CASE REPORT

Afebrile Kawasaki disease: A case report

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ABSTRACT

Objective: Kawasaki disease (KD) is an acute, self-limiting vasculitis and the most common cause of acquired heart disease in children under 5 years old. Despite over 50 years of research, the etiology and long-term prognosis of KD remain unclear.
Case presentation: A 2-year-7-month-old boy presented with all the typical KD symptoms, including conjunctival injection, cracked lips, strawberry tongue, and membranous desquamation of the fingers and toes, but without fever. The patient was admitted with a 9-day history of peeling skin on his fingers, and no history of illness or fever in the past two weeks. Laboratory tests revealed elevated platelet count. An echocardiogram showed normal coronary arteries. The patient was diagnosed with KD and treated with low-dose aspirin and dipyridamole. He was discharged on the third day and showed complete symptom resolution and normal coronary arteries at follow-up.
Conclusions: This case report describes an instance of afebrile KD, emphasizing the need for clinical vigilance and comprehensive assessment for diagnosis and treatment. Afebrile KD poses a diagnostic challenge as fever is a major diagnostic criterion. However, clinicians should consider afebrile KD when other clinical features and laboratory results suggest KD, and initiate appropriate treatment to prevent severe complications like coronary artery lesions. This case underscores the importance of early diagnosis, timely intervention, and multidisciplinary team collaboration for effective management of KD.

Key Words: Kawasaki disease, Early diagnosis

1. INTRODUCTION

Kawasaki disease (KD) is an acute, self-limiting vasculitis and the most common cause of acquired heart disease in children under 5 years old. It was first described by Dr. Kawasaki in Japan in 1967. Despite over 50 years of research, the etiology and long-term prognosis of KD remain unclear. While expert guidelines, such as those from the American Heart Association (AHA) and the American Academy of Pediatrics (AAP), provide a framework for diagnosis and treatment, variations in clinical presentation, particularly in atypical cases, suggest that further refinement and standardization of diagnostic and therapeutic approaches may still be needed. Typical KD is defined by fever and at least four of the following five major clinical features: non-exudative conjunctival injection; changes in the lips and oral cavity such as dry, cracked, and red lips, strawberry tongue, and diffuse erythema of the oropharyngeal mucosa; acute non-suppurative cervical lymphadenopathy (usually > 1.5 cm in diameter); polymorphous rash, including redness around the BCG scar; changes in the extremities, such as erythema of the palms and soles, hard edema of the hands and feet in the acute phase, and membranous desquamation of the fingers and toes during convalescence, with or without coronary artery lesions (CAL).[1] This case report describes a 2-year-

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7-month-old male patient who presented with all the typical KD symptoms, including conjunctival injection, cracked lips, strawberry tongue, and membranous desquamation of the fingers and toes, but without fever. This poses a diagnostic challenge and underscores the necessity of clinical vigilance and timely intervention to prevent coronary artery complications.

