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Kawasaki Disease at Chandigarh

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Kawasaki Disease (KD) is an acute febrile illness which mainly affects infants and children below 5 years of age and is characterized by a vasculitis involving the medium size arteries (1,2). It has been reported from all regions of the world (3,4) though reports from India have, till very recently, been few and far between (5-9). As there is no confirmatory laboratory test, the diagnosis may be missed if one is not familiar with the clinical features (3). The purpose of the present communication is to

share our experience with this condition over the last 3 years.

Subjects and Methods

The first patient with KD was diagnosed in May 1994 and since that time we have seen 9 children in whom a possibility of KD was kept in the differential diagnosis. These children were then kept under

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close observation. On follow-up a diagnosis of KD could be established in 6 of these 9 children using the standard diagnostic criteria⁽³⁾ (*Table I*).

All children were below 5 years, the mean age being 2.4 years (range 1.2 years-4.5 years). The male: female ratio was 2:1 (*Table I*). Clinical features seen in all included high grade fever for more than 5 days, extreme irritability, palmar erythema, truncal maculopapular rash (with marked perineal accentuation in 1 patient), changes in lips/oral mucosa and skin manifestations acrally distributed in the extremities. Of the latter, periungual desquamation of skin seen after the first week of illness was particularly characteristic. Four children had non-purulent conjunctival injection and an equal number had cervical lymphadenopathy. Two patients had prominent edema over dorsum of hands and feet. None of our patients had arthralgia/arthritis, hepatic dysfunction or diarrhea. Overt cardiac involvement was not seen in any of our cases.

Investigations showed a modest elevation of total leukocyte counts (mean 12850/cu mm, range 7000-21000/cu mm) with a polymorphonuclear predominance. Mean erythrocyte sedimentation rate at admission was 49 mm (range 45-55 mm) in 1st hour (normal 0-20 mm). C-reactive protein (CRP) was positive in all 5 children where

this test could be performed. All 6 children had elevated platelet counts during the acute phase, with the mean being 6.9 lakhs/cu mm (range 4.68-10.2 lakhs/cu mm; normal 1.5-4.0 lakhs/cu mm). Urinalysis did not show any abnormality in any of our children.

Electrocardiography and echocardiography were done in all children during the acute stage of illness and no significant abnormality was detected. Repeat echocardiographic examination at 1 year follow up is available in the first child only and that too does not show any coronary artery dilatation or aneurysm.

Treatment included use of intravenous immunoglobulin (IVIG) 0.4 grams/kg/day for 4-5 days, along with aspirin in high doses (100 mg/kg/day) which was continued till the acute stage was over. With resolution of fever and normalization of values of acute phase reactants, the dose of aspirin was decreased to 3-5 mg/kg/day.

All children showed prompt resolution of fever on initiation of IVIG therapy and are presently asymptomatic. The mean duration of follow-up is, however, only 9.5 months (range 2-27 months.)

Discussion

Kawasaki disease was first reported in 1967 from Japan and since then it has been

Table I- *Clinical features of Patients with Kawasaki Disease*

S. No.	Age (Yrs)/Sex	Fever for > 5 days	Conjunctival infection			Mucosal changes	Changes in extremities	Truncal rash	Cervical adenopathy
1.	1.5 M	+	+	+	+	+	+	-	
2.	4.0 F	+	-	+	+	+	+	+	
3.	1.3 M	+	+	+	+	+	+	+	
4.	1.2 F	+	+	+	+	+	+	+	
5.	2.2 M	+	-	+	+	+	+	+	
6.	4.5 M	+	+	+	+	+	+	+	

described from all over the world(1-4). It is by no means a rare disease (4). Though the highest reported incidence has been from Japan from where approximately 5000-5500 new cases are reported every year, comparable figures are available from any other countries (3,4). The disease has also been described to sometimes occur in epidemics (4).

This condition has been reported frequently from India and upto 1995 there were just 3 case reports of the disorder in the pediatric and medical literature from our country (5-7). Moreover in none of these 3 children was IVIG used during the acute stage. We have recently reported the first child in whom IVIG was successfully used during this stage for prevention of coronary aneurysms (8).

All children were below 5 years of age. KD is a disease of young children with more than 80% of reported cases being 5 years(3,4). The male-female ratio in our series is 2:1 which is comparable to that reported in the literature (1.4:1)(3,4). All our children had a truncal rash. However, it must be noted that this was never very prominent and is likely to be missed if not specially looked for. Perineal accentuation of this rash, which is known to occur in KD(3), was seen in only one case. Conjunctival injection in KD is non-purulent and can similarly be overlooked if the index of suspicion is not high. It should be noted that the minimum criterion for diagnosing cervical lymphadenopathy is one lymph node more than 1.5 cm in size(3). Sixty-six per cent of our children fulfilled this criterion. Cervical lymphadenopathy is the least specific of the KD criteria, being seen in only 50% of patients(3).

In our limited experience the most important clues to the diagnosis of KD are provided by the changes in lips/oral cavity

(*i.e.*, erythematous and fissured lips, injected pharynx, strawberry tongue) and acral skin manifestations in the extremities(3) (*i.e.*, edema/erythema of hands and feet, periungual desquamation in fingers and toes). Desquamation generally occurs only after the first week of illness and in the relevant clinical setting is very suggestive of KD (3,4).

Of the laboratory investigations though none is by itself confirmatory, thrombocytosis is quite characteristic in the acute phase of KD(4). Platelet counts may sometimes remain elevated for many weeks (4). Thrombo-cytosis is considered by many to be an acute phase phenomenon and is a useful marker of ongoing inflammation in KD, as it is in many other rheumatological conditions.

Many different treatment protocols for IVIG administration have been described in KD(3,4, 10-13). We have used the older schedule of IVIG administration for the treatment of our patients(3). More recently, a single dose administration of 2 g/kg of IVIG has been recommended(11). However, as this product is very expensive, many patients in our country may be unable to find sufficient resources for a single dose administration. Spreading the expenses over 4-5 days gives the family more time to arrange the necessary finances.

Although the follow-up period is limited/ none of our patients has so far developed coronary artery aneurysms as seen on echocardiography. Coronary angiography has not been done in our patients because it is no longer recommended if echocardiography is normal(3,4). None of our patients has so far had any recurrences of KD. High dose aspirin is given for its anti-inflammatory effect during the acute stage while low dose aspirin is continued thereafter for a few weeks for its anti-platelet effect.

Aspirin can be discontinued after a few months if there is no evidence of coronary artery abnormalities, as was the case in our patients.

It is our contention that most pediatricians (and physicians) in India are still not diagnosing KD because they think this condition occurs very rarely. This does not appear to be the case. There is no reason why KD should be rare in India when it is being so commonly reported from other countries[^]). Greater awareness about the constellation of symptoms and signs which constitutes this syndrome, would lead to more children being diagnosed and treated appropriately. In many countries KD is now the leading cause of acquired heart disease in children⁽⁴⁾.

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