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TWO CASES OF KAWASAKI-DISEASE WHO SHOWED CORONARY ARTERIAL DILATATION EVEN AFTER GOOD RESPONSE TO A SINGLE HIGH-DOSE INTRAVENOUS IMMUNOGLOBULIN INFUSION

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It is now clear that a high-dose intravenous immunoglobulin therapy (IVIG) in patients with Kawasaki disease (KD) is effective for reducing the incidence of coronary arterial lesions (CAL). But both the total dose and the dose regimens of IVIG are still controversial.In this study, we report two cases with CAL in KD. These two cases were treated with aspirin (30mg/kg/day) and a single high-dose IVIG (1g/kg/day) on 5th day of illness. IVIG reduced their body temperature immediately to less than 37.5°C on 6th day of illness, so we did not add IVIG. Although they did not get high fever, and both the counts of white blood cells and the levels of C-reactive protein decreased after IVIG, their coronary arteries dilated gradually. In one case, the diameter of anterior descending branch of left coronary dilated to 4.0mm. In the other case, that of left main coronary artery dilated to 5.8mm. We confirmed these changes regressed to normal by either 2DE or selective coronary angiography after three months of onset. These cases suggest that a single high-dose IVIG (1g/kg/day) can not prevent CAL in KD, even after good response to IVIG.

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ADULT PATIENTS OF KAWASAKI DISEASE WITH ACUTE MYOCARDIAL INFARCTION WHO HAD NOT BEEN POINTED OUT CORONARY ARTERY LESIONS PREVIOUSLY

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We experienced two adult patients of Kawasaki disease with acute myocardial infarction who had not been pointed out coronary artery lesions previously. [Case 1: 27years old. Male] He suffered from Kawasaki disease at the age of one year. He did not receive any treatment of intravenous gamma-globulin therapy or aspirin. The cardiac catheterization at the age of 7 years revealed no coronary artery lesion. Thereafter, he had received follow-up examination once a year until 17 years old. He came to our outpatient clinic again because of recent recurrent chest pain at 22 years old. Further examinations including the cardiac catheterization, the treadmill test and the myocardial scintigraphy did not show any abnormalities and we concluded that his chest pain did not result from the coronary artery lesions of Kawasaki disease. After 5 years from these examinations, he suffered from an acute myocardial infarction with total occlusion of the right coronary artery (AHA Committee Report: Segment 2). He recovered well after the emergent percutaneus transluminal coronary anigioplasty (PTCA). [Case 2: 33 years old. Male] He suffered from Kawasaki disease at the age of two years. Any coronary artery lesion was not detected at that time. He had not received follow-up examinations since a few years after the onset. He had a chest pain at 30 years after the onset and had an acute myocardial infarction next year. The cardiac catheterization revealed 99% stenosis at segment 11, and total occlusion with well developed collaterals at segment 2. He recovered well after the directional coronary atherectomy (DCA) for the lesion at segment 11. [Conclusions] The patients with Kawasaki disease at the early era have grown to the young adult. They did not receive an accurate evaluation for the coronary artery lesions by echocardiography at the acute phase of Kawasaki disease. They did not receive effective therapy of gamma-globulin either. Those patients may have concealed coronary artery lesions. These two cases demonstrate well the high risk of those patients for the sudden onset of ischemic heart disease.

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THORACIC AORTIC ANEURYSM ASSOCIATED WITH KAWASAKI DISEASE

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Case Report: A 2-year-old female was referred to our hospital because of fever, skin rash, conjuncvital injection, strawberry tongue and erythema and edema of the hands and feet. Under the diagnosis of Kawasaki disease, intravenous γ-globrin (400mg/kg/day, for 5 days) and high-dose of aspirin (50mg/kg/day, orally) were started. However, the disease was refractory, so additional intravenous γ-globrin had to be done two times. The febrile duration was 18 days. Chest x-ray on 27th day after the onset revealed a partial projection of the descending aorta, which was not detected on chest x-ray on admission. However, the projection of the descending aorta was not detectable using ultrasonography. Echocardiography performed 22 days after the onset showed mild dilatations in right and left coronary arteries. Although normal coronary arteries were seen on selective coronary angiogram, the thoracic aortic aneurysm which was 19mm in diameter was disclosed on aortogram. Therefore, the abnormal projection of descending aorta on chest x-ray indicated the aortic aneurysm. The size of the aneurysm on chest x-ray has not changed for 2 years. In Kawasaki disease, there are a few case reports on abdominal aortic aneurysm, but thoracic aortic aneurysm has not been described yet. It is difficult to detect thoracic aortic aneurysm using ultrasonography, although the abdominal aortic aneurysm is detectable. Conclusion: In patients with refractory Kawasaki disease, we should consider the complication of thoracic aortic aneurysm in addition to abdominal aortic aneurysm, and careful observations of the changes in the shape of the thoracic descending aorta on chest x-ray are important.

