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Kawasaki Disease: A case report

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Abstract

Kawasaki disease (mucocutaneous lymph node syndrome) is characterized by fever, rash, strawberry tongue and is generally unresponsive to traditional treatments for viral and bacterial infections. Kawasaki disease occurs mostly in children age 6 months to 5 years. It is important to diagnose and begin treatment early due to the significant potential for long-term sequelae, especially in the coronary arteries. However, early recognition can be challenging because of the non-specific symptoms and rarity of the disease. Physicians must maintain a high index of suspicion for Kawasaki disease in children presenting with a febrile illness and a rash that is non-responsive to traditional treatment. We present this case report in order to promote the need for early recognition and treatment in children who present with irritability and a febrile illness.

Keywords

kawasaki disease; vasculitis; aneurysms; mucocutaneous lymph node syndrome; intravenous immunoglobulins

Introduction

Kawasaki disease (KD) is one of the most common vasculitides of childhood, affecting primarily medium-sized arteries, and is of unknown etiology but appears to be related to genetic, environmental, and autoimmune factors. It is usually self-limited but can and often does have significant long-term complications, especially in the coronary vessels. It is difficult to diagnose early, not only because it is relatively uncommon but also because it presents with non-specific symptoms and is often confused with viral exanthems. It was first described in 1967 by Tomisaku Kawasaki in a cohort of Japanese children who had at least 5 days of fever unresponsive to antipyretics, and at least four of the following: bilateral conjunctival injections, oral mucocutaneous injections including fissured lips or strawberry tongue, changes to the extremities including erythema of the hands and feet, polymorphous rash, and cervical lymphadenopathy [1].

Kawasaki disease is important to recognize and treat as early as possible because of a high risk of cardiac complications including coronary artery aneurysms. Prompt treatment within the first 10 days decreases the incidence of coronary artery aneurysms fivefold [2]. In recent years it has replaced rheumatic fever as the most common cause of acquired cardiac disease in children [3]. Early recognition of Kawasaki disease allows for definitive treatment to begin as early as possible in order to minimize these late complications.

It is our hope that this case report will contribute to increased recognition and earlier diagnosis of Kawasaki disease so that treatment can be instituted earlier and complications averted.

Case Report

A 14-month-old male of South Asian descent was brought to the Pediatric clinic due to three days of high fever (102°F-104°F), irritability and decreased feeding. The patient was given Ibuprofen and Acetaminophen with only mild relief of his fever.

He was taken to an Urgent Care Center 1 day after the fever began, and he was diagnosed with an ear infection. He was treated with Amoxicillin and Acetaminophen and then discharged. After no improvement, he was returned to the Urgent Care Center and diagnosed with an unspecified viral infection. The antibiotics were discontinued due to no evidence of an ear infection, and he was instructed to follow up with his Pediatrician.

The patient was upto date on all age appropriate immunizations and had no recent sick contacts. However, he did have a visiting relative from South Asia. There was no family history of similar symptoms.

The patient presented to the Pediatric clinic on day 4 of his illness. He was afebrile but had bilateral non-exudative conjunctival injection, erythematous lips, and oropharynx with white slough on the tongue. He developed a diffuse, maculopapular erythematous rash on bilateral flanks and left arm that his mother had not noticed. Palmar and plantar erythema were also present. At this point, the differential diagnoses were Roseola, Kawasaki disease, Scarlet fever, Adenovirus infection or Measles. A Rapid Strep test was performed which was negative, and a throat swab was sent to be cultured. The patient's mother was instructed to continue supportive care and return to the clinic the next day.

The patient presented to the Pediatric clinic on day 5 of his illness. He was afebrile at presentation and over the past 24 hours. He was irritable, and his rash had spread to his proximal thighs. His oropharynx, and lips were still erythematous but the slough was no longer present, and he still had bilateral conjunctival injection. At this point, it was determined that the etiology was likely viral, and the mother was instructed to follow up in 2 days or sooner if he had no improvement.

The patient presented to the Pediatric clinic on day 8 of his illness, after canceling the appointment due to thinking his condition was improving. He was febrile and irritable once again. His oropharynx remained erythematous, but his conjunctiva was not injected. The rash, palmar and plantar erythema had subsided. The Throat culture returned negative for *Streptococcus Pyogenes* or any other organisms. Kawasaki disease

was highly suspected, and the patient's mother was instructed to take him to the Emergency Department for further assessment and administration of intravenous immunoglobulin (IVIG).

After arriving at the Emergency Department in the night, the patient was evaluated. They suspected that he has Kawasaki disease based upon the history provided and the physical examination performed. He was then given high dose aspirin (dosed at 100 mg/kg), IVIG (dosed at 2 mg/kg), and an echocardiography was performed.

The echocardiogram showed coronary artery dilation. The Left Main or Left Coronary Artery (LCA) was 2.8 mm (Z score 3), Left Anterior Descending (LAD) was 1.8 mm (Z score 1.2) and Right Coronary Artery (RCA) was 2 mm (Z score 1.2). The laboratory results showed evidence of systemic inflammation, and he was admitted to the inpatient pediatric floor for observation and treatment of Kawasaki Disease. The patient remained in the hospital for 2 days as his condition significantly improved.