2. Case presentation

The patient, a 2-year-7-month-old boy, was admitted with a 9-day history of peeling skin on his fingers. He had no history of illness or fever in the past two weeks. Physical examination upon admission showed a temperature of 36.6°C, pulse of 119 bpm, respiratory rate of 27 bpm, weight of 12.0 kg, and height of 89 cm. He was alert, well-developed, and well-nourished. His skin, mucous membranes, and sclerae were not jaundiced, and no rash was observed. His neck was supple without resistance, and several pea-sized, non-tender, movable lymph nodes were palpable in the bilateral cervical region. Both eyes exhibited slight conjunctival injection without eyelid edema. The nose appeared normal with minimal nasal secretions. His lips were less red, cracked, and a strawberry tongue was noted. The pharynx was erythematous with grade I bilateral tonsillar enlargement without purulent discharge. Lung sounds were coarse and symmetrical, with no obvious dry or wet rales. Heart sounds were strong and regular, with no pathological murmurs. The abdomen was soft without tenderness or rebound pain, and no hepatosplenomegaly or abdominal masses were detected. The extremities were warm, with no edema, and the fingers and toes showed membranous desquamation. Given the risk of coronary artery complications associated with untreated KD, the multidisciplinary team (MDT) decided that hospitalization was necessary for further evaluation and immediate treatment. Laboratory tests on March 15, 2024, revealed elevated white blood cell count (12.06×10^9/L), platelet count (565×10^9/L), and erythrocyte sedimentation rate (33 mm/h). Biochemical tests showed elevated creatine kinase-MB (6.95 ng/ml) and α-hydroxybutyrate dehydrogenase (386 U/L). C-reactive protein is 3.95 mg/L. Urine and stool tests were unremarkable (see Figure 1). Echocardiogram showed normal chamber sizes, normal interventricular and left ventricular wall thickness and motion, intact septa, and normal valve morphology and function. Coronary arteries appeared smooth with normal dimensions: left main coronary artery (2.3 mm), left anterior descending artery (1.5 mm), left circumflex artery (1.3 mm), and right coronary artery (2.2 mm).[2] Based on the typical KD clinical features, the KD MDT team diagnosed the patient with KD. Several differential diagnoses were considered, including scarlet fever, toxic shock syndrome, and viral exanthems. However, these were ruled out based on the absence of a clear source of infection, and the characteristic clinical presentation of KD. The echocardiogram showing normal coronary arteries at diagnosis supported the timely diagnosis, as coronary artery involvement can develop later in the disease course if untreated.The membranous desquamation, particularly in the hands and feet, is a hallmark of the convalescent phase of KD, making this diagnosis more definitive despite the atypical absence of fever. Treatment with low-dose aspirin and dipyridamole was initiated. The patient was discharged on the third day after admission and was closely monitored and followed up. Three month later, the patient's symptoms had completely resolved, and echocardiogram showed no abnormalities.

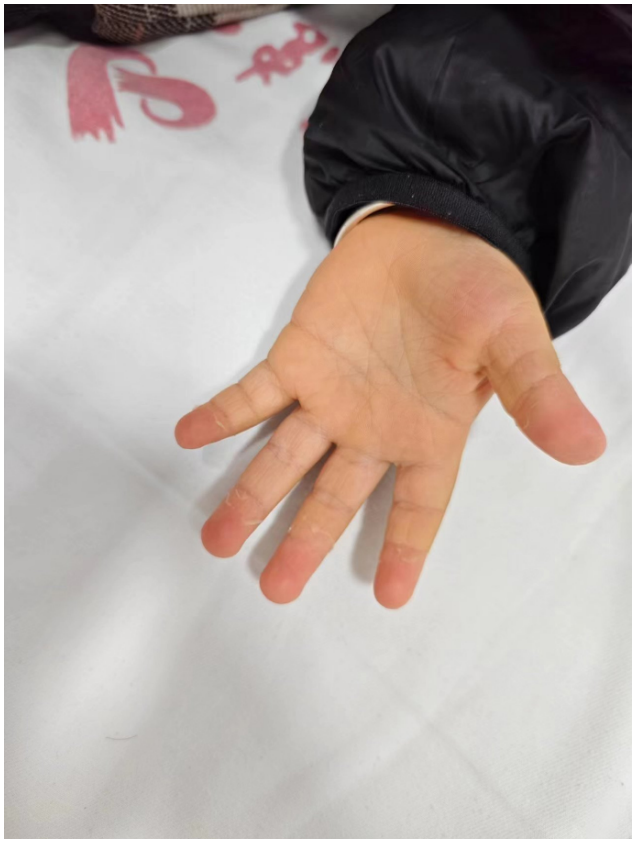

Figure 1. Membranous desquamation of the fingers

3. Discussion

This case report describes an instance of afebrile KD, which is relatively rare in clinical practice but crucial for a comprehensive understanding of KD. Typical KD diagnosis relies on fever and at least four major clinical features, but in this case, the patient exhibited all typical KD symptoms without fever, highlighting the need for clinical vigilance and the importance of comprehensive assessment for diagnosis and

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treatment of afebrile KD.[3] There are several recognized risk factors for missing a KD diagnosis, including age under 6 months or over 5 years, as KD is most commonly diagnosed in children aged 6 months to 5 years. Patients outside of this typical age range may present with incomplete or atypical symptoms, increasing the risk of misdiagnosis. Additionally, the absence of fever, as in this case, makes the diagnosis even more difficult, since fever is considered one of the hallmark features of KD.

