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Recurrence of coronary artery lesions after complete regression in a peculiar case of Kawasaki disease

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Brief Report

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Abstract

Kawasaki disease is a leading cause of acquired heart disease in children with serious repercussions of coronary artery lesions. Recurrences of the disease are relatively rare in clinical practice. We present a case of recurrent Kawasaki disease, wherein the coronary artery lesions which were documented during the initial illness demonstrated complete regression over the following months, but reappeared with recurrence of the disease.

Kawasaki disease or mucocutaneous lymph node syndrome is a leading cause of acquired heart disease in children, having cardiac sequelae of coronary artery lesions, the incidence of which has been reported to be as high as 20–25% in untreated cases.[1,2] However, if treated with intravenous immunoglobulin within the first 10 days of the illness, the incidence of coronary artery lesions has been shown to decrease to 4–8%.[2,3] Studies from Japan, where the incidence of Kawasaki disease is the highest, have shown the disease to be recurrent in about 2–4% patients,[4] with most having recurrence within 2 years from the initial episode. Recurrent disease is known to be associated with an increased incidence of coronary artery lesions; however, it is widely believed that coronary artery lesions of Kawasaki disease are not known to recur after complete regression demonstrated on an echocardiogram.[5,6] We present a peculiar case of Kawasaki disease wherein coronary artery aneurysms showed complete regression on follow-up echocardiograms. However, the child developed recurrent disease with reappearance of the coronary artery lesions.

Case report

A 9-month-old male child was admitted to our institution with complaints of high-grade fever of 10–12 days, swelling of the hands and feet, bilateral non-suppurative conjunctivitis, and oral erythema. His physical exam findings were significant for extreme irritability, and the above-mentioned features with no significant lymphadenopathy. On initial investigations, he was found to be anaemic (haemoglobin: 8.5 gm/dl {12–17}), had an elevated total leucocyte count (25,000 cells/μl {4000–11,000}), and had thrombocytosis (13.8 lakhs/cu.mm {1.5–4.5 lakhs}). The acute phase reactants were also significantly elevated with the erythrocyte sedimentation rate and C-reactive protein being 120 mm/hour (0–15) and 22 mg/L (0–5), respectively. Given his history, physical exam findings, and laboratory findings, viral syndromes (like adenovirus) and Kawasaki disease was considered as one of the most probable differential diagnoses, and an echocardiogram was performed on day 1 of admission, which demonstrated a structurally normal heart, normal biventricular function, and aneurysmal dilation of the proximal coronary arteries. The left main coronary artery aneurysm measured 3.2 mm (z score +4.2), left anterior descending was 2.95 mm (z score +4.9), and the right coronary artery was 3.56 mm (z score +5) (Figure 1). In view of the above findings, the patient was diagnosed with Kawasaki disease and given a 2 g/kg single dose of intravenous immunoglobulin over 24 hours and anti-inflammatory dose of oral aspirin was initiated simultaneously, in addition to other supportive measures. He responded well to treatment and demonstrated a decreasing trend in total leucocyte count, platelets, and acute phase reactants. He was discharged home on anti-thrombotic dose of oral aspirin. A repeat echocardiogram 2 weeks later demonstrated improvement in the proximal coronary artery aneurysm dimensions, with the left main coronary artery measuring 2.8 mm (z score +2), left anterior descending measuring 2.7 mm (z score +2.89), and right coronary artery measuring 2.7 mm (z score +2.3) (Figure 2). The coronary artery aneurysms continued to show regression on subsequent echocardiograms until the dimensions and z scores showed complete normalisation by the 5th month from the initial diagnosis of Kawasaki disease (left main coronary artery z score −0.05, left anterior descending artery z score +0.98, and right coronary artery z score +0.49).

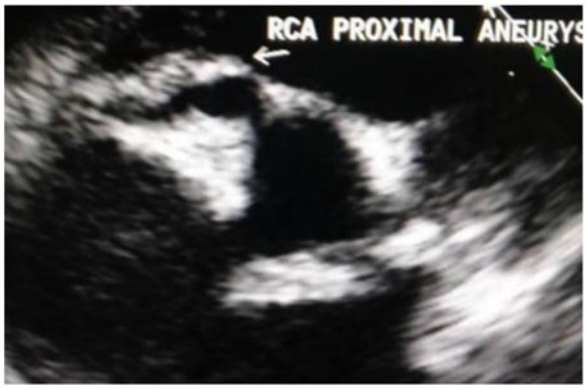

Figure 1. Right coronary artery aneurysm during the initial episode of Kawasaki disease.

