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Case Report

A Missed Kawasaki Disease Shock Syndrome Diagnosis Resulting in Coronary Complications

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Abstract

Background: Kawasaki disease (KD) is an acute inflammatory vasculitis that primarily affects children under 5 years of age. Kawasaki Disease Shock Syndrome (KDSS) is a rare complication characterized by hypotension and shock. Despite being the leading cause of heart disease in developed countries, diagnosing KD is challenging due to the lack of specific pathognomonic tests. Delayed treatment can increase the risk of long-term coronary artery abnormalities in up to 25% of patients. *Case presentation:* A 31-month-old boy initially presented with fever, lymphadenopathy, and pharyngeal congestion. The patient was treated with antibiotics due to positive Mycoplasma IgM and elevated inflammatory markers. Despite treatment, the patient a widespread rash, ulcers, lip peeling, and orbital swelling. The patient was transferred to the Pediatric Intensive Care Unit of a second hospital as a case of suspected septic shock. The patient received Intravenous immunoglobulin and corticosteroids without a diagnosis of KD. It was not until the 23rd day of illness that KD was diagnosed, and the absence of timely diagnosis resulted in aneurysmal dilation of all major coronary arteries with aneurysmal fusiform formation. *Conclusion:* The case highlights the importance of early recognition and diagnosis of KD and KDSS, recommending multidisciplinary evaluation. Clinicians should consider atypical KD in children who are unresponsive to antibiotics and have persistent fever. Positive infectious serology does not exclude the diagnosis of KD. In the absence of echocardiographic markers, early treatment should be prioritized to prevent long-term cardiac damage.

Keywords

Kawasaki Disease, Shock, Coronary Aneurysm, Acute Disease, Fever, Child

1. Introduction

Kawasaki disease (KD) is an acute systemic vasculitis that predominantly affects small to medium-sized arteries in children under 5 years. Despite being the leading cause of acquired heart disease in developed countries, KD has no pathognomonic diagnostic test [1]. Instead, the diagnosis of KD hinges on the following criteria: extremity changes, rash, bilateral conjunctivitis, oral mucosal changes, and cervical lymphadenopathy.

The diagnosis is classified into two categories: complete and incomplete KD. Complete KD requires five or more days of fever alongside four of the listed diagnostic criteria, while Incomplete KD requires at least five days of fever but only 2-3

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of the diagnostic criteria. In infants, a KD diagnosis can be concluded with at least seven days of unexplained fever supported by lab findings such as blood tests, echocardiography, and urinalysis. If untreated, Coronary artery abnormalities can appear in up to 25% of patients, requiring lifetime cardiology follow-up. However, this risk decreases significantly to 4% with prompt diagnosis and treatment [1]. Kawasaki Disease Shock Syndrome (KDSS) is a rare complication characterized by hypotension and shock [1]. Compared to KD, KDSS is associated with more severe systemic inflammation and a higher risk of coronary complications [2]. Recent data indicate that KDSS only appears in approximately 2.8-5.3% of KD cases [3]. The first-line treatment for KD includes intravenous immunoglobulin (IVIG) and high-dose aspirin [1]. Although the etiology of KD is unclear, research suggests that it arises from an abnormal immune response triggered by infection or environmental factors, as supported by a systematic review of genetic studies that discovered specific genetic markers that predispose to KD [4].

This case highlights the value of multi-specialty intervention along with the integration of recent biomarker research to improve early recognition and prevent the misdiagnosis of KDSS.

2. Case Report

A 31-month-old boy presented to the emergency department with his parents with three days of fever (maximum 39.6 ℃), cervical lymphadenopathy, neck stiffness, and pharyngeal congestion. A respiratory panel was conducted, and the patient was admitted for positive Mycoplasma IgM pneumonia. The patient was initially treated with Ceftriaxone and Azithromycin. However, he developed a new rash on his trunk, back, and groin the following day, and so he was switched to Tazocin due to a suspected adverse reaction or worsening of the Mycoplasma infection. Laboratory findings revealed high inflammatory markers and leukocytosis with neutrophilia (Table 1).

