IMAGES in PAEDIATRIC CARDIOLOGY

Micallef Eynaud S, Attard Montalto S, Grech V. A case of atypical Kawasaki disease with giant coronary artery aneurysm containing thrombus. Images Paediatr Cardiol 2016;18(3):9-15.
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Abstract

Introduction

Kawasaki disease (KD) is an acute febrile, systemic vasculitic syndrome of unknown etiology, occurring primarily in children younger than 5 years of age. Administration of IVIG within the first 10 days after onset of fever in combination with high dose aspirin reduces the risk of coronary artery damage in KD. Though rare, giant aneurysms of the coronary arteries may develop in untreated cases and prove extremely challenging to manage.

Case Presentation

A 9-month-old Caucasian boy presented to our paediatric emergency department with a 4-week history of intermittent pyrexia and irritability. Typical mucocutaneous signs of Kawasaki Disease were absent upon presentation. A trans-thoracic echocardiogram identified a giant aneurysm of the left anterior descending artery with thrombus formation in-situ and the child was managed with intravenous immunoglobulin, steroids, high dose aspirin therapy and later warfarinisation.

Discussion

Cardiovascular sequelae of Kawasaki disease include giant coronary artery aneurysms with thrombosis. Enlargement of a coronary aneurysm after the acute phase of Kawasaki disease is uncommon and the outcome of interventional approaches poorly studied.

Keywords

Kawasaki, Arteritis, Coronary, Giant Aneurysm, Thrombus.

Introduction

Kawasaki disease (KD) is an acute vasculitic syndrome of unknown etiology affecting small and medium-sized arteries in infants and young children.[1] It was described in 1961 by Tomisaku Kawasaki in a 4-year-old child with a fever and rash at the Red Cross Hospital in Tokyo.[2] 80% of patients are younger than 5 years of age and boys are more susceptible than girls with a ratio of roughly 1.5:1. The disease occurs most frequently in winter and spring. Young infants have the highest rate of aneurysm development and often exhibit incomplete clinical signs and symptoms. Giant coronary artery aneurysms (with internal diameters above 8mm) are seen in 0.5% to 1% of adequately treated patients.[3]

Patient

An 8.0kg, nine-month-old son of healthy Caucasian parents, born by normal vaginal delivery at forty weeks of gestation, presented to our Paediatric Emergency Department with a four-week history of intermittent pyrexia and irritability. Initial coryzal symptoms were attributed to a possible upper respiratory tract infection, with the subsequent development of an urticarial rash two weeks later thought to be the result of a food allergy. He subsequently developed occasional bouts of vomiting, pallor and a noticeable loss in weight, prompting his eventual referral. He had received a seven-day course of oral Augmentin to no effect.

Clinical examination confirmed a markedly irritable child that was pale and febrile at 38.5°C. His oxygen saturation was 95% in air, and his pulse rate stood at 152 beats per minute, with a grade 2/6 ejection systolic murmur heard at the lower left sternal edge. He was tachypnoeic with a respiratory rate of 65 breaths per minute, displayed bilateral subcostal recessions and a 4cm tender hepatomegaly. A full sepsis screen was carried out, and the child was commenced on intravenous ceftriaxone.

A complete blood count revealed a normocytic normochromic anaemia with a haemoglobin level of 7.6g/dl, a leucocytosis of 21.8×10^9/l with a lymphocytosis of 12×10^9 and a normal platelet count. Cerebrospinal fluid was sterile albeit a mild pleocytosis, with a normal protein concentration. His blood and urine cultures were sterile. A raised C-Reactive Protein of 144 and an Erythrocyte Sedimentation Rate of 115mm/hr indicated a marked inflammatory response. His liver and renal function tests were completely normal, and he had no evidence of disseminated intravascular coagulation. Abdominal ultrasound showed a homogenous hepatomegaly with no other focal abnormalities. The Chest X-ray showed a small right pleural effusion with no cardiomegaly.

