Case Report

Kawasaki Disease: A Case Report

Dr. Cheppalli Vani*, P. Salome Satya Vani[1], Mubeena shirin[2], U. Niharika[3]
Assistant professor*,[1], Pharm.D[2,3], Department of Pharmacy Practice,
Pulla Reddy Institute of Pharmacy, Domadugu (V) Gummadidala (M), Telangana, India.
***Corresponding author's E-mail:** cnvaniredddy@gmail.com

Received: 06-03-2020; **Revised:** 17-04-2020; **Accepted:** 24-04-2020.

ABSTRACT

Kawasaki disease also called as mucocutaneous lymph node syndrome. It is characterised by an acute febrile illness and acute multisystem vasculitis that occurs predominantly in infants and young children. The pathological changes in Kawasaki Disease affect medium-sized, extra-parenchymal muscular arteries and most commonly the coronary arteries. Acute phase of Kawasaki Disease is characterized with leukocytosis of immature and mature granulocytes, normochromic, normocytic anemia, and elevated proteins. The aetiology of Kawasaki disease is unknown but commonly described as an exaggerated immune response to environmental or infectious trigger in developmentally, immunologically and genetically susceptible children. Here is the case of 8year female patient was admitted in hospital with complaints of low grade fever, headache, macula-papulae rashes over both palms, foot over plantae surface associated with itching and redness all over upper and lower limbs since 3 days. This patient was diagnosed in early phase of Kawasaki disease and laboratory investigations were WBC: 15000/cumm, ESR: 46 mm/hr, CRP: 5.5 mg/L and Platelets: 4.97 Lakhs/cumm. The patient was treated by pediatrician with Intravenous immunoglobulin, Paracetamol and Ceftazidime. The symptoms were subsided after one week of treatment. The patient was discharged with prescription of antihistamines and vitamin supplements.

Keywords: Kawasaki disease, Mucocutaneous lymph node syndrome, Vasculitis.

INTRODUCTION

Kawasaki disease also referred to as Kawasaki syndrome or mucocutaneous lymph node syndrome affects children. The acute phase of the condition commonly lasts 10- 14 days or more. Most children recover fully. But in some cases, Kawasaki disease can lead to long term heart complications. The disease is named after Dr. Tomisaku Kawasaki, a Japanese paediatrician[1]. Kawasaki disease is the leading cause of acquired heart disease in infants and young children. Kawasaki disease causes inflammation in the walls of medium sized arteries throughout body[2]. It primarily affects children. The inflammation tends to affect the coronary arteries which supply blood to heart muscles. It affects lymph nodes, skin and the mucous membranes inside mouth, nose and throat.

Symptoms of Kawasaki disease appear in three phases. ***First phase***- A fever that is often higher than 102.2F (39˚C) and lasts more than 3 days, extremely red eyes (conjunctivitis) without thick discharge, a rash on main part of body (trunk) and in genital area, red dry cracked lips, an extremely red swollen tongue (strawberry tongue), swollen red skin on palms of hands and soles of feet, swollen lymph nodes in neck and irritability.

Second phase- Peeling of skin on hands and feet especially the lips, fingers and toes often in large sheets, joint pains, diarrhoea, vomiting, abdominal pain.

Third phase- Disease signs and symptoms slowly go away unless complications develop. It may be long as 8 weeks before energy level seems normal again[3].

Etiology: The exact cause of Kawasaki disease is still unknown. Researchers speculate that a mixture of genetics and environmental factors can cause KD. This may be due to the fact that cause KD occurs during specific seasons and tends to affect children[4].

Risk factors: Age: It is more likely between the ages of 1 year and 5 years. Gender: boys are more likely than girls to develop it. Genetics: if the parents had Kawasaki disease, their offspring may be more likely to have it, suggesting that it may be linked to an inherited gene. Environment: In the northern hemisphere, from January through March, the rate is 40 percent higher than in august through October. Some suggest it may be a reaction to some toxins or medications, but clinical evidence is lacking[5].

Complications: Patients with Kawasaki disease can develop heart complications such as mild dilation of the coronary arteries. These abnormalities usually diagnosed in the first two weeks of the illness, are likely to resolve spontaneously within a few weeks or months. Heart complications include: inflammation of blood vessels (vasculitis), usually the coronary arteries, hat supply blood to heart, Inflammation of heart muscle (myocarditis), heart valve problems. Inflammation of the coronary arteries can lead to weakening and bulging of the artery wall (aneurysm). Aneurysms increase the risk of blood clots

forming and blocking the artery which would lead to a heart attack or cause life-threatening internal bleeding[6,3].