Table 1. *Laboratory Results upon Admission to each Hospital.*

	H1	H2	H3	H4
White blood cell count $(\cdot 10^3/\mu L)$	23	22	-	12.6
Hemoglobin (g/dL)	-	-	-	8.0
Platelets $(\cdot 10^3/\mu L)$	-	-	-	596.0
Neutrophils (%)	-	-	-	60.0
Lymphocytes (%)	-	-	-	26.0
Monocytes (%)	-	-	-	12.0
D-dimer (μg/mL FEU)	-	2.58	-	2.35
C-reactive protein (mg/L)	134	201	-	116.8
Procalcitonin (ng/mL)	51.8	-	-	0.10
Erythrocyte sedimentation rate (mm/h)	-	-	-	24
Total serum protein	-	-	-	8.3
Albumin (g/dL)	Low	-	-	4.2

Two days after admission, the patient's status deteriorated, developing hypotension and persistent fever spikes. As a result, the patient was transferred to the Pediatric Intensive Care Unit (PICU) of Hospital 2 as a case of suspected septic shock. On the day of transfer (5th day of illness), the patient developed orbital swelling, peeling of lips, oral ulcers, and a rash on the trunk. His urine output was also noted to be low.

Upon admission, the patient was visibly sick, puffy, and flushed with signs of fever. Laboratory tests were significant for elevated inflammatory markers and D-dimer levels. Throughout his 10-day stay at the PICU, he was treated with Vancomycin, Ceftriaxone, Clarithromycin, and norepinephrine. A central venous catheter was inserted, and he was started on Inotropes. A 2-dose regimen of IVIG and methylprednisolone was administered empirically, yet no formal diagnosis of KD was made. The patient's chest X-ray and following CT scan confirmed bilateral pleural effusion, and a chest tube was inserted. While this led to an improvement in fever, his laboratory tests showed a persistent elevation in inflammatory markers.

The patient's first echocardiogram was performed on the 10th day of illness, showing mild dilation of all heart chambers. Following this, the patient continued Meropenem and Clarithromycin for 10 days and Linezolid for 7 days. Once symptoms improved, the patient was scheduled for discharge on the 16th day of illness, yet he developed a new fever on the same day. Despite this, the mother was advised to continue oral Linezolid and Cefuroxime at home. After discharge, the patient was seen at an outpatient clinic for generalized fatigue along with hand and foot swelling, where he was advised to switch from Linezolid to Clindamycin. After 5 days of discharge (21st day of illness), the patient returned to the emergency department of Hospital 3 with more frequent and intense fever spikes, where he was recommended for admission to a tertiary Hospital 4.

The following day (22nd day of illness), the patient presented to the emergency department of hospital 4 with unresolved fever and swelling in his hands and feet. He was then admitted for further evaluation and was started on antibiotics (Cefuroxime and Clindamycin) for one day. However, the treatment was discontinued as the infectious diseases team concluded that the illness was more likely inflammatory than infectious. Upon further examination, the infectious diseases team concluded a suspected diagnosis of KD. Cardiology was consulted, and an echocardiogram was performed, which showed a three-vessel coronary aneurysm with the results shown in Table 2.

Table 2. *Echocardiography Results.*

Coronary Artery	Size	Z-score
Left main coronary	8 mm	+12.5
Left anterior descending	11 mm	+35
Right coronary artery	11 mm	+21

As a result, the patient was confirmed as a case of refractory KD with involvement of all coronaries. The patient was then started on antithrombotic therapy (Enoxaparin and Aspirin) to reduce the risk of thrombus formation, and propranolol was administered to reduce myocardial oxygen demand. Moreover, echocardiograms were performed daily to monitor the progression of aneurysms. The rheumatology team reviewed the patient and advised two doses of IV Infliximab and a 5-day course of high-dose IV methylprednisolone followed by 7 days of low-dose oral prednisolone (2 mg/kg). As inflammatory markers continued to decrease, the patient was moved to a tapering dose of steroids. Due to low hemoglobin levels, he received a packed red blood cell transfusion without any complications. Ultimately, the patient was discharged from the hospital upon mutual agreement from the cardiology, rheumatology, and general pediatrics teams.

The patient continued follow-up at a pediatric cardiology outpatient clinic. During the most recent follow-up appointment, an echocardiogram was conducted and was significant for a persistent three-vessel giant aneurysm (Table 3).

Table 3. *Echocardiography Results on follow-up.*

Coronary Artery	Size	Z-score
Left main coronary	6.2 mm	+9.33
Left anterior descending	15.5	+48.91
Right coronary artery	12.3 mm	+30.02

A cardiac CT was also done and confirmed an aneurysm in the left main coronary artery, left circumflex, and right coronary artery. (Figures 1-4).

