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CASE REPORT

Kawasaki Disease in Somalia; A Case Report and Literature Review

Miski Abdullahi Roble[1], Amal Abdullahi Ali[1*], Aisha Mohamed Adan[1], Fardowsa Hassan Ahmed[1] and Mohamed Hussein Adam[2]

[1]Pediatric Department, Ministry of Health Recep Tayyip Erdogan University Training and Research Hospital: TC Saglik Bakanligi Recep Tayyip Erdogan Universitesi Egitim ve Arastirma Hastanesi Mogadishu, Somalia; [2]Department of Public Health, Faculty of Medicine and Health Sciences, SIMAD University, Mogadishu, Somalia

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Abstract: Kawasaki disease (KD) is a prevalent form of vasculitis in childhood, primarily affecting infants and young children. This leads to persistent cardiovascular complications resulting from coronary artery damage. Despite various hypotheses, the exact cause of this disease remains unknown. We present a case of Kawasaki disease in a one-year-old boy who exhibited one week history of high fever, rash, and conjunctivitis. The clinical manifestations progress, beginning with fever during the acute phase. Subsequently, non-purulent bilateral conjunctival redness developed, followed by changes in the mouth, such as red and cracked lips, along with a reddish tongue (strawberry tongue). This was achieved by one-sided cervical lymphadenopathy (with one node exceeding 1.5 cm in diameter), and within five days of fever onset, polymorphous rashes emerged. The patient was diagnosed with typical Kawasaki disease, and treatment was initiated with acetylsalicylic acid at a dose of 50 mg/kg/day and intravenous immunoglobulin (IVIG) at a dose of 2 g/kg.

The diagnostic criteria for Typical Kawasaki disease include both the Japanese working guidelines and the American Heart Association guidelines, which further categorize the disease as typical or atypical. Treatment with intravenous immunoglobulin (IVIG) and aspirin (ASA) during the acute phase significantly reduced the risk of coronary abnormalities by approximately 5%.

In summary, Kawasaki disease is infrequently documented in sub-Saharan countries. The uncommon presentation and rarity of the disease pose challenges for diagnosis, particularly in sub-Saharan Africa, notably Somalia, where no cases have been reported to date.

Keywords: Aspirin, Intravenous Immunoglobulin (IVIG), Vasculitis, Mucocutaneous lymph node syndrome, kawasaki disease.

1. INTRODUCTION

Kawasaki disease (KD) is a prevalent form of vasculitis in childhood, primarily affecting infants and young children. If not managed properly, it can result in lasting cardiovascular morbidity owing to coronary artery lesions. This condition is the primary cause of acquired heart disease among children in developed nations, with the highest incidence observed in Asia, where nearly 1 in 100 children in Japan experience the disease by the age of five [1, 2].

**Correspondence should be addressed to Amal Abdullahi Ali, Pediatric Department, Ministry of Health Recep Tayyip Erdogan University Training and Research Hospital: TC Saglik Bakanligi Recep Tayyip Erdogan Universitesi Egitim ve Arastirma Hastanesi Mogadishu, Somalia; E-mail: cafaaf199@gmail.com*

Kawasaki disease is predominantly prevalent in the Far East, particularly in countries such Japan, Korea, and China, but its occurrence is relatively rare in Africa, Europe, and the USA. In Africa, the first recorded cases emerged in South Africa in 1980, affecting Caucasian children, while the initial instance in an African child was documented on the Ivory Coast in 1981. While sporadic cases have been recorded in various African countries, primarily in North Africa and the West African region, comprehensive epidemiological data on Kawasaki disease in African nations are limited [4-6].

The cause of the disease remains unidentified, and various hypotheses have been proposed to elucidate its etiology. Epidemiological data indicate that the disease may be triggered by an infectious agent in individuals with a genetic predisposition [7].

According to the American Heart Association guidelines, the diagnosis of Classic Kawasaki Disease is established when there is a fever persisting for at least five days, with the onset day considered the first day of fever, along with the presence of at least four of the five principal clinical features. If there are at least four principal clinical features, particularly redness and swelling of the hands and feet, the diagnosis can be established after four days of fever (Table **1**). However, skilled clinicians well-versed in Kawasaki Disease may, in exceptional cases, diagnose a condition with 3 days of fever. Patients who do not exhibit complete clinical features of classic Kawasaki Disease are often assessed for incomplete KD. Diagnosis of Kawasaki Disease is typically confirmed when coronary artery abnormalities are detected [8].

Table 1. Diagnostic criteria of typical kawasaki disease outlined by the American Heart Association.

