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Coronary Artery Aneurysm in a 4-Month-Old Infant with Incomplete Kawasaki Disease

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Abstract
Kawasaki Disease (KD) is a type of inflammatory vascular disease that targets the medium vessels with a predilection for coronary arteries. The atypical presentation, called incomplete Kawasaki Disease, is common among children below six months. It presents as a fever with less than four of the KD clinical criteria. This report looks into a case of a four-month-old infant with features of incomplete KD who presented with a fever 38.7° C for four days, polymorphic skin rashes for 2 days, and diarrhea for 1 day. Initially, the child was being treated for urinary tract infection due to the presence of leukocytes on urinalysis. The case of incomplete KD was diagnosed upon echocardiographic confirmation of large and giant aneurysms on the right and left coronary arteries, respectively. The child improved with the administration of management for KD, including intravenous immunoglobulins and aspirin. This case shows the indiscernible presentation of incomplete KD, warranting a need to critically evaluate incomplete KD as one of the differential diagnoses when a child presents with an unexplained fever.
Categories: Pediatrics, Cardiology, Allergy/Immunology
Keywords: incomplete kawasaki disease in infants, aspirin therapy, intravenous immunoglobulins (ivig), fever, coronary artery aneurysm

Introduction

Kawasaki Disease (KD) is one of the vasculitic syndromes that affect the medium-sized vessels with a predilection for coronary arteries [1] . It commonly affects children above six months of age, with the common age of presentation being between nine and eleven months among the Japanese, 12 months among the Mexicans, and 29 months among the Koreans [2]. The clinical presentation before six months is rare and is usually associated with coronary artery aneurysms compared to children above six months [3]. One study noted that the risk of a coronary aneurysm in Kawasaki disease before six months was 20% and reduced to 5% [3] when the condition developed after six months.

A child is diagnosed with Kawasaki when they present with a fever for at least five days and an additional four or more principal clinical criteria, including a polymorphous rash, erythema and edema of the feet, erythematous lips and oral mucosa, bilateral non-purulent conjunctivitis, and cervical lymphadenopathy (≥1.5 cm) [4]. Suppose an individual presents with prolonged fever of unknown origin with less than four of those criteria but has typical echocardiographic findings or laboratory features; they are diagnosed with incomplete Kawasaki Disease [4]. Individuals with incomplete Kawasaki may further present with multi-systemic complications, including those of the gastrointestinal tract, such as abdominal pain, vomiting, diarrhea, and pancreatitis, and the genitourinary system, like urethritis [5].

Epidemiologically, the incidence of incomplete Kawasaki Disease based on a retrospective report among 242 Japanese children was 10% [6]. The incidence of this condition is higher among children below six months. This report explores a case of incomplete KD in a four-month-old infant.

Case Presentation

The case is for a four-month-old girl who presented at the emergency department with a fever of 38.7o C for four days, polymorphic skin rashes for 2 days, and diarrhea for 1 day. The fever was spiking more frequently in the last 2 days and did not improve with the intake of oral paracetamol. She did not receive any other medication apart from the oral paracetamol. The infant was active when the fever subsided. On physical examination, she looked stable with no dysmorphic features and no signs of dehydration. Notably, there were prominent polymorphic skin rashes all over the body. The heart rate was 143, blood pressure 78/64 mmHg, and the temperature 38.0 o C. The patient was admitted as a case of urinary tract infection due to a positive preliminary result of high white blood cells in urine dipstick and was started on intravenous ampicillin and cefotaxin.

The hematological investigations revealed a white blood cell count (WBC) of 19000 /mm3, a hemoglobin (Hb) level of 10.1 g/dl, and a platelet count (PLT) of 751,000/mm3. The neutrophil constituted 60% of the total white cell count, the c-reactive protein (CRP) was elevated at 97 mg/dl, the renal function tests were normal, and the urinalysis revealed 30 pus cells per high power field. On the third day of admission, she became irritable with high spikes of fever, reaching 39o C, which did not improve with paracetamol. Blood and urine cultures were negative and the antibiotics were upgraded because of suspicion of meningitis. A

lumbar puncture could not be done due to the refusal of the parent to consent to the procedure. On further assessment of the differential diagnoses, the prolonged fever, skin rash, and pus on urinalysis made us suspect the likelihood of Kawasaki disease. So, the primary team decided to do echocardiography (ECHO). (ECHO showed normal heart function with dilated left coronary artery(LCA) and right coronary artery (RCA)(Figure1,2).