A trans-thoracic echocardiogram identified a giant aneurysm of the left anterior descending artery with thrombus formation in-situ. The left main coronary was also dilated at 5mm immediately beyond its origin and the left circumflex artery dilated in a fusiform fashion at 9mm. A 2.5cm length of the right coronary artery was found to be dilated at 4mm at its proximal and 3mm at its distal end (figures 1-5).

Figure 1: Four chamber view showing left coronary artery dilatation with measurements.

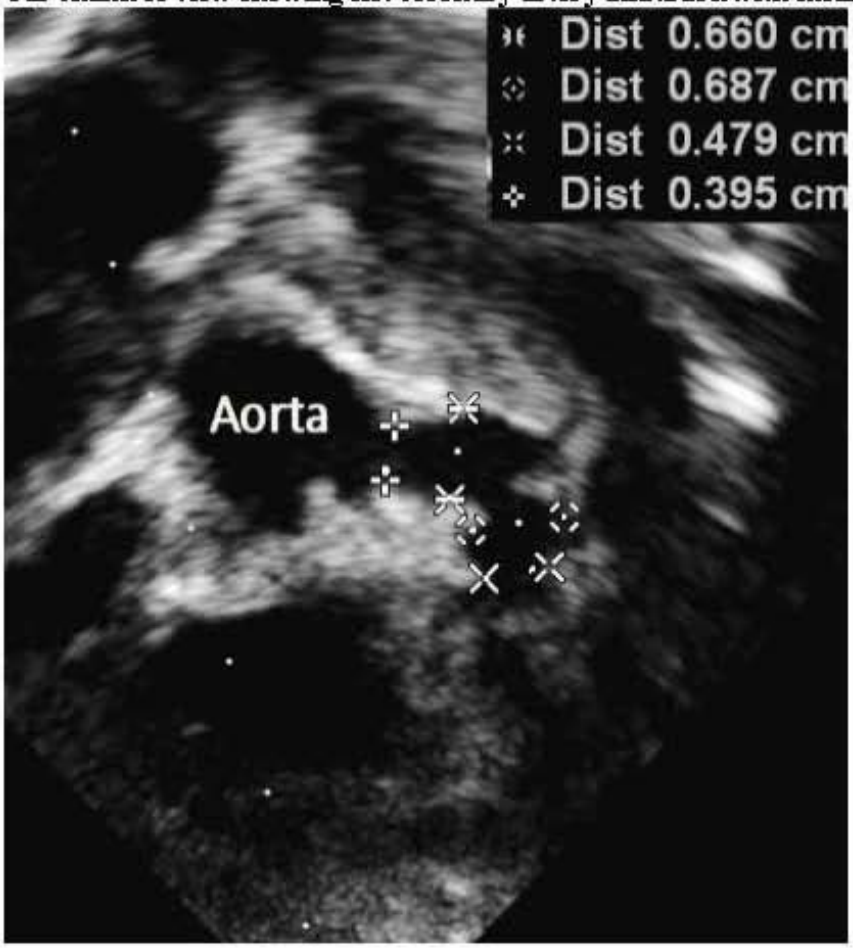


Figure 2: Parasternal short axis view showing a left coronary artery aneurysm, with arterial Doppler flow and with a clot (arrow).