Classic KD is diagnosed in the presence of fever for at least 5 d (the day of fever onset is taken to be the first day of fever) together with at least 4 of the 5 following principal clinical features.	1. Erythema and cracking of lips, strawberry tongue, and/or erythema of oral and pharyngeal mucosa
	2. Bilateral bulbar conjunctival injection without exudate
	3. Rash: maculopapular, diffuse erythroderma, or erythema multiforme-like
	4. Erythema and edema of the hands and feet in acute phase and/or periungual desquamation in subacute phase
	5. Cervical lymphadenopathy (≥1.5 cm diameter), usually unilateral

While Kawasaki disease typically resolves on its own, if left untreated, it can lead to the development of coronary artery ectasia and/or aneurysms in 15-25% of children [9,10]. Children who present with complete Kawasaki Disease (manifesting fewer than four of the five clinical characteristics) face an elevated risk of delayed treatment and the subsequent development of coronary artery disease [11,12].

Administering intravenous immunoglobulin (IVIG) and aspirin (ASA) during the acute phase of the illness significantly reduced the risk of coronary abnormalities by approximately 5%. These abnormalities in the coronary arteries pose a potential risk for future conditions, such as ischemic heart disease, myocardial infarction, and sudden death [15].

The epidemiology of Kawasaki disease in developing countries remains largely undefined, with only a few reported cases, creating the perception that the condition is rare. Measles, a common ailment among children with Somalia, poses a close differential to Kawasaki disease, suggesting the possibility of misdiagnosis in some instances. Additionally, the low index of suspicion among physicians and healthcare workers in Somalia may have contributed to underdiagnosis. Notably, there are no previously documented case reports of Kawasaki disease in Somalia. In this report, we present a case of Kawasaki disease observed at our hospital that met the diagnostic criteria outlined by the American Heart Association [3].

2. CASE REPORT

A one-year-old male presented with a one-week history of high-grade fever, rash, and conjunctivitis. His medical records indicated that he had undergone a four-day course of ceftriaxone therapy at a different hospital. There were no prior health issues and no family history of the disease. During the physical examination, he displayed signs of fatigue, had a sickly appearance, and recorded an axillary temperature of 38.5°C. Bilateral conjunctival redness was observed, along with hyperemic and crusted lips, red and painful

swollen distal and proximal interphalangeal joints in both upper and lower limbs, as well as both knees. Erythema and edema were noted on the palms and soles. Respiratory and cardiovascular system examinations yielded normal results, and bilateral cervical lymph node enlargement was observed. Abdominal examination revealed no distention, organomegaly, with normal peristalsis.

The results of the blood analysis were as follows: white blood cell (WBC): 23,000/mm*3, hemoglobin (HB): 9.5 g/dl, platelet: 372,000/mm*3 c-reactive protein (CRP): 19mg/dl, aspartate aminotransferase (AST): 22 u/l, alanine aminotransferase (ALT): 34 u/l, gamma-glutamyl transferase (GGT): 32 u/l, total bilirubin: 0.13 mg/dl, direct bilirubin: 0.08 mg/ dl, and albumin: 2,9 g/dl. The abdominal ultrasonography (USG) examination results were normal, Echocardiogram was done which reveals a normal results.

The patient was diagnosed with typical Kawasaki disease according to the diagnostic criteria of typical kawasaki disease outlined by the American Heart Association (Table **1**), and treatment was initiated with acetylsalicylic acid at a dose of 50 mg/kg/day and intravenous immunoglobulin (IVIG) at a dose of 2g/kg for 2 days, after 48 the fever decreased, and the aspirin dose was decreased to 5mg/kg/day. The patient took aspirin for 3 weeks, after normalization of erythrocyte sedimentation rate (ESR), aspirin was stopped, and the patient was discharged.

Follow-up assessments demonstrated a reduction in conjunctivitis and arthritis. At the three-week follow up the patient exhibited an absence of fever and joint pain. Additionally, the results of the complete blood count, C-reactive protein (CRP) level, and erythrocyte sedimentation rate (ESR) was within the normal range. Echocardiographic examination yielded normal results.

3. DISCUSSION

Kawasaki Disease is a worldwide illness, and its occurrence rates differ, mainly corresponding to the racial makeup of populations in different countries. The highest reported incidence of Kawasaki Disease is in Japan, showing a consistent rise, reaching an annual rate of 308.0 per 100,000 children under 5 years of age in 2014 [20]. The second-highest reported incidence occurred in South Korea in 2014, 21 with a rate of 199.7 per 100,000 children under 5 years of age, while Taiwan ranked third with a rate of 82.8 per 100,000 in 2010 [22]. In nations where the population is predominantly non- Asian, the typical annual rate is 10–20 per 100,000 children under 5 years of age [23]. The male-to-female ratio among Kawasaki Disease patients is approximately 1.5:1 in nearly all countries [20,23], and severe cardiac complications associated with Kawasaki disease is notably more prevalent in males. The reason behind this male preponderance is not clear but appears to be the parallel patterns observed in numerous infectious diseases.

