KAWASAKI SYNDROME IN MALAYSIA

ASMA OMAR

SUMMARY

19 cases of Kawasaki Syndrome were seen at the University Hospital, Kuala Lumpur between June 1979 and August 1984. The clinical features of the cases are reviewed in this paper. Kawasaki Syndrome is not an uncommon disease in Malaysia and clinicians should be aware of its presentation.

INTRODUCTION

Kawasaki Syndrome or the mucocutaneous lymph node syndrome (MLNS) was first described in Japan in 1967 by Kawasaki.[1] Since then this syndrome has been recognised all over the world including Malaysia.[2] This is a syndrome of unknown etiology. Its diagnosis is based entirely on clinical grounds and requires the presence of fever of at least five days duration plus four out of the five following criteria: rash, characteristic extremity changes; conjunctivitis; characteristic oral mucosal changes; cervical lymphadenopathy. This paper is a review of all cases which met these criteria seen at the University Hospital, Kuala Lumpur.

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MATERIALS AND METHODS

This is a retrospective study of case records of patients diagnosed as Kawasaki Syndrome in the University Hospital, Kuala Lumpur. All records were checked to see if the diagnostic criteria for Kawasaki Syndrome were met. Records of cases which met the diagnostic criteria were further analysed.

RESULTS

There were 20 cases of Kawasaki Syndrome diagnosed between June 1979 and August 1984. One case was rejected as it did not meet the diagnostic criteria outlined above.

The 19 patients comprised 12 Chinese, six Malays and one Indian. Their ages ranged from eight months to eight years with a median age of two years; 15 were males and four were females. 15 of these patients were resident in the Kuala Lumpur – Petaling Jaya area while the remaining four patients were from Klang, Tanjong Karang, Rawang, and Kuantan respectively. None of the 19 cases occurred between the months of May to August.

13 of the 19 cases were referred from general practitioners. The referral diagnoses were as follows: pyrexia of undetermined origin – 4; septicaemia – 2; Stevens-Johnson syndrome – 2; viral meningitis – 1; viral fever – 1; and Still's disease – 1. A correct

referral diagnosis of Kawasaki Syndrome was made in two cases.

There was no history of underlying illnesses in all cases. In one case, there was history of an attack of measles two weeks prior to admission to the hospital.

All 19 patients had fever which lasted longer than five days duration, as required by definition. The duration of fever ranged from seven to 40 days, with a mean of 13.6 days (SD = 7.8). Of the other diagnostic criteria, all 19 patients had rash, characteristic extremity changes (desquamation of the skin at the extremities, oedema of hands and feet and erythema of hands and feet) and characteristic oral mucosal changes (erythema and fissuring of lips, strawberry tongue and injected oropharynx). 16 of the 19 patients had conjunctivitis. Ten patients had all five criteria present.

Abnormal laboratory findings included leucocytosis and a raised ESR. The peak leucocyte counts ranged from 4000/mm^3 to 49,000/mm^3 with a mean of 18,400/mm^3 (SD = 11,100). The peak ESR values ranged from 5 mm/hr to 155 mm/hr with a mean of 57 mm/hr (SD = 41). Platelet counts done upon admission were within normal limits in all, except three patients where a thrombocytosis (285×10^3; 416×10^3 and 392×10^3) was seen. Urinalysis upon admission was also normal in all patients. Hydroxybutyric dehydrogenase levels were measured in 10 patients and were seen to be raised in nine ranging from 194–645 (Table I).

Blood cultures were negative in all patients. Throat swabs done in all patients also did not reveal any pathogens of clinical significance.

There were ECG changes in 12 patients and these consisted of elevated ST segments (five patients), abnormal Q waves (five patients) and atrial ectopics (one patient). Two patients had more than one ECG abnormality. Echocardiography was performed in seven patients and were normal in six patients. In one patient the echocardiogram showed a right coronary aneurysm and concomitant mitral valve prolapse. Coronary angiography was done in only one patient but did not reveal any abnormality. One patient developed bilateral axillary aneurysms.

TABLE I
LABORATORY TEST RESULTS FOR PATIENTS WITH KAWASAKI SYNDROME

Laboratory tests	Mean peak	Range	No. of cases analysed
Leucocyte counts (per mm^3)	18,478	4,000–49,000	19
Erythrocyte sendimentation rate ESR (mm/hr)	57	5–155	19
Platelet counts (mm^3)	226×10^3	46×10^3 – 416×10^3	19
HBDH (iu/L)	356	194–645	10

There was accompanying diarrhoea in six patients, aseptic meningitis and encephalopathy in three patients, arthritis/arthralgia in three patients, symptoms of upper respiratory tract infection in three patients and pneumonia in one patient.