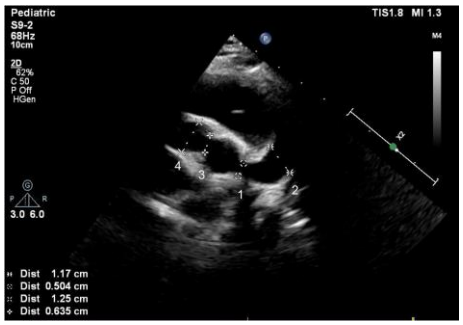

Figure 1. *Echocardiography showing 1) Main left coronary artery, 2) Left anterior descending, 3) Right coronary artery, 4) Proximal Right coronary artery.*

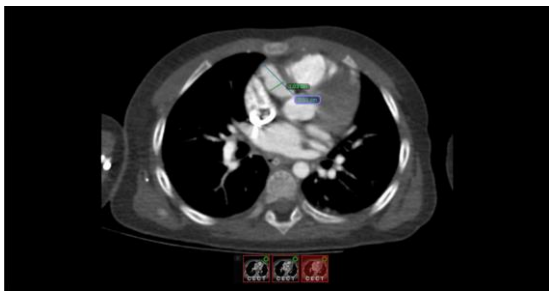

Figure 2. *Cardiac CT showing a right coronary artery aneurysm.*

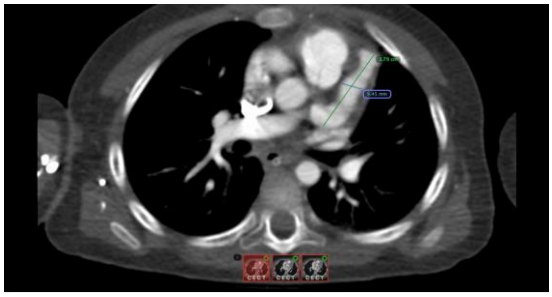

Figure 3. *Cardiac CT showing a left anterior descending aneurysm.*

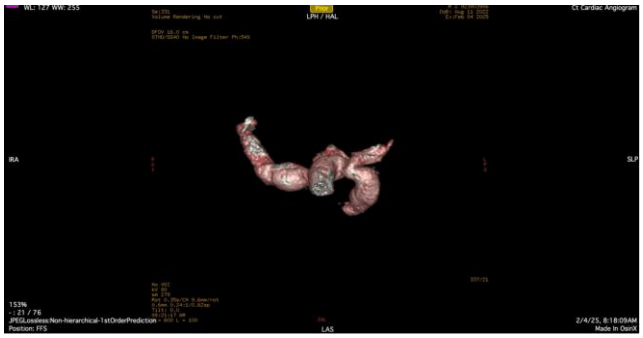

Figure 4. *Cardiac CT showing a right coronary artery aneurysm, left anterior descending aneurysm, and circumflex aneurysm.*

3. Discussion

This case highlights how overlapping clinical criteria can delay the timely treatment and diagnosis of KDSS. The dilemma of KD undoubtedly stems from the lack of pathognomonic tests. As a result, KD remains a diagnosis that relies on the clinical criteria outlined by the 2023 American Heart Association scientific statement [1]. These criteria include fever, unilateral lymphadenopathy, rash, bilateral nonexudative conjunctival injection, swelling and erythema of the hands and feet, and oropharyngeal findings including strawberry tongue and erythematous lips. A diagnosis can be made in the presence of at least 4 of these clinical findings, allowing for earlier identification than the commonly accepted 5 days of fever. Experienced clinicians may diagnose KD in the presence of just 3 days of fever [1].

The risk of KD is highest in infants, males, and patients of Asian descent [1]. This presentation met the AHA criteria for both early-phase incomplete KD and atypical KD. Incomplete KD poses a significant diagnostic challenge, often overlooked by clinicians. For incomplete cases with prolonged fever and only two to three of the defined features, performing a thorough laboratory evaluation and echocardiography is crucial. This case meets the criteria for incomplete KD, as the initial presentation of persistent fever, cervical lymphadenopathy, and mucosal changes, followed by rash and extremity swelling do not meet the full diagnostic criteria for KD. Here, the delay in diagnosis stemmed primarily from clinical criteria that closely overlapped with infectious etiologies, which resulted in several unnecessary antibiotic prescriptions before KD was suspected. Furthermore, the patient was positive for Mycoplasma pneumonia serology, which misled the diagnosis even further and delayed consideration of KD [5]. Elevated inflammatory markers that do not improve through antibiotic treatments should raise suspicion for coronary complications [6]. Early echocardiography is essential in such cases to detect coronary changes before they become irreversible. Studies show that infants below 6 months of age are at elevated risk for developing coronary artery aneurysms, with many presenting with a baseline echocardiogram with a Z-score of at least 2.5 [7]. However, a coronary artery aneurysm is a complication of KD and should not be used to diagnose it [1].

