

CASE REPORT

Kawasaki Disease in a 5-Year-Old Boy: A Rare Vasculitis Syndrome Presenting with Mucocutaneous Manifestations



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Abstract: Kawasaki disease (KD) is a rare, self-limiting vasculitis predominantly affecting children under the age of five. We report a case of a 5-year-old boy presenting with fever, generalized maculopapular rash, peeling of lips, erythematous oral mucosa, lymphadenopathy, and nail changes. The patient was admitted to the pediatric ward with a 3-day history of low-grade fever, rash, and vomiting episodes. Physical examination revealed bilateral lymphadenopathy in the cervical, axillary, and inguinal regions. Laboratory investigations showed elevated white blood cell count, neutrophilia, platelet count, alkaline phosphatase, aspartate aminotransferase, C-reactive protein, and erythrocyte sedimentation rate. The antistreptolysin O titer was positive, indicating recent streptococcal infection. Based on the clinical presentation and diagnostic criteria, a diagnosis of Kawasaki disease was established. The patient was treated with intravenous ceftriaxone, azithromycin, pantoprazole, and supportive therapy, including paracetamol, antihistamines, zinc, and topical calamine lotion. After 15 days of treatment, the patient showed improvement and was discharged with a planned follow-up. This case highlights the importance of early recognition and prompt treatment of Kawasaki disease to prevent potential complications, such as coronary artery aneurysms. Increased awareness among pediatricians is crucial for timely diagnosis and management of this rare vasculitis syndrome, especially in developing countries where cases may go undetected.

Keywords: Kawasaki disease; Vasculitis; Mucocutaneous manifestations; Pediatric; Lymphadenopathy

1. Introduction

Kawasaki disease (KD), also known as mucocutaneous lymph node syndrome, is a rare and potentially serious condition primarily affecting children under the age of five. Initially described in Japan in 1967 by Dr. Tomisaku Kawasaki, this acute self-limited vasculitis has since been recognized worldwide as a significant cause of acquired heart disease in children. Despite decades of research, the etiology of KD remains elusive, posing challenges for diagnosis and treatment. [1, 2] The prevailing hypothesis suggests that in genetically predisposed individuals, an aberrant immune response is triggered by one or more infectious agents, leading to systemic inflammation and vasculitis. However, no single causative pathogen has been identified to date. The clinical manifestations of KD are characterized by a constellation of signs and symptoms that appear in a typical temporal sequence. The hallmark feature is a high-grade fever lasting for five days or more, often accompanied by mucocutaneous inflammation, including non-exudative conjunctivitis, erythematous lips and oral mucosa, strawberry tongue, and polymorphous rash. Additionally, patients may experience cervical lymphadenopathy, swollen hands and feet, and desquamation of the skin in the convalescent phase. [3,4]

Early recognition and prompt treatment are crucial in managing KD, as delayed or inadequate therapy can increase the risk of potentially life-threatening complications, particularly coronary artery aneurysms. The diagnosis is primarily based on clinical criteria, as no single confirmatory laboratory test exists. The diagnostic process often involves ruling out other conditions that may present with similar symptoms. The standard treatment for KD involves intravenous immunoglobulin (IVIG) and aspirin, which have been shown to reduce the risk of coronary artery abnormalities when administered within the first ten days of illness. In cases of IVIG resistance or persistent inflammation, additional therapies, such as corticosteroids or other immunomodulatory agents, may be considered. While KD is relatively uncommon, with reported incidence rates ranging from 60 to 150 cases per 100,000 children under five years of age in certain countries, it remains a significant public health concern due to its potential for serious cardiac complications. In developing nations like India, the true burden of KD may be underestimated due to a lack of awareness among healthcare professionals and the potential for misdiagnosis or underreporting. [5, 6]

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2. Case presentation

A 5-year-old boy was admitted to the pediatric ward of a tertiary care hospital in Rajahmundry, India, with a three-day history of low-grade fever and a generalized maculopapular rash with mild scaling around the face, trunk, back, and limbs. The patient had experienced four episodes of vomiting over the preceding two days. Additionally, the parents reported early signs of perianal excoriation, peeling of the lips, erythematous oral mucosa, and bilateral lymphadenopathy. The patient's medical history was unremarkable, with no significant illnesses or hospitalizations in the past. He was up-to-date with routine immunizations and had no known allergies or family history of autoimmune or rheumatological disorders.