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INFLAMMATORY PULMONARY NODULES IN KAWASAKI DISEASE

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Significant pulmonary manifestations of Kawasaki disease (KD) are uncommon. However, epidemiologic data from KD epidemics reveal an association between KD and a recent respiratory illness, and peribronchial cuffing or interstitial infiltrates have been reported on chest X-rays in 15% of acute KD patients. Moreover, pneumonia has been documented in 86% of autopsied acute stage cases. We report two children with KD in whom pulmonary nodules developed. The first was a 4-month-old boy with fatal acute KD. This patient had symptoms of cough and nasal congestion, and autopsy revealed inflammatory nodules in the lung as well as large coronary artery aneurysms. The second patient was a 6-month-old boy with atypical KD complicated by a coronary aneurysm in whom three peripheral pulmonary nodules were noted on chest Xray and confirmed by chest CT scan. One nodule was biopsied, revealing a non-encapsulated fibrovascular nodule infiltrated by numerous mononuclear cells. The patient was treated for KD with intravenous gammaglobulin and aspirin therapy, and did well. To characterize further the histologic features and inflammatory component of these nodules, immunohistochemistry was performed on tissue sections using antibodies to common leukocyte antigen (LCA) and Factor VIII-related antigen. In both cases, there was marked LCA positivity within the lesions indicating the presence of inflammatory mononuclear cells, and marked Factor VIII-related antigen positivity within and around the nodules, indicating extensive angiogenesis. We propose that there is a spectrum of severity of pulmonary involvement in KD, ranging from subclinical or symptomatic interstitial micronodular infiltrates to larger inflammatory pulmonary nodules. These infiltrates and nodules are likely a host response to the etiologic agent and may resolve with the disease process. Recognition of this pulmonary complication of KD may enable cautious observation of such lesions for spontaneous resolution; biopsy may be unnecessary unless the lesions persist after acute KD has resolved.

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EPIDIDYMO-ORCHITIS IN A PATIENT WITH ATYPICAL KAWASAKI DISEASE - THE ROLE OF STEROIDS

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The clinical features of typical Kawasaki disease (KD) are well known and complications such as hydrops of the gallbladder, arthritis and aseptic meningitis have also been commonly described. Epididymo-orchitis is an extremely rare manifestation of Kawasaki Disease and there has only been, from our literature search, one report by Connolly in 1979. We describe the clinical presentation and management of a five-year-old boy with recurrent KD complicated by epididymo-orchitis. The patient was diagnosed with atypical KD and subsequently developed coronary aneurysms. He was treated with intravenous immunoglobulin, recovered well, and was placed on maintenance warfarin and aspirin. In the next two years, he developed two relapses. He presented four days after apparent remission of his 2nd recurrence with acute left testicular pain. This responded dramatically to intravenous hydrocortisone, and his symptoms abated within 24 hours. The occurrence of epididymo-orchitis in KD, along with the use of steroids in the treatment of extra-cardiac manifestations, is highlighted in this report. As this is a rare complication of KD, treatment protocols have not been widely formulated. Further studies to elucidate the immuno-pathogenesis of this manifestation would be necessary to define the role of steroids and to establish further treatment strategies for this interesting manifestation.

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ACUTE KAWASAKI DISEASE IN ASSOCIATION WITH BURN INJURY

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While the etiology of Kawasaki disease (KD) is unknown, a role for infectious agents, including common skin organisms, has been proposed. We report 2 cases of KD associated with burn injury. Case 1: A 2 year old boy was admitted with a first and second degree scald injury to more than 9% of his body. Initial leukocyte count (WBC) was elevated. He was discharged to home the next day, and on daily visits was noted to have decreased fluid intake with fever without an infectious source. A generalized macular rash developed and he was readmitted with persistent fever. He subsequently developed conjunctivitis, oral changes and unilateral cervical lymphadenopathy with erythema of his palms. He had elevated WBC, ESR and liver transaminases. Acute KD was diagnosed and he received a single dose of IVIG on day 8 of his illness, with good response. All culture results were negative. Initial and follow-up echocardiograms were normal. Case 2: A 2 year old boy was transferred to hospital with second degree scald injury to 20% of his body. Within 24 hours he developed fever and cough, which continued without an infectious source for the next 6 days. He developed conjunctivitis and oral changes, with normal WBC, elevated ESR and normal liver transaminases. A diagnosis of atypical KD was considered, and he subsequently developed a generalized rash and extremity changes. A single dose of IVIG was given on day 8 of his illness, with good response. Initial echocardiogram showed mild dilation of the left main coronary artery, which resolved by his 6 week assessment. All culture results were negative. We speculate that the association of KD with burn injury in these patients may relate to stress or injury-induced immunologic changes, skin or tissue antigen release, or weakened barrier for entry of skin and other organisms