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Case Report

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A case report on Kawasaki disease

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ABSTRACT

Kawasaki disease (KD) is a significant pediatric condition characterized by diverse clinical manifestations and potential cardiovascular complications. This case report examines a 4-year-old male child, third in birth order from a consanguineous marriage, presenting with a 14-day history of fever, strawberry tongue, peri-oral and peri-anal excoriation, and skin peeling on the upper limbs. Clinical examination revealed bilateral bulbar conjunctival injection and cervical lymphadenopathy. Laboratory findings indicated systemic inflammation, supporting the KD diagnosis according to established criteria. The child was treated with intravenous fluids (IVF), paracetamol, intravenous immunoglobulin (IVIG), aspirin, and topical calamine lotion. The treatment regimen led to favorable patient outcomes, although the consanguineous background suggests potential genetic predisposition. This case underscores the critical need for timely diagnosis and intervention in KD to prevent severe cardiac complications. It highlights the importance of adhering to diagnostic criteria and prompt therapeutic measures, contributing to improved understanding and management of KD in pediatric practice. Further research is recommended to explore genetic factors influencing KD.

Keywords: IVIG, Strawberry tongue, Consanguineous marriage

INTRODUCTION

Kawasaki disease (KD), initially identified in Japan in 1967 by Dr. Tomisaku Kawasaki, has emerged as the primary cause of acquired heart ailments among children in developed countries. It manifests as a brief but intense inflammation of blood vessels, mainly affecting the coronary arteries.[1]

Understanding the epidemiology of KD could provide valuable insights into its mysterious origins. Firstly, KD predominantly affects infants and young children, with 80% of cases occurring in those under 5 years old, although it can also appear in adolescence. This early onset hints at a potential association with the development of the immune system. Secondly, while KD has been observed globally and across all racial backgrounds, its incidence varies significantly among different populations.[2]

Etiology and pathogenesis

One proposed theory regarding the disease suggests that one or more widely distributed infectious agents trigger an abnormal immune response in individuals who are genetically susceptible. However, despite this theory, no specific causative agent has been identified.

KD is characterized by a systemic inflammation of blood vessels throughout the body, although the coronary arteries are predominantly affected. In the initial phase of presentation, the smooth muscle cells within the vessel walls demonstrate edematous dissociation. Additionally, there is swelling of the endothelial cells and

subendothelial edema, while the internal elastic lamina remains intact. Around seven to nine days after the onset of symptoms, there is an infiltration of neutrophils and lymphocytes, leading to the destruction of the internal elastic lamina and fibroblastic proliferation. Over the following weeks to months, the active inflammation gradually gives way to progressive fibrosis and scar tissue formation. This process may also involve arterial remodeling and revascularization, which can lead to the progressive narrowing of the affected vessels.[1]

Diagnosis

The standard method for diagnosing KD involves ruling out other possible illnesses and observing a fever lasting at least 5 days, along with the presence of at least four out of five main clinical features: bilateral non-pus-producing conjunctivitis, varied skin rashes, abnormalities in the lips or oral mucosa, abnormalities in the extremities, and swollen lymph nodes in the neck.[1]

Cardiac involvement is a significant aspect of KD during its acute phase and poses a risk for long-term complications. In this phase, various components of the cardiovascular system such as the pericardium, myocardium, endocardium, valves, and coronary arteries may be affected. Clinical examination of infants or children with KD typically reveals a hyperdynamic precordium, tachycardia, a gallop rhythm, and sometimes an innocent flow murmur due to concurrent factors like anemia, fever, and myocardial contractility issues secondary to myocarditis. Mitral regurgitation, if present, may manifest as a pansystolic regurgitation murmur. In severe cases, reduced myocardial function can lead to conditions like low cardiac output syndrome or shock.

Apart from cardiac manifestations, KD presents with various noncardiac symptoms. Irritability is common among affected children. Arthritis or joint pain often involving multiple joints, especially the knees and ankles, can arise within the first week of the illness. Gastrointestinal symptoms such as diarrhea, vomiting, and abdominal pain are reportedin about one third of patients. Hepatic enlargement and jaundice may also occur. Another notable noncardiac finding is acute acalculous distention of the gallbladder (hydrops), typicallyappearing in the initial two weeks of illness and detectable through abdominal ultrasound.[3]

In the acute phase of KD, treatment focuses on reducing inflammation in the coronary artery wall and preventing coronary thrombosis. Long-term therapy for individuals with coronary aneurysms aims to prevent myocardial ischemia or infarction.