Diagnosis: There is no specific diagnostic test, although laboratory and echocardiography findings (ex- elevated erythrocyte sedimentation rate and C- reactive protein level, hyponatremia, hypoalbuminemia, coronary aneurysms) may be helpful in evaluating suspected cases and differentiating Kawasaki disease from other conditions. Diagnosis largely is a process of ruling out diseases that cause similar signs and symptoms including: scarlet fever which is caused by streptococcal bacteria and results in fever, rash, chills and sore throat. Juvenile rheumatoid arthritis, steven – Johnson syndrome, toxic shock syndrome, measles. The doctor will do a physical examination and other tests include urine tests, blood tests. Testing for substance called B- type natriuretic peptide (BNP) that **is** released when heart is under stress. Electrocardiogram and echocardiogram is also done[7,4]**.**

Pathophysiology: In the earliest stages of disease the endothelial cells and the vascular media become oedematous, but the internal elastic lamina remains intact. Then approximately 7-9 days after onset of fever, an influx of neutrophils occurs which is quickly followed by a proliferation of CD8+ (cytotoxic lymphocyte and immunoglobulin A- producing plasma cells. The inflammatory cells secrete various cytokines (tumour necrosis factor, monocyte chemotactic and activating factor), interleukins (IL-1, IL-4, IL-6) and matrix metalloproteinase's (MMP3 and MMP9) and target endothelial cells that lead to fragmentation of internal elastic lamina and vascular damage[8]. In severely affected vessels, the media develops inflammation with necrosis of smooth muscle cells. The internal and external elastic laminae can split leading to aneurysms.

Prevention: Kawasaki disease cannot be prevented, but it can be managed. Certain foods may help promote healthy blood vessels- a diet rich in nitrates may help promote a healthy cardiovascular system. Commonly found in vegetables, nitrate turns into nitric oxide (NO) when digested, helping support normal endothelial function and protecting mitochondria. The best way to increase the production of nitric oxide in body is to consume leafy greens such as- basil, cabbage, spinach and broccoli. Exercise can also produce nitric oxide in body[9].

Treatment: Children affected by Kawasaki disease are hospitalised. Kawasaki disease is treated with high doses of aspirin (salicylic acid) to reduce inflammation and to mildly thin the blood to prevent blood clot formation. Also given with gamma immunoglobulin administered through vein (IVIG) together with fluids. This treatment has been shown to decrease the chance of developing coronary aneurysms in the coronary arteries, especially when used early in the illness. Some corticosteroids medications are given. Persisting joint pains are treated with anti-inflammatory drugs such as ibuprofen or naproxen. Plasma exchange (plasmapheresis) has been reported as effective in patients who were not responding to aspirin and gamma globulin. Plasmapheresis is a procedure where the patient plasma is removed from blood and replaced with protein containing fluids. By taking out portions of the patient plasma the procedure also removes antibodies and proteins that are felt to be part of the immune reaction that is causing inflammation of the disease. Kawasaki disease that is not responding to traditional treatment aspirin and gamma immunoglobulin infusions can be deadly[10].

CASE STUDY

A female patient of age 8 yrs was admitted in hospital with complaints of low grade fever since 3 days followed by headache, macula-papulae rashes associated with itching and redness all over upper and lower limbs as shown in the figure (1, 2). Her vitals were noted as Temperature was 99.2F, Blood pressure was 90/60 mm of Hg, Pulse rate was 110 per min, Respiratory rate was 22 per min, Spo_2 was 98%. Levels of serum acute phase reactants includes erythrocyte sedimentation rate (46 mm/h) and C-reactive protein (5.5 mg/dL) were high; no bacterial growth was detected in any of her culture. Her white blood cell count was 15,000/cumm; platelet count was 4.97 lakhs/cumm and the liver function tests were noted within the normal ranges. Her echocardiograph and ultrasonograph impression was normal and no significant abnormalities were detected.

