Super Giant Coronary Aneurysm in Kawasaki Disease

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Abstract

Background: Giant coronary artery aneurysms caused by Kawasaki disease are rare; however, they are one of the most serious complications and can be lethal.

Case Presentation: We report a 3.5-month-old boy referred to us because of high fever for fifteen days, generalized maculopapular rash, irritability and cough. Transthoracic echocardiography showed dilatation of right coronary (RCA) and left main coronary (LCA) arteries. Serial echocardiography revealed rapidly progressive dilatation of coronary artery aneurysms of RCA and LCA. We performed invasive cardiac catheterization with selective coronary angiography when the boy was 16 months old. Selective right and left coronary arteriography showed a super giant fusiform aneurysm of RCA and a diffuse giant aneurysm of the proximal LCA. Regression of coronary artery aneurysms was not observed during 6 years of follow up.

Conclusion: Pediatricians should be alert for possibility of incomplete Kawasaki disease in young infants with atypical presentation. They are at higher risk of coronary aneurysm formation. The diagnosis often was late with higher complication rate of coronary aneurysm. Echocardiography is an important tool for diagnosis of incomplete Kawasaki disease. Selective coronary angiography is the gold standard for diagnosis, and estimation of shape and size of aneurysms.

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Key Words: Incomplete Kawasaki; Kawasaki disease; Coronary aneurysm; Giant coronary-artery aneurysm; Young infant

Introduction

Kawasaki disease is an acute vasculitis syndrome of unknown etiology occurring in infants and young children and affecting mainly small and medium-sized arteries, particularly the coronary arteries [1,2,3]. It is the most common cause of acquired heart disease...
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in children in many parts of the world [4-7]. Patients who lack sufficient clinical signs to fulfill the classic criteria have been diagnosed as having atypical or incomplete Kawasaki disease [5,8]. Incomplete Kawasaki disease is more common in children younger than one year, who are unfortunately at greatest risk of developing coronary artery aneurysm [5,8,9,10].

Coronary artery dilatation or aneurysm occurs in 15% to 25% untreated children [5,8,11,12,13]. Large coronary aneurysms can be termed giant (internal diameter of 8 or 9 mm) and super giant (internal diameter greater than 10 mm) [14]. Large coronary artery aneurysms are seen in 0.5% to 1% of adequately treated patients [15]. We report a 3.5-month-old male infant with atypical or incomplete Kawasaki disease and super giant coronary artery aneurysm.

**Case Presentation**

A 3.5-month-old boy was referred to our hospital with 15-day duration of fever. In the first day of his illness, he had high fever (40.5°C), generalized maculopapular rash, irritability and cough. He was treated with several antibiotics by his pediatrician and in another hospital.

Physical examination on day 15 showed body temperature of 38.8°C, irritability, and desquamation of finger, toes and perianal area. In laboratory findings, there was marked leucocytosis. Leukocyte count rose from 17.66x10^9/L to maximum 44.2x10^9/L on day 15 of fever. The platelet count was normal during the initial early phase of illness. Initial platelet count was 380x10^9/L on day 5 and rose on day 9 from 459x10^9/L to maximum of 1080x10^9/L on day 15 in the course of illness. Erythrocyte sedimentation rate (ESR) was 78 mm/hr on day 5 and rose to 135 mm/hr on day 15 of fever and then it declined gradually. CRP was negative and hemoglobin 8.1 g/dl on day 15 of illness.

Sepsis workup including blood and urine cultures was negative. Widal and Wright tests were also negative. Chest X-ray in anteroposterior view showed perihilar fine reticulonudular infiltration suggestive of viral pneumonia. Echocardiography on day 15 of illness was recommended by pediatrician. Echocardiography showed diffuse dilatation of the right coronary (RCA) and proximal dilatation of the left main coronary arteries (LCA). RCA measured 9.5 mm at origin while LCA measured 6.5 mm at origin. No impairment of the left ventricular function or pericardial effusion was found. The diagnosis was incomplete Kawasaki disease complicated by coronary artery aneurysm. Therapy was initiated with single dose intravenous gamma globulin (2 gm/kg) injections and high-dose aspirin (100 mg/kg/day) for two weeks and was subsequently reduced to 5 mg/kg/day on day 30 of disease. Fever gradually subsided on day 17.

Follow up echocardiography, 6 weeks later, revealed a super giant fusiform aneurysm of RCA and a giant aneurysm of the proximal left main coronary artery. (RCA 12.5mm, LCA 9mm). Subsequent echocardiogram after 6 months showed progressively enlarging aneurysm of both coronary arteries. RCA measured 18 mm at origin while LCA measured 10 mm at origin. Initial and repeated surface electrocardio-gram was normal. Cardiac enzymes remained normal. We started ASA (5 mg/kg/day) and warfarin, the dose was adjusted to maintain international normalized ratio (INR) on about 2.5. Despite standard doses of aspirin and gamma globulin during hospitalization, and maintenance doses of aspirin and warfarin in outpatient clinic, the size of coronary aneurysm increased on echocardiogram during a follow up of 13 months.

