Case Report

# Adult kawasaki disease

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## ABSTRACT

Kawasaki disease was first described from Japan in 1967. Most of the patients reported have been under 3 years of age. We are reporting a typical case of Kawasaki disease in an adult Yemeni male, who had all criteria for diagnosis. Additionally our patient had tricuspid regurgitation as well as tricuspid valve endocarditis, which is only rarely reported in literature. We also noted a rise in monocyte and eosinophil count up to 10%, which may have pathophysiologic significance in this disease. At two year follow up our patient had persistent right bundle branch black which he had developed during the course of his illness. Mortality of the disease is related to cardiac complications especially coronary arteritis and aneurysms in children. In adults too once diagnosis is established patients should be followed for a long period of time to look for any possible cardiac complications.

**Keywords:** Adult kawasaki disease, tricuspid regurgitation, tricuspid valve endocarditis.

**Saudi Medical Journal 2000; Vol. 21 (11): 1078-1080**

Kawasaki disease, first reported from Japan,[1] is an acute febrile multisystem illness of unknown etiology, which mainly affects infants and young children. So far only a few cases have been reported in adults. The disease involves the heart in around one fourth of patients. We report a 23 year old Yemeni male who had typical features of Kawasaki disease. This case confirms the global distribution of the disease and its existence in Middle East in adults. The patient had endocarditis of the tricuspid valve, which is reported rarely in literature. We noticed gradual rise in peripheral blood monocytes and eosinophils. This may have significance in the pathogenesis of the disease and may have diagnostic value.

Case Report. A 23 year old male patient from Yemen came to the emergency room of Najran General Hospital on 13.11.1418H with four days history of high-grade fever, anorexia, sore throat, dry cough, joint pains and persistent vomiting. No significant information could be gathered from the personal and family history especially as regards use of drugs, recent travel, sexual contact or previous illness. On examination he was conscious but restless and looked ill with temperature of 39°C, pulse 120/min and BP of 140/90mmHg. He had injected conjunctivae, congested throat, strawberry tongue and angular stomatitis and the entire oral mucosa and lips were markedly inflamed with few erosions (Figure 1). Facial erythema and a faint morbilliform exanthema was seen on the trunk. Examination of chest and cardiovascular systems was normal. Abdomen showed palpable liver from the 3rd day onwards. Joint examination was normal. Investigations revealed WBC 6700, DLC polymorphs 88%, lymphocytes 10%, hemoglobin 14.4gm/dl, platelet 228, ESR 70mm in the first hour, peripheral blood film for malarial parasite negative, RBS 6.6mmol/L, urea 4.5mmol/L, creatinine 60umol/L, Na 129, K 3.1, AST 98.5, ALT 245.8, ALP 125, bilirubin 63.6umol/L, total protein 68.2gm/

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Received 29th March 2000. Accepted for publication in final form 14th June 2000.

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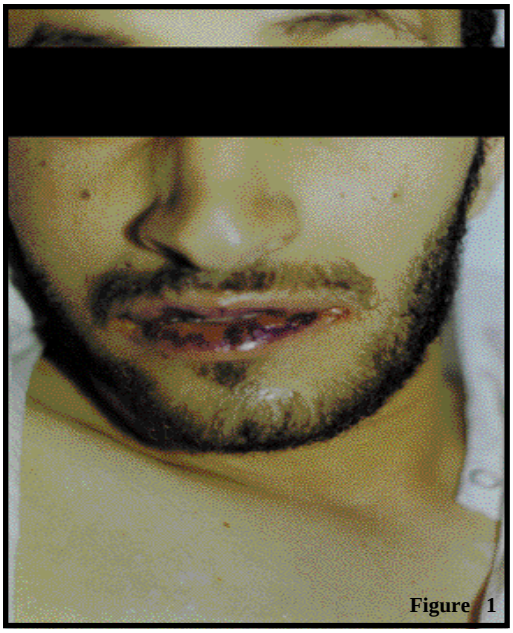
L, albumen 40.2gm/L. Urine examination revealed 15-18 WBCs, and no protein or casts. Paul-Bunnel test was negative, Brucella agglutination and Widal test, Toxoplasma and Bilharzial serology, VDRL, ANA, HIV, HbsAg, anti-HCV, anti-HAV IgM were all negative. Throat swab culture, sputum culture, blood, urine, stool and bone marrow cultures were all negative for pyogenic organisms. Coagulation profile showed PT 16 sec (control 14), APTT 35 (35), INR 1.2. X-ray chest and ECG were normal. Ultrasound abdomen showed mild hepatomegaly. Patient was initially put on cefalothin but did not respond.

