Quality care foster a positive outcome in infant with Kawasaki disease-case report

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KEYWORDS

Kawasaki disease, Infant, Coronary artery aneurysms, IVIG Therapy

ABSTRACT:

Background: Kawasaki disease (KD) is a rare, acute, multisystem vasculitis affecting medium and small sized vessels, typically occurring in children under 5 years of age. First described in 1961 by Dr. Tomisaku Kawasaki, the disease was initially termed "Mucocutaneous Lymph Node Syndrome."Now a days Kawasaki disease is known for its occurrence in small epidemics, especially in closed communities and has a higher incidence in Asian populations. The etiology remains unclear but environmental factors, infections, and genetic predisposition are believed to play a role. Kawasaki disease can cause serious cardiovascular complications such as coronary artery aneurysms leading to ischemic heart disease or even sudden death if not treated promptly.

Case Presentation: We report the case of 1-year-old female infant who developed KD and small coronary artery aneurysms despite timely treatment with intravenous immunoglobulin (IVIG). The patient presented with high-grade fever, rash, conjunctival congestion, strawberry tongue and mild bilateral cervical lymph-adenopathy. Despite receiving IVIG, small coronary artery aneurysms were noted on echocardiogram.

Conclusion: The case report underlines the importance of early diagnosis, monitoring the child, treatment in preventingsevere cardiovascular complications and providing health care in Kawasaki disease. Even with timely IVIG therapy, the development of coronary artery aneurysms remains a significant concern, especially in younger patients with incomplete or atypical forms of the disease. This case also underscores the need for ongoing vigilance in monitoring these patients for potential long-term cardiovascular issues and positive outcome of the Kawasaki diseas.

Keywords: Kawasaki disease, Infant, Coronary artery aneurysms, IVIG Therapy

1. Background

Usually affecting infants and children under five years old, Kawasaki disease (KD) is an acute multisystem necrotising vasculitis of medium and small-sized arteries with an unclear aetiology.[1] Tomisaku Kawasaki initially described KD in 1967 under the name "muco-cutaneous lymph-node syndrome.[2,3] It is now well-known for occurring in tiny outbreaks, particularly in isolated groups, and for being more common among Asian people.[4,5]

A high fever lasting five or more days combined with at least four of the other five symptoms—bilateral conjunctival hyperaemia, lip ulcerations and oral cavity inflammation, polymorphous rash, oedema and desquamation of the extremities, and cervical lymphadenopathy—or fever accompanied by fewer than four diagnostic criteria and coronary artery echocardiogram abnormalities is the basis for diagnosing classic KD.[6,7] Twenty-five to thirty percent of children who do not receive treatment may develop coronary artery aneurysms, which can result in myocardial infarction (MI), ischaemic heart disease, or even sudden death.[8]

While the long-term objective of treatment, especially in patients with coronary ectasias or aneurysms, is to minimize myocardial damage, the acute phase of treatment aims to reduce inflammation in the wall of the coronary artery and prevent coronary thrombosis.[9,10]

KD is still an illness with a number of issues nowadays. The three biggest challenges for doctors are treating refractory forms, preventing cardiovascular consequences, and making an accurate diagnosis in a timely manner. The number of refractory forms has been steadily rising, and two significant risk factors appear to be the patient's youth and the delay in beginning treatment.[11,12]

We describe a case of a 1-year-old male infant with KD who developed severe coronary artery aneurysms despite an early diagnosis and a timely administration of intravenous immunoglobulin (IVIG).

Case presentation

A 1-year-old female infant who developed KD and small coronary artery aneurysms despite timely treatment with intravenous immunoglobulin (IVIG). The child presented with high-grade fever, rash, conjunctival congestion, strawberry tongue, and mild bilateral cervical lymphadenopathy. Despite receiving IVIG, small coronary artery aneurysms were noted on echocardiogram.

Figure 1

Clinical Course: The child was admitted with a 24-hour history of high-grade intermittent fever, which was unresponsive to acetaminophen. She also presented with an erythematous rash that initially appeared on her back and spread to the chest and legs, accompanied by vomiting and poor oral intake. She had a history of urinary tract infection (UTI) treated with cefixime 10 days prior to this admission.