After obtaining informed consent from parents we performed invasive cardiac catheterization with selective coronary angiography when the boy was 16 months old. Selective right coronary arteriography showed a super giant fusiform, and diffuse aneurysm measuring 19 mm at proximal RCA (Fig 1-A).
Giant fusiform aneurysm of left main coronary artery measuring 9 mm was confirmed by selective left coronary arteriography (Fig 1-B). There was no thrombus detected inside the coronary artery aneurysm. The coronary aneurysm involution by repeated echocardiography was not observed. The stress test was performed when the boy was 6 years old. The stress test was normal.

Discussion

Incomplete Kawasaki disease refers to patients who do not fulfill the classic criteria of at least four of the five findings. Incomplete Kawasaki disease is more common in children younger than one year, in whom the rate of coronary artery aneurysm is paradoxically higher if not treated; therefore, establishing the diagnosis and initiating treatment are essential. The diagnosis of Kawasaki disease can be difficult because many features mimic common childhood illnesses (eg, adenovirus, scarlet fever) and drug reactions. Therefore, physicians need to keep Kawasaki disease in their differential for children who have prolonged fever without clear etiology, because the consequences of missed diagnosis can be serious morbidity or, in rare cases, death [3]. Giant coronary artery aneurysms are seen in 0.5-1% of adequately treated children with Kawasaki disease [16].

Cardiovascular sequelae of Kawasaki disease include asymptomatic coronary artery ectasias or aneurysm formation, giant coronary artery aneurysms with thrombosis, myocardial infarction, and sudden death. They have been reported in 15-25% of untreated children [5,8,11,12,13,17]. With treatment using IVIg within first 10 days of the disease, the incidence drops down to about 5% [17].

Coronary artery dilatation can be seen as early as 4 days after the first appearance of fever. It peaks at approximately 4 weeks after the onset of illness [17]. In our patient, the coronary artery involvement was detected about 15 days after the onset of symptoms.

The most common sites of aneurysms in the order of frequency include the proximal LAD, proximal RCA, followed by the LMCA, LCX, and finally the distal RCA [17]. The sites of aneurysms in our patient were right coronary and left main coronary arteries.
The risk of aneurysms is increased in patients with fever lasting for more than 16 days, male sex, age less than one year, hemoglobin less than 10 g/dl, ESR higher than 100 mm/h, platelets equal or higher than 900×10^9/L as was seen in our patient and thrombocytopenia[17,18]. Among children with coronary lesions, 56% presented an incomplete form of the disease[19].

A previous study demonstrated 85% of Kawasaki infants younger than 6 months of age developed coronary artery aneurysms[18]. The diagnosis was often delayed as pediatricians are less likely to consider Kawasaki disease with atypical presentation.

This case and other investigations illustrate the fact that atypical Kawasaki disease is often a late consideration, especially when the symptoms of the classical form are absent. This condition should be considered in every infant presenting with long-lasting unexplained fever with associated features, risk factors of coronary artery involvement and inflammatory tests. Because young infants with Kawasaki disease are at an extremely high risk of developing coronary arterial abnormalities, early diagnosis and appropriate therapy are particularly important.

Hence pediatricians should be alert for the possibility of incomplete Kawasaki disease in young infants with fewer clinical findings. Therefore young infants of six months or younger with fever for at least seven days and no clear etiology should have a laboratory assessment even if no features of Kawasaki disease are present; echocardiography should be performed if evidence of inflammation is found. All patients with Kawasaki disease should undergo echocardiography on diagnosis and 6-8 weeks after the onset of the disease. Those with giant aneurysms may require a stress test and possibly coronary angiography to identify stenotic lesions[20].

The addition of warfarin to aspirin therapy has been recommended for those with giant aneurysms as was done in our patient. Coronary artery bypass grafts using the internal thoracic artery has been performed with some success on small number of cases [6,8,21]. Attempts at excision or plication of the aneurysm have not been successful and have even caused death. The arterial graft patency rate in later adult life is still unknown[17]. Further studies and long term follow up are required to evaluate the long-term outcomes of bypass graft in patients with giant aneurysms secondary to Kawasaki disease[6].

Percutaneous coronary intervention with placement of a covered stent was not feasible in our patient in view of small vessel size and potential for the artery to grow. In view of these limitations and in the absence of any obvious evidence of myocardial ischemia and to avoid thrombus formation in the giant aneurysm of the right and left coronary artery we decided to treat the patient with long term aspirin together with warfarin. The outcome following 6 years of follow up has been very surprising and favorable to date. Elective follow up catheterization is planned in the ensuing years.

**Conclusion**

Pediatricians should be alert for the possibility of incomplete Kawasaki disease in young infants with atypical presentation. They are at higher risk of coronary aneurysm formation. The diagnosis often was late with higher complication rate of coronary aneurysm.

Pediatric clinician should have a high index of suspicion in evaluating young infants presenting with fever of unknown origin with abnormal inflammatory laboratory results without infection. Serial echocardiography is an important non-invasive tool for diagnosis and following up of incomplete Kawasaki disease. Selective coronary angiography is the gold standard for diagnosis, and estimating shape and size of aneurysms.

**References**


