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Case Report

A case report on Kawasaki disease

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ABSTRACT

Kawasaki disease (KD) is an acute vasculitis of children that leads to coronary artery aneurysms in ≈ 25 of untreated cases. It has been reported worldwide and is the leading cause of acquired heart disorder in children in developed countries. The diagnosis of KD is made on basis on the clinical findings. Atypical KD includes patients who don't meet all the criteria for opinion. The main complication of Kawasaki complaint is coronary aneurysm, and the treatment is intravenous immunoglobulin and aspirin. Another dose of immunoglobulin is given if the patient doesn't ameliorate, and several other treatment options have been proposed over the last many years as alternate and third line options. The AHA criteria, which incorporate suggestions for laboratory tests and early echocardiography, are helpful for diagnosing incomplete KD. Diagnosis is based on the presence of fever lasting longer than 5 days and four of five specific clinical criteria. In Japan, at least five of six criteria (fever and five other clinical criteria) should be fulfilled for the determination of KD. From the Japanese circulation society joint working groups criteria (JCS 2008, Guidelines for diagnosis and management of cardiovascular sequela in KD), KD can be diagnosed indeed when fever lasts lesser than 5 days. Though, according to the American heart association (AHA) criteria, fever lasting more than 5 days is essential for KD diagnosis. The use of intravenous immunoglobulin is well established in KD. Aspirin has been used in KD for anti-inflammatory effect, and low-dose aspirin is recommended to reduce the risk of thrombosis.

Keywords: KD, Coronary artery aneurysm, Aspirin, Intravenous immunoglobulin

INTRODUCTION

Kawasaki disease (KD) is a rare condition that causes inflammation or swelling of the blood vessels throughout the body. It's generally self-limited and the highest incidence occurs in children under five years of age.¹ This causes high fever and shelling of skin. It's very rare (lesser than 1 lakh cases per year in India) and it can be last for several days or weeks.

Exact cause isn't known, current proof strongly suggests that genetic factors have a more important part in the incident of KD. Polymorphisms of the IgG receptor can increase the susceptibility of children to KD and increase the threat of developing coronary artery aneurysm.²

Symptoms includes- Fever, vomiting, abdominal pain, red eyes, cracked lips, an extremely red and swollen tongue, red skin on the palms of the hands and the soles of the feet and swollen lymph nodes in the neck.

Treatment

KD is treated with intravenous immunoglobulin (IVIG) and high dose aspirin.^{3,4} A meta-analysis studies showed that the use of corticosteroids with IVIG as initial treatment has a better effect on reducing the threat of coronary artery abnormalities compared to the use of IVIG itself.⁵ It's highly recommended to start the treatment instantly if the patient meets the clinical criteria for KD. The IVIG can prevent the development of coronary artery aneurysms.

Aspirin is believed to modify the inflammatory state present in KD and prevent the threat of thrombosis.⁶ It's also recommended to continue the use of aspirin if there are any persisting coronary abnormalities. Patients with refractory KD and mild to moderate aneurysm can be treated with aspirin alone or along with antiplatelet agents. Larger aneurysms can be treated with heparin and warfarin as needed.

CASE REPORT

A 2-year-old male child presented in emergency on 23rd February 2022 with the following complaints of high fever (104°F) for past 6 days, poor oral intake, oral ulcer for 5 days. He had 2 episodes of loose stools, vomiting and history of rash with red eyes and mouth for past 4 days. His initial investigations showed that maculopapular rashes presented whole over the body, bilateral non purulent conjunctival congestion, left sided cervical lymphadenopathy of size 2.1 cm, cracked lips, erythematous tongue and perianal excoriation.

After basic investigations such as urine routine, liver transaminases were shown normal range. He had no history of perinatal issues and no significant past illness. Fever persists more than five days of duration, bilateral bulbar conjunctivitis, oral mucositis, right sided lymph node enlargement and skin rashes overwhelmingly in favour of KD.

Thrombocytosis and leukocytosis were noted on 24th February 2022. C-reactive protein level were raised (55 mg/dl), increased T4 and LDH level concluded the diagnosis of KD. In view of KD, ECHO was done which showed dilated coronaries with functionally and structurally normal heart which raises the red alert to giving the treatment with Intravenous Immunoglobulin (2 gm/kg) and oral aspirin (50 mg/day).

