Kawasaki Disease: An Epidemiological and Clinical Study

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Abstract- To review children with Kawasaki disease admitted in 17-Shahrivar hospital of Rasht from 1999 to 2007. We reviewed retrospectively 64 children with Kawasaki disease between 1999-2007 admitted in 17-Shahrivar Hospital of Rasht. Frequency distributions of variants including age, sex, season, clinical and laboratory manifestations, response to treatment and complications of the patients were abstracted using SPSS 14. Patients' age ranged from 2 month to 12 years (median: 41.5 months). The male / female ratio was 1:0.78. The most cases were admitted in the autumn and then spring. Fever in 100%, changes in lip and mouth in 92.1%, rash in 87.5%, Conjunctivitis in 82.8%, changes in extremities in 67.1%, and cervical lymphadenopathy in 59.3% were present. Coronary aneurysm was found in one case. The most common extracardiac complication in this study was vomiting (85.7%). With respect to Kawasaki disease differential diagnosis and its treatment, Better knowledge can help us to make diagnosis more accurately.

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Key words: Epidemiology; mucocutaneous lymph node syndrome

Introduction

Since Dr Tomisaku Kawasaki characterized the complex pattern of illness known as Kawasaki disease (KD) in 1967 in Japan, it has been found to be the leading cause of pediatric acquired cardiac disease worldwide, especially in developed countries (1). It is an acute systemic vasculitis of infancy and childhood. Despite extensive investigation, the cause(s) of this disease remains a unknown. This acute illness presents as systemic inflammation and occurs as fever lasting >5 days, with conjunctival and oral mucosa changes, fissured lips, cervical lymphadenopathy, skin rash, and palm/sole erythema/induration. The most serious complication of KD is coronary artery aneurysm (CAA). Children <5 years of age are the most susceptible population, with a higher incidence reported for boys (2-4). Most experienced clinicians have no problem making the diagnosis of Kawasaki disease when it presents in its classic form; however, many times its presentation is subtle. This is especially true when it presents atypically. The term "atypical Kawasaki disease" was initially coined to describe patients with coronary artery abnormalities whose illness did not meet the strict criteria for classic Kawasaki disease.

It is critical to identify these children and choose appropriate treatment before they develop coronary artery damage. Therefore, most clinicians today use the term "atypical or incomplete Kawasaki disease" to describe children who fail to meet the case definition for classic Kawasaki disease but have compatible laboratory findings and no other explanation for their illness. This approach allows for the prompt initiation of immunoglobulin therapy and, hopefully, a decrease in the occurrence of coronary artery complications.

We conducted this study to review demographics of patients and characteristics of Kawasaki disease among children admitted in 17-Shahrivar hospital of Rasht from 1999 to 2007.

Patients and Methods

We conducted a retrospective study and included all 64 children with Kawasaki disease who admitted in 17-Shahrivar Hospital of Rasht. The diagnostic criteria were as follows: Fever lasting for at least 5 days in addition with presence of at least 4 of the following 5 conditions: bilateral conjunctival injection; changes of the mucosae of the oropharynx, including injected pharynx, injected and/or dry fissured lips, or strawberry tongue; changes in the peripheral extremities, such as edema and/or erythema of hands and/or feet, desquamation usually beginning periungally; polymorphous, but nonvesicular, rash; cervical lymphadenopathy greater

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than 1.5 cm. Illness should not be explained by other known disease processes.

Demographics of patients and characteristics of disease including season, clinical and laboratory manifestations, response to treatment and complications were abstracted using SPSS 14. Data are expressed as mean (SD), median (range), or percentage (number).

Results

We included 64 patients with Kawasaki disease who met the mentioned criteria. Patients' age ranged from 2 month to 12 years (median: 41.5 months). The male / female ratio was 1:0.78. The location of residence was urban in 79.7% and rural in 20.3%. Demographics were detailed in table 1.

