

ATYPICAL KAWASAKI DISEASE WITH CORONARY ARTERY INVOLVEMENT

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Kawasaki disease (KD), an inflammatory vasculitis of unknown etiology, is a well-established clinical entity affecting mainly infants and young children. It was initially described in Japan, and is now being seen with increasing frequency in many countries worldwide.¹ A specific diagnostic test is not available and the diagnosis is based on the presence of characteristic clinical findings.^{2,3} Although these criteria are extremely useful for establishing the diagnosis, the strict use of such criteria may result in atypical cases being unrecognized.⁴ We report a young girl in whom significant coronary artery dilatation developed during an illness that was consistent with some of the clinical features of KD.

Case Report

A previously healthy three-year-old black girl presented with a 10-day history of fever of 38.5°C, sore throat, bilateral conjunctivitis with severe photophobia, and generalized erythematous rash. On the fourth day of the illness, she had been treated with oral antibiotics, but due to lack of response to the antibiotics she was admitted to the hospital for observation and further investigations. There was no history of any joint pain, obvious swelling of her extremities, gastrointestinal symptoms or neck swelling.

Physical examination revealed a pale, irritable girl with a fever of 38°C, but with other vital signs stable. She had dry, cracked lips with mild bleeding, and erythematous pharynx without exudate or petechiae. Slit lamp examination was consistent with anterior uveitis. Examination of the skin revealed generalized maculopapular rash. There was no cervical adenopathy or extremity changes, the abdomen was normal and the remainder of the examination was unremarkable.

Laboratory investigations showed WBC to be $8.3 \times 10^9/L$, with 72% neutrophils and 15% lymphocytes,

while hemoglobin was 9.6 g% and platelet count was $331 \times 10^9/L$. The sedimentation rate was 110 mm/hr., and urinalysis was normal. Blood, urine, throat and eye swab cultures were negative. Chest x-ray and electrocardiogram were normal, however, two-dimensional echocardiogram showed 4 mm dilatation of the left main coronary artery alone, consistent with those seen in Kawasaki disease. The diagnosis of atypical Kawasaki disease was made based on the presence of coronary artery dilatation, and four out of the six criteria of Kawasaki disease.

The patient was started on a single dose of intravenous immunoglobulin (IVIG) at 2 g/kg over 12 hours, and aspirin therapy 100 mg/kg/day in four divided doses. The next day she was afebrile, with improvement in her eye and skin symptoms. The aspirin was reduced to 5 mg/kg daily. On follow-up, two weeks later, she was noted to have membranous desquamation of fingers and toes. Repeat echocardiogram on the third week of illness showed regression in the coronary artery dilatation (3 mm). The patient has continued to do well on low-dose aspirin for her coronary artery dilatation.

Discussion

It is generally agreed that a diagnosis of KD can be made only when a patient develops prolonged fever (>5 days) and fulfills four of the remaining criteria: 1) oropharyngeal changes (erythema, fissuring and crusting of the lips, strawberry tongue or diffuse oropharyngeal erythema); 2) bilateral non-purulent conjunctivitis; 3) peripheral extremity changes (erythema of palms and soles, swelling and induration of hands and feet); 4) erythematous rash; and 5) enlarged cervical lymph node (>1.5 cm in diameter).³

With the increasing incidence of KD there will be greater numbers of children with incomplete clinical features of the disease.⁴ Atypical Kawasaki disease (AKD) includes those patients who do not fulfil the classic criteria, but have some of the clinical features of KD, in particular, coronary artery involvement.

Our patient did not fulfill the classical criteria for the diagnosis of KD, however, she developed prolonged fever, erythematous skin rash, bilateral conjunctivitis, anterior uveitis and mild oropharyngeal changes. She subsequently

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developed significant left coronary artery dilatation. Fukushige et al.⁵ reported that of 25 patients who demonstrated incomplete Kawasaki disease, 68% had four, while 28% had three of the six principal criteria.

Unfortunately, the true frequency of KD in Saudi Arabia is not known, but it is known that KD is more common in Asians than in Caucasians.⁶ There are no studies of the comparative incidence of KD in Saudi Arabia and other countries, and our feeling is that the disease is underdiagnosed in Saudi Arabia. To our knowledge, no similar case has been previously reported in Arab children.

The purpose of this report is to demonstrate the possible presence of coronary artery involvement in patients who did not fulfil the classical criteria of KD. Although KD is generally self-limiting, the prognosis is directly related to the potentially serious sequelae of coronary artery involvement.⁷ Because of this, increased clinical awareness and early recognition of this disease and prompt treatment are essential to ensure a successful outcome.⁸ We, therefore, recommend the close observation of patients who present with symptoms of the disease but do not fulfil the classic diagnostic criteria. High white

blood cell count, platelet count and elevated sedimentation rate are useful investigative tools for the diagnosis and monitoring of Kawasaki disease. Evaluation by two-dimensional echocardiogram in such patients is also recommended.

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