***Evolution.*** Patient continued to be febrile and started to have exfoliation of the skin of the face, crusting on the lips and angles of mouth. A lymph node appeared on the left side of the neck on the fourth day. TLC rose to 24,400 with a monocytosis of 10%. ESR was 93 and platelet 738 at the end of first week. Echocardiography showed tricuspid regurgitation with vegetations on the tricuspid valve. ECG showed complete right bundle branch block (RBBB**)**. A diagnosis of infective endocarditis was entertained and patient was started on penicillin and gentamicin. Meanwhile repeated blood and urine cultures continued to be sterile. Four days later patient started with desquamation of the skin of hands and feet (Figure 2). A diagnosis of Kawasaki disease was made. All antibiotics were stopped and he was put on high dose aspirin (100mg/kg/day) in divided doses. Patient showed rapid defervesence within two days. He was discharged from the hospital on the 19th day on low dose aspirin (100mg/day). On follow up patient was seen in the outpatient two weeks later. At this time he had started developing Beau's lines on the nails and new skin had replaced the desquamated one. On checking the investigations, ESR came down to 20mm/first hour and echocardiography showed minimal TR but normal ejection fraction and no coronary aneurysms. Follow up at six months, at one year and at two years revealed no abnormality with a normal echocardiography. However RBBB persisted in the ECG.

Discussion. Kawasaki disease (KD) is an acute systemic vasculitis of the small and medium sized arteries. Children of Asian countries are more commonly involved than those in the west. Although many hypothesis have been proposed its etiology is still unknown. For diagnosis five out of the following six criteria have to be present: fever, injected eyes, oral mucositis, cervical lymphadenitis, hand and foot changes, and polymorphous rash.[2] The case we are presenting satisfies all the six criteria required and also has some additional features of KD like leucocytosis, raised ESR, late thrombocytosis, sterile pyuria and albuminuria as well as hepatitis. All these manifestations confirm the diagnosis of KD in this case. To date no specific diagnostic test is known for KD, hence the diagnosis remains clinical.[2] The refractory fever for more than five days that hugs 40°C is the hallmark of this disease. It is usually dry, sweat-less fever and is resistant to antipyretics and antibiotics. Fever persisted for 18 days in our patient. Interestingly the erythema and desquamation



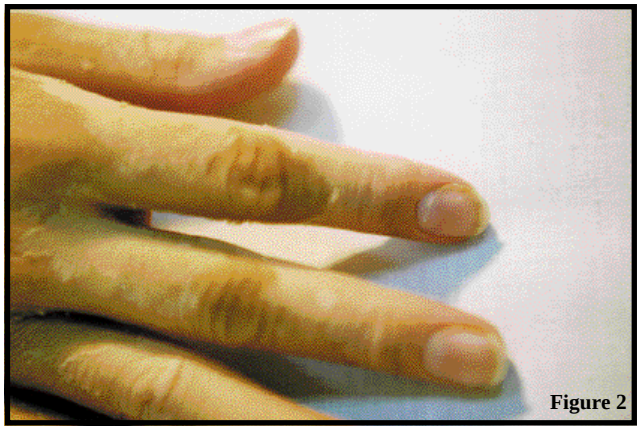




**Figure 1** - Adult kawasaki disease. Angular stomatitis and fissured lips seen in first week.

**Figure 2** - Adult kawasaki disease. Exfoliation of hand seen in the thrid week.

of hands and feet were not associated with edema in our case. Possibly, in contrast to children, acral edema may not be a feature of KD in adults. The cardiac findings were sinus tachycardia (a sign of myocarditis), RBBB (a sign of defect of conduction), and vegetations of the TV with moderate TR (a sign of endocarditis). All this resolved in the convalescent stage except RBBB. The negative blood cultures carried out 17 times at spikes of fever and the fact that patient improved without antibiotics exclude septicemia as well as infective endocarditis. Toxic shock syndrome is a multisystem disease like KD but is more common in women during periods of menstruation and is associated with hypotension and no lymphadenitis. Staphylococcus aureus is isolated from the vagina, nose or a skin wound in such cases.[3] Leptospirosis, rickettsial diseases, scarlet fever, drug reaction and viral exanthems due to cytomegalovirus should also be considered in the differential diagnosis. However none of these will satisfy all the six criteria mentioned above. Culture of the organisms, specific serologic tests and history of drug use would help to differentiate these.

Review of literature in the last 10 years shows that endocarditis of the TV was reported only once before.[4] However we found TR as the most common valvular lesion reported in KD.[5] Thus the rarity of TV endocarditis compared to the common occurrence of TR may be due to underdiagnosis of the transient inflammatory lesion. We have noticed gradual increase of monocytes (from 1% on day 2 to 9% on day18) and eosinophils (from 0.6% on day 2 to 10% on day 20) in the peripheral blood. The values returned to baseline in the recovery phase. This may be of importance as the histologic picture of vessels in KD shows perivascular monocytic and granulocytic infiltrate in the first week of illness but a predominant monocytic invasion in the 3rd to 5th week.[6] Interestingly, monocyte was the focus of research recently and was found to be markedly activated in KD.[7] This is the first case of KD reported in a Yemeni from Najran and along with the cases reported in Saudi Arabs,[8] it confirms its occurrence worldwide. KD in adults is more benign than in children and cardiac complications have been rarely reported,[9] but should be ruled out by ECG and echocardiography in all cases of KD. We recommend that all cases should be followed and screened for long-term cardiac complications involving all three layers of the heart.

Acknowledgment. We wish to express our thanks to Sister Aleah A. Saclote for her secretarial services.

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