

Original Articles

A study of children with Kawasaki disease

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Sri Lanka Journal of Child Health, 2003; **32**: 40-3

(Key words: Kawasaki disease, coronary arteritis, atypical or incomplete Kawasaki disease)

Abstract

Objectives To study demography, presentation, treatment, complications and prognosis of children with Kawasaki disease (KD).

Design A descriptive ongoing case study.

Method All children clinically suspected to have KD, following admission to professorial unit at Lady Ridgeway Hospital for Children or when seen in the private sector, from November 2001 to September 2002, were included in the study. The demographic details, presenting features, treatment and complications were recorded. Children with coronary arteritis were reviewed periodically to analyse long-term effects.

Results 19 children were suspected to have KD. Mean age was 3.9 years. 53% were males. Fever, conjunctivitis and mucocutaneous lesions were the commonest presenting features. 14 (74%) had coronary artery changes. 5 (26%) patients fulfilled the criteria for diagnosis. Intravenous immunoglobulin was used as treatment in 8 (42%) cases.

Introduction

Kawasaki disease (KD), also called mucocutaneous lymph node syndrome, was first reported by Kawasaki in 1967¹. Criteria for diagnosis of KD include fever of 5 days duration with 4 out of the 5 following features viz. conjunctivitis, lymphadenopathy, polymorphous rash, changes in mouth, and changes in extremities¹. Presentation of KD can be atypical and in some instances this may be misleading in management. Successful resuscitation of a cardio-respiratory arrest in a child ready to go

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(Received on 12 May 2003)

home, with subsequent investigations showing a dyskinetic myocardium due to myocardial infarction, prompted us to look at the presenting features of KD and the need to investigate early.

Objectives

To study the demography, clinical presentation, treatment, complications and prognosis of children with KD.

Design

A descriptive ongoing case study.

Method

All children, clinically suspected to have KD following admission to professorial unit at Lady Ridgeway Hospital for Children and the private sector in Colombo, from November 2001 to September 2002 were included in the study. The demographic details, presenting features, treatment and complications were recorded. Children with coronary arteritis were reviewed periodically to analyse long-term effects.

Results

There were 19 cases in all. Ages ranged from 1-8 years with a mean of 3.9 years. 53% were males. The presenting features of these patients, in accordance with the diagnostic criteria of KD, are shown in Table 1. Whilst fever was present in all 19(100%) cases, conjunctivitis and mucocutaneous lesions were present in more than 50% of patients.

Table 1
Presenting features of Kawasaki disease

<i>Diagnostic criterion</i>	<i>No. of cases < %</i>
Fever	19(100)
Mucocutaneous lesions	12(63)
Conjunctivitis-non purulent	10(53)
Peripheral lesions	09(47)
Rash	09(47)
Cervical lymphadenopathy	07(37)

Case	Age(Yrs)	Criteria	ESR	Platelet count (10 ⁹ /l)	2D Echo	IVIg	Persistence of dilated coronaries
1	4.5	F + 2	121(4)	318(4)	**** (4)	No	Yes =1Yr
2	2.0	F + 3	104(2)	625(1)	* (2)	Yes	Resolved <1 mth
3	1.0	F + 3	130(2)	675(3)	*** (3)	No	Yes =1Yr
4	2.0	F + 1	77(1)	410(2)	* (1)	No	Resolved <1 mth
5	1.5	F + 4	126(1)	680(2)	Normal	No	
6	3.5	F + 4	110(1)	510(2)	* (2)	Yes	Resolved <4 mths
7	3.0	F + 3	23(1)	507(1)	Normal	No	
8	2.5	F + 1	110(2)	436(2)	* (2)	Yes	Resolved <2 mths
9	3.0	F + 2	130(2)	340(2)	*** (2)	No*	Resolved <3 mths
10	8.0	F + 3	100(1)	270(1)	** (2)	Yes	Yes =1Yr
11	5.0	F + 3	80(1)	310(1)	Normal	No	
12	3.5	F + 2	96(1)	1365(2)	* (2)	No	Resolved <1 mth
13	4.2	F + 4	127(1)	529(4)	Normal	Yes	
14	8.0	F + 4	120(1)	865(3)	Normal	No	
15	4.8	ND	ND	ND	*** (2)	??	Yes <1 Yr
16	2.7	F + 2	110(1)	527(1)	* (1)	Yes	Resolved <4 mths
17	4.5	F + 4	125(2)	520(2)	* (1)	Yes	Resolved <3 mths
18	8.0	F + 2	105(2)	549(2)	** (2)	Yes	Yes <6 mths
19	1.8	F + 1	142(3)	625(2)	*(3)+Pul arteritis	Yes	Resolved <4 mths

Note a) Numericals within brackets indicate the week of illness since onset of fever

b) Asterisks denote coronary artery dilatation

* = 2-3 mm, ** = 3-4 mm, *** = 4-8 mm, **** = >8 mm

Mean erythrocyte sedimentation rate was 107.5 mm (range 23-130 mm) in the first hour. Highest platelet count recorded from each patient varied from 270-1,365x10⁹/L. Coronary artery changes were evident in 14 (74%) patients. Intravenous immunoglobulin (IVIg) was used as treatment in 8 (42%) cases. The patient profile is shown in Table 2. Cases 1, 6 and 17 are further described to illustrate the varying presentations and complications.