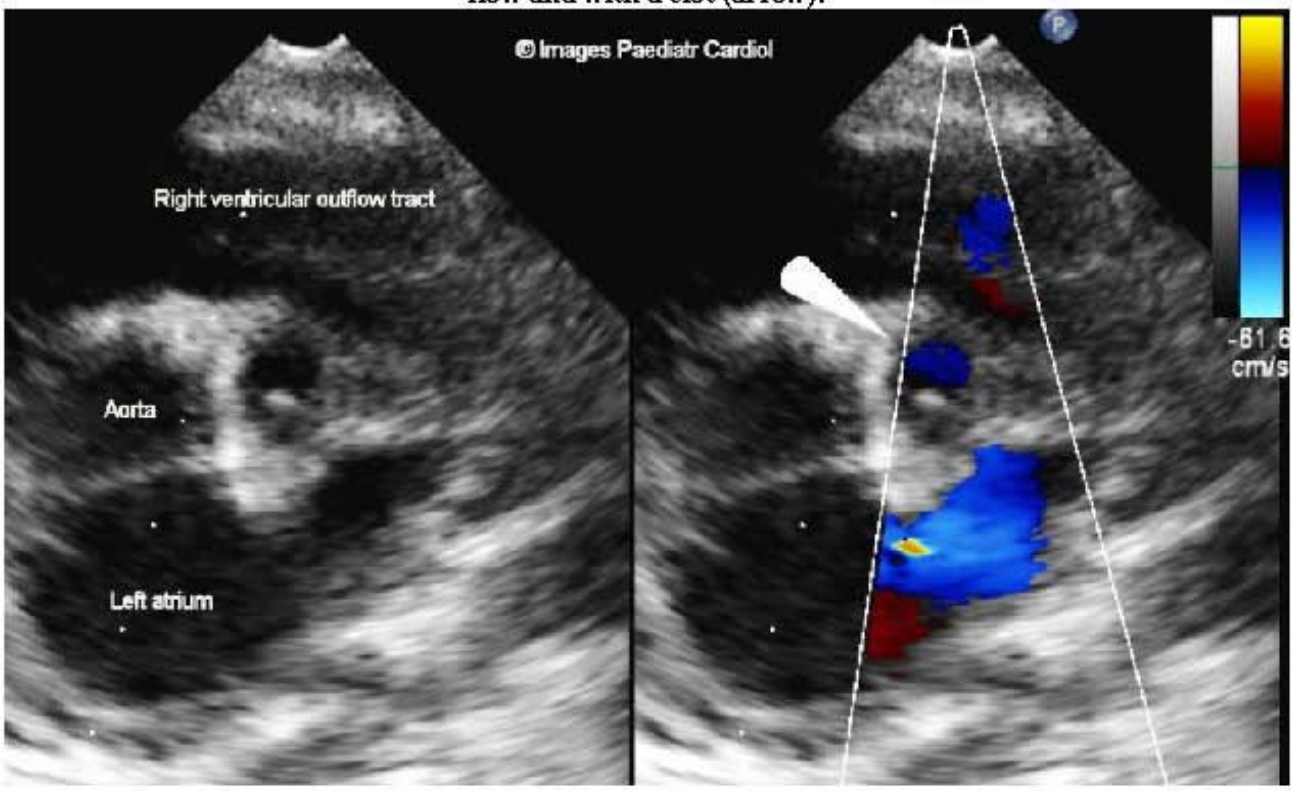


Figure 3: As figure 2 showing a left coronary artery aneurysm, with arterial Doppler flow and with a clot (arrow).

