CASE REPORT

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A 1 YEAR 2 MONTHS OLD CHILD WITH KAWASAKI DISEASE - A CASE REPORT

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ABSTRACT

Kawasaki disease (KD) is a self-limiting and acute medium vessel vasculitis the causes of which are unknown and takes place predominantly in infants and children. The disease in its acute phase is self-limited and the diagnosis may be missed; if untreated, KD can result in aneurysm of the coronary artery in 25% of patients. Principal criteria for diagnosing the disease is fever of more than 5 days and presence of a minimum of the following 4 principal features: erythema and cracking of lips, strawberry tongue, bilateral non-purulent conjunctival injection, rash, erythema and edema of the hands or feet, cervical lymphadenopathy >1.5cm, usually unilateral. Our patient presented with all the above features.

Additional features like irritability, arthritis, desquamation, vomiting, diarrhea, abdominal pain, Bacillus Calmette-Guérin (BCG) induration.Diagnosis of KD is mainly clinical, but anemia, neutrophilic leukocytosis, thrombocytosis, raised Erythrocyte Sedimentation Rate (ESR) , raised aminotransferase, raised C-Reactive Protein (CRP), reduced serum albumin, pyuria may be present.

The investigation findings of this patient indicated towards the diagnosis of KD. Echocardiogram was done and coronary artery dilatation was noted. The patient was managed using a single dose of intravenous immunoglobulin(IVIG) 2g/kg administered over a period of over 10 hours which was followed by aspirin 75mg/kg until the patient became afebrile. This dose of aspirin was continued for another 3 days then 5mg/kg of aspirin was given for 8 weeks. During follow-upthe patient'scondition was uneventful.

Keywords: Self-limiting disease, Acute phase Vasculitis, Coronary aneurysms

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INTRODUCTION

Kawasaki disease, formerly known as mucocutaneous lymph node syndrome and infantile polyarteritis nodosa, is a disease that is acute in nature with fever occurring in children of <5 years of age in 80% of cases. It occurs mainly in Japan and other countries of Asia, but those belonging to ethnicity other than that may also suffer[1].

Etiology of KD is unknown, however, it is considered as an abnormal response of the immune system due to some infectious trigger. A syndrome, Multisystem Inflammatory Syndrome in Children (MIS-C), with some similarities to KD has been, in recent times, reported as a complication of COVID-19 infection in children[2].

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Every two years, surveys have been done in Japan for monitoring trends of incidence of KD.The record reached its highest in 2010 when KD incidence was observed for every 100,000 children (ages 0 to 4 years) to be 239.6 with the highest rate in children of 6-11months age. Infants <6 month and children >5 year were noted to be prone to highest risk of developingcoronary artery abnormalities (CAA) in the latest Japanese survey.

Approximately 20%-25% of children who remained untreatedsuffered from development of CAA including aneurysms whereas <5% of children treated with IVIG develop CAA[3].

Predictors of poor outcome across several studies include male gender, young age, persistent fever, poor response to IVIG, and laboratory abnormalities including neutrophilia, thrombocytopenia, raised transaminases, hyponatraemia, hypoalbuminemia, elevated levels of N-terminal- pro brain natriuretic protein (NT-ProBNP) and elevated CRP levels[3].

PATHOLOGY

KD is a vasculitis that predominantly affects the medium sized arteries. The coronary arteries are involved most commonly, even though other arteries, like the popliteal and brachial arteries can also develop dilation. Neutrophilic necrotizing arteritis is found in KD. Subacute/chronic vasculitis characterized by presence of lymphocytes, plasma cells, and eosinophils (lasting weeks to years)results in fusiform aneurysms.This inflammation of blood vessel leads to conversion of smooth muscle cells into myofibroblast resulting in the development of progressive stenosis. Thrombi may also form in the lumen and obstruct blood flow[1].

Epidemiologic definition (Classic clinical criteria).

Fever continuing for a minimum of 5 days.

Existence of at least four of the following principal features:1)Bilateral non-purulent conjunctival injection.2) Changes of the mucosa of the oropharynx, including injected pharynx, injected and/ or dryfissured lips.3) Strawberry tongue. 4) alterations of the peripheral extremities, such as edema and/or erythema of the limbs, desquamation, usually beginning periungually. 5) Rash, primarily truncal: polymorphous but not vesicular. Cervical lymphadenopathy >1.5 cm; mainly unilateral.6) Illness not explained by other known disease process.

Patients (having fever for a minimum of 5 days, and less than 4 main criteria)may be diagnosed with KD when CAA are found by two-dimensional angiogram or echocardiogram.

The diagnosis is clinical but following laboratory investigations suggest KD (Table 1).

Table 1: Laboratory investigations findings suggestive of KD

Laboratory investigations suggestive of KD
Elevated Erythrocyte Sedementation Rate
Elevated C reactive protein.
Elevated NT-ProBNP (B-type natriuretic peptide)
Anemia
Abnormal plasma lipids
Hypoalbuminemia

Table 1: Continued

Laboratory investigations suggestive of KD
Thrombocytosis after 1 week
Sterile purpura
Elevated serum transaminase
Pleocytosis of cerebrospinal fluid
Leukocytosis in synovial fluid
Echocardiography showing coronary artery dilation
Leukocytosis with neutrophilia and immature forms[4]

TREATMENT

Administration of IVIG 2g/kg over 10 hours (single dose) to decrease incidence of coronary artery complications. Administration of Aspirin 75-80 mg/kg/day in 3-4 divided doses until the child becomes afebrile. Later on, Aspirin is continued in doses of 5mg/kg/day in divided dose daily as an anti-thrombotic agent for 6-8 weeks[4].