While fever is indeed a hallmark of KD diagnosis, there are documented cases of afebrile KD, although they are rare. For example, Atsunori Yoshino et al. reported the first cases of afebrile KD in Japan. One case involved a 7-month-old Japanese girl who presented with bulbar conjunctival injection, diarrhea, skin erythema, and redness around the Bacillus Calmette-Guérin (BCG) inoculation site. Thirteen days after the onset of symptoms, an echocardiogram (UCG) showed bilateral coronary artery dilatation, which had completely resolved five months later. Another case involved a 13-month-old Japanese boy, where the echocardiogram also revealed bilateral coronary artery dilatation 22 days after symptom onset, with mild dilatation in the proximal coronary arteries remaining.[4] In addition to these atypical KD patients, there have also been reports of afebrile KD in neonates in China, adding complexity to the diagnosis of KD. In this case, a newborn female was hospitalized due to elevated peripheral blood leukocyte count, initially diagnosed with neonatal sepsis and bacterial meningitis. Although the infant did not have a fever after admission, she developed a facial rash by day 7, and desquamation of the distal extremities on day 11. By day 15, an ultrasound revealed non-suppurative cervical lymphadenopathy, and an echocardiogram showed coronary artery aneurysms in both coronary arteries. The patient was ultimately diagnosed with incomplete Kawasaki disease (IKD). Follow-up echocardiograms three months later showed that the coronary arteries had returned to normal.[5]

In this case, despite the absence of fever, the patient exhibited multiple classic KD symptoms, such as conjunctival injection, cracked lips, strawberry tongue, and membranous desquamation of the fingers and toes. The MDT recognized the atypical nature of this presentation but based the diagnosis on the presence of these characteristic features, which align closely with the diagnostic criteria for KD. Furthermore, even in the absence of coronary artery abnormalities at the time of diagnosis, the risk of CAL remains significant if untreated. The diagnosis of KD in this case, particularly in the absence of fever, involved careful consideration of other potential differential diagnoses. The multidisciplinary team evaluated a range of conditions that can present with similar symptoms, including scarlet fever, toxic shock syndrome, Stevens-Johnson syndrome, and viral exanthems. Scarlet fever was considered due to the presence of a rash and desquamation; however, the absence of a primary streptococcal infection source and a distinct pattern of rash helped rule it out. Toxic shock syndrome was considered but excluded due to the lack of systemic shock symptoms, such as hypotension. Stevens-Johnson syndrome was ruled out because the patient did not have mucosal erosions or the characteristic target lesions on the skin. Viral exanthems were also considered, but the presence of key KD symptoms, such as conjunctival injection, cracked lips, strawberry tongue, and extremity changes, strongly supported the diagnosis of KD. Ultimately, the diagnosis was made based on the combination of clinical features characteristic of KD and the exclusion of other potential diagnoses. The decision to initiate anti-platelet therapy early, despite the absence of fever or coronary abnormalities at presentation, was guided by the understanding that untreated KD can lead to serious complications such as coronary artery lesions. The team prioritized early treatment to mitigate this risk.[6, 7]

4. Conclusion

This case report of afebrile KD highlights the importance of clinical vigilance, comprehensive evaluation, and timely intervention to prevent coronary artery complications. By discussing this case, we aim to increase awareness of afebrile KD and promote early diagnosis and standardized treatment in clinical practice.

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Authors contributions

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DATA SHARING STATEMENT

No additional data are available.

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