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MANAGEMENT OF A CASE OF KAWASAKI DISEASE

General Medicine

Dr. Geetika Dandamudi Resident in the department of General Medicine

Dr. Nivedita D Moulick* Professor in the department of General Medicine *Corresponding Author

Dr. Harshal Bisen Resident in the department of General Medicine

ABSTRACT

Kawasaki disease (KD) is an acute systemic vascular disease that affects mostly medium sized and small vessels [1]. It is usually a self-limited disease with the highest incidence occurring in children under five years of age. We present a case of a 14 year old boy who presented with fever and rash in the lower part of the trunks and legs. Clinical examination revealed the presence of cervical lymphadenopathy, conjunctivitis and strawberry tongue. After making a clinical diagnosis of Kawasaki disease, the patient was successfully treated with intravenous immunoglobulin and aspirin. Proper diagnosis based on clinical findings, proper investigations and follow-ups are important to treat and prevent associated complications of the disease especially coronary aneurysm.

KEYWORDS

Kawasaki Disease (KD), Immunoglobulin, Aspirin, Coronary aneurysm.

INTRODUCTION:

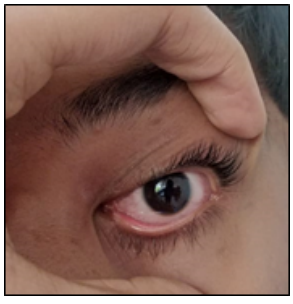
Kawasaki disease (KD), also known as mucocutaneous lymph node syndrome and Kawasaki syndrome, is an acute febrile illness of early childhood characterized by vasculitis of the medium-sized arteries. The diagnosis is clinical which is made by the presence of fever along with 4 of the 5 following criteria: (A) Conjunctivitis, (B) Cervical lymphadenopathy, (C) Skin rash commonly maculopapular, (D) Changes to oropharyngeal mucous membranes, including injected and/or fissured lips, strawberry tongue and enanthema, and (E) Palmar and plantar erythema in acute phase and periungual desquamation in convalescent stages. Generally, inflammatory changes to arterial vessels of all body regions can be present; however, coronary arteries are most commonly affected [2]. Globally, KD is the most common primary childhood vasculitis. In central Europe and North America, it is the second most common form. To date, KD is considered the most common acquired cardiac condition in childhood in developed countries [3, 4]. In cases of delayed treatment, missed diagnosis, or in treatment refractory cases, aneurysms can result and cause severe sequelae, including cardiac infarctions.

CASE REPORT:

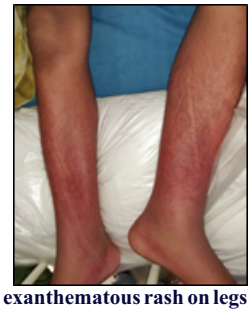
A 14 year old male presented to our hospital in the emergency department with the chief complaints of high grade fever with chills since 3 days. It was associated with a generalised non itchy body rash over the legs and lower trunk since one day. On presentation, the patient had fever with temperature of 101F associated with tachycardia. Blood pressure and oxygen saturation levels were normal. On clinical examination there was bilateral eye conjunctivitis and left cervical lymphadenopathy. Examination of the oral cavity revealed the presence of strawberry tongue associated with generalised erythema of the oral mucosa. Also there was erythema of the legs along with a generalised exanthematous rash seen over the legs and lower part of the trunk region. With a clinical suspicion of Kawasaki disease made based on the above clinical features, blood tests were sent for the patient.

Since cardiac complications especially coronary artery aneurysms and consequent myocardial infarction and coronary thromboembolism are the major causes of morbidity, a detailed echocardiographic examination was performed on the patient which revealed a normal study. Laboratory investigations revealed the presence of leucocytosis with neutrophilic predominance with normal hemoglobin and platelet counts. There was elevation of acute phase reactants i.e. erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP). Anti-Streptolysin titers and throat swab cultures were sent for the patient to rule out scarlet fever which is the closest differential diagnosis for Kawasaki disease.