Case History

A 1 year 2 months old female child fully immunized as per Expanded Program on Immunization (EPI) schedule, hailing from Dhaka got admitted on 10.09.2024 in a private hospital of Dhaka city with the complaints of fever for 5 days, rash for 1 day.

The patient was suffering from a high grade continuous fever that was not associated with chills and rigors and did not subside by taking acetaminophen. Rash had also developedon the chest for 1 day. There was no history of vomiting, headache, burning sensation of micturition and travelling history to malaria and kala-azarendemic zone.

On examination: Patient was febrile (temperature :103^0 F), maculopapular rash on the trunk, the tongue was the color of strawberry, Bilateral non purulent conjunctival injection was noted in the eye. Lips were cracked (Figure 1); erythema and oedema of hands and feet were observed. Cervical lymph node (left) were enlarged (about1.5 cm), was non tender, and not suppurative. No organomegaly was found. Signs of meningial irritation were absent.

Investigation findings:
Complete blood count showed
Hemoglobin%: 8.7 g/dL.
ESR- 143 mm in 1st four,
WBC -16700/cumm,
Neutrophil-74%,
Platelet Count: 444 k/L,
CRP- 201.4 mg/L,
Serum Glutamic Pyruvic Transaminase
(SGPT)- 135 u/L,
Serum albumin- 19.6 g/L,
Serum electrolytes: Normal,
Urine: Routine Microscopic Examination:
Pus cells. 35-40/High Power Field

Echocardiograph shows dilated distal right coronary artery, left anterior descending artery, right coronary artery is prominent, left anterior descending arterylumen irregular and blurred.

Probable diagnosis:

All findings were suggestive of Kawasaki disease.

Figure 1: Irritable Child having cracked lip and strawberry tongue

Aspirin 75 mg/kg until fever subsided. After subsiding of fever, Aspirin 5 mg/kg (another 8 weeks) was given. After 6 weeks few investigations were done like CBC, CRP, Which was almost normal; follow up ECHO at 6 weeks shows, Seen post Kawasaki disease (borderline distal RCA). Follow up ECHO after 3 months which was normal. During second follow up patient is apparently normal and healthy.

DISCUSSION

KD is a medium vessel vasculitis first described by Japanese pediatrician Tomi Saku Kawasaki in 1967. Although it is considered to be a rare condition, KD has become the most common reason for development of heart disease (acquired); the cause of KD still remains unknown but may be triggered by environmental or infectious agent (viral and/or bacterial)[5]. Asian children, especially of Japanese ancestry have the relative greater risk for developing KD. The incidence rate and number of patients with KD in Japan continue to increase. Epidemics of KD primarily occur in the spring and late winter at two to three year intervals[6,7]. In Bangladesh KD is increasing day by day.

Our patient suffered from KD during the spring season. KD is diagnosed in the existence of fever for a minimum of 5 days together with at least 4 of 5 principal clinical features in absence of other known diseases; erythema and crackling of lips, strawberry tongue, bilateral bulbar non purulent conjunctival injection, rash, erythema and oedema of the limbs, cervical lymphadenopathy 1.5 cm(usually unilateral); and illness not explained by other known disease. Our patient had all the above features. Liu et al. noted irritability, convulsion, headache, lethargy that are the additional features of KD[8]. Our patient had irritability at initial period. Wang et al. reported rash (90%), conjunctivitis (90%), pyuria (50%) lymphadenopathy (25-30%) in 110 KD cases[9]. Our patient had features of rash,

conjunctivitis, pyuria and lymphadenopathy.Uehara et al. reported and proposed that redness at the BCG vaccination site is a valuable indicator for diagnosis of KD[10]. New burger et al. observed anemia, platelet count >=450,000, albumin level <=3g/dl, elevated alanine transaminase, white blood cell count >=15,000/mm^3,urine white blood cells >=10/ high powered field and echocardiogram findings that indicated towards KD[11].Our patient had anemia, neutrophilic leukocytosis, raised transaminase, pyuria and coronary artery dilatation.

Chaiyarak et al. found ESR level > 40mm in 1st hour which exhibited a sensitivity of 90.5%, but a specificity of merely 66.67% among 114 KD patients[12].Tsai et al.noted CRP exceeding 24 mg/L and ALT exceeding 30 U/L [13].Our patient had CRP 201.16 mg/dl and ALT 135 U/L. There are many associated manifestations for KD like meatitis, perineal erythema and desquamation, arthralgia, arthritis, abdominal pain,diarrhea, hepatitis, obstructive jaundice, hydrops of the gall bladder, pulmonary infiltrates, pleural effusions, uveitis, sensory hearing loss, and cardiovascular manifestations[14.15].

Management of the disease in its acute phase is directed at limiting inflammation. Treatment with IVIG in different regimens has been found to significantly reduce myocardial inflammation and incidence of CAA formation, as well as lead to a rapid defervescence and more rapid normalization of acute phase reactants [16,17].

We managed the patients with IVIG 2g/kg over 10 hours; associated with aspirin 75mg/kg in divided dose for 3 days; then 5mg/kg for 8 weeks. During follow up patient remained afebrile; Vital signs were within normal limit and there were no cardiac complication.

CONCLUSION

KD is an self limited acute vasculitis that typically occurs in young children. KD targets the coronary arteries and other cardiovascular structures. Approximately 1 in 5 children who do not receive IVIG during the disease's acute phase, develop CAA. Recurrence is unusual; in Japan the recurrence rate is 3% and 1% in North America. With prompt treatment, the prognosis is good.

CONFLICT OF INTEREST

There is no conflict of interest.

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