Journal of Pharmaceutical Advanced Research
(An International Multidisciplinary Peer Review Open Access monthly Journal)
Available online at: www.jparonline.com

CASE STUDY JPAR 2023

A case report on incomplete Kawasaki disease – Autoimmune Mucocutaneous Syndrome

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Received: 01.06.2023 Revised: 12.06.2023 Accepted: 20.06.2023 Published: 31.06.2023

ABSTRACT:
Kawasaki disease was initially described by Dr. Tomisaku Kawasaki in 1967 in Japan and was set up to be an important cause of cardiac complaints in children shortly thereafter. More than 1,15,000 cases were reported in Japan .Kawasaki disease (KD)can also be called Kawasaki syndrome and mucocutaneous syndrome which includes skin, mouth, lymph nodes and inflammation of blood vessels. It was an acute, self-limited vasculitis of unknown etiology that occurs generally in infants and young children. Here we have presented a clinical case from Pediatric Resuscitation Emergency Medicine Department – 1 year old male child was admitted to the hospital with complaints offever for 12 days, high grade ,intermittent , associated with chills, and rigor occasionally (2 times). Rashes all over the body for 10 days, started as fine maculopapular rashes then progressively increased and spread all over the body. The drug plan for this patient was Paracetamol, Ranitidine, IVIG, Aspirin, and Azithromycin.

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Keywords: Incomplete Kawasaki disease, IVIG, Vasculitis, Auto immune disease, Aspirin.

INTRODUCTION:

Kawasaki disease was originally described by Dr. Tomisaku Kawasaki in Japan in 1967 and was considered to be an important cause of cardiac complaints in children shortly later. More than 1, 15, 000 cases were reported in Japan [1]. Kawasaki disease (KD) can also be called Kawasaki syndrome and mucocutaneous syndrome which includes skin, mouth, lymph nodes and inflammation of blood vessels. It is an acute, self-limited vasculitis of unknown etiology that occurs generally in infants and young children. It was manifested initially by high fever, mucocutaneous inflammation, and cervical lymphadenopathy, KD

targets the coronary arteries and other cardiovascular structures. Approximately 1 in 5 children who were not treated with intravenous immunoglobulin (IVIG) in the acute phase of illness developed coronary artery aneurysms (CAA). Indeed, KD has replaced rheumatic fever as the leading cause of acquired cardiac disease in children in the World [2]. It occurs more often in boys than in girls, with a ratio of about 1.5:1. About 80 % of cases occur in children less than 5 years of age, and most of the children are under age 2. The onset of the disease is rare after 8 years of age. Less than 2% of patients have recurrences [3]. Here we are presenting a case study on Incomplete Kawasaki disease with clinical important details, diagnostic and treatment approaches of the concerned case discussed below. The objectives of the study are to describe an individual's clinical situation in detail, to identify the key issues of the case, and to analyze the case using relevant theoretical concepts.

CASE REPORT:

In the Pediatric Resuscitation Emergency Medicine (PREM) department – a 1-year-old male child was admitted to the hospital with complaints of Fever for 12 days, high grade, intermittent fever, associated with chills, rigor occasionally (2 times). Rashes all over the body for 10 days started as fine maculopapular rashes then progressively increased and spread all over the body (started in the face first), h/o vomiting for the past one week (4 to 5) episodes/ day., h/o cough and cold for 2 days, h/o reduced for food intake. After admission inward child undergoes General physical examination**:**

General appearance shows the child was alert, awake, Febrile, with Irritable pallor, Pedal edema minimal, mild itching, oral mucosa erythema, and child had intermittent fever, and the high grade is about 102.1 °F, Rashes all over the body.

HEAD TO FOOT EXAMINATION:

- Head - Normal
- Hair - Black in color soft in texture
- Eye - Normal
- Tongue - Normal
- Lips - Mucosal peeling + over the lips and around the mouth
- Limbs - Minimal edema + over limbs both LL
- Genitalia - Normal

Table 1. The other investigations report.

