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KAWASAKI DISEASE: TWO CASE REPORTS FROM SECONDARY LEVEL HEALTH CARE FACILITY

Pediatrics

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ABSTRACT

Kawasaki disease (KD) is an acute febrile vasculitis. In the differential diagnosis KD should be considered as fever for at least several days of duration, nonpurulent conjunctivitis, and rashes especially in children younger than 1 year old and in adolescents, in whom the diagnosis is missed most frequently. Here we described the case 1 of an 11-month-old female presented in January 2019, with a 5 days history of fever which was low grade intermittent initially and was relived with antipyretics while three days later fever became high grade, intermittent nature associated with marked irritability and not responding to antipyretics and case 2 of A 15-month-old male presented to the paediatric department with a 10-day history of high-grade fever up to 103o F. Initially the patient was admitted to a private hospital and received IV antibiotics there, and due to no improvement patient was referred to regional hospital Bilaspur. In case 1 infantile KD was diagnosed and in case 2 the diagnosis was classical Kawasaki disease. Any child presenting with a acute high grade fever with irritability and mucocutaneous involvement should promptly be evaluated and managed accordingly.

KEYWORDS

Kawasaki disease, secondary level health care facility

INTRODUCTION:

Kawasaki disease (KD) is an acute febrile vasculitis. The peak incidence of KD is in the age group of a half year to 2 years, which includes around half of all KD patients [1,2]. KD is the predominant reason for paediatric acquired heart disease in developed countries [3]. Cardiac lesions, like coronary artery aneurysms, are a hallmark of KD [4–6]. If the disease is left untreated, the coronary artery complications may develop in up to 25% of affected cases, which leads to mortality in approx. 2% of the cases [7–10]. Prevention of aneurysms is the primary target for paediatricians treating patients with KD. Aneurysms prevention is the primary target for paediatricians treating patients with KD. Coronary artery abnormalities can be reduced from 25% to 5% by prompt diagnosis and the administration of intravenous immunoglobulin (IVIG)[11]. In the differential diagnosis, KD should be considered as fever for at least several days of duration, non-purulent conjunctivitis, and rashes especially in children younger than 1-year-old and in adolescents, in whom the diagnosis is missed most frequently.

"atypical KD" is the term which is interchangeable with "incomplete KD", has recently been used to describe patients with an incomplete presentation of the disease, regardless of the presence of coronary complications [12,13]. Classic (typical) Kawasaki disease is diagnosed on the basis of the presence of fever lasting five or more days, accompanied by four out of five findings: oral changes such as cracked and erythematous lips and strawberry tongue, bilateral non purulent conjunctival injection, cervical lymphadenopathy, extremity changes such as erythema or palm and sole desquamation, and polymorphous rash.

The diagnosis of KD is difficult in infants because of its atypical presentation mostly and similar to that of other diseases. The most challenging thing is to distinguish between KD and enteroviral meningitis in febrile infants with cerebrospinal fluid pleocytosis [14]. Elevation in inflammatory markers including ESR, CRP, and platelet count is frequently associated with KD. Other findings like low sodium levels, high white blood cell (WBC) count (neutrophilic type), nonseptic pyuria, hypoalbuminemia, or elevated liver enzymes may supplement the diagnosis.

Some researchers have reported that infants younger than a-half-year old take the longest time to diagnose, have the least favorable laboratory results, and are the least likely to fulfill the major clinical criteria, all of which are risk factors for developing coronary artery abnormalities[14–16].

Case presentations

Case 1:

An 11-month-old female presented in January 2019, with a 5 days history of fever which was low grade intermittent initially and was relived with antipyretics while three days later fever became high grade, intermittent nature associated with marked irritability and not responding to antipyretics. On the third day of illness redness of lips and tongue, reddish rashes over the body, and swelling over hand and feet were noticed by her parents. The patient also has a history of decreased oral intake, marked irritability, and a history of redness in eye and discharge. The patient was initially taking treatment on an OPD basis, but due to no improvement, the child was admitted to the general pediatric ward of govt hospital and treated like acute febrile illness/ acute upper respiratory infection. The patient also had a history seizure triggered by fever. The patient was initially treated with antibiotics and anti-allergic with antipyretics. However, as there was no improvement, the child was shifted to govt pediatrics HDU. She was admitted there from 22-1-19 to 24-01-19 and suspected to have Kawasaki disease based on clinical presentation which were further supported by laboratory parameters.

On general physical examination, the Patient had unilateral left cervical non-tender lymphadenopathy of size 0.5 cm * 1 cm. The patient had oedema over the dorsal aspect of palm and soles, strawberry tongue and red cracked lips, conjunctival injection with sparing of the limbus. A polymorphic rash was present over trunk and limbs. Chromonychia was also present.

When the child was admitted to paediatric HDU patient she had leucocytosis with neutrophilia, normocytic normochromic anaemia's, high ESR and CRP, sterile pyuria, and hyponatremia. The patient had classical signs of Kawasaki disease. A diagnosis of Kawasaki disease was made therefore intravenous immunoglobulin (IVIG) at 2gm per kg and tablet ecspirin was given as per protocol.

