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Atypical Infantile Kawasaki Disease with Rare Presentation of Purulent Conjuctivitis: Case Report

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Abstract
Kawasaki disease (KD) is a medium vessel vasculitis which presents as an acute febrile illness affecting young children with 80% affecting less than 5 years and with slight male preponderance. KD remains purely a clinical diagnosis rests on the recognition of a typical temporal sequence of a constellation of clinical features. The main concern about the KD is development of coronary artery abnormalities (CAA) in 15-25% of affected individuals. The diagnosis of infantile KD can be particularly difficult due to obscure clinical manifestations and presentation is usually incomplete or atypical. Adenovirus poses the greatest diagnostic dilemma which presents with many of the features consistent with the atypical KD. We are illustrating a case of infant with features temporally correlating with Kawasaki disease but some atypical features like purulent conjunctivitis.

Keywords: *Kawasaki disease, Atypical, Purulent conjunctivitis, Mucocutaneus lymph node syndrome.*

Introduction

Kawasaki disease (KD) was first reported from Japan in 1967 by, Tomisaku Kawasaki at Red cross hospital in Tokyo. He described 50 children who appeared to have a unique set of clinical features which he called the 'Mucocutaneous lymph node syndrome'[1],[2],[3]. Kawasaki disease (KD) is a medium vessel vasculitis which presents as an acute febrile illness affecting young children with 80% affecting less than 5 years and with slight male preponderance[1],[2],[3]. KD remains purely a clinical diagnosis rests on the recognition of a typical temporal sequence of a constellation of clinical features, with none of the features taken individually being of any diagnostic significance whatsoever. Moreover, these clinical features may change from day to day, the spectrum evolves over a period of time i.e 1-3 weeks and the entire clinical spectrum is not seen at any one particular point of time. There is no laboratory test or marker which is pathognomonic of the condition. In the absence of a specific laboratory test for KD, a set of clinical criteria have been established to assist the physician in arriving at a diagnosis[2],[3],[4]. (Table 1)

Table 1: Diagnostic criteria for kawasaki disease

1. Fever of at least five days duration.
2. Presence of any four of the following 5 features:
• Changes in extremities (periungual desquamation, indurated edema)
• Polymorphous exanthema(maculopapular rash)
• Bilateral conjuctival injection (non purulent with limbal sparing)
• Changes in the lips and oral cavity
• Cervical lymphadenopathy (unilateral >1.5 cm)
3. Exclusion of other diseases with similar findings

The main concern about the KD is development of coronary artery abnormalities (CAA) in 15-25% of affected individuals that may lead to significant long term cardiac morbidity, if not diagnosed and treated in time[1],[2]. The differential diagnoses of KD are viral infections, group A streptococcus infection, Ebstein-barr virus, measles, collegen vascular disorders and drug reactions. When a patient has clinical features not commonly associated with this condition, a diagnosis of "atypical" KD can be made[2],[3],[4]. Adenovirus poses the greatest diagnostic dilemma which presents with many of the features consistent with the atypical KD. We are illustrating a case of infant with features temporally correlating with Kawasaki disease but some atypical features like purulent conjunctivitis.

Case Report

Nine month old infant presented to the outpatient department with complaints of high grade fever for 5 days along with generalized maculopapular rash, irritability and poor oral acceptance. Initial possibility of infective aetiology kept, relevant investigations (Table 2) were sent and started empirically on 3rd generation cephalosporins. In view of persistent fever spikes antibiotics were escalated to meropenum and vancomycin. On day 9 of illness patient developed indurated oedema of dorsal aspect of bilateral hands and feet, angular chelitis (Fig. 2), bilateral purulent conjunctivitis (Fig. 2) and perianal desquamation (Fig 3). Though there were some atypical clinical feature but, Keeping in view the temporal sequence of clinical findings, some suggestive investigations (table 1) and fever not responding to antibiotics possibility of atypical KD was kept. Rapid direct fluorescent antigen test to rule out acute adenoviral infection was done as the clinical feature of purulent conjunctivitis is more commonly Associated with it, which was unremarkable. Intravenous immunoglobulins and aspirin were stared which was followed by prompt resolution of fever spikes. In the second week of illness patient developed periungual desquamation (Fig. 1), leucocytosis, thrombocytosis and suggestive echo with minimal pericardial effusion but with no coronary involvement even on long term follow up of 6 months.

Table 2: Investigation chart

S.NO.	INVESTIGATIONS	DAY 5	2ND WEEK
1.	Hemoglobin	8.8 g/dl	
2.	Total leucocytes count	5380 thou/µl	21530 thou/µl
3.	platelets	180000 thou/µl	434000 thou/µl
4.	ESR	48 mm 1st hr	
5.	CRP	Positive	
6.	Total protein	6.5 g/dl	
7.	Albumin	3 g/dl	
8.	SGOT	34 u/l	
9.	SGPT	22 u/l	
10.	Urine routine/Microscopy	Normal	
11.	USG abdomen	Normal	
12.	ECHO		Minimal pericardial effusion with normal coronary arteries.