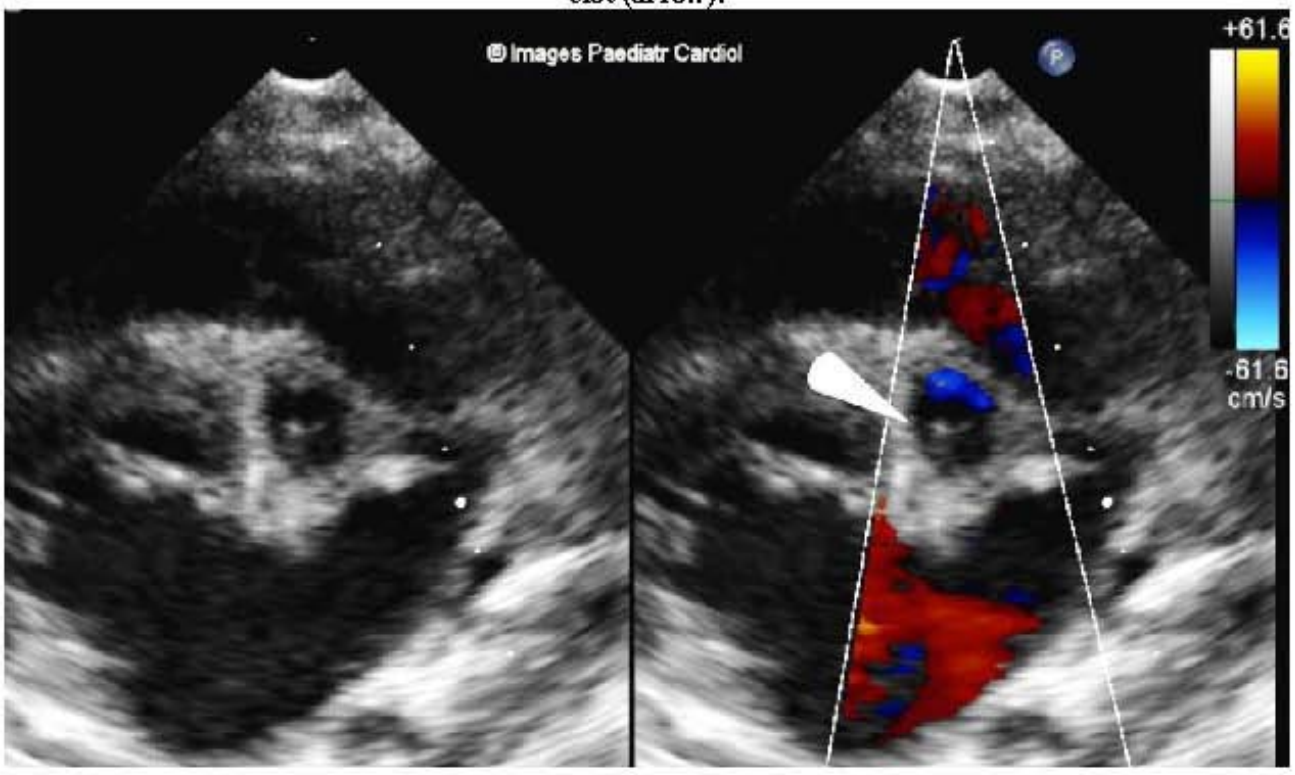


Figure 4: Dilated right coronary artery with measurement.

