International Journal for Multidisciplinary Research (IJFMR)
E-ISSN: 2582-2160 ● Website: www.ijfmr.com ● Email: editor@ijfmr.com

Kawasaki Disease – A Case Overview

Rajalakshmi R.[1], Sharanabassappa Dambal[2]

[1]M.Sc.(N), M.H.Sc.CCD, [2] M.Sc.(N)
[1, 2] Nursing Officer
AIIMS, Raipur, Chhattisgarh, India

Abstract:
Kawasaki disease, a medium vessel vasculitis, is seen commonly in children below 5 years of age. In developing countries Kawasaki disease is being increasingly reported, and is one of the emerging leading causes of acquired heart disease in children. Diagnosis of this case is often missed in this age group as it is not considering an upfront clinical possibility in a febrile child which often leads to delay in diagnosis and initiation of therapy. A 2 years old male child got admitted on 24/08/2022 under PICU, AIIMS, Raipur with chief complaints of cough and cold for 7 days, fever for 5 days, redness of eyes for 2 days, 3 days back, rash for 3 days, vomiting after meals for 2 days, fast breathing for 1 day, redness of palm and soles for 1 day, neck swelling for 1 day. The patient underwent basic investigations to rule out the diagnosis and differential diagnosis and was treated with oxygen support, IVIg, aspirin, antibiotics and supportive management. There was significant improvement in the child's condition and was discharged with follow up advise.

Keywords: Kawasaki Disease, Vasculitis, Mucocutaneous Lymph Node Syndrome

Introduction

Kawasaki disease, also called mucocutaneous lymph node syndrome is an acute systematic vasculitis which causes inflammation (i.e., swelling and redness) in the walls of small and medium sized blood vessels in the body; swelling in glands (lymph nodes), mucous membranes of mouth, nose, eyes and throat. This condition most often affects children of less than 5 years of age. Children with this disorder will show signs and symptoms of high fever, rash over body and extremities, redness and swelling of hands and feet, skin peeling in hands and foots in the second and third weeks, and red eyes and tongue. Kawasaki disease is usually treatable, and most children recover without serious problems if they receive treatment within 10 days of onset. Early treatment to prevent cardiovascular complication is important.

Kawasaki disease is the leading cause of acquired heart disease in children. In as many as 25 percent of children with Kawasaki disease, the heart becomes affected without proper treatment. Kawasaki disease commonly leads in inflammation of coronary arteries.

Case History

Informant: Child's Mother

A 2 years old developmentally normal male child got admitted on 24/08/2022 under PICU, AIIMS, Raipur with chief complaints of cough and cold for 7 days, fever for 5 days, redness of eyes for 2 days, 3 days

back, rash for 3 days, vomiting after meals for 2 days, fast breathing for 1 day, redness of palm and soles for 1 day, neck swelling for 1 day.

History of Past Illness

Diagnosed case of cyanotic congenital heart disease, child was in usual state of health and started having dry cough without any diurnal or postural variation and not severe enough to disturb his sleep along with cold which was later followed by fever which was documented, low grade, intermittent, without any chills and rigor with child being active during the inter-febrile period without other foci initially. 2 days later child developed painless redness of both eyes associated with matting of eyelids in morning on waking up without any purulent discharge that self-resolved after two days. On day 4 of fever child developed erythematous maculopapular rash over the trunk, extending up to dorsum of hand and sparing face, palm and sole. Child had non projectile, non-bilious/blood-stained vomiting after feeds 4 days prior to presentation for 2 days. On the day of admission, mother noticed the child to have redness of palms and soles, swelling of neck along with increased efforts of breathing for which they brought the child to AIIMS, Raipur. There has been no history of any drug intake, insect bite, contact with person with similar illness.

Past History: No similar history in the past. No history of fever with rash in the family or community. Detected to have cyanotic congenital heart disease-mitral atresia with non-restrictive ASD, large inlet VSD with bidirectional shunt, severe PAH and had undergone cath study at our institute by dept of cardiology.

Birth and Immunisation History: Uneventful**.**

Mother could recall up to 6 weeks and after those two vaccines at 1 and 1/2 years of age.

No vaccination card available.

Developmental History: Developmentally normal child as per information provided.