After IVIG dose there was no recurrence of fever child activities improved. Inflammatory markers were on a declining trend. At the time of discharge, the patient had periungual peeling. On USG abdomen no evidence of GB hydrops was present. Echocardiography was normal. At discharge, the child was better and was on low dose ecospirin as per protocol.

Table : 1 Significant laboratory findings

S. No	Investigations	Day 1 of admission in general ward	Day 1 in HDU	Last day in HDU
1	Hb (gm/dl)	9.9	7.5	9.1
2	TLC	12600	18100	14900
3	Platelets	266000	254000	532000
4	ESR (mm/1st hr)	65	115	108
5	Na	134	135	135
6	K	4.1	4.82	5.7
7	CRP (mg/dl)	24	24	12
8	Urine R/E M/E	3 to 5 pus cells	NAD	NAD

Case 2

A 15-month-old male child presented to the paediatric department with a 10-day history of high-grade fever up to 103° F. Initially the patient was admitted to a private hospital and received IV antibiotics there, and due to no improvement patient was referred to regional hospital Bilaspur.

From the last 5 days, the patient developed redness of lips and tongue. Since the last 2-4 days non-purulent conjunctivitis, peeling of skin over the perioral area, swollen fingertips, and excessive irritability was observed. On examination the patient was febrile, irritable, erythematous rash over the trunk and body, strawberry tongue, and skin peeling was present in the perioral and periungual region.

The patient was given IVIG at 2g/kg dose and ecospirin was given 6 hourly as per protocol. Echocardiography was normal. The patient was discharged from the hospital in good condition on 5th day on low dose aspirin as per protocol.

Table no: 2 Significant laboratory findings

S. No	Investigations	Day1	Day2	Last day
1	Hb	9.6	9.5	9.4
2	TLC	16300	14200	16000
3	platetes	685000	528000	400000
4	ESR (mm/1st hr)	105	90	92
5	Na	128.7	135	139
6	K	4.23		
7	CRP (mg/dl)	148	24	6
8	Urine R/E M/E	Sterile pyuria present	NAD	NAD

DISCUSSION:

In our study, the 1st case was 11-month-old infant and 2nd case was 15 months old, 80% of patients with Kawasaki disease are under the age of 4 years, and with the peak incidence occurring at 9 to 11 months of age [17]. The atypical presentations (longer duration of disease before diagnosis, lower incidence of rash, lower incidence of extremity change, lower incidence of conjunctivitis, and lower C-reactive protein) in infants are common, that can lead to a delay in diagnosis and effective treatment[18].

The children had classical mucocutaneous features that are key to making the diagnosis. In case 1, the infant was showing redness of lips, tongue, and eye, reddish rashes over the body, and swelling over hand and feet was noticed by her parents. In case 2 the infant was having fever from 10 days, Redness of lips and tongue from 5 days, Non purulent conjuctival discharge, peeling of the skin, swollen fingertips, and excessive irritability

Hematologic parameters can assist to confirm or support the diagnosis however they are not diagnostic of Kawasaki disease. Both our children were admitted with normal platelet counts which became elevated subsequently. This was the common finding in the second week of illness and was used as an adjunct in making a diagnosis of incomplete Kawasaki disease [9]. The CRP was raised in both the cases as expected and eventually reduced after the initiation of aspirin. Both the cases were treated with aspirin and IVIG. In the current senerio, the IVIG is an expensive drug that is not readily available or affordable to most families in our population. This hampers effective treatment which is essential to reduce cardiovascular sequelae.

Corticosteroids could be used as other treatment options. In a review of 7 trials showed that using steroids as an adjunct therapy to IVIG early in the course of illness could benefit in reducing cardiac complications [19]. In low resource settings steroids could be used to reduce the cardiovascular morbidity if IVIG is not available, [20]. Other agents like infliximab have shown promising results when used as adjuncts to IVIG but are also not widely available [21].

The echocardiograms for both children were normal at baseline and on subsequent follow-ups. This was reassuring since the patients who are not treated with IVIG have a high incidence of coronary abnormalities (25–30%) [22]. For follow up the cardiac sequelae in children with Kawasaki disease, echo is a vital modality. However, it is not widely available in most resource-limited settings and requires specialized training to operate.

These 2 cases were seen over a 1.5 year period at a secondary level health care facility. This raises concerns about how many missed cases there may be which are not diagnosed or managed effectively since the fever and mucocutaneous changes eventually resolve within 3 weeks even without specific treatment. In children with a missed diagnosis of this condition the subsequent devastating cardiovascular morbidity remains unknown.

CONCLUSION:

In case 1 infantile KD was diagnosed and in case 2 the diagnosis was classical Kawasaki disease. A diagnosis of Kawasaki disease is entertained for any febrile illness lasting longer than 5 days and presenting with mucocutaneous findings. Any child presenting with a prolonged fever should promptly be referred and evaluated. Doctors and other care providers should be made familiar with this condition in order to diagnose, treat, and prevent cardiovascular morbidity and mortality.

Abbreviations

CBC: Complete blood count
IVIG: Intravenous immunoglobulin
WBC: White blood cells

Declaration of patient consent:

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity.

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