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A CASE ON KAWASAKI SYNDROME IT'S DIAGNOSIS AND MANAGEMENT

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ABSTRACT: Kawasaki Syndrome (KS) can be a widespread vasculitis affecting many systems, and some of the other recognized complications are discussed, including those affecting the skin, nervous system, gastrointestinal tract, musculo-skeletal system, kidneys, lungs, eyes and haematological effects. There is no specific diagnostic test or pathognomonic clinical features; those previously mentioned clinical criteria have been established to assist physicians in diagnosing Kawasaki disease (KD). We are reporting a case of 1 year 3 months child who has successfully relieved by the IgG and Aspirin therapy. A greater awareness on the part of paediatricians is now contributing to increased recognition of this entity even in India. We suggest that physicians should be cognizant of the fact that they must individualize every patient's management to the best of their knowledge and judgment, rather than merely going by the guidelines.

INTRODUCTION: Kawasaki disease (mucocutaneous lymph node syndrome) was first described by Dr. Tomisaku Kawasaki in 1967. Since then, more than 100,000 cases have been reported worldwide, the majority occurring in Japan. The disease predominantly affects children under 5 years of age and it is now the commonest cause of acquired heart disease in children in developed countries. Occasionally Kawasaki disease is reported in neonates, adolescents and adults^{1, 2, 3}. Kawasaki disease is a condition that causes inflammation in the walls of medium-sized arteries throughout the body, including the coronary arteries, which supply blood to the heart muscle⁴.

The aetiology of Kawasaki disease is unknown, but epidemiologic data suggest that the mechanism involves an immune response to a predisposing infection in genetically predisposed people.⁵ The diagnosis is based on the presence of at least five of the following six clinical features like A) persistent fever, B) polymorphous rash, C) characteristic changes in the extremities (erythema of the hands and feet followed by desquamation of the fingers and toes), D) bilateral conjunctivitis, E) cervical lymphadenopathy, F) pharyngeal changes including 'strawberry' tongue (prominent lingual papillae); dry, erythematous or cracked lips, and erythema of the oropharyngeal mucosa⁶.

Histologically, KD is characterized by vasculitis involving the muscular arteries. Endothelial cells express markers of activation (HLA class II antigens, Endothelial Leucocyte Adhesion Molecule [ELAM1] and Intercellular Adhesion Molecule [ICAM1]). As a result, levels of proinflammatory cytokines interleukin (IL)-1, IL-6, Tumour Necrosis Factor (TNF)-a, neopterin and g-

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interferon (IFN) will be increased that characterize the acute phase of this illness. High levels of serum IgE also noticed which represents the allergic nature of the disease. The combined effect of all this may increase the thrombogenicity of the vesselwall².

Serious complications of KS include coronary artery dilatations and aneurysms, coronary artery thrombosis or stenosis, myocardial infarction, and sudden death^{6,7}.

This case illustrates the Kawasaki Syndrome in a young child treated with IgG and Aspirin without any complications.

Case Report:

A 1 year 3 months old female child weighing 9.4kg was admitted in a tertiary care hospital with complaints of intermittent fever for 5 days not associated with rigors/chills, redness in both eyes, strawberry tongue and decreased food intake. Her immunisation was complete and up to date and she had not received any vaccination prior to admission. On examination she was very irritable with a temperature of 38.8°C and heart rate was found to be 126/min. Systemic examination was unremarkable while the eye represented with bilateral conjunctivitis and lips are in reddish colour. Abnormal laboratory findings revealed low Haemoglobin 9.7g/dl (Normal range-12-15g/dl), Raised white cell count $18.1 \times 10^9/L$ (Normal range- $4.3-11 \times 10^9$), with a predominant thrombocytosis, platelet count of $518 \times 10^3/L$ (Normal range $150-400 \times 10^3 U/L$), C-reactive protein (CRP) and Erythrocyte Sedimentation Rate were elevated 124mg/dl and 101 mm/hr (Normal range 0-11mg/dl and 0-20mm/hr) respectively. The chest X-ray was normal. Blood, urine and throat swab cultures were negative.

Based on the symptoms and clinical findings, the physician diagnosed as Kawasaki disease and the child was treated with Intravenous immunoglobulin (IVIG -2 gm/kg), Aspirin (150 mg/kg/day) QID and Nexpro Junior (Esomeprazole 10mg) OD, her fever subsided dramatically after the IVIG administration. While discharging the ESR-17 mm/hr, CRP-6 mg/L, Platelet count- $388 \times 10^3/L$ was normal and the patient was clinically stable.

The patient was advised to regular follow-up for one week and the drug aspirin was continued till 8 weeks.

DISCUSSION: Kawasaki's Disease was normally confirmed after tracing the fever spiking with high temperature and rashes all over the body with strawberry fissured tongue, redness of eyes, and also reddish colour lips along with abnormal laboratory values like ESR and C - reactive protein, Hb, TWbc, and Red blood cell count which was reflected in our case. Unless early detected, 20% to 25% of patients may develop cardiac complications such as coronary artery aneurysms, decreased myocardial contractility, congestive heart failure (CHF), arrhythmias, and myocardial ischemia; with treatment, the incidence decreases to 4%. Hence, early recognition and treatment has to be initiated as soon as the diagnosis is made and should involve the administration of intravenous immunoglobulin (IVIG) and high-dose aspirin which significantly reduces the incidence of these complications⁸

The American Heart Association recommends that patients should be treated with single infusion of IVIG over twelve hours at a dosage of 2 g/kg within ten days of fever onset, along with an anti-inflammatory 100mg/kg/day dose of aspirin (acetylsalicylic acid) spread out over 4 doses until the child is afebrile⁹. Hence, IVIG administration has drastically reduced not only the incidence of coronary artery disorders but also the mortality rate due to KD. The action mechanism of IVIG is variegated, but the fever, plasma C-reactive protein and inflammatory cytokines level in acute-stage KD patients reduce early after IVIG treatment. It is also known that the activation of the immunocompetent cells is inhibited by IVIG therapy¹⁰.

According to standard therapy, 80-90% of treated patients shows clinical and biochemical remission; where as in the remaining percentage (10%) of patients, persistent fever even 48 hours after administration of the drug, represents a sign of unresponsiveness to IVIG, which is the major risk factor for the development of coronary artery lesions. Such patients are said to have resistant disease and may require additional (one or two) doses of IVIG. Intravenous methyl prednisolone

can also be considered in such cases. Recently, the tumor necrosis factor alpha (TNF α) antagonist, infliximab, has been recommended for use in such children. Some children may need long term management with aspirin or additional anticoagulation^{2, 4}. In our case, early detection, standardize treatment was ensured to the patient with Intravenous immunoglobulin (IVIG -2 gm/kg), Aspirin (150 mg/kg/day) QID and Nexpro Junior (Esomeprazole 10mg) OD, got relieved without any further complications. At the time of discharge, all the laboratory reports like ESR-17 mm/hr, CRP-6 mg/L and Platelet count- 388x 10³/L were normal and patient was clinically stable.

CONCLUSION: This case report highlights the importance of treating Kawasaki syndrome prompt diagnosis and standard treatment with early administration of Intravenous Immunoglobulin (IVIG) and aspirin in preventing coronary complications.

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