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KAWASAKI-LIKE SYNDROME IN AN ADULT AIDS PATIENT

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A 42-year-old white male with HIV infection and Kaposi's sarcoma presented with a 4 day h/o fever, rash, arthralgias, red eyes, and headache. Physical examination revealed T102.6F, bilateral conjunctival injection with limbal sparing and no exudate, an erythematous oropharynx and fissured lips, and an erythematous, maculopapular rash on the trunk and extremities with a violaceous desquamating rash of the scrotum. Multiple, tender 1 cm anterior cervical lymphnodes were palpated bilaterally. Cardiac and abdominal exam were unremarkable.The hands and feet were edematous with red palms and soles. There was warmth, swelling, and tenderness of both ankles. Extensive evaluation including head CT scan, LP, serologic tests for Cryptococcus, Coccidioidomycosis and Treponema pallidum, cultures for virus, bacteria, and PCR for parvovirus were negative. The patient was treated initially with intravenous immunoglobulin (IVIG) and high-dose aspirin (ASA) and defervesced with resolution of his rash and peripheral edema. His conjunctival injection improved only minimally. His CD4 count was 10 cells/mL and his HIV load was >750,000 copies/uL . He was started on antiretroviral therapy. Within the next week, he relapsed with fevers, peripheral edema and worsening of conjunctival injection. Again, he was treated with IVIG and high dose ASA with resolution of the edema and fever. An initial echocardiogram revealed dilated (5-6 mm)arteries. Transesophageal echo after the second IVIG dose revealed 5 mm coronary arteries. After a third relapse, the patient was treated with plasmapharesis followed by IVIG with complete resolution of his symptoms. We reviewed 13 English language case reports (11 adults) of HIV-infected patients who developed a Kawasaki-like syndrome. Most patients experienced typical signs and symptoms of KD and the illness resolved spontaneously with no CAA, even without specific therapy. These reports raise interesting questions about the possible infectious nature of KD and the occurrence of a pediatric illness in immunosuppressed adults.

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CORONARY DILATATION IN EPSTEIN-BARR VIRUS INFECTION

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BACKGROUND: Epstein-Barr virus infection shows variety of clinical features. Chronic fashion of the disease, called chronic Epstein-Barr virus infection, and acute aggressive manifestation of the disease, Epstein-Barr virus-associated hemophagocytic syndr ome sometimes show coronary dilatation resembling that of Kawasaki disease. In this study, we demonstrated coronary arterial lesions in patients with Epstein-Barr virus infection. METHOD: Four patients with Epstein-Barr virus infection were selected in t he study. Clinical characteristics, laboratory data, and echocardiograhpic findings were studied according to the hospital records. RESULT: Patients were aged from 1 to 15 years old. The distribution of gender was three girls and one boy. Clinical diagn osis was chronic Epstein-Barr virus infection in two patients and hemophagocytic syndrome in the remains. All four patients suffered coronary arterial dilatation which were demonstrated by echocardiography. The shape of coronary artery is linear. Corona r y aneurysm like Kawasaki disease were not observed. A 15 year-old boy with chronic Epstein-Barr virus infection suffered coronary dilatation and dilated cardiomyopathy. The patient died of cardiac failure. His autopsy revealed prominent lymphocytic inf ilt ra tion in the myocardium but no inflammatory cells in coronary artery. Ten year-old girl with chronic Epstein-Barr virus infection complained chest pain and showed coronary dilatation. She died of aggressive onset of hemophagocytic syndrome. Six year-old gi rl and one year-old girl with hemophagocytic syndrome also showed coronary dilatation at the onset. One year-old girl died of interstitial pneumonia. CONCLUSIONS: Epstein-Barr virus infection sometimes shows coronary artery dilatation like Kawasak i diseas e. The echocardiographic morphology of coronary lesion is not aneurysm but dilatation in our cases. Autopsy findings did not suggest acute inflammation around coronary artery. Coronary lesions may advance silently. Once the coronary lesions obser v ed, close cardiac follow up should be held.

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THE EFFECTIVENESS AND SAFETY OF EARLY INTRAVENOUS IMMUNE GLOBULIN TREATMENT FOR KAWASAKI DISEASE

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The diagnosis of Kawasaki disease (KD) is based on 5 characteristic clinical symptoms with fever persisting at least 5 days. Recently, early diagnosis and early intravenous immune globulin (IVIG) treatment are often reported. The aim of this study is to evaluate the effectiveness and safety of early IVIG treatment. One hundred thirty nine patients (pts.) were studied. The Harada's score was used to evaluate the severity of KD. In severe cases (Harada's score >3), pts. were administered high dose IVIG(2g/kg). In other cases (Harada's score <4), pts. were administered low dose IVIG (1g/kg). Group A, which was consisted of 77 pts. were received early IVIG treatment within 4 days. Group B, which was consisted of 62 pts. ,were received conventional IVIG treatment since 5 days. All patients were treated by 10days of illness. Twenty pts. in Group A(26%) and 13 patients in Group B(21%) were not clinical responders to the initial IVIG treatment (P=NS). No significant differences were found in incidence of coronary artery lesions between Group A (5/77, 6.5%) and Group B (5/62, 8.1%). The total duration of fever in early treatment group (Group A: 5.7±2.3days) was significantly shorter than that of conventional treatment group (Group B: 7.8±3.6days)($p<0.05$). Conclusions: Early IVIG treatment was effective to improve KD symptoms earlier and didn't increase the complications of coronary artery lesions. The results in this study may demonstrate the merit of the further investigation of the KD as a source of baseline information for the design of future IVIG therapeutic protocols for KD.

