Case Report

KAWASAKI'S DISEASE

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ABSTRACT

Kawasaki Disease is a medium vessel vasculitis. It occurs commonly in children under 5 years of age and has a propensity of affecting the coronary arteries. The etiology and the pathogenesis of this disease are not known yet, but the major contributors are genetic factors, immune reactions and infections. The diagnosis of Kawasaki disease is based on clinical manifestations that appear on a temporal sequence. This disease is self-limiting in most cases, but it can lead to severe complications such as coronary artery aneurysms or thrombo-embolic events in up to 25% of the cases. Once the disease has been acquired prompt recognition and early treatment can be a lifesaver for the patients. Intravenous immune globulin (IVIG) and aspirin remained the treatment of choice for such patients.

Key Words: Coronary aneurysms, Lymph node, Vasculitis

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INTRODUCTION

Kawasaki disease (KD) is also known as "Mucocutaneous lymph node syndrome" as one of its clinical presentations is the swelling of lymph nodes and mucous membranes inside the mouth, nose, eyes, and throat. The incidence of KD in children under five years of age is reported in the United States as 25/100,000 children and has varied to approximately 250/100,000 children in Japan.

Kawasaki disease presents with acute febrile illness associated with systemic vasculitis. It affects young children mostly under 5 years of age and can be complicated by the development of, coronary artery aneurysms and other long-term cardiovascular sequels. An increased risk of developing Kawasaki disease is observed in Asian male children under 5 years of age.

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Japanese and Korean population has shown a high incidence of this disease as compared with global statistics. Seasonal increases in incidence have been observed in winter in North America.

CASE DESCRIPTION

A 6-year-old girl who was previously healthy presented in the Pediatric OPD of Farooq Hospital on 27th May, 22 with a 7 days history of high-grade fever and a 2-day history of arthralgia.

On the second day of her illness, she had left-sided cervical lymphadenopathy. A day after, a maculopapular rash appeared on her neck and trunk while sparing her extremities, which spontaneously resolved within two days. In addition, the patient had a bilateral conjunctival infection and dried, cracked lips, and swollen red tongue. During this period, she was given antibiotics, antivirals, and antipyretics; however, her symptoms were not resolved. The arthralgias were initiated in her ankle joint and then advanced in an ascending manner involving all the large and small joints impeding her ability to move. There was no significant history of dyspnea, orthopnea, peroxymal norturnal dyspnea

(PND), chest pain, or palpitations. She was referred to us for the evaluation of prolonged fever. Upon examination, the child was irritable, but alert, weighing 25kg and height of 124 cm (both above the 90th centile). Her vitals were in the normal range. She had bilateral congestion of the eyes without any discharge (conjunctivitis), along with dried lips and swollen tongue (strawberry tongue). The cervical node of 1.5 x 1.5 cm was also palpable unilaterally. A maculopapular eruption of rash on the neck and trunk was also observed. There were no positive findings of edema and erythema of the hand and feet and no perineal and periungual desquamation in this child.

On systemic review, no visceromegaly was observed, bowel sounds were audible, first and second Heart sounds were audible with no added sounds, her chest was bilaterally clear, and reflexes were elicited.

Her Laboratory investigations further revealed normochromic normocytic anemia with leukocytosis and neutrophilia. Her ESR and CRP levels were raised to 100min/1st hour and 288.5 mg/L respectively. The autoimmune antibodies came negative. Other labs were also insignificant. Upon arrival, the ECG showed sinus tachycardia that subsequently disappeared. Her echocardiography indicated an aneurysm in the left coronary artery.

After admitting to the hospital, she was treated with IVIG along with aspirin and clopidogrel for 2 days and after 48 hours the patient was clinically improved with major declines in her ESR and CRP levels i.e., 98min/1st hand 75mg/L respectively. Under observation for 5 days her examination findings became insignificant and she was discharged on aspirin and clopidogrel with the follow-up of repeat baselines and echocardiography.

DISCUSSION

Kawasaki disease (KD) is a multisystem febrile vasculitic disease of children described for the first time in 1974 by Kawasaki et al.[1] Along with rheumatic carditis Kawasaki disease is the leading cause of acquired cardiac disease in children.[2] A lot of geographical, seasonal, and ethnic variation is seen in KD.[3] Thus, several research articles suggest different underlying etiologies like viruses, genetic predispositions, and immune variations.[4] Till today autoimmune basis of KD has the most strong evidence for underlying etiological factors[5] supported by a high level of eosinophil and Interleukin5 at the site of lesions.

The major persistent impact of KD is on the heart of children in the form of coronary artery lesions (CAL). It ranges from fistula and dilatation to aneurysm formation in coronaries. Lesions may be transient for 6-8 weeks to permanent in nature depending upon the timing and appropriateness of treatment offered to the baby.[6]

Diagnosis of KD is still today is clinical one as suggested by the Japan circulation society (JCS) and the American heart society (AHA). Fever lasting more than 5 days is essential for diagnosis while conjunctivitis, cervical lymphadenopathy, oral mucosal lesions, and edema of hands and feet or peeling rash are other requisite for diagnosis.[7] In some patients, all 5 criteria are not fulfilled but the urgency of treatment demands labeling of diagnosis to avoid the impending complication of CAL. Such patients are labeled as Atypical KD or Incomplete KD.[8] AHA in such patients also recommends support of supplementary laboratory criteria for establishing the diagnosis. This includes serum albumin, platelet count, ALT, urine white cells, and anemia.[9] Fulfilling all essential 5 criteria is also not necessary if coronary changes are detectable at the time of admission.[10]

Our patient M age 6 years admitted with a high fever of 7 days and difficulty in moving about at the time of admission. She had eye congestion and cervical lymphadenopathy. Because of missing essential criteria help of echocardiography was taken which revealed aneurysmal dilatation of the left coronary artery. 100 mm ESR and 288 mg/L CRP supplementary supported for diagnosis. The introduction of treatment IVIG and aspirin

not only relieved her fever and body aches and pains but an indicator of inflammation also returned to normal.

Treatment of KD is debated regularly by different guidelines regarding the choice of drugs, time of therapy, and duration of therapy.[11] IVIG is the mainstay of treatment to avoid CAL. Earlier immunoglobulin (2gm/kg) was given to lessen the chance of developing coronary complications. Aspirin is very effective in subsiding inflammation and fever (70-100 mg/kg/day). Duration of aspirin therapy is variable but the dose should be reduced to a minimum as the fever and inflammatory markers reduce to normal.

CONCLUSION

Kawasaki disease is common in Asian Population. It affects male gender and children under five years of age.

AUTHOR'S CONTRIBUTION

NA: Critical review
MBA: Introduction writing
WZ: Discussion writing
MA: Introduction writing
ASA: Supervision and conception of idea

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