After discharge, the patient's mother followed up with the Pediatric clinic as instructed; He was examined and noted to be in significantly better health with all symptoms resolved. The patient was then given an influenza vaccine to prevent him from developing a viral syndrome while on aspirin and referred to a Cardiologist to obtain a follow-up echocardiography.

The echocardiogram was performed about 13 days after discharge and showed coronary artery dilation. The LCA was 2.8 mm (Z score 2.82), LAD was now 2 mm (Z score 1.86), RCA was now 2.5 mm (Z score of 2.6), and the Circumflex artery was 1.4 mm (Z score 0.22) with no evidence of true aneurysms. The patient has to follow up with the Cardiologist in 6 months for another echocardiography and has to remain on aspirin (40 mg/day) until his coronary arteries return to the normal size. No other anticoagulation therapy was not necessary as there were no evidence of true aneurysms.

Discussion

Kawasaki disease (KD) is a clinical diagnosis and requires a high index of suspicion. It typically presents with high fever unresponsive to antipyretics, irritability, decreased appetite, a strawberry tongue, and often a non-specific polymorphous rash. The presentation is non-specific, therefore it is often times initially treated as an acute viral or bacterial illness. It most commonly occurs in children of East Asian descent under five years old, and it affects males more than females. KD has also been reported in people of all ethnic and age groups, rarely including adults [4].

The differential diagnosis can include viral illnesses with exanthem, drug reaction, idiopathic juvenile arthritis, toxic shock syndrome, scarlet fever, Stevens-Johnson syndrome, and scalded skin syndrome. Some of which are much more common than KD and may delay definitive diagnosis. Irritability is an important sign which is nearly always present, although interestingly not included as one of the diagnostic criteria [17,18]. Laboratory studies are non-specific and include elevated erythrocyte sedimentation rate and C-reactive protein. Many patients also have moderate leukocytosis and thrombocytosis.

There is no single laboratory test for confirmation of the diagnosis of KD. Laboratory markers rarely provide conclusive evidence for the diagnosis of KD. Clinical laboratory investigation may be used to support the diagnosis of KD, especially in children with incomplete or atypical KD and to assess the intensity of inflammation. Thrombocytopenia in the acute stage of KD can be a marker of macrophage activation syndrome [5,6]. Low platelet count has also been found to correlate with the development of coronary aneurysms and such patients often have severe forms of the disease [7].

McCrindle et al [5] and Manlhiot et al [8] have proposed the classification scheme based on z score for severity of coronary artery abnormalities, which has been adapted and recommended by AHA 2017 guidelines. It is mandatory that body surface area-adjusted 'Z' scores be used to grade the severity of coronary artery involvement so that objectivity can be maintained and results can be compared with other studies [5]. Echocardiography findings in KD other than coronary artery ectasia, dilatation, and aneurysm, include lack of tapering of coronary arteries, myocardial dysfunction, pericardial effusion, aortic root dilatation and valvular regurgitation [5,9,10].

As a vasculitis that affects medium-sized arteries, its major complication is coronary artery aneurysm. This complication occurs in up to 25% of patients not treated with IVIG but decreases to 1%- 4% in treated cases [11]. Despite the low percentage in treated patients, the consequences can be devastating and include myocardial infarction and death [12]. Therefore, serial echocardiography is indicated in all patients with Kawasaki disease.

The primary goal of treatment in Kawasaki disease is to prevent complications. Intravenous immunoglobulin (IVIG) remains the standard of care based on objective evidence collated from prospective studies and meta-analyses [5]. For IVIG to be most effective, it should be given in the first few days of the illness [13]. However, if the child presents after day 10 of fever, IVIG should still be given if the acute inflammatory parameters are high [13]. It has also been suggested that administration of IVIG before day 5 of fever may inadvertently increase the need for further IVIG therapy and also increase the chances of developing a refractory state [14]. Aspirin at a dose of 80–100 mg/kg/day in the United States and 30–50 mg/kg/day in Japan and Europe is recommended during the acute phase of the illness, as this may be better tolerated than higher doses in terms of gastrointestinal and other side effects. The dose should be reduced to an antiplatelet dose of 3–5 mg/Kg once fever and inflammation have subsided [15].

Management after the acute phase of KD includes risk stratification. Risk stratification of coronary artery abnormalities is of primary importance for the long-term follow-up and management of patients with KD [16]. Patients with coronary dilatation that persists beyond 6 weeks need to be kept on low dose aspirin for longer periods of time. For patients with large and giant aneurysms, frequent echocardiographic assessment should be continued. Such patients may also require CT coronary angiography at periodic (approximately 3-5 yearly) intervals. Statins have also been recommended in these situations. Thromboprophylaxis can be maintained with antiplatelet drugs (*e.g*., aspirin/dipyridamole used singly or in combination) and anticoagulants (*e.g*. heparin/warfarin) [5].

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