Typical Kawasaki Disease in a Pediatric Patient with Pre-existing Atopic Disorder

Rimisha Thomas*, Arshamol Varghese, Aaja Alosious, Tany Maritta George, Jobin Kunjumon Vilapurathu

Department of Pharmacy Practice, Nirmala College of Pharmacy, Muvattupuzha, Kerala, INDIA.

ABSTRACT

Kawasaki disease (KD), is an acute febrile illness that is known to be a leading cause of cardiovascular morbidity, if not adequately treated. The association between preexisting allergic diseases and Kawasaki disease has been the subject of mounting clinical interest. We herein report a case of typical Kawasaki syndrome in a seven-year-old child with preexisting atopic disorder. Once the diagnosis of Kawasaki was established, intravenous immunoglobulin and oral aspirin were initiated at a dose of 2 g/kg body weight and 100 mg/kg/day respectively. Response to treatment was remarkable and the he was discharged on the fifth day post admission with aspirin to be continued at maintenance dose. This report highlights the importance of prompt diagnosis and management of Kawasaki and the need for future investigations on the risk of this condition in children with history of immune disequilibrium.

Keywords: Kawasaki disease, Atopic disorder, Vasculitis, Immunoglobulin.

INTRODUCTION

Kawasaki disease (KD) is a self-limiting, systemic vasculitis disease with a predilection to coronary artery involvement.[1] It is frequent in children under 5 years old, though children up to the age of 13 can sometimes be affected.[2] The first case was reported in an African child in Ivory Coast in 1981.[3]

Accumulating evidence has suggested the possibility of a reciprocal link between KD and allergic diseases. While KD is a complex condition, an underlying allergic immune response plays a significant role and hence children with a history of immune disequilibrium, are more likely to get affected.[4]

This case report sheds light onto the positive influence of pre-existing atopic diseases on the development of KD and the fact that they may share common early-life determinants.

CASE SUMMARY

A 7-year-old boy, with a past medical history of asthma and eczema since the age of one presented to the pediatric ICU with chief complaints of non-resolving high-grade fever of 2 weeks duration, followed by skin peeling and scaly lesions over extremities, face and genital area. He was referred from a local hospital for evaluation of symptoms that persisted despite antimicrobial therapy. The child had a history of MRSA positive otitis media 4 months back and was fully immunized.

Upon physical examination, the child was found to have a typical strawberry tongue, non-purulent conjunctivitis and cervical lymph node enlargement. He was febrile to 38.9°Celsius (102.02°Farenheit). Preliminary investigations indicated hemoglobin 10.6 g/dl, total leucocyte count 20470/mm^3 with 65% polymorphs, 23% eosinophils, 11% lymphocytes and 1% monocytes. Thrombocytosis was evident with an initial platelet count of 594000 cells/mm3 and

DOI: 10.5530/ijopp.15.4.60

Address for Correspondence:
Ms. Rimisha Thomas,
Nirmala College of Pharmacy, Muvattupuzha-522503, Andhra Pradesh, INDIA.
Email id: rimishathomasa1@gmail.com

www.ijopp.org

the ESR and CRP levels confirmed a significant state of inflammation. Blood biochemistry indicated normal serum electrolytes, renal and liver function. Blood, urine and CSF cultures were sterile and viral studies were negative, ruling out an infectious etiology. Upon cardiology consultation, an ECHO and ECG were taken and were within normal limits with the exception of sinus tachycardia. Based on the overall clinical picture and the current criteria, a diagnosis of KD was confirmed.

The patient was started on intravenous immunoglobulin (IVIG) at 2 g/kg body weight over 12 hr and oral aspirin at a dose of 100 mg/kg/day divided every six hours while febrile. A therapeutic challenge in this case was the pre-existing eczema in this patient as there were previous reports of extensive eczematous reactions following immunoglobulin administration. But the patient tolerated the therapy and showed remarkable clinical response with subsidence of fever in 24 hr after IVIG. On the subsequent days, the skin lesions started resolving, inflammatory markers and hematological parameters were normalized and the child was shifted to room on the third day. He was discharged on the fifth day post admission at a maintenance dose of aspirin 4 mg/kg/day to be continued for 6 weeks.

DISCUSSION

KD is an acute systemic vasculitis of small and medium sized blood vessels.[1] The innate as well as the acquired immune system holds a significant role in the pathophysiology of KD. The disease usually occurs as a result of an immunologic response that is triggered by different microbial agents, leading to subsequent cytokine cascade stimulation and activation of endothelial cells[1,5]

The diagnosis of KD is based on the clinical symptoms. (Table 1).[5-7] Fever is the mandatory criterion and four of the remaining five criteria are required to confirm the diagnosis. Being an inflammatory process, elevation of ESR, CRP and white cell count should be considered.[8] Echocardiographic findings may be helpful in evaluating suspected cases and differentiating KD from other conditions with similar presentation.[5,7,8]

Though the key to prevent the potentially dangerous cardiac involvement is not known, resolution of systemic inflammation as soon as possible is the target of the treatment.[5] The mainstay of therapy is the immediate administration of immunoglobulin 2g/kg once and aspirin at a dose of 50-100 mg/kg/day. A second dose of immunoglobulin 2g/kg should be considered for individuals who are not responsive to the dual therapy.