Clinical Examination and Relevant Findings

Vital Signs on Admission

- Temperature: 99.6 f
- Heartbeat Rate: 140 beats/mt
- Respiratory Rate: 44 breaths/mt with nasal prongs
- Spo2: 86% on RA

Anthropometric Measurements

- Weight: 8 kgs (-3.72 SD)
- Height: 84 cms (-1.28 SD)
- Head Circumference: 43 cms

General Examination

Pallor	**Icterus**	**Cyanosis**	**Clubbing**	**Lymphadenopathy**	**Edema**
Absent	Absent	Present: Acrocyanosis	Present: Grade II	B/L Cervical and Submandibular	Absent
Ear	**Nose**	**Throat**	**BCG Scar**	**Skin**	**JVP**
Normal	Normal	Normal	Present	Normal	Normal

- Skull and spine normal.
- No dysmorphism.
- Genitalia: Normal
- Skin: Maculopapular rashes over anterior and posterior aspect of trunk, bilateral upper and lower limbs sparing palm and sole.
- Erythematous palm and soles.
- Perineal desquamation present.
- Bilateral significant cervical lymphadenopathy present. Throat congested with grade III tonsils. Tongue congested. Posterior pharyngeal wall could not be seen as child is irritable.
- Lip cracking present. Angular stomatitis present.

Systemic Examination

RS	Inspection: B/L equal chest rise with sub-costal, inter-costal and supra-sternal retractions.
	Palpation: B/L chest expansion equal.
	Percussion: Resonant note.
	Auscultation: B/L equal air entry with stridor heard without stethoscope.
CVS	S1 normally heard. Loud P2. No murmur audible.
P/A	Soft and non-tender. No organomegaly.
CNS	E4V5M6, Cranial nerve normal, Tone normal in all 4 limbs, Power > 3/5 in all limbs, Reflexes normal in upper limb and lower limb, B/L plantar flexor. Meningeal signs absent, Cerebellar signs absent, Autonomic system normal, Skull/Spine normal

Diagnostic Evaluations

Type		**24/08/2022**	**27/08/2022**	**28/08/2022**	**3/09/2022**
Hematogram	Hb (g%)	14.8	13.0	12.7	11.7
	TLC (/mm^3)	9.44	19.97	28.06	26.21
	P/L/M/E (%)	-/-/10.2/0.2	41.3/53.9/4.3/0.1	44.3/51.1/4.1/0.1	27.7/68.7
	Platelets(/mm^3)	170,000	216,000	254,000	418000
	MCV (fl)	97.3	76.1	78.2	83
	MCH (pg)	26.6	24.7	24.5	26
	MCHC (%)	27.3	32.4	31.4	31.3
	RDW – cv (%)	23.9	18.0	18.1	19.9
RFT	**Urea/Uric Acid**		14/4.4	17/3.8	18/5.4
	Creatinine		0.42	0.45	0.50
	Na/K/Cl		131/4.19/92	135/4.45/94	137/4.44/101
LFT	**TOT/DIR Bilirubin**		0.63/0.12		0.63/0.19
	AST/ALT		50/17		44/16
	ALP/GGT		119/14		111/19
	A/G		3.1/5.2		3.2/5.7
ESR		45 MM			
HS-CRP Quantitative		54.30		17.16	
Dengue (IgM/IgG/NS1 Antigen)			**NR/NR/NR**		
Procalcitonin				1.40	
Blood C/S				**Growth of Enterococcus faecalis after 24 hours.**	**No bacterial growth after 24 hours.**
Urine R/M			Specific Gravity: 1.000, Protein/Glucose: -/-, Pus Cells: 10-20 cells/HPF, RBC: Not detected, Occasional epithelial cells.		Specific Gravity: 1.020, Protein/Glucose: -/-, Pus Cells: 5-10 cells/HPF, RBC: Not detected, Epithelial cell: Not detected
Urine C/S				**No bacterial growth after 24 hours.**	

Course of Illness and Management

2 years old developmentally normal, inadequately immunized third order male child born out of non-consanguineous marriage diagnosed to have cyanotic congenital heart disease 1 month back presented with acute febrile illness associated with rash, significant lymphadenopathy and respiratory distress.

Course in PICU

Child was received in a state of irritability with tachypnea and cyanosis noted on initial assessment. Primary survey revealed tachypnea with increased efforts for which oxygen support via nasal prongs was provided initially and later shifted to HFNC. The child had following issues:

1. **Acute Febrile Illness:** Child had low grade fever for more than 5 days with red, cracking lips with erythematous palm and soles, cervical lymphadenopathy with maculopapular rash and congested and hypertrophied tonsils V (grade 3). Possibilities of diseases like Measles, EBV, Kaswasaki were ruled out. Since, age group was not appropriate for EBV; Measles and Kawasaki disease were kept as close differentials. Points against measles were appearance character of the rash, absence of enanthem and severe prodromal cough. criteria's in favour of Kawasaki were met along with raised acute phase reactants and ESR. So, IVIg was given on admission following which significant improvement was noted in form of disappearance of palm and sole erythema, rash and red, cracked lips. Fever resolved and child remained afebrile for 48 hours.