Case 1 A 4 ½ year old from Kurunegala was transferred with fever and a provisional diagnosis of leptospirosis. As the fever was not settling, he was investigated extensively. This included an ultrasound scan of abdomen and an explorative laparotomy. His fever subsided and he was about to be discharged from hospital when he had a cardiorespiratory arrest.

He was resuscitated and managed in the intensive care unit for a day and was noted to have a triple rhythm. 2-dimensional echocardiogram (2D echo) revealed evidence of anteroseptal myocardial ischaemia with a dilated right coronary artery equal to root of aorta. The diagnosis was revised to KD and he was started on warfarin, dipyridamole, captopril and isosorbide dinitrate.

Case 6 This was a 3 ½ year old from Kurunegala with fever and 4 other features, which completed the diagnosis of KD, and a 2D echo showed a dilated left coronary artery. This child was treated with IVIg and aspirin and at present is well with normal coronary arteries.

Case 17 A 4 ½ year old girl was transferred from Negombo with fever of 7 days duration, rash, conjunctivitis with subconjunctival haemorrhage and cervical lymphadenopathy. She complained of headache, itchy palms and diarrhoea. 2D echo showed a dilated left coronary with arteritis of right coronary artery. She was treated with IVIG and aspirin. Next day she developed papilloedema and the computed tomogram showed presence of cerebral oedema. She was given IV mannitol for 48 hours. One week after treatment she developed fever with red eyes and recrudescence was of concern.

Discussion

It has been shown that in KD there is 20-25% chance of developing coronary arteritis². Over the years there have been many reports on incomplete and atypical KD in which some have progressed to show the full spectrum later on^{3,4}. The main concerns for the paediatrician are twofold:

- Whether to investigate children with incomplete diagnostic criteria and
- When to do the first 2D echo and when to repeat it.

In our series only 5 patients (26%) fulfilled the criteria for diagnosis. 2 (40%) of this group had coronary artery changes. IVIG therapy was given to the 2 children with coronary artery changes and 1 other child, who was managed in the private sector, and whose parents paid for the treatment. There were 14 (74%) cases with incomplete diagnostic criteria and 12 (86%) of them had coronary artery changes. Due to constraints in purchase of IVIG in the public sector, and late diagnosis, only 6 (50%) of these 12 cases were treated with IVIG. Aspirin was commenced in all patients at onset. Aspirin (3-5 mg/kg/day) and/or dipyridamole were continued until coronary artery changes had returned to normal. Aspirin has never been subject to a randomised controlled trial alone but has been studied with or without IVIG. Advantages of IVIG therapy have been promising^{5,6}. Meta-analysis has not shown significant advantage of IVIG with high dose aspirin (80-120mg/kg/day) over IVIG with moderate dose aspirin (30-50mg/kg/day) in preventing coronary aneurysms in acute phase of illness⁵. 2 patients developed elevated serum alanine aminotransferase (ALT) levels and were managed on dipyridamole only. One of them was drowsy, and resembled Reye syndrome⁷.

All patients have been followed up for 6 months to a year and our first patient is still on warfarin, dipyri-

damole, isosorbide dinitrate and captopril. There were insufficient cases to look for statistical significance in resolution of coronary artery dilatation between the IVIG treated and untreated group. Cases with incomplete criteria (74%) outnumbered cases with complete criteria (26%). Coronary artery dilatation > 4mm was seen in the 'incomplete criteria' group during or after second week due to delay in suspecting KD and the dilatation is still persisting, except in one case. Cases diagnosed late were not given IVIG as fever had settled, indicating that the acute inflammatory process was settling. This reinforces the need to investigate 'incomplete criteria' cases during the first week on suspicion and if the coronary arteries are normal, a repeat assessment becomes mandatory within 3-5 days.

In view of these findings revised criteria for diagnosis of KD becomes essential. The KD research group (KDRG) in United Kingdom have redesigned inclusion criteria for diagnosis by reducing one criterion in the presence of coronary arteritis i.e. fever, presence of 3 criteria and evidence of coronary arteritis⁸.

In conclusion, we emphasize that KD is the commonest cause of acquired coronary artery disease in children. A high degree of suspicion is needed to diagnose 'incomplete criteria' cases. Revised criteria would help to make the diagnosis early. This, in turn, would help in early resolution of arterial changes because IVIG therapy and aspirin would be started early.

Assessment of coronary arteries is the mainstay of diagnosis in atypical cases and an experienced cardiologist is a prerequisite in this situation. Availability and pricing of IVIG is equally important to arrest progression of coronary arteritis on which atypical cases are diagnosed. The need for life long follow up in children with KD, who have coronary artery involvement, is being debated⁹.

Acknowledgements

We acknowledge Dr S. Narenthiran, Consultant Cardiologist, Lady Ridgeway Hospital, Colombo for his invaluable help in identifying the coronary artery lesions.

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