A Young Patient With History of Kawasaki Disease Presenting with Triple Vessel Disease

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SUMMARY

Kawasaki disease is primarily a condition that affects young children and it is associated with cardiac morbidity and mortality. This disease has been known to cause coronary artery aneurysms which occurs as a sequelae of vasculitis. The progression of triple vessel disease in adult which results from cardiac complications from Kawasaki disease is rare. We report a case of a young man with history of Kawasaki disease at infancy presenting with triple vessel disease requiring cardiac bypass surgery at the age of 20 years old.

KEY WORDS:
Kawasaki disease; coronary artery bypass surgery; cardiac complications; coronary artery aneursyms

INTRODUCTION

Kawasaki disease was first discovered by Tomisaku Kawasaki in January 1961 in Japan[1]. Although the cause remains unknown, this disease has became the leading cause of acquired heart disease among children. This condition is diagnosed using clinical criteria which incudes fever for five days or longer and at least four of the following: (1) non-exudative conjunctival injection; (2) oral involvement including strawberry tougue, mucosal hyperemia, and cracked or erythematous lips; (3) changes in the peripheral extremities, including edema or desquamation; (4) polymorphous rash and (5) cervical adenopathy1 . Cardiac sequelae contributes to majority of the morbidity and mortality related to Kawasaki disease[2] . Most of the pathology of this disease is induced by medium vessel arterial vasculitis which can lead to the development of coronary aneurysms. The natural history of coronary aneurysms in Kawasaki disease varies from complete resolution to fatal myocardial infarction[2]. The standard treatment of Kawasaki disease involves acetylsalicylic acid and intravenous immunoglobulin which have been shown to reduce the incidence of coronary complications remarkably.

CASE HISTORY

This patient is a 20 year old man who was initially diagnosed with Kawasaki disease at four months of age. He presented to the district hospital in Kuching, Sarawak with history of prolonged fever for three weeks which was not relieved with antipyretics and antibiotics. His other symptom was generalised body flushes. He was transferred to a tertiary hospital in Kuching for second opinion when his condition did not improve despite treatment and the diagnosis was inconclusive. He was nursed in intensive care unit for a week and was investigated further. He was finally diagnosed to have Kawasaki disease when he was found to have severe coronary aneurysms. His parents were counseled that his mortality risk was high. His condition improved remarkably after he was given intravenous immunoglobulin. He was discharged well after about a month's stay in the hospital and he underwent regular follow up under the care of the paediatric cardiology team.

He remained asymptomatic until the age of 16 years old when he had sudden onset of shortness of breath during sports. An electrocardiogram (ECG) was done and it showed ST depression at lead 1 and AVF and slight ST elevation on lead III. However, cardiac enzymes were not raised. Angiogram was done and it showed an aneurysm of left main coronary artery, proximal left circumflex artery (LCx) and proximal right coronary artery (RCA). There was stricture of the left main coronary artery and total occlusion of the RCA. A repeat angiogram was done in two years later. It showed collapsed of two coronary arteries with one natural bypass of the artery. No active surgical intervention was planned but patient was followed up regularly to assess symptoms and for continuation of medications. His serial echocardiogram (ECHO) showed good ejection fraction in the range of 68-75%.

At 20 years old, his condition worsened and he was then symptomatic with Canadian Cardiovascular Functional Classification of Angina (CCS) II-III with reduced effort tolerance. He also had on and off palpitations at rest. A repeat angiogram was done and he was found to have triple vessel disease with 100% occlusion of proximal RCA, 100% occlusion of ostial left anterior descending (LAD) and 50% occlusion of proximal LCx. There was also coronary artery aneurysm of the proximal RCA and proximal LCx. He was referred to cardiothoracic unit for coronary artery bypass graft (CABG) . His pre-operative ECHO showed good ejection fraction of 65% and his valves were of normal morphology. Patient was electively admitted for CABG on 8th December 2010. On admission, his vital signs were stable with blood pressure of 124/66 and heart rate of 59 beats per minute. His weight was 109.5kg and height was 178cm. His calculated body mass index (BMI) was 34.6 kg/m2. He subsequently underwent CABG on 10th December 2010. Intra-operative findings were cardiomegaly and scarring of inferior right

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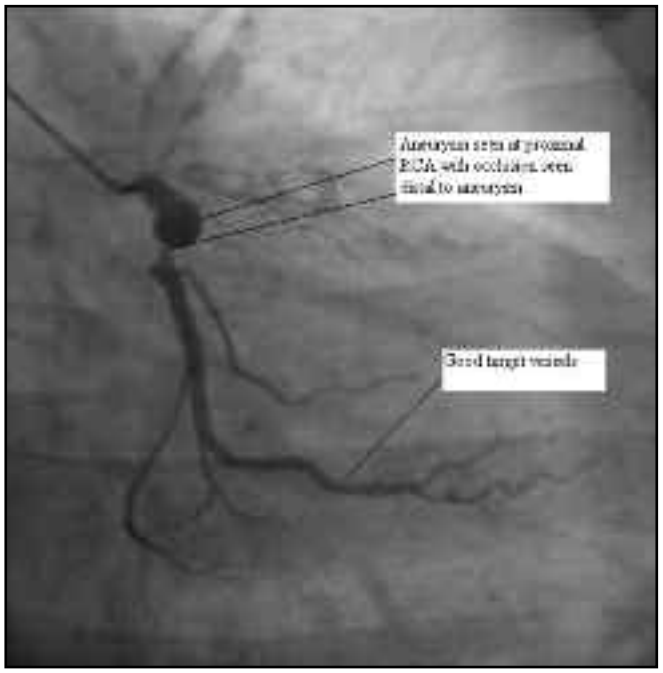

Fig. 1: Angiogram findings of patient with Kawasaki disease at age 20 years.