Echo report	Coronary artery dilated LVEF-60 %
Usg abdomen	Mild hepatosplenomegaly Gall bladder wall edema
Peripheral smear report	Moderate Microcytic Hypochromic Anemia
Culture report	Salmonella typhi
COVID report	Negative

Table 2. The various test report.

Tests	Observed value	Reference value
ESR	16 ?	3- 13mm/h
CRP	20.0 ?	< 10mg/L
Hemoglobin	1st day – 7.3 5th day – 6.9 6th day – 7.0 15th day – 6.1 25th day – 8.5	9.5 - 14 g/dL

TREATMENT:

Initially, the child was treated with IVF.DNS @ 36 ml/h, Injection of Cefotaxime 300 mg IV TDS for, Injection of Ranitidine 0.4cc IV BD for 10 days, and then injection Rantiditine changed into tablet form for 8 days with the supplement of Paracetamol 150 mg given for 2 days and then changed into syrup Paracetamol Injection IVIG 1st dose 2 g/kg over 18 h (20 ml/h on flow) again treated with 2nd dose of IVIG 2g/kg over 18 h (20 ml/h on flow) Tablet of Aspirin 150 mg for 8 h Syrup of Azithromycin 5 ml OD. The patient was discharged with a Tablet of Aspirin 150 mg OD for 6 weeks.

Outcome and follow-up:

The child's condition was stabilized with supportive management with more precaution. Parents were advised to review after 1 month in the Outpatient department.

DISCUSSION:

Kawasaki disease (KD) also known as Mucocutaneous lymph node syndrome is an acute febrile illness characterized by inflammation of blood vessels throughout the body that primarily affects young children and infants under 5 years of age. It is an autoimmune disease. KD is a systemic necrotizing vasculitis affecting the medium and small-sized arteries. It is not only the common cause of vasculitis disorder in children but also the common cause of acquired heart disease in children [4].

Causative agent:

The etiology of KD remains obscure. It is believed that KD may be triggered by a streptococcal/ staphylococcal toxin which acts as a superantigen and causes widespread immune activation. Kawasaki disease may be associated with living bodies of water nearby or exposure to house dust mites or recently shampooed carpets, rugs, shampoo, or mercury poisoning [5]. Critical data to substantiate these possible relations are lacking. Although an infectious etiology has been suggested, person-to-person transmission has not been documented, even in day-care centers, a common source for outbreaks has not been defined, and cases among siblings are very rare [6].

CLASSIFICATION:

It is classified based on diagnostic criteria;

- Complete Kawasaki disease.
- Incomplete Kawasaki disease.

COMPLETE KAWASAKI DISEASE SYMPTOMS:

Diagnosed based on the;

Clinical diagnostic criteria:

- FEVER – duration of 5 days and more plus of the following;
- Eye – Conjunctivitis.
- Changes in the extremities – edema, erythema, desquamation.
- Cervical lymphadenopathy - Swollen glands in the neck.
- Lips - Fissuring of lips and over the cavity.
- Tongue – strawberry.
- Rash - all over the body.

Supplementary laboratory criteria:

- Albumin :< 3 g/dL
- C- reactive protein :> 3mg/dL
- Erythrocyte sedimentation rate :> 40mm/h.
- Elevated alanine aminotransferase level.
- Leukocytes level: 15000 /mm.
- Normochromic, normocytic anemia for age.

Incomplete Kawasaki disease:

It occurs in persons with a fever lasting 5 or more days and with 2 or 3 of these above-mentioned findings. Diagnostic criteria to differentiate the two types are slightly different. But treatment is similar for both types. The case presentation is diagnosed by considering this category.

According to the IAP (Indian Academy of Pediatrics) Guidelines:

- In the acute phase ANEMIA is common – it takes 6-8 weeks to get return to normal values.