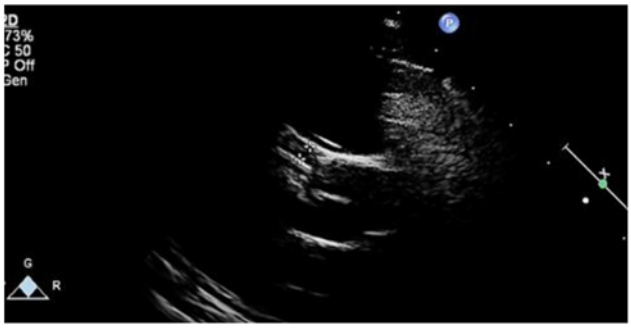

Figure 2. Right coronary artery aneurysm showing regression during subsequent echocardiograms.

The patient remained clinically well on follow-up visits up to 1 year after treatment at which time he was readmitted with complaints of 4 days of fever and bilateral non-suppurative conjunctivitis. Due to his prior history, a high index of suspicion for recurrent Kawasaki disease was entertained and an echocardiogram was performed, which demonstrated a recurrence of aneurysm in the right coronary artery with the proximal measurements of left main, left anterior descending, and right coronary arteries being 2.9 mm (z score +1.29), 2.1 mm (z score +0.76), and 3.2 mm (z score +3.87), respectively (Figure 3). A diagnosis of recurrent Kawasaki disease was made, and patient's parents were advised to repeat a course of intravenous immunoglobulin, but due to parental denial and leaving against medical advice, treatment could not be initiated and the patient was subsequently lost to follow-up.

Discussion

Kawasaki disease is an acute systemic vasculitis affecting young children, and as yet, its aetiology remains unknown. It has developed into a leading cause of acquired heart disease in children, especially prevalent in the developed world. The diagnosis of Kawasaki disease is mainly clinical as per the published criteria by the Japanese Kawasaki Disease Research Committee and the American Heart Association.[6] A typical case of Kawasaki disease is diagnosed when a child presents with fever of more than 5 days and has at least four of the following criteria: non-exudative conjunctival injection; oral mucosal changes (strawberry tongue, mucosal erythema, and cracked and erythematous lips); changes in peripheral extremities (oedema or desquamation); polymorphous rash; and significant cervical lymphadenopathy.[6] The diagnosis becomes difficult when patients do not meet all the above-mentioned criteria and requires a high index of suspicion, along with echocardiographic evidence of coronary artery lesions, and is labelled as incomplete Kawasaki disease.

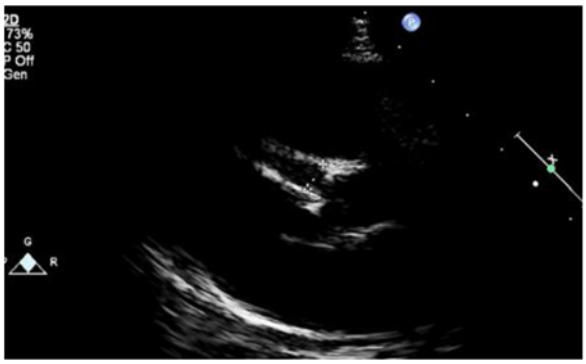

Figure 3. Recurrence of right coronary artery aneurysm during recurrence of Kawasaki disease.

Recurrence of Kawasaki disease has been previously noted with reported rates about 3% in Japan.[7] Factors which predict recurrence include age <2 years at the initial diagnosis, male gender, longer duration of fever, lower haemoglobin levels during the initial episode, and presence of coronary artery lesions during the first episode,[4,8] all of which were present in our case.

Coronary artery complications are the most serious sequelae of the disease, and some studies have shown that the risk of coronary artery lesions increases with recurrence of the disease.[9,10] While many aneurysms appear to resolve spontaneously, long-term morbidity can result from scaring of cardiac tissue, which in about 1% cases affects the heart valves as well.[11]

Our patient did not present with all the classical diagnostic features of Kawasaki disease and thus was labelled as a case of incomplete Kawasaki disease. He also presented with a recurrence of the disease 1 year from the initial diagnosis and demonstrated coronary artery lesions during the first as well as the second episode, which reappeared after they had shown complete regression on echocardiogram. Literature review demonstrates several case reports and articles which have mentioned recurrence of Kawasaki disease as well as the higher risk of coronary artery lesions associated with recurrence of disease; however, we have not come across any report which mentions the reappearance of these lesions once they have regressed.

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Conflicts of Interest. None.

Ethical Standards. This article does not involve experimentation on humans or animals.

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