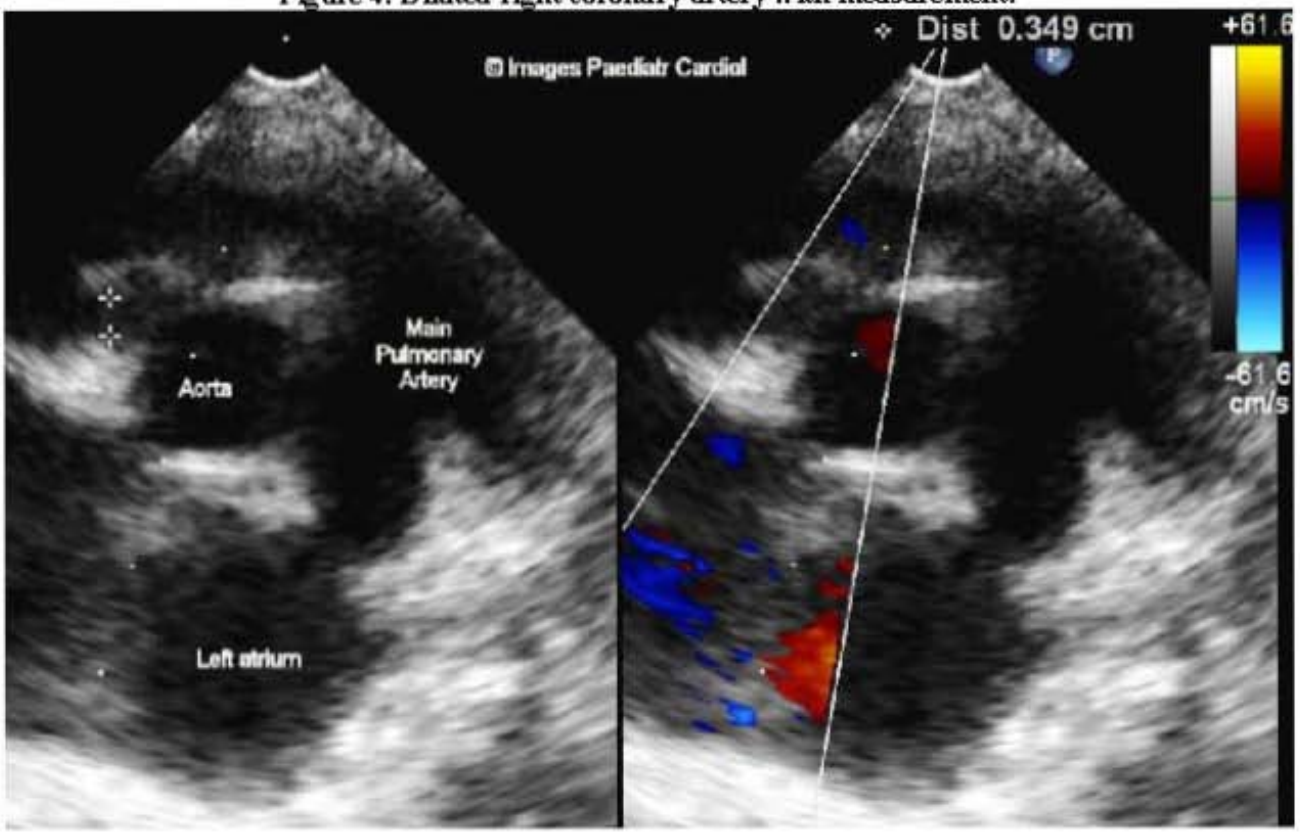
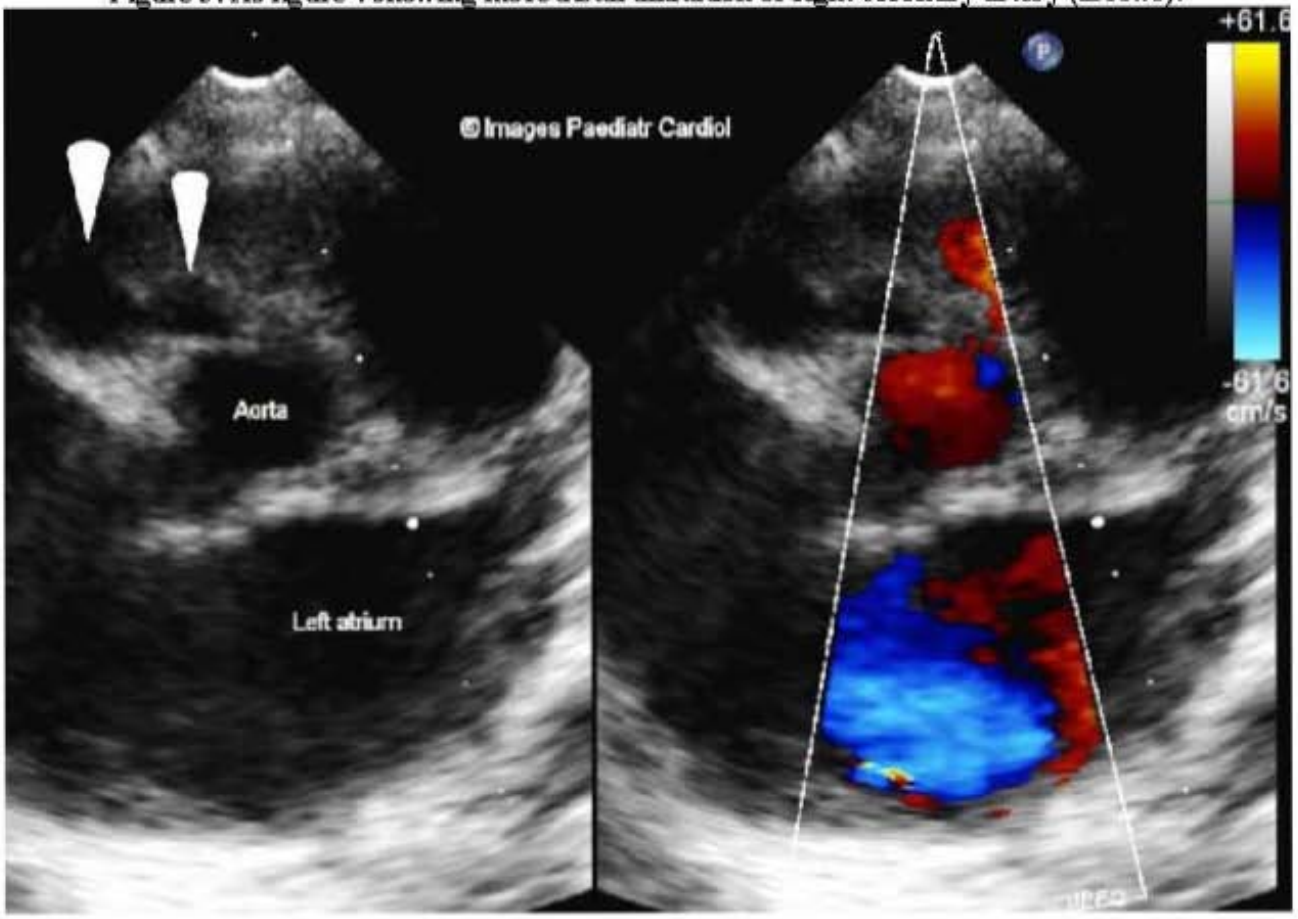


