never been published elsewhere in any form or language.

Informed consent Informed consent has been taken from the parent(s) to publish this case report.

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