The patient was started on an intravenous immunoglobulin (IVIG) infusion 2g/kg, and high dose of aspirin 10 mg/kg every six hours, and an intravenous heparin infusion. The fever improved after 24 hours, skin rashes improved on the second day of IVIG, and the patient's general condition improved as she became more active and playful. Blood investigations were retaken on the fourth day after starting IVIG, and the findings were: WBC: 14,566 /mm3, Hb: 10.4 g/dl, PLT: 546,612 /mm3, CRP: 24 mg/dl. Antibiotics were stopped, and the patient was discharged home after 24 days of admission with aspirin 5 mg /kg orally and subcutaneous enoxaparin with pediatric cardiology clinic follow-up in two weeks.

Echocardiography was done before the discharge and showed no change in coronary artery size nor evidence of thrombus formation. There was also no pericardial effusion, and the cardiac function was generally good. An electrocardiogram (ECG) showed normal sinus rhythm with no evidence of ischemic changes. The patient was booked for angiography for more visualization of the coronary arteries.

Discussion

The case of incomplete Kawasaki disease is likely to be underdiagnosed because of the nature of presentation that mimics other systemic diseases. Besides, the failure to meet the full criteria for KD can often make clinicians less likely to focus on the likelihood of being an atypical presentation of Kawasaki. The case of the four-month-old girl above presented with features that do not meet the full criteria of Kawasaki. The unexplained fever for four days, followed by polymorphic skin rashes for two days, and the echocardiographic with bilateral coronary z-scores exceeding +5.0 indicate an atypical form of Kawasaki Disease. The coronary artery Z-score between +5.0 and +10.0 is a large aneurysm while exceeding +10.0 is considered a giant aneurysm [7]. The case of the patient above, with a left coronary artery Z score of 10.47 and a right coronary artery Z score 7.97, indicates that she has a giant aneurysm on the left coronary and a large aneurysm on the right coronary. The finding is congruent with study findings that show children younger than six months who develop KD are likely to develop giant coronary aneurysms even after treatment in the first 10 days of presentations with acute KD [8].

Management

The first-line treatment for incomplete Kawasaki Disease is high-dose intravenous immunoglobulins (IVIG) in a single infusion of 2g/kg [9] of body weight. The IVIG reduces the risk of formation of new coronary artery aneurysms and helps reduce vasculitis and fever [9]. The IVIG should be administered before day 10 of the illness. However, administration before the 7th day is preferred since the risk of developing aneurysms is highest on days 8-9 after the onset of the illness. The IVIG is accompanied by aspirin at a high dose of about 80mg/kg/day divided into four doses. The aspirin is then reduced to a low dose of 3-5 mg/kg/day in a single dose 48 hours after defervescence [9] and maintained for six to eight weeks due to its antiplatelet effect.

Failure of the effectiveness of the two drugs can allow for the use of corticosteroids like oral prednisolone as a second line at a dose of 2 mg/kg/day [10]. Anticoagulants are also indicated in cases of giant aneurysms, as in the four-month-old infant. Warfarin at a dose of 0.05-0.12 mg/kg/day once daily with a target international normalized ratio (INR) of between 2.0 and 3.0 is given. Alternatives like low molecular heparins like enoxaparin (100 IU/kg/dose every 12 h) are used in pediatrics [9].

Conclusions

Incomplete Kawasaki Disease appears to be more common among children below six months. It is associated with cardiac complications in this cohort, as shown by a study that revealed the likelihood of developing coronary aneurysms as being 20% among children below six months than those above six months . The presentation of incomplete KD can be confusing for clinicians due to its multisystem clinical features, including the genitourinary and gastrointestinal manifestations. Findings based on the clinical criteria of unexplained fever for five days with a polymorphous rash and cardiac involvement on echocardiography should offer clues on the need to start them on IVIG and aspirin

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