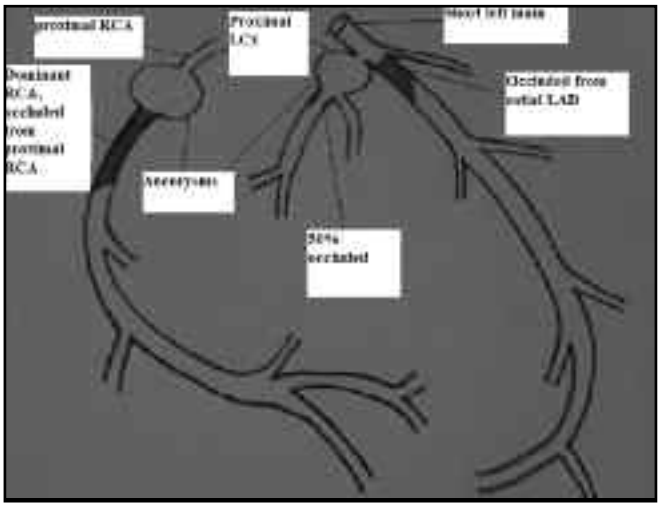

Fig. 2: Diagramatic presentation of his angiogram findings:

ventricular wall. Patient was planned for grafts from both internal mammary artery (IMA), however the right IMA (RIMA) was not harvested as the length was insufficient and radial artery was not taken in view of negative Allen's test of bilateral radial arteries. Three grafts were inserted: Left internal mammary artery (LIMA) to LAD, saphenous grafts for obtuse marginal brannch (OM1) and RCA-posterior descending artery (PDA). Post bypass surgery, this patient is managed with early education of cardiac rehabilitation and further reinforcement of compliance medications and breathing exercises. He recovered well and was discharged at day 12 post operatively. He is now back pursuing further studies in a private college.

DISCUSSION

With early diagnosis and prompt treatment with intravenous immunoglobulin and aspirin, only about 3-5% of children eventually develop cardiac complications as a sequelae of Kawasaki disease[1]. The risk factors for coronary artery involvement include an age of less than 6 months or greater than 7 years, treatment with intravenous immunoglobulin after eight days of illness, recurrent disease and male sex[3]. The rates of disease are highest in children under one year old and boys are more commonly affected with a ration of 1.4:1[2]. It is also documented that children less than 6 months old were observed to have a higher incidence of developing giant coronary aneurysms and the preventive therapy with immunoglobulin is low if it is administered at day 8 of illness or later[3]. The major setbacks for the development of coronary complications in this patient seems to be the late detection of Kawasaki disease which most likely attributed to the atypical presentation of only prolonged fever and generalised body rashes in this patient. He did not have other symptoms like cervical lymphadenopathy or changes in peripheral extremities which would greatly aid the diagnosis of Kawasaki disease. Therefore, the commencement of aspirin and immunoglobulin was delayed, in this case is after three weeks of fever. This increases the risk of development of cardiac complications.

The pathology of coronary aneurysms can develop as early as within 10 days which involves periarteritis and endarteritis followed by a panvasculitis and perivasculitis with formation of aneurysm or stenosis due to intimal proliferative inflammation[2] . This patient is fortunate to have survived the acute phase of Kawasaki disease. It was documented that the rates of the disease and mortality is very high in patients who develop Kawasaki disease below 1 year old[2] . The prognosis is even poorer should these patients develop myocardial infarction. The mortality rate in children with myocardial infarction induced by Kawasaki disease is about 22% after the first infarct, 66% after the second infarct and 87% after the third infarct[5]. The best way to prevent the development of cardiac sequelae is still early introduction of gamma-globulin therapy[3].

Should these patients survive the acute phase of Kawasaki disease, they should be monitored closely and parents should be counseled about the long term care and outcome of the patients. The importance of compliance to medications and follow up should be stressed and parents should be informed about the possible symptoms that their child might suffer from should any cardiac events set in. By any chance should their child feels unwell, immediate medical consultation should be sought.

In the long run, patients with a history of Kawasaki disease who develop cardiac sequelae can progress from complete resolution of the cardiac complications to development of myocardial infarction which could be fatal. For patients who develop coronary complications during the acute phase of the disease like this patient, the development of coronary artery

stenosis is about 20%[1]. Most of these children will need coronary artery bypass surgery. The long term outcomes of myocardial revascularisation was studied over a period of nearly 12 years in Japan and it showed that internal thoracic(mammary) artery grafts has significantly better long term patency than saphenous vein grafts in patients with Kawasaki disease and the use of internal thoracic artery grafts reduced the incidence of late cardiac deaths [5]. There is significantly higher rates of survival ($p<0.05$) in the internal thoracic artery group at 90 months post surgery [5]. This patient was planned for internal mammary harvest bilaterally. However in view of insufficient length of the RIMA, only left IMA was used. Radial arteries were not harvested in view of negative Allen's test.

After bypass surgery, these patient should be followed up at regular intervals life long to ensure that other comorbidities are optimised and healthy lifestyle enforced. The outcome of those who survived bypass surgery is generally good should other risk factors controlled and proper education and awareness informed to patient.

By and large, early diagnosis and prompt treatment of Kawasaki disease is vital to prevent the onset and progression of cardiac complications. Detailed investigations is needed once diagnosis is made and patients should be monitored and followed up regularly [1]. Patients and parents should also be counselled early about the cardiac complications of Kawasaki disease and be advised to seek medical consultations should patient develop any symptoms. Further investigations and studies are required to identify a possible treatable underlying cause and define the management of coronary complications in both children and adults in the long term.

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