Aspirin is commonly employed to alleviate inflammation and inhibit platelet aggregation in children with KD. However, it does not appear to reduce the incidence of coronary abnormalities in patients.[4]

IVIG

The effectiveness of IVIG administered during the acute phaseof KD in reducing the occurrence of coronary artery abnormalities has been well documented. The mechanism through which IVIG operates remains uncertain, although various theories have been proposed. These include actions such as cross-linking of FcγII andFcγIII receptors on macrophages, induction of immune inhibitory receptors, inhibition of interaction between endothelial cells and natural killer cells, enhancement of T-cell suppressor activity, suppression of antibody production, neutralization of bacterial superantigens or othercausative agents, and provision of anti-idiotypic antibodies.[4-9]

CASE REPORT

A 4-year-old male child 3rd in birth order born out of 3rd degree contagious marriage brought with the complaints of fever for 14 days, strawberry tongue (Figure 1), peri oral excoriation for 8 days, peri anal excoriation for 5 days, peeling of skin on both upper limb for 6 days. No past history of similar illness in the past. On examination child is moderate active, heart rate, respiratoryrate-24/min, pallor (+), icterus (-), cyanosis (-), koilonychia (-), clubbing (-), lymphadenopathy (+). B/L upper jugular lymph nodes measuring 2 cm, nontender.

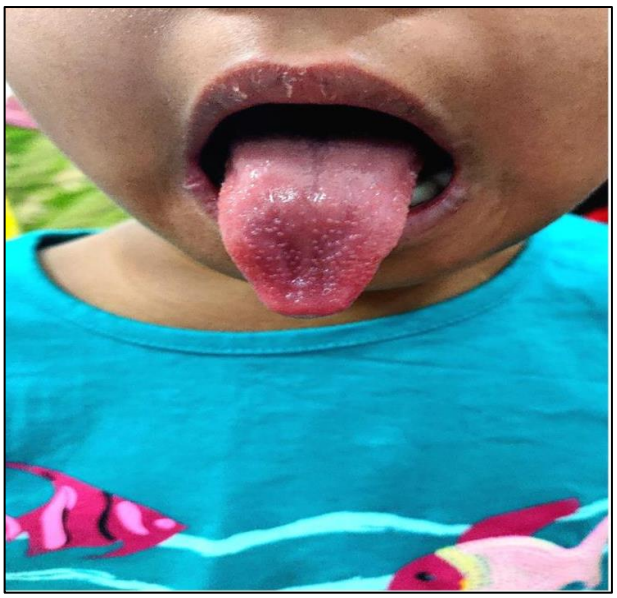

Figure 1: Strawberry tongue.

The investigations include echocardiogram in which no coronary artery aneurysm or artery dilations are found. CRP was noted as 24 mg/dl, WBC-21000 cells, platelets-5.2l, neutrophils-60.8%, HCT-29.5%.

KD presents a diagnostic and therapeutic challenge for pediatricians worldwide due to its diverse clinical manifestations and potential complications, especially involving the cardiovascular system.[3]

Table 1: Diagnostic criteria for KD.

Diagnostic criteria	Present or absent
Fever>5 days	Present
B/L bulbar conjunctival injection (non-purulent)	Present
Oral mucosal membrane changes (strawberry tongue) (fissured lips)	Present
Polymorphous rash	Absent
Cervical lymphadenopathy (LN>1.5 cm)	Present

The diagnostic criteria for KD, as outlined in (Table 1), were met by the patient, confirming the diagnosis. Fever lasting more than 5 days, bilateral bulbar conjunctival injection, oral mucosal changes (strawberry tongue), and cervical lymphadenopathy were all present. Although a polymorphous rash was absent, its variable presentation does not exclude the diagnosis.

Based on all above aspects the pediatricians diagnosed case as KD.

The patient received a comprehensive treatment regimen, as detailed in (Table 2). IVF were administered to maintain hydration, while paracetamol helped alleviate feverand discomfort. IVIG therapy, a cornerstone in KD management, was initiated promptly to reduce the risk of coronary artery abnormalities. Additionally, aspirin therapy was commenced to mitigate inflammation and thrombosis, while calamine lotion provided topical relief for skin manifestations.

Table 2: Treatment.