Figure 5: As figure 4 showing more distal dilatation of right coronary artery (arrows).



He was otherwise determined to be situs solitus, have levocardia and concordant atrioventricular connections. The inferior vena cava and superior vena cava both drained into the right atrium, and all four pulmonary veins drained into the left atrium. He had a left-sided aortic arch with no coarctation or patency of the ductus arteriosus. In addition, a small pericardial effusion, 6mm at its widest diameter, moderate mitral regurgitation with some dilatation of the left atrium, mild pulmonary and tricuspid regurgitation, and an incidental patent foramen ovale were identified.

With a preliminary diagnosis of atypical Kawasaki disease, the patient was started on 2g/kg of intravenous immunoglobulin (IVIG) and high dose aspirin therapy. A supporting transfusion of crossmatched packed red cells was given due to his worsening anaemia. With a Hb of 6.5g/dl just prior to transfusion, he was transferred to our Neonatal Intensive Care Unit for stringent monitoring. He remained febrile despite being treated with IVIG, ultimately responding to methylprednisolone. A magnetic resonance angiographic study excluded concomitant aneurysms in the Circle of Willis or renal vasculature. He was started on regular diuretics, aspirin, and heparinisation, before being weaned onto warfarin, with his clinical condition resolving steadily. After a total hospital stay of eighteen days, he was deemed fit to be discharged home. His treatment regimen consisted of Prednisolone 12mg daily, Aspirin 80mg every 6 hours, Furosemide 8mg twice a day, Spironolactone 8mg twice a day and Warfarin 1mg daily, adjusted for his INR.