FIRST LINE THERAPY - INTRAVENOUS IMMUNOGLOBULIN (IVIG):

- 2 g/kg as a single IV infusion.
  IVIG should be given within the first 10 days of illness but should also be given to children diagnosed after 10 days if there is evidence of ongoing fever.
- 2nd dose of IVIG 2g/kg should be given to children who do not respond to the 1st dose, as demonstrated by persistent or recurrent fever within 36 hrs after the end of the 1st IVIG infusion.
- If the patient is resistant to 2nd dose IVIG may be treated with methylprednisolone 30 mg/kg/day for 1-3 days.

Aspirin:

- Given in doses 80 to 100 mg/kg daily in 4 divided doses in the acute phase and then continued in the smaller antithrombotic dose of 3 to 5 mg/kg orally as a daily dose for 6 weeks until the ECHO report was found to be normal.

SECOND LINE THERAPY – INFLIXIMAB:

About 5 mg/kg given Intravenously over 2 hrs for children with refractory Kawasaki disease

General management:

- Monitor temperature frequently.
- Cardiac monitoring – frequently check ECG, ECHO, heart rate, Spo_2 level.
- Assess extremities for edema, redness, and desquamation.
- Examine eyes for conjunctivitis.
- Monitor mucous membrane changes for inflammation.
- Give soft foods and liquid foods.
- Monitor intake and output chart and weight of the child daily.
- Administer IVIG, Aspirin [7].

Parent education/ Instructions:

- Explain regular follow-up care and its importance.
- Explain the persisting signs and symptoms after acute illness treatment.

- Irritability may last for 2 months after the onset of symptoms.
- After discharge, it may take 3 to 4 weeks for the complete recovery of the child.
- Don't be alarmed if children get some peeling of the skin on their hands or feet – this is very common.
- Kawasaki disease – need to have long-term follow-up.
- After 6 to 8 weeks – ECHO TEST should be taken. If the report is normal, it is unlikely that further changes to the coronary arteries will occur.

CONCLUSION:

In this study we discussed a clinical case of incomplete Kawasaki disease, IVIG 2 doses and Aspirin were given for this patient helped in subsiding the condition. Parents were advised to come hospital after 6 to 8 weeks for ECHO TEST should be taken. If the report is normal, it is unlikely that further changes to the coronary arteries will occur.

ACKNOWLEDGEMENT:

We would like to express our deepest gratitude to hospital authorities for providing us access to patients' medical records.

REFERENCES:

1. Kawasaki T. Acute febrile mucocutaneous syndrome with lymphoid involvement with specific desquamation of the fingers and toes in children. Arerugi, 1967; 16: 178-222.
2. Kato HS, Koike S, Yamamoto M, Ito Y, Yano E. Coronary aneurysms in infants and young children with acute febrile mucocutaneous lymph node syndrome. J Pediatr, 1975; 86(6): 892-898.
3. Dajani AS, Taubert KA, Gerber MA, Shulman ST, Ferrieri P, Freed M, *et al*. Diagnosis and therapy of Kawasaki disease in children. Circulation, 1993; 87(5): 1776-1780.
4. Yanagawa H, Nakamura Y, Yashiro M, Hirose K. Results of 12 nationwide surveys of Kawasaki disease. Arch Pediatr Adolesc Med, 1995; 149(7): 779-783.
5. Singh S, Vignesh P, Burgner D. The epidemiology of Kawasaki disease: a global update. Arch Dis Childh, 2015; 100(11): 1084-1088.
6. Nair PA, Diwan NG. Kawasaki disease-two case reports from a rural set up in Gujarat. Indian J Paediatr Dermatol, 2015; 16(3): 155-158.
7. Abrams JY, Belay ED, Uehara R, Maddox RA, Schonberger LB, Nakamura Y. Cardiac complications, earlier treatment, and initial disease severity in Kawasaki disease. J Pediatr, 2017; 188: 64-69.

Conflict of Interest: None
Source of Funding: Nil
Paper Citation: Kowsalya DS*, Medona JM. A case report on incomplete Kawasaki disease – Autoimmune Mucocutaneous Syndrome. J Pharm Adv Res, 2023; 6(6): 1873-1876.