Treatment given	Doses
IVF DNS	20 ml/hour
Injection paracetamol	10 mg/kg/dose (14.7 ml/iv/QID)
Injection IVIG	2 gm/kg/day over 10-12 hour (3 days)
Tablet aspirin	3 mg/kg/day
Calamine lotion	Topical use

Follow-up

The patient was monitored closely over the next several weeks. At the 2-week follow-up, feverand mucocutaneous symptoms had resolved, and the patient was in good general health. A repeat echocardiogram at 4 weeks confirmed the absence of coronary artery abnormalities. Continued follow-up at 3 months showed no recurrence of symptoms and normal cardiovascular findings.

DISCUSSION

KD is an acute, self-limited vasculitis of unknown etiology that predominantly affects children under 5 years of age. First described by Dr. Tomisaku Kawasaki in in 1967.[1]

KD has become the leading cause of acquired heart disease in children in developed countries, characterized by a brief but intense inflammation of blood vessels, predominantly affecting the coronary arteries.[1] The disease presents with various noncardiac symptoms. Affected children commonly exhibit irritability. Arthritis or joint pain, particularly in the knees and ankles, may occur within the first week of the illness. Approximately one-third of patients experience gastrointestinal symptoms such as diarrhea, vomiting, and abdominal pain. Additionally, hepatic enlargement and jaundice can develop. Acute acalculous distention of the gallbladder (hydrops), usually detectable through abdominal ultrasound, oftenappears within the first two weeks of the illness.[3] Aspirin is frequently used to reduce inflammation and prevent platelet aggregation in children with KD.[4] This caseinvolves a 4-year-old male presenting with a 14-day history of fever, strawberry tongue, perioral and perianal excoriation, and skin peeling on the upper limbs. The patient met severaldiagnostic criteria for KD, including prolonged fever, bilateral conjunctival injection, oral mucosal changes, and cervical lymphadenopathy. Laboratory findings supported the diagnosis, showing elevated CRP, WBC, platelets, and neutrophils. An echocardiogram revealed no coronary artery abnormalities. In a study by McCrindle et al a 5-year-old girl presentedwith fever for 7 days, conjunctival injection, strawberry tongue, rash, and lymphadenopathy.[10] Unlike our case, this patient developed coronary artery aneurysms despite prompt IVIG treatment. This highlights the variability in disease severity and the importance of early and aggressive treatment to prevent cardiac complications. Another report by Singh et al described a 3-year-old boy with incomplete KD, presenting primarily with fever, irritability, and gastrointestinal symptoms, but lacking several classic KD signs such as conjunctival injection and lymphadenopathy.[11] This contrasts with our case, which exhibited clear mucocutaneous symptoms and lymphadenopathy. The variability in presentations underscores the diagnostic challenge of KD, especially in incomplete cases. A case described by Nakamura et al involved a 4-year-old boy with KD who presented with neurological symptoms including seizures and irritability, in addition to the typical KD symptoms.[12] Our patient did not exhibit neurological symptoms, which are rare but possible in KD, further illustrating the disease's diverse clinical manifestations. Our patient received a comprehensive treatment regimen including IVF, paracetamol, IVIG, aspirin, and calamine lotion. This aligns with the standard treatment protocols aimed at reducing inflammation, preventing coronary artery complications, and providing symptomatic relief. IVIG and aspirin are particularly crucial in KD management. Studies have shown that IVIG administered within thefirst 10 days of illness significantly reduces the risk of coronary artery abnormalities.[13] Thiscase underscores the

importance of recognizing the diverse clinical presentations of KD. Prompt diagnosis and treatment are essential to mitigate the risk of serious complications, particularly coronary artery aneurysms. Comparison with other case reports highlights the variability in KD manifestations and outcomes, emphasizing the need for heightened clinical awareness and early intervention.

CONCLUSION

In conclusion, the case study of the 4-year-old boy with KD highlights the complexities and challenges inherent in diagnosing and managing this condition. Despite its rarity, KD demands swift recognition and intervention to mitigate potentially severe cardiac complications. This case underscores the importance of thorough clinical assessment, adherence to diagnostic criteria, and timely initiation of treatment, including IV immunoglobulin and aspirin therapy. By emphasizing the significance of early intervention, this case report contributes to our understanding of KD and underscores the importance of continued vigilance in pediatric practice. Further research and clinical awareness are imperative to enhance diagnosis, treatment, and long-term outcomes for children affectedby this condition.

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