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Original Article

Kawasaki Disease (KD) in Iran: A Report of 85 Cases

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ABSTRACT

Background*: Kawasaki disease (KD) is a systemic vasculitis that occurs most commonly in children les than 5 years. We assessed 85 cases of the disease (KD) in Iranian society of pediatrics from March 2004 to March 2005.*

Materials and Methods*: We retrospectively analyzed 85 medical records of children with KD; SPSS software Version 13.0 for windows was used for this statistical analysis. The distribution for clinical manifestations and laboratory findings were observed by using data collected from questionnaire forms enrolled all over Iran.*

Results*: In our report, 35 cases fulfilled all diagnostic criteria of KD whilst 36 cases had incomplete criteria (atypical form). 14 cases that enrolled were not fulfilled those criteria. Our criteria for case definition were according to* ***Rowley's*** *and Newburger's studies.*

Conclusion*: We think that the number of atypical cases appears to be increasing.*

Keywords: *Kawasaki disease, Children, Iran*

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INTRODUCTION

In 1967 Tomisaku Kawasaki developed diagnostic criteria for an apparently new illness. These criteria continue to be useful in identifying patients with Kawasaki disease (KD) (1). However, in the 1970s and 1980s, report of severe morbidity or mortality death during childhood following illnesses with some, but not all features of KD began to emerge from both the US and Japan. Since these early reports of incomplete KD had been recognized worldwide as a major pediatric clinical diagnostic dilemma (1-5). It has become the leading cause of aquired heart disease among children in North American and Japan and despite of recent advances in treatment and research exploring its etiology, is still unknown (5-7). In this study, we presented and assessed 85 patients with potential KD.

MATERIALS AND METHODS

A survey questionnaire form and diagnostic guidelines for KD were sent to pediatric physicians of referral centers all over the country. The questionnaires and diagnostic guidelines were provided in Tehran by the KD research committee of Iranian society of pediatrics.

Patients with KD were identified by the *discharge diagnosis code* in medical records. Physicians were asked to review the medical records and report patients with KD diagnosed during one year period from March 2004 to March 2005. All physician who participated in the survey were senior pediatricians or subspecialists of pediatric infectious disease, pediatric cardiologists or rheumatologists. Two suspecialists of pediatric infectious disease were designated to ensure investigator compliance with the study protocol.

Questions in the questionnaire form were included: Date of admission; date of birth; gender; address; clinical findings; cardiac involvement; type of treatment and laboratory findings.

Definition and inclusion criteria

Patients were included in the study if had at least five of the following six clinical signs:

1. Fever persisting 5 days or more
2. Changes of peripheral extremities
3. Polymorphous exanthema
4. Bilateral conjunctival congestion
5. Changes of lips and oral cavity
6. Acute nonpurulent cervical lymphadenopathy.

Clinical and laboratory findings that should prompt consideration of incomplete (atypical) disease include:

Daily high spiking fevers, especially for 5 days and-particularly in infants without evidence of a bacterial infection, with or without: 1. one or more other diagnostic criteria for KD, especially conjunctival injection, oral mucosal changes and/ or rash or 2 anterior uveitis on slit- lamp examination. Laboratory findings included:

1. Markedly elevated ESR and/ or C reactive protein.

2. Elevated peripheral white blood cell count or normal white blood cell count with neutrophil predominance and immature forms on differential

3. Thrombocytosis one week after fever with or without sterile pyuria, elevated alanine aminotransferase, aseptic meningitis, anemia, hypoalbuminemia and echocardiogram changes showing pericardial effusion. Patients were also included if they had any above signs with coronary abnormalities documented by echocardiography (1-5).

Statistics parameters were expressed as median, mean and mode and a software SPSS version 13.0 for windows was used for the analysis. $P<0.05$ was considered as statistically significant.

RESULTS

A total of 85 potential patients were reported. In 35 patients the onset was complete (typical KD) and 36 cases had incomplete (atypical KD) according to Rowley's and Newburger's definition (1,5). The male to female ratio was 1.36; the median age at diagnosis was 32 months.

The majority of cases occurred in children <5 years (77%). Median of fever was 38.5 °C. The most clinical findings of KD patients were changes in lips and mouth (70%), strawberry tongue (58%), lip fissure (50%), skin erruptions (56%) as maculopapular rash, conjunctivitis without discharge (45%), changes of extremities (49%). Cervical lymphadenopathy was the least common presentation among five clinical findings (43%) that was bilateral and painless. Cardiac findings were 21% and

aneurysm was seen 7.3%. Other clinical manifestation included gasterointestinal symptoms such as diarrhea, vomiting, ileus and icter.

Median of erythrocyte sedimentation rate (ESR) that performed before treatment was 90 mm/1h. Median of haemoglobin (Hb) level was 10.8 g/L and median of platelet count was 464500/ mm3.

Aspirin (50-80 mg/kg/day), during the acute phase and then 3-5 mg/kg/day and high dose (2 g/kg intravenous gamma globulin (IVIG) were given. Coronary artery abnormalities including aneurysms and dilatation were reported respectively 7.3% and 21% (12 males and 5 females) in whom who had received IVIG.

Table 1. Type of Kawasaki disease

Type	Frequency	Percent
Atypical (incomplete)	36	42.3
Suspicious	14	16.5
Typical	35	41.2
Total	85	100.0

DISCUSSION

36 of 85 potential patients (42.3%) had fewer than 4 to 5 clinical signs. In Falcini report 83.6% fulfilled all diagnostic criteria of KD whilist 41.250 (16.4%) had incomplete disease. 11.25 patients (4.4%) had atypical onset (1, 2). Most patients had fever of more than 5 days with minimum duration of 2 days and maximum of 60 days. There were 49 boys and 36 girls with a *male/ female ratio* of 1.36/1 and in references was 1.7/1 in previous reports (1-5). The age of onset ranged from 2.5 months to 13 years (median, 32 months) in refenreces 2.3 years). In our study, patients below 5 years accounted 77.3% and in references was 85%).

The clinical manifestations of KD were similar to those in previous reports. Fever longer than 5 days was the most common finding, followed by red lips and oral changes, strawberry tongue and skin exanthema as maculopapular rash and like other studies, the cervical lymph node enlargement was the least frequent sign.

In our references oral manifestations were common (5). During the 5-year period changes in peripheral extremities have occurred less frequently in 1995 and 1997 and more common in 1999. The incidence rate of cervical lymphadenopathy decreased gradually from 1995 to 1999. In our study lymphadenopathy was the least common presentation. There was 21% positive cardiac findings and 7.3% aneurysm of coronary arteries. Other reports had showed 20% cardiac involvement (6,7). Blood analysis showed high ESR (median 90 mm/1sth and mode 107 mm/1sth). C-reactive protein was 3-4+ in 30%. Leukocytosis (median 14100/mm3) with polymorphonuclear predominance and thrombocytosis (median 464500/mm3) was seen. Median of haemoglobin (Hb) levels was 10.8 g/I and in Falcini's report was 9.2 g/dl (2). In the time of management all other diseases were excluded for these patients.

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