The patient was monitored clinically and by repeated trans-thoracic echocardiography in an outpatient setting. He continued to show normal development and at the age of two his developmental milestones were satisfactory and his weight was 12.0 kg. His serial echocardiograms were far less reassuring. His most recent scan at two years of age showed an increase in the size of the aneurysm at the junction of the left main and left anterior descending coronary arteries to 11mm by 30mm. The Montreal Z-Score was 28.1. The left main coronary artery was dilated at 4mm with a Z-Score of 5.6. The right main coronary artery was 2.5mm in diameter, Z-score 2.1, and a proximal aneurysm measuring 6.5mm, Z-score 13.6mm. A good systolic function was demonstrated, as well as a decent pulsatility of the abdominal aorta with no diastolic tail or reverse flow. The interventricular septum was intact. The coronary dilatation is confirmed on high resolution CT scan (figures 6 and 7).

Figure 6: Anterior view of a CT-reconstructed coronary angiogram, showing aneurysms.

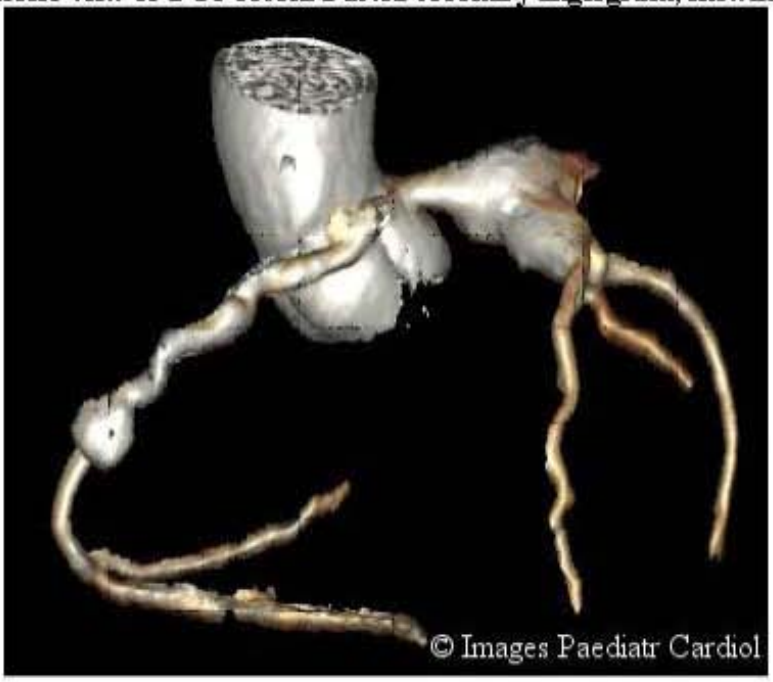
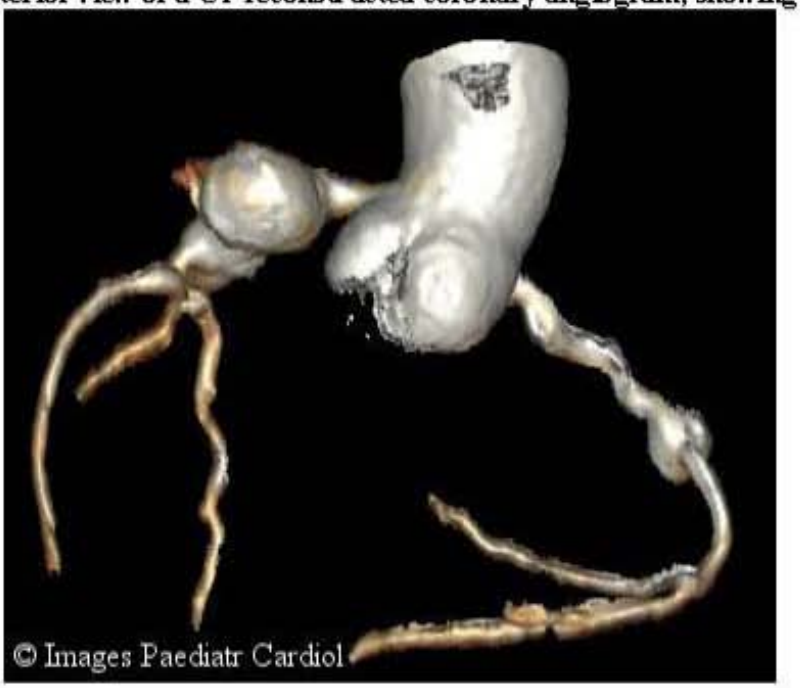


Figure 7: Posterior view of a CT-reconstructed coronary angiogram, showing aneurysms.