3. Clinical examination

Upon physical examination, the patient appeared ill but was alert and responsive. His vital signs revealed a blood pressure of 92/57 mmHg, a pulse rate of 152 beats per minute, an oxygen saturation of 92% on room air, a respiratory rate of 22 breaths per minute, and a temperature of 101°F (38.3°C). A comprehensive examination revealed bilateral lymphadenopathy involving the cervical, axillary, and inguinal regions. The lymph nodes were firm, non-tender, and mobile. Mucocutaneous manifestations were evident, including peeling of the lips, erythematous oral mucosa, and a strawberry-like appearance of the tongue. Furthermore, the patient exhibited leuconychia (white discoloration of the nails) and a generalized maculopapular rash with mild scaling on the face, trunk, back, and limbs.

Cardiovascular and respiratory examinations were unremarkable, with no evidence of murmurs, added sounds, or adventitious breath sounds. The abdomen was soft, non-tender, and non-distended, with no palpable organomegaly or masses. The neurological examination was normal, with appropriate responses to verbal commands and age-appropriate reflexes and coordination. Based on the clinical presentation and the constellation of signs and symptoms, a preliminary diagnosis of Kawasaki disease was suspected, prompting further diagnostic evaluations to confirm the diagnosis and assess potential complications.

4. Diagnostic evaluation

Given the clinical suspicion of Kawasaki disease, a comprehensive diagnostic workup was initiated to confirm the diagnosis and assess potential complications. Laboratory investigations were ordered, including a complete blood count (CBC), inflammatory markers, liver function tests, renal function tests, and a streptococcal antibody titer test. The CBC revealed leukocytosis (11,400 cells/cumm) with neutrophilia (62%) and thrombocytosis (459,000/cumm), which are common findings in Kawasaki disease due to the underlying inflammatory process. Additionally, the erythrocyte sedimentation rate (ESR) was elevated at 65 mm/hr, and the C-reactive protein (CRP) level was significantly elevated at 43.4 mg/L, indicating a heightened inflammatory state. Liver function tests showed mildly elevated alkaline phosphatase (144 IU/L) and aspartate aminotransferase (SGOT) (42 IU/L) levels, which can occur in Kawasaki disease due to the involvement of the hepatobiliary system. Renal function tests, including serum creatinine and blood urea, were within normal limits. The antistreptolysin O (ASO) titer test, which detects antibodies produced in response to a recent streptococcal infection, was positive. While not specific for Kawasaki disease, a positive ASO titer can support the diagnosis by ruling out other potential etiologies. To evaluate for potential cardiac involvement, which is a major concern in Kawasaki disease, an electrocardiogram (ECG) and echocardiogram were ordered. The ECG showed no significant abnormalities, while the echocardiogram revealed mild dilation of the left coronary artery, a common finding in the acute phase of Kawasaki disease.

5. Treatment and management

With the clinical presentation and diagnostic findings supporting a diagnosis of Kawasaki disease, prompt treatment was initiated to reduce the risk of cardiovascular complications and promote recovery.[7,8] The patient was started on intravenous immunoglobulin (IVIG) therapy, which is the standard first-line treatment for Kawasaki disease. IVIG is believed to modulate the aberrant immune response and reduce the risk of coronary artery aneurysms when administered within the first ten days of illness. The recommended dose of 2 g/kg was administered over 12 hours. In addition to IVIG, the patient was prescribed high-dose aspirin therapy. Aspirin serves a dual purpose in Kawasaki disease treatment: during the acute phase, high doses (80-100 mg/kg/day) are administered for its anti-inflammatory and anti-platelet effects, while during the convalescent phase, low-dose aspirin (3-5 mg/kg/day) is continued for its anti-platelet effect until the coronary artery abnormalities resolve or the patient has been afebrile for at least six to eight weeks. To manage the fever and associated discomfort, the patient was prescribed paracetamol (acetaminophen) as needed. Additionally, intravenous ceftriaxone and azithromycin were administered to provide broad-spectrum antibiotic coverage and reduce the risk of secondary bacterial infections. Supportive care included intravenous pantoprazole for gastrointestinal protection, hydroxyzine hydrochloride (an antihistamine) to alleviate pruritus associated with the rash, and topical calamine lotion for symptomatic relief of the rash. Regular monitoring of vital signs, fluid balance, and clinical response was undertaken throughout the hospital stay. Repeat laboratory tests were ordered to assess the inflammatory markers and adjust the treatment regimen accordingly

