

Multiple Giant Succular and Fusiform Right and Left Coronary Artery Aneurysms after Early and Adequate Treatment of Atypical Kawasaki Disease with Unusual Presentation

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Abstract- The major complication of Kawasaki disease is coronary artery dilatation and aneurysm. It occurs in approximately 15-25% of untreated children with Kawasaki Disease. Early diagnosis and treatment with Intravenous immune globulin (IVIG) and aspirin (ASA) can reduce the incidence of coronary artery abnormality to 2%-5%. We report one case of Atypical Kawasaki Disease with Multiple giant coronary artery aneurysms despite early adequate treatment with IVIG and ASA.

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Introduction

Kawasaki disease is an acute systemic and diffuse vasculitis of unknown etiology that occurs in infant and children (1). It is one of the most acquired cardiac diseases in children (2). Although Kawasaki disease (KD) is a self-limited condition, however the disease may be complicated by the development of coronary artery dilatations or aneurysms. In untreated patients, the incidence of coronary artery aneurysms is about 25 percent, and it may lead to myocardial infarction, sudden death and ischemic heart disease (3-8).

Giant coronary artery aneurysms are seen in 0.5%-1% of adequately treated patients with KD.

Here, we describe a 10 year-old girl with incomplete Kawasaki disease who had an atypical presentation.

Case Report

A 10-year old girl was referred to hospital because of high fever for seven days, malaise bilateral hips, ankles and knees arthralgia bilateral wrist arthritis, and neck pain. She was treated with multiple oral and intravenous antibiotics by her pediatrician and in another hospital.

Physical examination showed body temperature of 40°C, bilateral wrist arthritis, and bilateral arthralgia of hips, knees and ankles without sings of Arthritis, neck

pain without inflammation or adenopathy and malaise without muscle tenderness.

In Laboratory findings, there were marked leukocytosis (20200/ml), neutrophilia (86%), anemia (Hb11.5/dl), Normal thrombocyte count (394×1000ul), elevated erythrocyte sedimentation rate (ESR= 95), elevated CRP (++) , low titer ASO (166 Todd), negative ANA (0.25), Wright, Widal and RF Latex Test. Chest X-ray in anteroposteriorly was normal. Echocardiography on day 8 fever showed diffuse dilatation of the proximal right and left main coronary arteries.

The diagnosis of Atypical Kawasaki was suspected and then early treatment was initiated with single dose Intravenous Immunoglobulin (2g/Kg) and high dose aspirin (75 mg/ Kg/ day).

The high dose Aspirin continued for one week and was subsequently reduced to 100 mg (3 mg/Kg). Fever subsided 48 hours after beginning of treatment. Follow up echocardiography, two week later revealed giant aneurysms of the proximal right and left main coronary arteries. Serial echocardiography at 6 WK, three mo, six mo and 12 mo showed no regression of coronary aneurysm; therefore, coronary angiography was planned. After obtaining informed written

Consent from parents, selective coronary angiography was performed.

Selective coronary angiography showed giant

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fusiform aneurysms of right coronary artery and diffuse fusiform and saccular of left coronary arteries. (Figures 1, 2) To avoid thrombus formation in the giant coronary artery aneurysm, we decided to treat the patient with long term, Aspirin, and warfarin.

The patient refused to give warfarin due periodic blood sampling for prothrombin time and International normalized ratio (INR).

Therefore, she put on aspirin, dipyridamole and Plavix®.

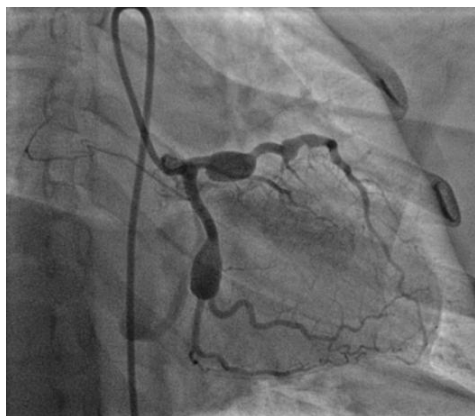


Figure 1. Left coronary angiogram in right anterior oblique view with cranial angulation showing multiple saccular aneurysm

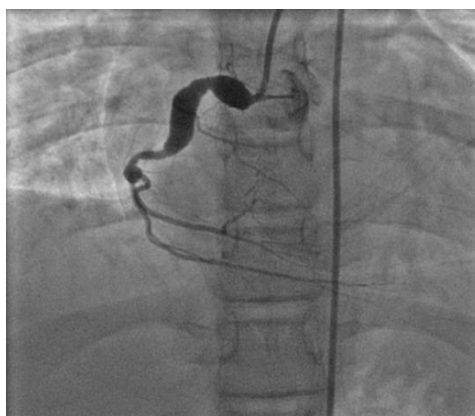


Figure 2. Right coronary angiogram in left anterior oblique view showing giant fusiform coronary aneurysm

Discussion

Kawasaki disease is rare in adolescent age group. Adolescent with Kawasaki disease accounts for about 1% of all the cases (9).

The term Atypical KD describes the condition of children who lack sufficiently clinical signs of fulfill the classic criteria, but have convincing laboratory finding (5).

The diagnosis of KD in older children is often difficult and delayed because of unusual presentation (10).

Cardiovascular complication of KD include coronary artery dilatation, aneurysm formation, giant coronary artery aneurysms, myocarditis, myocardial infarction, sudden death, congestive heart failure, pericarditis with pericardial effusion, Mitral regurgitation, Aortic

Insufficiency and Arrhythmia during the early course of the disease (11). Coronary artery ectasis can be detected as early as four days after the onset of fever (12). In our patient, coronary artery dilatation was seen about eight days after the appearance of symptoms.

Our patient represents atypical case by age of presentation. The diagnosis was often delayed as clinicians are less likely to consider KD at extremes of age with atypical presentation (13,14). Incomplete KD seems to be associated with an increased risk of coronary artery aneurysms (14,15).

In adolescents, rarity of this disease, which frequently presents in the incomplete form with lack of clinical signs, mean that, diagnosis is usually delayed. It is recommended that treatment be started before the disease has prolonged for 10 days. Our case is of interest because of the inadequate diagnostic clinical criteria as well as the unfavorable outcome.

Ghelani *et al.*, (16) reported after publication of the 2004 American Heart Association Guideline, diagnosis of incomplete KD and Laboratory use increased. Potential reason for increased diagnosis of incomplete KD is the use of echocardiography and Z-scores to determine coronary artery dilation and also increased use of laboratory value as supplemental criteria (16).

In a study of Song *et al.*, (17) both patient age groups younger than 1 year and older than 5 years had higher incidences of KD-associated coronary artery aneurysms.

Giant coronary artery aneurysms were reported, despite early diagnosis and adequate treatment with IVIg and Aspirin (11,18-20).

In our case despite early diagnosis and adequate treatment with IVIg, ASA, multiple rights and left coronary artery aneurysms developed as soon follow-up echocardiography two weeks later.

Kawasaki disease presenting with prolonged fever and arthralgia is rare. Adolescent with Kawasaki disease may have an unusual presentation and inadequate criteria for diagnosis of incomplete Kawasaki disease. Clinician should have high levels of suspicion in evaluating patients presenting with unexplained fever, with abnormal acute phase reactants (ESR, CRP). There is a higher risk of coronary giant aneurysm formation in adolescent age group. Despite immediate diagnosis and adequate treatment with IVIg and Aspirin, Multiple giant coronary artery aneurysms may be developing.

Selective coronary angiography is the gold standard for diagnosis, severity, shape and estimating size of Aneurysms.

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