



Typical and Incomplete Kawasaki Disease in Infants: A Case Series Highlighting Diagnostic Challenges and Cardiac Outcomes

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ABSTRACT

Background

Kawasaki disease (KD) is an acute, self-limiting vasculitis predominantly affecting children under five years of age and is a leading cause of acquired heart disease in children. It is characterized by prolonged fever and mucocutaneous inflammation, with a risk of developing coronary artery abnormalities if left untreated. We report two cases of infantile Kawasaki disease with variable clinical presentations. The first case involved a 1-year-old female presenting with prolonged fever, mucocutaneous features, and elevated inflammatory markers, consistent with typical KD. The second case involved a 4.5-month-old infant with incomplete clinical features but significant coronary artery aneurysmal dilatation on echocardiography. Both patients were treated with intravenous immunoglobulin (IVIG) and aspirin, resulting in clinical improvement. These cases highlight the importance of early recognition of both typical and incomplete Kawasaki disease, especially in infants, to prevent cardiac complications.

Keywords: Kawasaki Disease; Infant; Coronary artery aneurysm; Vasculitis; Intravenous immunoglobulin.

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INTRODUCTION

Kawasaki disease (KD) is an acute, self-limiting medium-vessel vasculitis that predominantly affects infants and young children. First described in Japan in 1967, it is now recognized worldwide, with a higher incidence in Asian populations. Approximately 80-90% of cases occur in children younger than five years of age.^{2,5}

The etiology of Kawasaki disease remains unknown; however, it is believed to result from an abnormal immune response to infectious triggers in genetically predisposed individuals. Several genetic factors such as ITPKC, ANGPT1, CCR5, and ABCC4 have been implicated in its pathogenesis.^{3,6}

Diagnosis is primarily clinical and requires fever persisting for at least five days along with at least four of the following principal features: bilateral non-exudative conjunctivitis, oral mucosal changes, peripheral extremity changes, polymorphous rash, and cervical lymphadenopathy greater than 1.5 cm. Incomplete Kawasaki disease occurs when fewer criteria are present but is supported by laboratory findings and echocardiographic abnormalities.^{1,4}

Coronary artery abnormalities develop in approximately 25-30% of untreated patients, making early diagnosis and treatment essential. Intravenous immunoglobulin (IVIG) significantly reduces the risk of coronary complications.

CASE PRESENTATION

Case 1

A 1-year-old female child presented in May 2023 with a 7-day history of rhinorrhea and 4 days of high-grade fever, along with two episodes of vomiting. She was admitted to the Pediatric Intensive Care Unit.

On examination, she was irritable and febrile. Clinical findings included cracked and erythematous lips, bilateral conjunctival congestion, periungual desquamation of the index finger, and a maculopapular rash over the face measuring approximately 0.5-1 cm. A small, firm, and mobile cervical lymph node was palpable.

Laboratory investigations revealed leukocytosis with a total leukocyte count of 28,030/mm³, hemoglobin of 11.2 g/dL, and platelet count of 410,000/mm³. Inflammatory markers were elevated with C-reactive protein of 79.4 mg/L and erythrocyte sedimentation rate of 40 mm/hr. Coagulation profile showed mildly prolonged activated partial thromboplastin time. Infectious workup, including COVID-19, dengue, and scrub typhus, was negative.

Initial echocardiography demonstrated normal coronary artery dimensions with left ventricular ejection fraction of 55% and mild tricuspid regurgitation.

The patient was initially treated with empirical intravenous antibiotics. Due to persistent fever up to 104°F despite treatment, Kawasaki disease was suspected. High-dose aspirin (30 mg/kg/day) was initiated, and intravenous immunoglobulin (IVIG) was administered on the third day of admission.

Repeat echocardiography showed mild left atrial dilatation and mild biventricular systolic dysfunction with an ejection fraction of 52%. Following IVIG therapy, fever subsided, and clinical condition improved significantly. Aspirin was later reduced to low-dose (3-5 mg/kg/day) after resolution of fever. The patient was discharged in stable condition. On follow-up after one week, inflammatory markers normalized, and platelet count showed reactive thrombocytosis.



Figure 1: Bilateral non-exudative (non-purulent) conjunctivitis



Figure 2: Periungual desquamation

Case 2

A 4.5-month-old female infant presented in June 2023 with a 5-day history of fever (maximum temperature 103°F), along with cough and multiple episodes of loose stools.

On examination, the patient was irritable with bilateral non-exudative conjunctivitis, erythema of the lips and oral mucosa, and cracked lips. Rash, strawberry tongue, and cervical lymphadenopathy were absent.

Laboratory investigations showed leukocytosis (18,040/mm³), hemoglobin of 9.9 g/dL, and thrombocytosis (501,000/mm³). Inflammatory markers were markedly elevated with CRP of 90 mg/L and ESR of 46 mm/hr.

Echocardiography revealed severe aneurysmal dilatation of both the left and right coronary arteries with preserved cardiac function, consistent with Kawasaki disease.

The patient was initially managed with broad-spectrum antibiotics. Due to persistent fever and echocardiographic findings, a diagnosis of incomplete Kawasaki disease was made. High-dose aspirin was initiated, and intravenous

immunoglobulin (IVIG) was administered on the sixth day of illness.

Following IVIG therapy, fever resolved and clinical condition improved. The patient was discharged on low-dose aspirin therapy with advice for regular cardiology follow-up.



Figure 3: Strawberry tongue (prominent red papillae on an erythematous tongue)



Figure 4: Dry cracked lips

DISCUSSION

Kawasaki disease is a systemic vasculitis with diverse clinical manifestations, which can make diagnosis challenging, particularly in infants. Incomplete Kawasaki disease is more frequently observed in younger infants and is associated with a higher risk of coronary artery complications due to delayed diagnosis.^{1, 5, 6}

In the first case, classical clinical features allowed early diagnosis and treatment, resulting in favorable outcome without coronary artery involvement. In contrast, the second case presented with incomplete clinical features and required echocardiographic findings for diagnosis, highlighting the importance

of imaging in such cases.

Elevated inflammatory markers such as CRP and ESR are important supportive findings in both typical and incomplete Kawasaki disease^{1,4} According to the American Heart Association guidelines, intravenous immunoglobulin (IVIG) and high-dose aspirin remain the mainstay of treatment. Early administration of IVIG significantly reduces the incidence of coronary artery aneurysms.^{1,7}

These cases emphasize the need for high clinical suspicion, especially in infants presenting with prolonged fever and atypical features.

CONCLUSIONS

Kawasaki disease should be considered in any child presenting with prolonged unexplained fever, even in the absence of complete clinical criteria. Incomplete Kawasaki disease is more common in infants and carries a higher risk of coronary artery complications. Early echocardiographic evaluation and prompt initiation of IVIG therapy are essential to prevent long-term cardiovascular morbidity.

Limitations

The cross-sectional design limits the ability to establish causal relationships between sleep quality, gender, academic phase, and physical activity levels. Data were collected using self-administered

questionnaires, which may be subject to recall bias. The study was conducted in a single medical institution, which may limit the generalizability of the findings to other medical schools or student populations.

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Authors' contributions

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