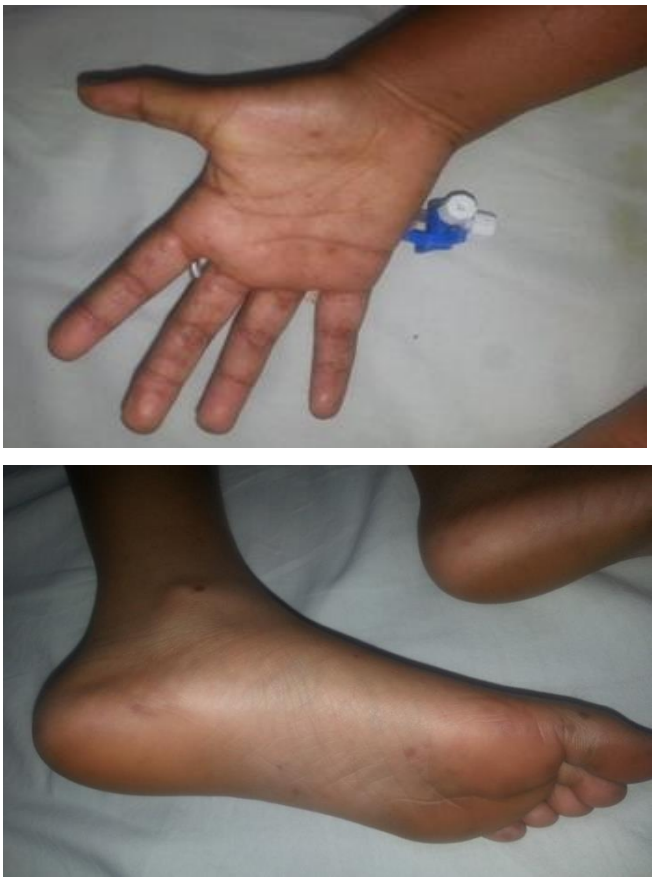

Figure (1, 2): Maculopapular rashes over hands and plantar surface of foot

The child was treatment with intravenous immunoglobulin (IVIG) 2g/kg as a single infusion decreases the risk of developing coronary artery abnormalities when given early in illness, Injection Paracetamol 360mg IV TID was given to reduce fever, Injection Ceftazidime 750 mg IV BID was prescribed to treat bacterial infections or hospital acquired infections Injection Dexamethasone 3 mg IV BID was used to treat allergy. Her fever disappeared after the dose of IVIG therapy and acute phase reactants like ESR and CRP were decreased at the end of the week. The patient was discharged from the hospital with Tablet Fexofenadine was used for allergy, Tablet Vitamin C helps in maintenance of healthy body tissue and immune system and Syrup multivitamin 10 ml BID works by protecting cells from damage.

DISCUSSION

Kawasaki disease is an acute multisystem inflammatory disease of blood vessels (vasculitis) that commonly affects infants and young children's. The disease may be characterised by a high fever, inflammation of the mucous membranes of the mouth and throat, a reddish skin rash and swelling of lymph nodes (lymphadenopathy)[2]. In addition individuals with Kawasaki disease may develop inflammation of arteries that transport blood to heart muscle (coronary arteritis), associated widening or bulging (aneurysms) of the walls of affected coronary arteries, inflammation of heart muscle (myocarditis). Although cause is unknown it is widely thought due to infectious agent in an immunologically susceptible individual the causative agent remains elusive. Some of the infectious agents proposed are parvovirus, staphylococcus aureus, Epstein- Barr virus, Chlamydia and mycobacterium. In infants, the atypical presentations longer duration of illness before diagnosis, lower incidence of conjunctivitis, lower incidence of rash, lower incidence of extremity change and lower C – reactive protein) are common and may result in delay in diagnosis and effective treatment. Delayed diagnosis in KD will be a significant risk factor for development of coronary artery abnormalities[4]. In this study, the patient has complaints of low grade fever with headache, *Macula papulae* rashes over both palms foot over plantae surface associated with itching and redness over upper and lower limbs. The patient was treated with intravenous immunoglobulin, antibiotics to avoid bacterial infections and corticosteroids were prescribed to treat itching and rashes.

CONCLUSION

Kawasaki disease is rare and early detection and treatment of KD results in prevention of coronary ectasia. The child with fever and classic clinical and laboratory findings of KD, treatment with IVIG on or before 5 days of fever resulted in better coronary outcomes and decreased the total length of time of clinical symptoms.

REFERENCES

1. Kawasaki T. Acute febrile mucocutaneous syndrome with lymphoid involvement with specific desquamation of the fingers and toes in children. Arerugi, 16, 1967, 178-222.
2. Naoe S, Takahashi K, Masuda H, Tanaka N. Kawasaki disease with particular emphasis on arterial lesions. Acta Pathol Japonica, 41, 1991, 785-97.
3. Son MBSRP. Textbook of Paediatric Rheumatology: Kawasaki disease. Philadelphia: Elsevier; Volume 7, 2016, 467-83.
4. Newburger, J.W, Takahashi M, Gerber M.A, Gewitz M.H, Tani L.Y et al. Diagnosis, treatment and long-term management of Kawasaki disease: A statement for health professionals from the committee of Rheumatic Fever, Endocarditis and Kawasaki disease, Council on Cardiovascular Disease in the young, American Heart Association: Paediatrics, 114(6), 2004, 1708-33.
5. Holman R.C, Curns A.T, Belay E.D, Steiner C.A, Schonberger L.B. Kawasaki syndrome hospitalizations in the United States. Paediatrics, 112(3), 1997, 2000, 495-501.
6. Kijima Y, Kamiya T, Suzuki A, Hirose O, Manabe H. A trail procedure to prevent aneurysm formation of the coronary arteries by steroid pulse therapy in Kawasaki disease. Japanese Circulation Journal, 46, 1982, 1239-42.
7. Tremoulet A.H, Jain S, Chandrasekar D, Sun X, Sato Y, Burns J.C. Evolution of laboratory values in patients with Kawasaki disease. Paediatric Infec Dis J, 30, 2011, 1022-26.
8. Anne H Rowley, Stanford T Shulman. Pathogenesis and management of Kawasaki disease. Expert Rev Anti Infect Ther, Feb:8(2), 2010, 197-203.
9. Jane W, Newburger MD, Masato Takahashi MD, Burns MD. Kawasaki Disease. Journal of the American College of Cardiology, 67(14), 2016, 1738-1749.
10. Athappan G, Gale S, Ponniah T. Corticosteroid therapy for primary treatment of Kawasaki disease- weight of evidence: A meta-analysis and systemic review of the literature. Cardiovasc J Afr, 20(4), 2009, 233-236.

Source of Support: Nil, **Conflict of Interest:** None.