13 of the 19 patients were treated with aspirin with doses ranging from 40 mg/kg/day to as high as 100 mg/kg/day. The rest of the patients were treated symptomatically. No patients were on steroids. All patients were well at time of discharge. One patient had a relapse of the syndrome after four months, while another (the patient with the coronary aneurysm) had two relapses after five and nine months respectively.

Eight patients did not return for follow-up. Three patients were followed up for a month, five for two months, two for five months and one is still being seen at regular intervals. This is the patient with coronary aneurysm. A repeat echocardiogram performed a month after the second admission showed the disappearance of the aneurysm but the mitral valve lesion remained. All other patients were well at their last follow-up visit.

DISCUSSION

Kawasaki Syndrome, since it was first described in 1967 has been recognised worldwide. Melish has written a comprehensive review of the subject and

readers are requested to refer to this article.[3] The incidence varies with ethnic origin and among Asian children in the United States the incidence was 4.07 per 100,000 children. Children of Asiatic origin had a higher incidence than Caucasian children.[4] The true incidence of the disease is not known in Malaysia, and there is a possibility that the disease is underdiagnosed as a result of a lack of awareness of the disease. This is reflected in the referral diagnoses which were seen. In Malaysia there appears to be a predominance of cases among the Chinese. The male to female ratio was 3.75:1. This is a very marked male predominance. In the United States the male to a female ratio has been reported to be only 1.6:1.

Seasonal distribution has been noted in Japan and the United States.[3] In Japan, there is a slight increase in the summer months, whilst in the United States the peak incidence occurs between February and May. An increase in incidence was observed between the months of May to August. However the series is too small to make any definite conclusions about seasonal distribition. Also the authors did not see any cases which could be related to an outbreak.

Conjunctivitis was seen in 96% of all cases in the United States.[4] Conjunctivitis was noted in only 13 out of 19 cases (68%). This could be because conjunctivitis occurs early in the disease and could have subsided by the time the patient was seen by the doctor. Lymphadenopathy is the least constant of the principal features and in our series occurred in 84% of the cases. The corresponding figure in the United States was 82%.

Leucocytosis and a raised ESR were seen in most patients. However the platelet count was within normal limits in the majority of our patients. Thrombocytosis has been reported as a universal laboratory abnormality in Kawasaki Syndrome.[3] This discrepancy could be explained by the fact that thrombocytosis is seen after the second week of disease and many of our platelet counts were performed upon admission.

Cardiac disease is a common complication in Kawasaki Syndrome. More than half of our patients had ECG abnormalities other than sinus tachycardia. No patient had obvious cardiac failure but the raised hydroxybutyric dehydrogenase levels in nine out of ten patients suggest that myocardial involvement is common. Coronary aneurysm was detected by echocardiography in only one patient. Coronary angiography in unselected patients has revealed coronary aneurysms in as many as 20% of patients.[5] Two-dimensional echocardiography has been shown to be as sensitivie as coronary angiography in detecting coronary aneurysms.[5] Unfortunately two dimensional echocardiography was available in our unit only after 1982. Had this been done on all patients we may have been able to detect more cases. Most of these aneurysms regress spontaneously as was seen in the single patient we had. Peripheral vascular abnormalities are rare and brachial and femoral aneurysms had been reported.[6] We have been unable to come across any documentation of bilateral axillary aneurysms as occurring in one of our patients.

Diarrhoea, arthritis and arthralgia and central nervous involvement are recognised associated features of Kawasaki Syndrome. These features may be so prominent or severe as to mislead the physician attempting a diagnosis. This was seen in some of the referrals. Hepatitis and gall bladder involvement has also been described but this was not seen in any of our patients.

Fatalities in Kawasaki Syndrome are uncommon. In the United States a case-fatality rate of 1.2% has been reported.[4] There were no deaths in this series.

Kawasaki Syndrome is not a uncommon disease in Malaysia. Its diagnosis is important because of the associated cardiac involvement. There is no specific treatment but aspirin has been found to be useful.[3] Much of Kawasaki Syndrome is yet to be understood and there is scope for further studies in this disease in Malaysia.

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