Over a period of 12 hours after IVIG, He was afebrile, rashes vanished, oral intake was better. His vitals become normal {HR (120/min), RR (24/min), BP (94/62 mmHg), SpO₂ (98%)}. His vitals, intake and output were serially monitored throughout hospital stay.

Syrup P250 (Paracetamol 250) was given as antipyretic and analgesic use. IV antibiotic inj. ceftriaxone 100 mg/kg/day were started as prophylaxis treatment for infections. Calosoft lotion was given for local application over the skin for relieving the symptoms of erythematous rashes over the body. He was started on IV fluids DNS to maintain the calories requirement. Two episodes of vomiting was managed by antiemetic's inj. emeset. To maintain and normalize the gut organisms and flora, syrup Nutrolin B was given as oral on 24th February 2022.

Intravenous immunoglobulin 20 grams (2 gm/kg) were given as continuous infusion over 24 hours. Oral steroids (Methyl prednisolone-1 mg/kg/dose) and low dose aspirin therapy (50 mg/day) were also started with intravenous

immunoglobulin (IVIG) and instructed to continue by tapering the dose after discharge. The parents were also counseled regularly about the condition of the ongoing treatment regimens. On further investigations, it was seen that complete blood count, urine culture, liver function tests were normal range. He was improved clinically better and hemodynamically stable. Hence he was discharged with advice on 26th February 2022.



Figure 1: Extremely red and swollen tongue.

DISCUSSIONS

KD is an acute febrile, systemic vasculitic syndrome of an unknown etiology that primarily occurs in children less than five years of age.^{1,15} It occurs more frequently in boys than in girls. Eighty percent of cases occur in children lesser than 5 years of age, and most are in children under the age of 2.⁷ It's the most common cause of acquired heart disorder in young children.^{3,10}

Fever for 5 days of duration and presence of four following principal features such as changes in extremities, polymorphous erythema, bilateral conjunctival injection, changes in the lips and oral cavity. Cervical lymphadenopathy are the diagnostic criteria for KD which was approved by AHA (American heart association).^{2,14} Inflammation and swelling in medium-sized arteries, particularly the coronary arteries is one of the important clinical feature of this disease.¹¹

KD also causes swelling in glands (lymph nodes), hence lymph node enlargement supports the determination of KD.⁸ A typical initial laboratory evaluation may include the complete blood count (CBC), renal function testing, liver enzymes, erythrocyte sedimentation rate (ESR), C-reactive protein (CRP), and urinalysis.⁹ Echocardiography is generally used to assess for CAA's, in both completely manifested and suspected incomplete cases of KD.²

Treatment with intravenous immunoglobulin (IVIG) and high doses of aspirin is strongly recommended if the patient meets the clinical criteria for KD.⁴ High-dose aspirin and IVIG appears to possess an additive anti-inflammatory effect which is helpful for reducing the complications of KD.⁴ The use of corticosteroids with

IVIG as initial treatment has a better effect on reducing the threat of coronary artery abnormalities compared to the use of IVIG alone.⁶ The efficacy of IVIG administered in the acute phase of KD in reducing the prevalence of coronary artery abnormalities is well-established.¹³

Corticosteroids also have been used to treat patients with KD with an improvement from symptoms and the absence of a significant progression in coronary artery abnormalities or adverse effects.^{5,12} Low-dose aspirin is continued till the inflammatory markers return to the normal range and the echocardiogram doesn't show any abnormalities.⁶

It's important to identify early provisional diagnosis and to initiate the treatment instantly. Early and suitable treatment of KD may reduce the life-long sequela of coronary artery lesion in children.

CONCLUSION

KD is a rare disease that causes inflammation or swelling of the blood vessels throughout the body especially for children less than 2 years of age. KD will become serious if it wasn't diagnosed earlier. Earlier diagnosis can be effective in treating the patients with guidelines of drugs having for KD. Most of the cases well responds to IVIG and Aspirin therapy along with steroid therapy. This treatment strategy will be more beneficial too. Thus, adding other vitamin supplements will be more effective in the treatment adherence for the patient. In this case, the treatment strategies used as well and patient too responded well.

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