Kawasaki Disease exhibits a notable age distribution, with nearly all cases affecting children. Approximately 80% of these cases are found in children under 5 years of age, and approximately 50% occur in children under 2 years of age [20].

The cause of Kawasaki Disease is still not well understood, and there is a possibility of an unknown infectious agent, given the clinical and immunological similarities between Kawasaki disease and disorders mediated by staphylococcal and streptococcal superantigens [24].

Moreover, due to the elevated incidence of Kawasaki Disease in the Asian population, its persistence even after migration, and various genetic investigations, genome-wide association studies are recommended. These studies offer the advantage of identifying genes associated with the disease [25,26].

In many low- and middle-income countries, diagnosing Kawasaki Disease is challenging because the resemblance between prevalent infectious diseases and Kawasaki Disease. Diagnostic criteria play a crucial role in identifying Kawasaki Disease, incorporating the Japanese working guidelines and the American Heart Association guideline [27], which further categorizes the disease into typical and atypical forms. In the typical presentation of Kawasaki Disease, the clinical manifestations progress with the onset of fever during the acute phase. Subsequently, non-purulent bilateral conjunctival redness observed. Following this, changes manifest in the oral cavity, characterized by red and cracked lips and a reddish tongue (strawberry tongue) This is followed by one-sided cervical lymphadenopathy, where one node exceeds 1.5 cm in diameter, and then polymorphous rashes emerge within 5 days of the onset of fever.

Additionally, there were alterations in the extremities, including induration and/or erythema of the palms and soles of the feet. Around three weeks after onset, desquamation of the fingers and toes may occur. In the typical course of Kawasaki Disease, clinical characteristics progress with the onset of fever during the acute phase. Subsequently, non-purulent bilateral redness of the conjunctiva developed. Following this, alterations manifest in the oral cavity, including red and cracked lips and a reddish tongue (resembling a strawberry). This is followed by one-sided cervical lymphadenopathy, where one node exceeds 1.5 cm in diameter, and polymorphous rashes emerge within 5 days of fever onset. Changes in the extremities include induration and/or erythema of the palms and soles of the feet. Around three weeks after onset, desquamation of the fingers and toes may occur, marking the sub-acute phase. Kawasaki Disease may also present with other systemic manifestations, such as rheumatologic symptoms (joint pain and swelling), respiratory symptoms (cough and rhinorrhea), and gastrointestinal symptoms (vomiting, diarrhea, and abdominal pain) [27].

Kawasaki disease is categorized as atypical if there is fever persisting for ≥5 days with only ≥2 diagnostic clinical features, and this variant is more frequently observed at the extremes of childhood. In our case, the patient was diagnosed based on these criteria, which included a one-week high-grade fever along with bilateral non-supportive conjunctival injection, dry fissured lips, red tongue, edema, redness of extremities, maculopapular desquamating rash, and right-sided cervical lymphadenopathy.

Treatment of KD involves the use of IVIG and high-dose aspirin, Timely identification is essential, as it directs the clinician to commence swift and targeted treatment, effectively averting the occurrence of complication(s) [28-30]. Our patient was managed with aspirin(50mg/kg/day) and intravenous immunoglobulin (IVIG).

KD complications include carditis (myocarditis and pericarditis), congestive heart failure, coronary arteritis, and sudden death [29,32]. Fortunately, our patient did not exhibit coronary artery complications, as confirmed by echocardiography performed 3 weeks post treatment.

CONCLUSION

In summary, Kawasaki disease is infrequently documented in sub-Saharan countries. The uncommon presentation and rarity of the disease pose challenges for diagnosis, particularly in Sub-Saharan africa, notably Somalia, where no cases have been reported to date. It is recommended to maintain heightened awareness of Kawasaki disease in cases of prolonged febrile illnesses lasting more than 5 days with mucocutaneous manifestations. Healthcare professionals should familiarize themselves with this condition to effectively diagnose, treat, and prevent severe cardiovascular complications and mortalities. Additionally, there is a need to make intravenous immunoglobulin (IVIG) more accessible at a subsidized and affordable cost, particularly to benefit patients with Kawasaki disease and other autoimmune conditions where its use is indicated.

AUTHORS' CONTRIBUTIONS

The author confirms sole responsibility for the following: study conception and design, data collection, analysis and interpretation of results, and manuscript preparation.

ETHICS APPROVAL

Ethical approval was not required for the case study at Mogadishu Somali Turkey, Recep Tayyip Erdogan Training and Research Hospital.

CONSENT FOR PUBLICATION

Written informed consent was obtained from the patient's parents/legal guardian for publication and any accompanying images. A copy of the written consent is available for review by the Editor in Chief of this journal on request.

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CONFLICT OF INTEREST

The author confirms that this article's content has no conflict of interest.

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