Figure 2

On physical examination: she manifested Congested throat, Erythematous rash over the body, Right-sided cervical lymphadenopathy (1x2 cm),Strawberries tongue, Bilateral mild oral congestion, Lip erythema, Mild conjunctival congestion.

Figure 3

Laboratory results showed:**Table:1**

Initial findings

Sl.no	Laboratory findings	Patient value
1	Hemoglobin	8.3 g/dL,
2	Platelet count	582,000/mm³
3	White blood cells	22,900/mm³ (neutrophils70%, toxic changes)
4	C-reactive protein (CRP)	190 mg/L (elevated)
5	Albumin	2.9g/dL (hypoalbuminemia)
6	Sodium	133mEq/L (hyponatremia)

Laboratory Findings:

Table:2 Comparing laboratory findings day-1,day-3,day-6.

DAY	Hemoglobin (g/dL)	Platelets (x10⁶/mm³)	WBC (x10³/mm³)	Neutrophils (%)	CRP (mg/L)	Sodium (mEq/L)	Albumin (g/dL)
1	9.7	583,000	22,900	74%	190	133	2.9
3	8.3	405,000	9,100	58%	25.8	133	2.9
6	7.7	301,000	7,900	58%	1726	133	2.9

Echocardiogram Findings:

Echocardiogram showed the small aneurysms in the LMCA(Left Main Coronary Artery) (2.6 mm), LAD (2.4 mm), and LCX (2.3 mm).

No significant increase in z-scores of LMCA and LAD after 10 days.

Treatment:

Despite the high suspicion of KD, the childs's condition remained unresponsive to antibiotics, and fever spikes persisted. Given the features of strawberry tongue, erythema of the oral and pharyngeal mucosa, and bilateral conjunctival congestion, a diagnosis of KD was confirmed. IVIG was administered at 2g/kg over 12 hours. After IVIG administration, the patient's fever resolved, and there were no adverse reactions to the treatment.

On the 6th day of illness, an echocardiogram revealed small coronary artery aneurysms involving the left main coronary artery, left anterior descending artery, and left circumflex artery. The patient was initiated on low-dose aspirin at 5 mg/kg/day.

Quality care and nurses role:

1. Performed a comprehensive evaluation of the child's medical history, taking into account the presence of related symptoms, the length of the fever, and the beginning and progression of symptoms.
2. Evaluation of heart function, which includes keeping an eye out for myocarditis symptoms such chest pain, irregular heartbeats, and variations in electrocardiogram (ECG) values.
3. Evaluation of systemic involvement, which includes mucous membrane alterations, rash, conjunctival injection, and hand and foot redness or swelling.
4. Tracked laboratory results, including liver function tests, C-reactive protein (CRP), erythrocyte sedimentation rate (ESR), and complete blood count (CBC)..
5. Regularly monitored vital signs, including temperature, blood pressure, heart rate, and respiratory rate, to assess for changes and potential cardiovascular complications.
6. Examined the skin for characteristic findings, such as rash, erythema, and peeling, which are common in Kawasaki disease.
7. . Periodically and occasionally evaluated cardiac function by auscultating the heart for abnormal sounds or murmurs and keeping an eye out for heart failure symptoms.
8. **Administration of Intravenous immunoglobulin (IvIg):** 2 g/kg as a single IV infusion on diagnosis given

    - IVIg protocol followed throughout the given period(the first 10 days of illness the child on continuous monitoring of fever or inflammation.
    - Children who do not respond to the first dose, as evidenced by a persistent or recurrent fever 36 hours after the end of the first IVIg infusion, should receive a second dose of 2 g/kg IVIg.
    - One rare but known side effect of IVIg infusion, especially in children getting numerous doses, is haemolytic anaemia. It usually happens up to a week following the administration of IVIg.
    - After IVIg vaccination: according to the National Immunisation Handbook, live vaccines (such as varicella and measles) should be postponed. Vaccinate the child and then re-vaccinate them after the proper time if they are at high risk of contracting measles.

Complications:

Kawasaki illness has the potential to seriously harm the heart muscle and coronary arteries if treatment is not received.[13-16].