Discussion

Cardiovascular complications of Kawasaki disease include silent coronary artery ectasia or aneurysm development, giant coronary artery aneurysms with thrombus formation, myocardial infarction, and sudden death. The incidence is 15-25% in untreated children,[4] dropping down to 5% if patients receive intravenous immunoglobulin (IVIg) within 10 days of disease onset.[5] In a study by Kato et al. on the long-term effects of cardiovascular sequelae in Kawasaki disease the incidence of coronary aneurysm formation in acute cases was 25%, half of which showed regression. Ischemic heart disease developed in just under 5% and myocardial infarction in 2% of these patients during the follow-up period. Death occurred in 0.8%.[6] Coronary artery dilatation can be appreciated as early as 4 days after the first appearance of fever. Aneurysm formation peaks at approximately 4 weeks after the start of illness[6] that is roughly the time interval taken between symptom onset and presentation in our patient. The administration of steroids improves symptomatology and absence of progressive coronary artery abnormalities in those who fail to respond to IVIg.[6] The development of myocarditis, pericarditis, CHF, mitral or aortic insufficiency, and arrhythmias tends to occur during the early phase of the disease.

Aneurysms occur during the early stage of Kawasaki disease and are known to involve the most proximal segments of the large coronary vessels. The commonest sites of aneurysm formation include the proximal Left Anterior Descending, proximal Right Coronary Artery, followed by the Left Main Coronary, Left Circumflex, and the distal Right Coronary Artery. The aneurysms are classified as small (<5-mm internal diameter), medium (5-8 mm inner diameter), or giant (>8-mm inner diameter).[7] Approximately 50% of these lesions resolve within 5 years. In the majority of those with small coronary artery aneurysms (3-4 mm), regression occurs within 2 years.[8] The risk of an aneurysm developing increases in patients with a persistent fever of over two weeks, those who have recurrent pyrexia after an afebrile period of at least 48 hours, and those younger than 1 year. Thrombocytopenia, a low haematocrit and elevated neutrophil/band counts are individual predictive markers for aneurysm development.[9]

All patients with Kawasaki disease should undergo an echocardiogram as a first line investigation and six to eight weeks after making the diagnosis. A stress test and coronary angiography may be employed to identify stenotic lesions.[10] Our patient was too young to perform a stress test, and we proceeded with coronary angiography due to the increasing size of the aneurysm as was evident on serial echocardiograms. The resolution of coronary artery aneurysms depends upon their initial size, with those under 5 mm more likely to resolve than those over 8 mm in diameter. The age of the patient at the onset of disease is another important factor. Aneurysms have a greater chance of resolving in those under one year age. Those with a fusiform morphology, as opposed to saccular, are also more likely to resolve.[11]

Warfarinisation is recommended for those with giant aneurysms. Percutaneous coronary intervention with placement of a covered stent is often not feasible due to the small caliber of these vessels and the potential for the arteries to grow. Attempts at excision or plication of an aneurysm are rarely successful and carry a high risk of mortality. The arterial graft patency rate in adult life is still unknown. Management of such patients remains extremely challenging due to the rarity of giant aneurysms and the very limited data on the surgical and endovascular interventions available.

Conclusion

Our case report highlights the paradigm of early treatment and a low index of suspicion for atypical Kawasaki Disease to prevent its potentially fatal cardiovascular sequelae.

Acknowledgements

Christian Axiak, RIS PACS Manager, Mater Dei Hospital, Malta.

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