Table 1. Patients' demographics

Demographics	Value
Age	
< 5 years	49 (76.6%)
> 5 years	15 (23.4%)
Sex	
Male	36 (56.3)
Female	28 (43.7%)

Table 2. Patients' clinical manifestations

clinical manifestations	Frequency
Fever	64 (100%)
Conjunctivitis	53 (82.8%)
Skin rash	56 (87.5%)
Cervical lymphadenopathy	38 (59.3%)
Extremities symptoms & signs	
Edema	26 (40.6%)
Erythema	15 (23.4%)
Scaling	15 (23.4%)
Lips and mouth changes	59 (92.1%)
GI symptoms & signs	
Vomiting	24 (37.5%)
Diarrhea	15 (23.4%)
Abdominal pain	7 (10.9%)
Hepatosplenomegaly	4 (6.25%)
Joints symptoms & signs	
Arthralgy	12 (18.75%)
Polyarthritis	3 (4.7%)
Monoarthritis	1 (1.6%)
Neurologic symptoms & signs	
Neck rigidity	4 (6.25%)
Others (Meningitis, Seizures,…)	4 (6.25%)

The most cases were admitted in the autumn and then spring (37.5% and 31.3%, respectively). In our study, fever in 100%, changes in lip and mouth in 92.1%, rash in 87.5%, Conjunctivitis in 82.8%, changes in extremities in 67.1%, and cervical lymphadenopathy in 59.3% were present. Clinical manifestations were abstracted in table 2. The total frequency of each of signs were higher than 100 percents due to occurance of more than one sign in extremities (67%) or gasterointestinal complications (50 %) in some cases.

Leukocytosis in 25%, polymorphonucleosis in 63% and thrombocytosis in 54.6% were found. ESR in 56.3% was more than 50 mm/h and in 32.8% was more than 100 mm/h. Poleocytosis in CSF was reported in any one of cases that LP was done for them (5 cases). Leucocyturia observed in 34.4% of cases. Abdominal sonography in 57 cases was done and hydrops in gall bladder was reported in 15.7%, other abnormalities (hepatomegaly or spelenomegaly) was reported in 12% and 73.6% of cases were normal. Treatment with IVIG (2g/kg) and aspirin (80-100 mg/kg) was done in 87.5% of cases and 16% of them were required second dose of IVIG. Coronary artery aneurysm was found in one case (1.6 %). The most common extracardiac complication in this study was vomiting (37.5%).

Discussion

Like other studies, we found a higher incidence of KD in boys, during certain seasons, and in children <5 years of age (1-12). Taiwan and Korea have the highest incidence of KD in the summer, Beijing and Hong Kong in the spring and summer, and Japan in January and summer (2-6). Studies from Europe and Canada report a higher incidence in the winter months (7,12). Although the period may not be the same, there is seasonal clustering. In a 14-year study in Japan, Burns et al(13) clearly demonstrated the seasonality and temporal clustering of KD. Such seasonality and temporal clustering suggests that different infectious diseases or other environmental factors might trigger this clustering presentation.

Analysis of different clinical manifestations and lab data showed similar results to other studies (14,15). Coronary artery aneurysm was found in one case (1.6 %). In large-scale studies in East Asia, the percentage of coronary artery dilatation is reported to be around 18% (3), and the complication rate of CAA ranges from 2.5% to 7.2% (3-5). In the early course of KD, cardiac involvement results in a dilatation of the coronary artery that in most cases is transient under treatment and

will not proceed to CAA formation. If CAA formation is found, long-term follow-up is necessary. A 10- to 21-year follow-up study reported that 55% of coronary aneurysm showed regression, although ischemic heart disease developed in 4.7%, myocardial infarction in 1.9% and death occurred in 0.8% of patients (16).

Disorders that are frequently cited in the differential diagnosis of Kawasaki disease include presumed viral infections, group A streptococcal infections, Epstein-Barr virus infections, measles, collagen vascular disorders, and drug reactions. Better knowledge about Kawasaki disease can help us to make diagnosis more accurately.

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