The following are possible side effects:

- Aneurysms (weak, protruding patches in blood vessels)
- Arrhythmias, or irregular heartbeats

- The potential for heart attacks due to blood clots
- Heart valve issues; coronary arteritis, or inflammation of the coronary arteries
- Myocarditis, or heart muscle inflammations

Discussion:

While KD is usually a self-limiting vasculitis, approximately 25–30% of patients who do not receive treatment may develop coronary artery aneurysms. For children with acquired heart disease, this illness is the leading cause of death and the most important risk factor for poor prognosis.17. Since coronary anomalies are more prevalent in the subacute stage of the disease, the best predictive risk indicator for identifying them is the Harada and Beiser scores.[18]

One of the most important therapeutic skills for preventing cardiac problems is prompt diagnosis and therapy initiation.[19,20] In our instance, we saw an early change in the RCA and LAD walls six days after the fever started, and an increase in the RCA diameter of 0.41 cm during the next few days. KD typically occurs most frequently in children under five, whereas it is extremely uncommon in those under three months old (1.6%).[21]

Although the exact cause of KD is unknown, it is thought to be caused by a combination of microbial infection, the immune system, or genetic predisposition. This is similar to what has been proposed for atopic diseases, and a lot of attention has been paid to the role of several genes linked to inflammation.[5,22]

The clinical and epidemiological features of KD also suggest that infectious agents may be the cause of the disease's development, despite the fact that no specific pathogens have been identified and recent research has demonstrated a significant contribution of the innate immune system to the pathophysiology of the acute phase of the disease.[23] Infants six months of age or younger frequently have incomplete clinical characteristics, and a larger percentage of coronary artery abnormalities is associated with this phenotype. Clinical and biochemical remission occurs in 80–90% of patients receiving standard therapy; for the remaining patients, a persistent fever is a sign of IVIG non responsiveness, the main risk factor for the development of coronary artery lesions.[24]

Three different grading systems were created by Egami and Sano to identify which patients are most likely to not react to IVIG therapy.[25] In accordance with these ratings, our patient's value was higher than the threshold for each. It is yet unknown how IVMP is used and how effective it is in refractory forms. Some writers claim that using IVMP appears to be linked to a reduction in fever. Although some other writers proposed a preventive function if IVMP is taken in the early stages of the disease, its impact on the onset of coronary artery abnormalities is yet unknown.[26]

The clinical evolution of our patient indicates that Kawasaki Disease (KD) presents a significant risk of severe manifestations and lack of therapeutic response prior to three months of age. In fact, in our

patient an early diagnosis and a timely starting of IVIG therapy was ineffective in preventing coronary artery aneurysms.

Although the early resolution of systemic inflammation appears to be the therapy's objective, the secret to preventing this deadly cardiac involvement is still unknown. Taking into consideration the above, we would even at the early stages of the disease recommend IVMP linked to IVIG, if the score indexes are predictive, as there is no standardized regimen yet for resistant forms with a high chance of developing coronary artery abnormalities. Moreover, since the development of KD seems to be greatly influenced by the inflammatory and immunologic processes of the innate immune system, even infusion of infliximab or plasma exchange can be important rescue techniques for kids who are resistant to IVIG.[28-30]

Conclusion

The case report underlines the importance of early diagnosis, monitoring the child, treatment in preventing severe cardiovascular complications and providing health care in Kawasaki disease. Even with timely IVIG therapy, the development of coronary artery aneurysms remains a significant concern, especially in younger patients with incomplete or atypical forms of the disease. This case also underscores the need for ongoing vigilance in monitoring these patients for potential long-term cardiovascular issues and positive outcome of the Kawasaki disease.

Consent

Written informed consent was obtained from the parents of the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the editor-in-Chief of this journal.

Abbreviation

KD: Kawasaki Disease,IVIG: Intravenous Immunoglobulin,LMCA: Left Main Coronary Artery,LAD: Left Anterior Descending Artery,LCX: Left Circumflex Artery,RCA: Right Coronary Artery

Author’s contributions

All the authors participated sufficiently in preparation of this manuscript followed the child in clinical course and revised the literature and final analysis with critical revision .All authors gave final approval for manuscript for publication.

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