6. Outcome and follow up

Following the initiation of treatment with intravenous immunoglobulin (IVIG) and high-dose aspirin, the patient's clinical condition gradually improved. The fever subsided within 48 hours, and the mucocutaneous manifestations, including the rash, conjunctivitis, and oral lesions, began to resolve over the subsequent days. Regular monitoring of inflammatory markers revealed a progressive decline in the levels of C-reactive protein (CRP) and erythrocyte sedimentation rate (ESR), indicating effective control of the underlying vasculitis. Repeat echocardiography performed one week after the initial evaluation showed no progression of the coronary artery dilation, suggesting a favorable response to treatment.

The patient was discharged after 15 days of inpatient care, with instructions to continue low-dose aspirin therapy and scheduled follow-up appointments at the outpatient clinic. During the convalescent phase, the patient experienced the expected desquamation of the skin on the fingers and toes, a characteristic feature of Kawasaki disease. At the first follow-up visit, one month after discharge, the patient's physical examination was unremarkable, and laboratory tests showed normalization of inflammatory markers. A repeat echocardiogram revealed complete resolution of the coronary artery dilation, indicating successful treatment and recovery.

The patient was advised to continue low-dose aspirin therapy for an additional two months and to report any concerning symptoms promptly. Subsequent follow-up visits were scheduled at regular intervals to monitor for potential long-term cardiovascular complications, although the risk was considered low given the prompt diagnosis and appropriate treatment.

7. Discussion

Kawasaki disease (KD) is a rare and potentially serious condition that primarily affects young children.[9,10] Despite its relatively low incidence, early recognition and timely treatment are crucial to prevent life-threatening complications, particularly coronary artery aneurysms. In the presented case, the patient exhibited the classic clinical features of KD, including prolonged fever, mucocutaneous manifestations (rash, conjunctivitis, oral lesions, and desquamation), cervical lymphadenopathy, and changes in the extremities (swelling and nail abnormalities). [11, 12] The diagnostic criteria for KD were met, and the positive antistreptolysin O (ASO) titer supported the diagnosis by ruling out other potential etiologies. The prompt initiation of treatment with IVIG and high-dose aspirin played a crucial role in the patient's favorable outcome. IVIG has been shown to significantly reduce the risk of coronary artery abnormalities when administered within the first ten days of illness. The anti-inflammatory and immunomodulatory effects of IVIG are believed to dampen the aberrant immune response underlying KD, thereby mitigating the vasculitis and associated complications.

Aspirin, in addition to its anti-inflammatory properties during the acute phase, also serves as an anti-platelet agent, reducing the risk of thrombosis and potentially preventing the formation or progression of coronary artery aneurysms. It is important to note that while KD is relatively rare, it remains a significant public health concern due to the potential for serious cardiac complications if left untreated. In developing countries like India, where this case was reported, the true burden of KD may be underestimated due to a lack of awareness among healthcare professionals and the potential for misdiagnosis or underreporting.[13, 14] Increased awareness and education among pediatricians and other healthcare providers are crucial for early recognition and appropriate management of KD. Prompt diagnosis and timely initiation of treatment can significantly improve outcomes and reduce the risk of long-term cardiovascular complications.

8. Conclusion

Kawasaki disease is a rare but potentially serious condition that requires prompt recognition and treatment to prevent life-threatening complications. This case highlights the importance of early diagnosis, appropriate management with IVIG and aspirin therapy, and close follow-up to monitor for potential long-term cardiovascular implications. Increased awareness and education among healthcare professionals are crucial for improving the outcomes of children affected by this condition.

Compliance with ethical standards

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Conflict of interest statement

The authors declare no conflict of interest.

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Author's short biography

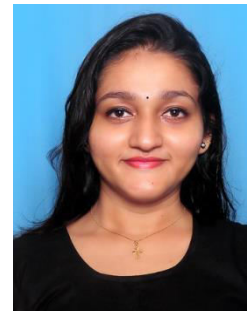
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