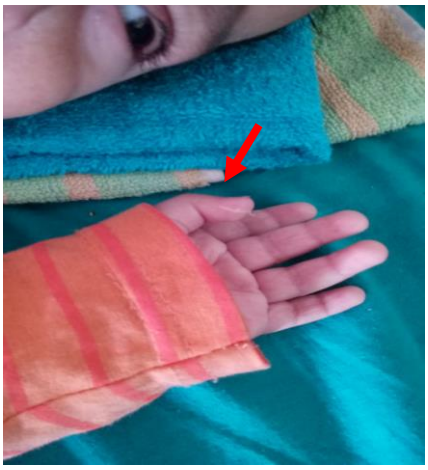

Fig 1 Perungual desquamation

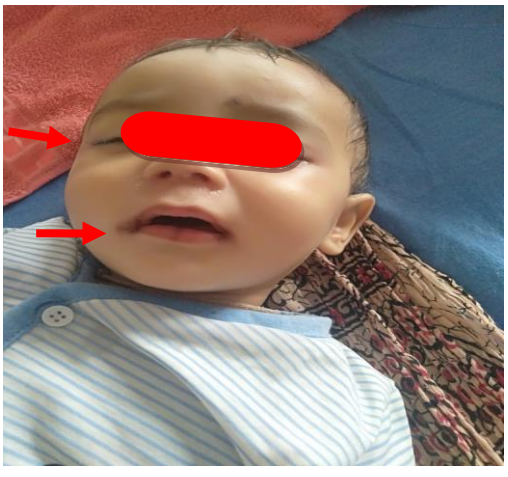

Fig. 2 Angular chilitis with purulent conuctivitis

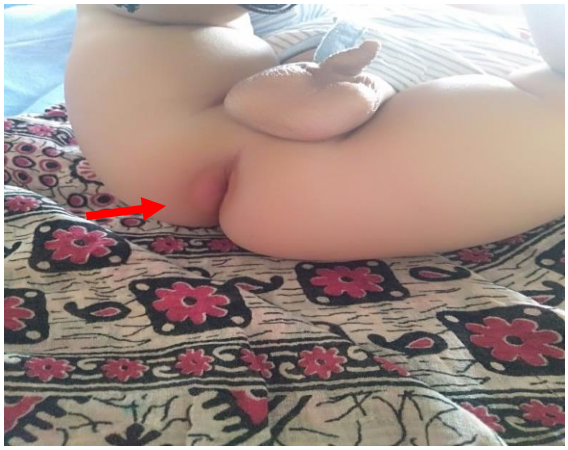

Fig. 3 Perianal desquamation.

Discussion

Kawasaki disease (KD) is an acute febrile vasculitis. The peak incidence of KD is from 6 months to 2 years of age. KD is the predominant cause of paediatric acquired heart disease. Cardiac lesions, such as coronary artery aneurysms, are a hallmark of KD[5],[6]. Prompt diagnosis and the administration of intravenous immunoglobulin (IVIG) can reduce the incidence of coronary artery abnormalities from 25% to 5%[7]. The diagnosis of infantile KD can be particularly difficult due to obscure clinical manifestations and presentation is usually incomplete or atypical. Some studies have reported that infants younger than 6 months old take the longest time to diagnose, are the least likely to fulfil the major clinical criteria, and have the least favourable laboratory results, all of which are risk factors for developing coronary artery abnormalities [8],[9].

Results of the study done by Shulman et al are consistent. They described 36 KD patients, who were less than l year of age during the pre-IVIG era and found that CAA developed in 31% compared with 18% in those who were 1 to 2 years of age and 10% who were more than 2 years of age[10]. The clinical challenge lies in distinguishing cases of infantile KD that do not fully meet the diagnostic criteria from those that strongly resemble a variety of common childhood disorders. Thus, have a high index of suspicion should always be kept while evaluating infantile KD, especially under 6months, with unexplained fever for more than 5 days, especially if unresponsive to antibiotics.

Current literature reports that infants below 12 months of age have a higher prevalence of incomplete and atypical KD (40%) compared to older patients (10–12%)[11]. Our case had a atypical presentation of purulent conjunctivitis. Study done by Stephen R barone et al demonstrated a statistically significant higher incidence of purulent conjunctivitis in acute adenoviral infection (43%) as compared to atypical KD (8%)[12], but the latter was ruled out by rapid direct fluorescent antigen assay in our case. Coronary artery aneurysm KD is frequently associated with elevation of inflammatory markers including ESR, CRP, and platelet count. Other laboratory findings such as high white blood cell (WBC) count (neutrophilic type), sterile pyuria, low sodium levels, hypoalbuminemia, or elevated liver enzymes may supplement the diagnosis. Delayed diagnosis and treatment, higher incidence of coronary arteries abnormalities, frequently occur in infantile KD, but thorough appropriate investigations must always be considered along with temporal sequence of a constellation of clinical features to supplement the diagnosis and exclusion of other disease with similar clinical picture to prevent unnecessary administration of immunoglobulin therapy.

Conclusions

KD clinical diagnosis below 1 year of age can be very challenging since patients may not have classic signs and symptoms, and individual manifestations may be subtle. Therefore, in young infants with unexplained fever lasting more than 5 days, a clinical possibility of KD must be considered and appropriate investigations performed. An early (<7days) intravenous immunoglobulins must be considered keeping high index of suspicion, after exclusion of the other common childhood diseases with similar manifestations.

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