- Recurrence of fever after 48 h occurred, so two possibilities were kept. Hospital acquired infection or IVIg resistant Kawasaki disease. So, repeat dose of IVIg @ 1 g/kg was given and empirically Inj. Piptaz had been started.
- The blood culture on admission grew enterococcus faecalis sensitive to ampicillin so antibiotic was changed to inj. ampicillin. The child responded to the treatment and been afebrile for more than 72 hours in ICU.

2. **Respiratory Distress:** Child had increased work of breathing for which nasal prongs support was provided and later shifted to HFNC and child was weaned off to nasal prongs in subsequent days.

3. **Pulmonary Hypertension:** Child had been started on Sildenafil and Bosentan in view of severe PAH which was continued but on day 3 of admission child had hypotension episodes so sildenafil was withheld and was re-started after 7 days at low dose. There was no PAH crisis episodes during PICU stay. CTVS opinion was taken but operability was told be limited and advised to continue on medical management for PAH.

Therapeutic Interventions

The child received combination of therapeutic management in view of underlying condition, supplementation and antibiotics in view of blood culture report. The child has been treated with the following medications:

1. Inj. Ampicillin 400 MG @ 200 MG/KG/Day for 10 Days
2. Tab. Aspirin 75 MG 1/2 Tab PO OD
3. Tab. Bosentan 8 MG PO BD
4. Tab. Sildenafil 50 MG PO BD

5. Syp. PCM 125 MG / 5 ML PO SOS
6. Zinc Oxide Ointment LA TDS
7. Furoped Drops 0.8 ML PO BD
8. Syp. Vit. A 2 Lakhs IU PO OD
9. Inj. Amoxyclav 120 MG IV TDS
10. Inj. Lasix 8 MG @ 2 MG/KG/Day
11. Tab. Enalapril 2.5 MG 1/2 Tab + 5 ML DW @ 0.1 MG/KG/Day PO OD
12. Adrenaline Nebulisation (1:1000) 2 ML + 4 ML NS STAT
13. IVIg 5% 2 G/KG (15 G) (300 ML) over 24 Hours @ 12.5 ML/HR

Course in Ward

Once child become hemodynamically stable and considering the prognosis in child's condition child has been shifted to pediatric ward from PICU on 27/08/22. In ward Oxygen was tapered and stopped. Antibiotics were continued for 10 days and stopped. Child was afebrile throughout the course of ward and no new complaints has been noted. Since the child was hemodynamically stable and hence being discharged on 29/08/22.

Assessment on Discharge

RS	Inspection: B/L equal chest rise.
	Palpation: B/L chest expansion equal.
	Percussion: Resonant note.
	Auscultation: B/L equal air entry.
CVS	S1 normally heard. Loud P2. No murmur audible.
P/A	Soft and non-tender. No organomegaly.
CNS	E4V5M6, Cranial nerve normal, Tone normal in all 4 limbs, Power > 3/5 in all limbs, Reflexes normal in upper limb and lower limb, B/L plantar flexor. Meningeal signs absent, Cerebellar signs absent, Autonomic system normal, Skull/Spine normal

Discharge and Follow Up

Child was properly assessed every day for prognosis and review of treatment. Childs condition had a marked improvement and underlying symptoms reduced and need for oxygen reduced considerably over the course of management. Parents were satisfied with the management from the health care team. The patient was discharged after explaining the danger signs and to continue medication - Tab Aspirin 75 mg 1/2 tab Po BD, Tab Bosentan 8 mg PO BD, Tab Sildenafil (1 tab = 50 mg) 20 ml DW give 1 ml, Furoped drops (10 mg/ml) 0.8 ml, Zinc oxide ointment L/A TDS, Syrup Vit D (400 IU/ml) 1 ml PO OD, Syrup IFA (15 ml = 38 ml iron) 5 ml PO BD, Syrup PCM (125 mg / 5 ml) 5 ml PO SOS, Follow up in CTVS OPD, Review SOS in pediatric OPD.

The parents were counselled about the need for nutritional supplementation as the child is malnourished. The child was referred to dietician in view of malnourished and has underlying heart disease, and was advised about the steps to improve the intake of nutritious food suitable to the economic conditions and dietary patterns of the patient. The child was advised to come back after 10 days of discharge for a follow up on the improvement in-patient's health.

Discussion

After collecting history based on the inputs provided by the informants, presenting chief clinical signs and symptoms and conducting requisite tests on the child, diagnosis has been made as Kawasaki disease with blood stream infection (enterococcus). The child was given with appropriate treatment including oxygen therapy, IVIg, aspirin and supportive management and helped the child to get back health on track. Parents were advised and created the sense of awareness on need for long term follow up, medications, proper care of hygiene, healthy lifestyle, diet patterns etc. providing all the information and symptoms of Kawasaki disease so that, similar cases don't occur in their immediate family. A long term follow is important in such case as the child is having underlying congenital heart disease and Kawasaki disease will impose more complications on heart and heart vessels.

