

Kawasaki disease: A case report

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Abstract

Kawasaki disease is a vasculitis of medium sized arteries which is diagnosed by exclusion and by use of diagnostic criteria. It needs a high index of suspicion to diagnose and should be suspected in any child with history of fever for more than a week with rash and lymphadenopathy. Such patients should be treated with intravenous immunoglobulin and Aspirin to prevent complications. Here we report a case of Kawasaki Disease who presented with prolonged fever, rash and lymphadenopathy and made an uneventful recovery.

Key words: Kawasaki disease, Vasculitis, Rash

Introduction

Kawasaki disease is a vasculitis syndrome mainly affecting children younger than 5 years of age. It is also called as mucocutaneous lymph node syndrome. This condition was first described by Dr. Tomisaku Kawasaki in 1967¹.

The cause of Kawasaki Disease is unknown. Its incidence varies with geographical location and race. Japanese population has incidence of 80-100 per 100,000 children below five years of age. In the United States its incidence is about 8 per 100,000 under five children whereas in European under 5 children is even lower ranging from 3-6^{2, 3, 4}. There are very few cases of Kawasaki disease reported in Nepal. Here we are discussing a rare case of Kawasaki disease.

Case report

A three year old female child from Panauti, Kavre presented with history of high grade fever for 7 days, swelling in neck for five days and rash all over the body for three days. She was investigated and treated by local practitioner for initial three days and subsequently got admitted in local hospital and treated without improvement for which she attended outpatient department of Kathmandu Medical College Teaching Hospital (KMCTH).

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Examination at KMCTH revealed fever with generalized polymorphous erythematous maculopapular rash, significant tender lymphadenopathy over posterior triangle of left side of the neck, cracked lips with strawberry tongue, enlarged bilateral tonsils with congestion of faucial pillars, uvula and posterior pharyngeal wall, a few enanths in oral mucosa, bilateral nonexudative painless conjunctivitis, desquamation over peri-anal region and also over hands with swelling. She also developed arthritis after second day of admission.

Investigations showed total WBC count of 17,300/mm³ with 64% neutrophils, elevated ESR (40 mm at the end of 1 hour) and positive C-reactive protein, and elevated platelet count (3,24,000/mm³). Her antistreptolysin-O titres were normal. Anti nuclear antibody was negative. Throat swab culture was sterile. Widal test was negative. Liver enzymes were normal except for total serum albumin which was 2.3gm/dL. Urine routine examination and culture was negative. Her electrocardiogram and echocardiograph were also normal. This diagnosis was made as per the Japanese worker's criteria⁵.

She was started with Aspirin (100 mg/kg/day). Her symptoms subsided within 24 hours of starting aspirin and she was discharged in the same dosage for next 7 days followed by 5 mg/kg/day for the next 6 weeks. On a follow up after one week she was asymptomatic except for exfoliation over her hands and feet. Repeat blood investigations, after one week of aspirin, revealed normal WBC counts and ESR. However the platelet count was still high (4,00,000/mm³).



Fig. 1: Pictures showing cracked lips, exanthematous rash and periungual exfoliation

Discussion

Kawasaki disease is the second most common cause of vasculitis in children after Henoch Schonlein purpura^{2,3,4,6}. Early diagnosis and treatment of KD is of utmost importance because of dreadful complications during acute illness which include myocarditis, pericarditis, valvular heart disease and coronary arteritis. Studies have shown that coronary aneurysm can occur in 20% if left untreated⁷. The diagnosis of KD is basically clinical and it is a diagnosis of exclusion. There are diagnostic criteria for diagnosis of KD. In our case we used Japanese worker's criteria⁵. Our patient had all the criteria for the diagnosis of KD. One of the KD diagnostic guidelines includes fever lasting for at least 5 days along with presence of at least 4 of the following 5 signs— (1) bilateral conjunctival injection, (2) generally non purulent, changes in mucosa of oropharynx, including injected oropharynx, injected and dry fissured lips, strawberry tongue, (3) changes of peripheral extremities such as edema and/or erythema of hands or feet in acute phase, (4) rash, primarily truncal, polymorphous but non vesicular, (5) cervical lymphadenopathy (more than or equal to 1.5 cm in diameter), usually unilateral. Illness is not explained by other known disease process⁸.

In Kawasaki disease prompt treatment is important to prevent coronary aneurysm. Studies have shown that immediate treatment reduces the risk of such complications⁹. The drug of choice in such situation will be a single dose of intravenous immunoglobulin (IVIG) 2 g/kg and 100 mg/kg per day for 14 days followed by 3–5 mg/kg per day for 6 weeks¹⁰. Because of economic constraints parents in developing countries cannot afford IVIG. There are reports of aneurysms being detected within three days of onset of illness but more commonly these occur 10 days to four weeks after the onset of symptoms⁷.

Conclusion

Kawasaki disease is a rare but dreadful vasculitis in child. Any child with fever of more than a week should be investigated for KD and one should have a high index of suspicion in diagnosing it. Prompt diagnosis and early institution of intravenous immunoglobulin and aspirin is of utmost importance in preventing coronary complications.

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