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EARLY TREATMENT WITH INTRAVENOUS IMMUNOGLOBULIN IN PATIENTS WITH KAWASAKI DISEASE

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OBJECTIVES: To determine if a shorter interval between Kawasaki Disease (KD) treatment with intravenous immunoglobulin (IVIG) and fever onset results in increased treatment failures, need for adjunctive therapy (ADJ) or development of coronary artery lesions (CAL). **STUDY DESIGN:** 178 KD patients diagnosed between 1987-1999 were included in this case-control study. All patients had fever plus > 4 out of the 5 clinical criteria for KD. Eighty-nine patients who received treatment at day 5 or earlier were matched to 89 patients treated at days 6-9 of fever according to the closest date of KD diagnosis to the index case. Compiled data from a detailed chart review included demographics, clinical features, fever duration, investigations, disease course and response to therapy. Differences between matched case and control pairs were analyzed using t-tests and McNemar tests. **RESULTS:** No demographical differences were noted between the 2 groups. Patients treated on 5 days of fever had a shorter total fever duration (5.2±1.9 days vs 8.0±1.8 days, $p<0.0001$), longer fever post IVIG treatment (1.5±1.9 days vs 0.8±1.3 days, $p=0.008$) and less coronary artery ectasia at 1 year post KD onset (4% vs 16%, $p=0.02$). There was no significant difference between cases and controls in the number of patients with KD recrudescence, need for repeat courses of IVIG, need for corticosteroids, length of hospitalization or development of coronary artery aneurysms within the first 3 months. Patients who were treated at day 5 of fever, had higher levels of serum albumin (36±5 g/L vs 33±5 g/L, $p<0.01$) and serum ALT (115±155 U/L vs 46±49 U/L, $p<0.001$), as well as a lower platelet count (354±131 vs 403±166, $p=0.02$) than controls during the acute phase. **CONCLUSIONS:** Early treatment of KD resulted in less coronary ectasia at one-year post KD onset but was not associated with a quicker resolution of fever, an increased number of treatment failures, an increased need for ADJ, length of hospitalization nor development of CAL. In children with fever and classical clinical and laboratory findings of KD, treatment with IVIG prior to 6 days of fever resulted in better coronary outcomes and decreased the length of time of clinical symptoms.

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RISK FACTORS FOR CORONARY ARTERY ABNORMALITIES IN PATIENTS WITH KAWASAKI DISEASE TREATED WITH ADDITIONAL INTRAVENOUS GAMMA-GLOBULIN

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Objective: Although an additional intravenous gamma-globulin (IVGG) treatment is commonly used for patients with Kawasaki disease (KD) who do not respond to the initial IVGG treatment, the efficacy of this strategy for preventing coronary artery abnormalities remains unclear. Our objective was to determine the risk factors associated with the development of coronary artery abnormalities in patients with KD treated with additional IVGG. *Methods:* We retrospectively assessed the risk factors for coronary artery abnormalities among 68 patients with KD treated with additional IVGG between 1993 and 1999 at 12 clinical centers in Japan. *Results:* According to univariate analyses, the risk factors for coronary artery abnormalities consisted of the duration of the initial IVGG treatment (>2 days), the duration of the fever before (>10 days) and after (>2 days) the start of the additional IVGG treatment, the white blood cell count (>15,000 /μl) and the C-reactive protein level (>5 mg/dl) before the additional IVGG treatment, and the use of steroid therapy. According to multivariate analyses, the risk factors for coronary artery abnormalities consisted of the duration of the fever before (>10 days; odds ratio, 8.51; 95 percent confidence interval, 2.13 to 33.9; P = 0.002) and after (>2 days; odds ratio, 4.66; 95 percent confidence interval, 1.25 to 17.3; P = 0.02) the start of the additional IVGG treatment. *Conclusions:* In patients with KD who do not respond to the initial IVGG treatment, the additional IVGG treatment within 10 days of the fever and resulting in the cessation of the fever within 2 days of administration reduce the risk for coronary artery abnormalities.

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A STUDY OF PATIENTS WITH KAWASAKI DISEASE WHO REQUIRED AN ADDITIONAL IMMUNOGLOBULIN THERAPY

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The subjects were 134 patients who received an initial immunoglobulin (IVIG) therapy in the acute phase of the disease from January 1990 to December 2000; 26 cases (19.4%) were prescribed an additional dosage of IVIG. The definition of the additional administration was either one of the followings: (1) when a patient received an initial IVIG therapy continuously for more than 5 days; (2) when the dosage of IVIG was increased during prescription; (3) when a patient received re-IVIG after the initial therapy. The coronary arterial lesion (CAL) was classified into four groups: no coronary lesion (N), mild coronary dilatation (Dil), middle-sized aneurysm (ANm), large aneurysm (ANl) based on echo-cardiography around the 30th day of illness. (1) The frequency (%) of CAL with and without additional IVIG was N: 65.4 vs 89.8, Dil: 3.8 vs 5.6, ANm: 19.2 vs 2.8, and ANl: 11.5 vs 1.9. (2) Percentage of cases who received additional IVIG in each category: (a) gender: male 18.4% and female 20.7%; (b) age at onset: subjects under 1 year of age or older than 4 years or more exceeded 20% in each age group; (c) start for initial IVIG therapy was as follows: the 2nd to 4th day of illness: 41.7%, the 5th to 7th day: 10.4% and the 8th day or more: 14.3%; (d) the first dosage (mg/kg) of IVIG were 100~350: 50.0%, 351~900: 18.2% and 901 or more: 8.3%.The two therapeutic characteristics obtained from the observations of the patients who required an additional IVIG were: (1) the start of the initial IVIG within the 4th day of illness and (2) the lower initial dosage of IVIG. In conclusion it was advisable to start the therapy from the 5th to 7th day of illness and to give a larger initial dose (more than 901mg/kg).