Conclusion

Kawasaki disease is clinically stated as systemic vasculitis of mainly medium sized arteries. While Kawasaki disease remains as poorly understood disorder, delayed and missed diagnosis can result in severe complications in children, while timely diagnosis and prompt treatment initiation and long term follow up could immensely result in good prognosis and also lowers the complications associated with Kawasaki disease. Awareness of the diseases should be created by educating the parents/public at large regarding the symptoms, major causes, importance of early diagnosis and treatment to prevent long term complications, follow up requisites and how these can be countered. Such an widespread awareness will help in early diagnosis/treatment, thereby bringing the situation under control without causing major damage to the children physically, mentally and economically; not just for the child but for their entire family and society.

Statement of Human and Animal Rights

All procedures performed in human participants were in accordance with the ethical standards of the institutional research committee. Rights and privacy of the patient maintained. This article does not contain any studies with animals performed by any of the authors.

References

1. Marilyn J. Hockenberry, David Wilson. Wong's Essentials of Pediatric Nursing. 1st South Asia Ed. Elsevier. (2013). 745-748.
2. Kawasaki T. Acute febrile mucocutaneous syndrome with lymphoid involvement with specific desquamation of the fingers and toes in children. Arerugi [Allergy] 1967; 16: 178–222.
3. Kamleshun Ramphul, Stephanie Gonzalez Mejias. Kawasaki disease: A comprehensive review. Arch Med Sci Atheroscler Dis. 2018; 3: e41–e45.
4. Elisabeth Binder, Elke Griesmaier, Thomas Giner, Michaela Sailer-Höck, Juergen Brunner. Kawasaki disease in children and adolescents: clinical data. Pediatric Rheumatology. (2014). 12(37).

5. Eleftheriou D., Brogan P.A. Therapeutic advances in the treatment of vasculitis. Pediatr Rheumatol Online J. (2016) 14: 26. https://doi.org/10.1186/s12969-016-0082-8
6. Batu E.D., Ozen S. Pediatric vasculitis. Curr Rheumatol Rep. (2012) 14: 121–129. https://doi.org/10.1007/s11926-011-0232-4
7. Tremoulet A.H., Pancoast P., Franco A., Bujold M., Shimizu C., Onouchi Y., et al. Calcineurin inhibitor treatment of intravenous immunoglobulin-resistant Kawasaki disease. J Pediatr. (2012) 161: 506–512.e1. https://doi.org/10.1016/j.jpeds.2012.02.048
8. Eleftheriou D., Brogan P.A. Therapeutic advances in the treatment of vasculitis. Pediatr Rheumatol Online J. (2016) 14: 26. https://doi.org/10.1186/s12969-016-0082-8
9. Yang H.M., Du Z.D., Fu P.P. Clinical features of recurrent Kawasaki disease and its risk factors. Eur J Pediatr. (2013) 172: 1641–1647. https://doi.org/10.1007/s00431-013-2101-9
10. Kusakawa S, Tatara K. Efficacies and risks of aspirin in the treatment of the Kawasaki disease. Prog Clin Biol Res. (1987) 250: 401–413.
11. Christian M. Hedrich, Anja Schnabl, Toni Hospach. Kawasaki disease. Front. Pediatr., 2018 Sec. Pediatric Immunology. https://doi.org/10.3389/fped.2018.00198
12. Shafferman A, Birmingham JD, Cron RQ. High dose Anakinra for treatment of severe neonatal Kawasaki disease: A case report. Pediatr Rheumatol Online J. (2014) 12: 26. https://doi.org/10.1186/1546-0096-12-26
13. Ankur Kumar Jindal, Rakesh Kumar Pilania, Sandesh Guleria, Pandiarajan Vignesh, et al. Kawasaki Disease in Children Older Than 10 Years: A Clinical Experience From Northwest India. Front. Pediatr., 2020. https://doi.org/10.3389/fped.2020.00024
14. https://ped-rheum.biomedcentral.com/articles/10.1186/1546-0096-12-37
15. https://www.ncbi.nlm.nih.gov/books/NBK537163/
16. https://www.ncbi.nlm.nih.gov/pmc/articles/PMC6374576/
17. https://www.mayoclinic.org/diseases-conditions/kawasaki-disease/symptoms-causes/syc-20354598
18. https://kidshealth.org/en/parents/kawasaki.html

Acknowledgement

We take this opportunity to thank the management of Paediatric Division of AIIMS, Raipur, Chhattisgarh, India, for giving me the permission to do a documented case study about the above mentioned case. I deeply appreciate the care and attention provided by the treating/consulting doctors and the medical staffs in providing the treatment and healthcare services to the child and their family satisfaction. I take this opportunity to thank to the parents of child for giving consent
in sharing the information.

Conflict of Interest

The authors declare no conflict of interest.