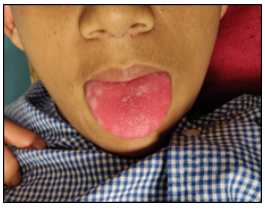
Figure 1: Clinical manifestations in our patient before immunoglo bulin therapy



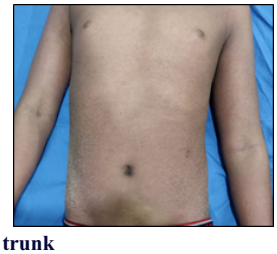
(a) Conjunctivitis



(b) Erythema with exanthematous rash on legs



(c) Strawberry tongue



(d) Rash over the trunk

Table 1: Laboratory reports of the patient:

Hemoglobin	12.4 g/dl	CRP	186.59
WBC counts	**20,800/ul**	**ASO titers**	**52.8**
Platelet counts	**2,92,000/ul**	**Urine examination**	**Normal with no RBCs or pus cells**
ESR	**45mm/hr**		

The patient was immediately started on intravenous immunoglobulin therapy at a dose of 2g/kg body weight in divided doses for three days. The patient showed dramatic improvement with the treatment. The fever subsided and there was a marked reduction in the oral mucosal erythema and strawberry tongue. The exanthematous rash over the legs and trunk subsided associated with subsequent desquamation by day 4 of treatment. Also there was thrombocytosis in the blood picture. The above changes suggest the convalescent phase of Kawasaki disease and clinical improvement. He was also started on aspirin at a dose of 30mg/kg/day. The patient was monitored carefully during the course of the treatment and discharged after 10 days of hospital stay. He was advised to continue aspirin at 3mg/kg daily for 6 weeks and asked for follow up for a review echocardiographic study for coronary aneurysm after 4weeks of discharge.

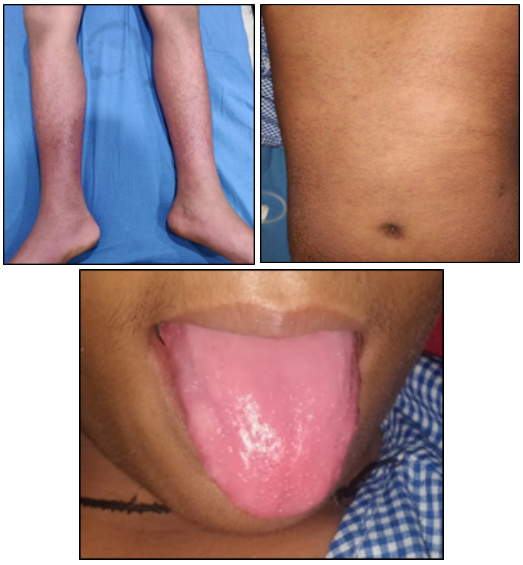


Figure 2: Clinical improvement in the patient following immunoglobulin therapy.

The diagnosis of Kawasaki disease is clinical and the presence of the classical clinical features in our patient made it easier for the diagnosis and early initiation of the treatment. KD is usually treated with intravenous immunoglobulin (IVIG) and high dose aspirin therapy. Immunoglobulin therapy has a dose-dependent effect. It is administered at a dose of 2g/kg body weight within 10 days of illness or later if the patient has signs of inflammation, persistent fever or aneurysm on echocardiography. The IVIG can influence the T cell activity and reduce the production of antibodies and cytokines responsible for the symptoms of KD [5-8]. It can lower the risk of giant aneurysm to 1% and coronary artery aneurysms from 25% to less than 5% [9, 10]. It is highly recommended to start the treatment immediately if the patient meets the clinical criteria for KD. Aspirin is believed to modify the inflammatory state in KD. It also prevents the risk of thrombosis. As per the AHA guidelines recommendation aspirin is administered at a dose of 30-80mg/kg/day in 4 divided doses till the patient is afebrile for a period of 48-72hours. Later on it is tapered to 3-5mg/kg/day for at least 6-8 weeks after the onset of the disease. It is also recommended to continue the use of aspirin if there are any persisting coronary abnormalities [9, 10]. Patients who are refractory to the above treatment may require second dose of immunoglobulins, corticosteroids, cyclosporine A and cytokine blocking strategies [11-13]. Since arterial aneurysms, particularly coronary aneurysms, develop within in the first few weeks after the onset of KD, coronary ultrasound should be performed within and at the end of this period i.e. 4–6 weeks after first treatment. Follow up echocardiography at 4 weeks in our patient revealed a normal study with no aneurysms.

CONCLUSION:

Although Kawasaki disease is a poorly understood condition, it can be managed well with a proper treatment and regular follow up. This case is presented to sensitize the clinicians regarding the clinical features, quick diagnosis and initiation of prompt treatment which can prevent the complications associated and improve the prognosis of Kawasaki Disease.

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