Table 1: Diagnostic criteria for classic Kawasaki disease.[7]

Fever for at least five days with at least four of five principal clinical features:

- Oral cavity and lip changes: Strawberry tongue cracked and erythematous lips
- Polymorphous rash: Erythema multiforme–like, maculopapular, or scarletiniform rash, involving trunk, extremities and perineal regions
- Non-purulent bilateral conjunctivitis
- Extremity changes (such as erythema of the feet and hands, desquamation of the toes and hands in weeks 2 and 3)
- Unilateral cervical lymphadenopathy (> 1.5 cm in diameter)

Alternative diagnostic criteria for classic Kawasaki:

- Fever for at least 5 days and two or three principal clinical features or coronary artery abnormalities on transthoracic echocardiography

The dose of aspirin needs to be reduced to 3–5 mg/kg once the fever and inflammation subsides.[6-8]

Some studies have suggested that there exists a reciprocal link between KD and atopic disorders. This is evident from the fact that the occurrence of KD is more frequent in children at risk of immune disequilibrium. It initially presents as an abnormal inflammatory response and eventually progresses with more allergic manifestations.[4,9,10] In this case, the child had a known history of asthma and eczema for a long time, which might have been a predisposing factor for the development of KD. Hence, there is a growing interest in deciphering this potential link between atopic disorders and KD, given the increasing epidemiologic evidence for the same.

CONCLUSION

Our case is notable as it suggests the possibility of a higher risk of KD in children with history of atopic disorders. Further studies that focus on this aspect are warranted to establish the link between these conditions. This can aid in prompt diagnosis and initiation of therapy so as to prevent the potential cardiovascular sequelae of KD.

CONFLICT OF INTEREST

The authors declare that there is no conflict of interest.

ABBREVIATIONS

KD: Kawasaki Disease; **ICU:** Intensive Care Unit; **MRSA:** Methicillin Resistant *Staphylococcus Aureus*; **ESR:** Erythrocyte Sedimentation Rate; **CRP:** C Reactive

Protein; **CSF:** Cerebro Spinal Fluid; **ECG:** Electro Cardio Gram; **IVIG:** Intra Venous Immuno Globulin.

REFERENCES

1. Ren Y, Zhang C, Xu X, Yin Y. A case report of atypical Kawasaki disease presented with severe elevated transaminases and literature review. BMC Infect Dis. 2021;21(1):415. doi: 10.1186/s12879-021-06101-y, PMID 33947336.
2. Stanfordchildren's. Health Communications [internet]; 2022. Stanfordchildrens.org[citedFeb232022]. Available from: https://www.stanfordchildrens.org/en/topic/default?id=kawasaki-disease-in-children-90-P01801.
3. Noorani M, Lakhani N. Kawasaki disease: Two case reports from the Aga Khan Hospital, Dar es Salaam, Tanzania. BMC Pediatr. 2018;18(1). doi: 10.1186/s12887-018-1306-5.
4. Huang PY, Huang YH, Guo MM, Chang LS, Kuo HC. Kawasaki disease and allergicdiseases. Front Pediatr. 2020;8:614386. doi: 10.3389/fped.2020.614386, PMID 33490002.
5. Leonardi S, Barone P, Gravina G, Parisi GF, Di Stefano V, Sciacca P, *et al*. Severe Kawasaki disease in a 3-month-old patient: A case report. BMC Res Notes. 2013;6(1):500. doi: 10.1186/1756-0500-6-500, PMID 24294914.
6. Kontopoulou T, Kontopoulos DG, Vaidakis E, Mousoulis GP. Adult Kawasaki disease in a European patient: A case report and review of the literature. J Med Case Rep. 2015;9(1):75. doi: 10.1186/s13256-015-0516-9, PMID 25890055.
7. Saguil A, Fargo M, Grogan S. Diagnosis and management of Kawasaki disease[internet].org. Am Fam Physician.2022;91(6):365-71. PMID 25822554.
8. Eleftheriou D, Levin M, Shingadia D, Tulloh R, Klein NJ, Brogan PA. Management of Kawasaki disease. Arch Dis Child. 2014;99(1):74-83. doi: 10.1136/archdischild-2012-302841, PMID 24162006.
9. Wei CC, Lin CL, Kao CH, Liao YH, Shen TC, Tsai JD, *et al*. Increased risk of Kawasaki disease in children with common allergic diseases. Ann Epidemiol. 2014;24(5):340-3. doi: 10.1016/j.annepidem.2014.02.003, PMID 24613197.
10. Liew WK, Lim CW, Tan TH, Wong KY, Tai BC, Quek SC, *et al*. The effect of Kawasaki disease on childhood allergies - a sibling control study. Pediatr Allergy Immunol. 2011;22(5):488-93. doi: 10.1111/j.1399-3038.2011.01149.x, PMID 21443753.