Atypical KD refers to patients who display clinical features outside of the established diagnostic criteria [1]. This patient presented several atypical features, including bilateral pleural effusions, oral ulcers, and hemodynamic instability, which are consistent with Kawasaki disease shock syndrome (KDSS). KDSS is a rare but severe form of the illness characterized by vasodilatory shock, hypotension, and poor perfusion [1, 8]. In this case, KDSS was initially misdiagnosed as septic shock due to their many shared features including fever, rash, elevated inflammatory markers, and hypotension [8, 9]. Recent reports have described similar diagnostic challenges, where patients were misdiagnosed with toxic shock syndrome prior to being confirmed with KDSS [9, 10].

To combat misdiagnosis, recent research highlights several biomarkers that may aid in reliably differentiating KDSS from sepsis, namely C-reactive protein, procalcitonin, and low albumin [3, 9, 11]. As presented in Table 1, CRP (134-20 mg/L) remained persistently elevated while procalcitonin dropped dramatically from 51.8 ng/L to 0.10 ng/L. While the decline in procalcitonin normally indicates an improvement in symptoms, the patient still experienced fever and inflammation. Low albumin, another recognized predictor of KDSS due to its involvement in capillary leak and inflammation, was also prevalent in this case. This along with the persistently high CRP and procalcitonin suggests that while the initial bacterial infection was resolved, a non-bacterial systemic inflammation consistent with KDSS remains. Additionally, this patient also presented with high D-dimer levels (2.35 (μg/mL FEU), which is shown by modern research to have a strong correlation with KDSS, making it another key indicator for differentiating KDSS from sepsis [3, 12].

Early diagnosis of KD within the first 10 days of illness is crucial to prevent long-term coronary abnormalities [1]. The current first-line standard of treatment recommends IVIG and high-dose aspirin. High-risk patients with IVIG treatment resistance should be carefully monitored and thoroughly evaluated. For refractory cases, a second dose of IVIG as well as second-line agents such as corticosteroids and Infliximab are recommended to reduce cardiac risk [13, 14]. However, a recent case report has described successful treatment of a giant aneurysm with Infliximab ad Enoxaparin [15]. In this case, IVIG and corticosteroids were administered empirically without a diagnosis of KD, which proved to be ineffective due to incomplete treatment planning and inadequate follow-up [16]. This highlights the importance of confirming a diagnosis early on, as it allows for thorough monitoring of coronary status along with sustained treatment for maximum benefit [16]. The importance of early recognition also lies in the increased risk of coronary artery complications over time, which persist even after treatment is initiated [12]. After diagnosis, the patient was discharged on antithrombotic therapy (enoxaparin and aspirin), consistent with recommendations for those at elevated thrombotic risk [6].

Patients suspected of having KD should receive regular

echocardiographic imaging regardless of initial normal results, as coronary artery changes may appear later as the disease progresses. These patients must be evaluated using risk stratification based on Z-scores and consistent follow-up [7]. Multi-specialty intervention including Cardiology, Rheumatology, and Infectious Diseases, is highly recommended for the precise diagnosis and treatment of patients with prolonged fever.

4. Conclusions

Early suspicion and multi-specialty intervention are essential for the diagnosis of KD before it leads to any long-term complications. Furthermore, incorporating independent predictors such as D-dimer, CRP, procalcitonin, and albumin into clinical practice is valuable for reducing the risk of misdiagnosis of KD with sepsis. Having a positive serology for an infectious agent does not exclude the diagnosis of KD.

Atypical KD should always be in the differential diagnosis for febrile patients who do not respond to antibiotic treatment. If KD is suspected, treatment should not be delayed, regardless of normal echocardiography results.

Abbreviations

KD	Kawasaki Disease
KDSS	Kawasaki Disease Shock Syndrome
IVIG	Intravenous Immunoglobulin
PICU	Pediatric Intensive Care Unit
CT	Computed Tomography
IV	Intravenous
Z-score	Standard Deviation Score
H1	Hospital 1
H2	Hospital 2
H3	Hospital 3
H4	Hospital 4

Author Contributions

Milad Sulaiman: Project administration, Writing – original draft.

Heba Ghazy: Investigation, Resources.

Yara Sulaiman: Writing – review & editing.

Alena Sulaiman: Visualization.

Salwa Gendi: Supervision, Validation.

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Data Availability Statement

Not applicable.

Conflicts of Interest

The authors declare no conflicts of interest.

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Biography

Milad Sulaiman is a first-year pediatric resident at Ain Shams university hospital who started off his journey in medicine in 2017 at Poznan University of Medical Sciences. He then did one year of internship in Jordan before pursuing in pediatrics.

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