A Case of Incomplete Kawasaki Disease without Fever

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Abstract

Kawasaki disease is the second most common vasculitis syndrome of childhood. Fever is accepted as sine qua non for the diagnosis. Herein, we report a 17 month-old boy diagnosed as afebrile incomplete Kawasaki disease. Although he had 4 features of Kawasaki disease, his mother denied presence of any fever. The diagnosis was made by clinical criteria, elevated erytrocyte sedimentation rate and CRP levels and by exclusion of other possible etiologies. This case is presented since afebrile Kawasaki disease is a very rare entity.

Key words: Afebrile, kawasaki disease, incomplete

Introduction

Kawasaki disease (KD) is an acute self-limiting vasculitis of childhood which primarily affects coronary arteries. At least 75% of cases are occurring in children under 5 years of age. The diagnosis is still made by clinical criteria. Presence of fever lasting at least five days with four of the five principal features of non-purulent conjunctivitis, polymorphous skin rashes, abnormalities of lip or oral mucosa, abnormalities of the extremities, and cervical lymphadenopathy with exclusion of the other diseases were needed for the diagnosis. Especially in younger children due to the paucity or variability of symptoms, the diagnosis may be challenging. In the literature, there are exceptional reports regarding afebrile KD; we herein report an unusual case of KD manifesting without fever.
Case
A 17-month-old boy was admitted to our hospital with complaints of painful and swollen hands with desquamation of the fingertips. From history, it was learnt that he had had redness at both of his eyes and over his lips nearly 20 days ago. The desquamation had begun a week before attending the hospital. His mother had told that he was completely afebrile in this period. On physical examination, there were cracks above the lips and desquamation was present all over the fingertips. He had edema over both of his hands. There were no hepatosplenomegaly and lymphadenopathy. His body temperature was normal. At his laboratory work-up, his C-reactive protein was 54 mg/L, erythrocyte sedimentation rate was 75 mm/hr, and his complete blood count (CBC) revealed Hb 11.9 gr/dl, wbc 15,700/mm³, and platelets 419,000/mm³. Urinalysis was normal. ASO titer, viral serologies, and blood, throat and urine cultures were negative. A few days later, his platelet count had increased to 749,000/mm³ at the second CBC. Chest X-ray, echocardiography and abdominal ultrasonography were normal. His acute-phase reactants continued to increase. No etiology clarifying these laboratory tests and clinical features was found. This patient was diagnosed as having incomplete Kawasaki disease, with IVIG from 2 gr/kg/day being administered with 80 mg/kg/day of aspirin. On the 10th day of therapy, he was discharged from the hospital with a normal sedimentation rate and CRP levels and without any clinical manifestations. His control echocardiography did not reveal any coronary artery dilatation.

Discussion
Kawasaki disease is a systemic vasculitis and its diagnosis is still based on clinical criteria. But some cases that don’t fulfill the classical clinical criteria are present and they are named as incomplete KD [1]. These patients carry the risk of delayed diagnosis and coronary arterial involvement. Fever is the most common presenting feature in KD, whereas in the literature a few cases of afebrile KD had been reported. Kato et al. had presented a 2-year-old boy with afebrile incomplete KD having coronary arterial ectasia, who was treated by aspirin but not IVIG [2]. They had seen deterioration of the coronary arterial ectasia during follow-up. Hinze et al. had published a 3-month-old boy with afebrile KD and coronary aneurysms [3]. One patient with afebrile KD but without coronary involvement was published from Costa Rica by Uloa-Gutierrez et al. retrospectively [4]. Similarly, our patient had the features of conjunctivitis, oral mucosal changes, extremity changes, and peeling of both fingertips of hands and feet, accompanied by the elevated inflammatory markers and thrombocytosis that cannot be explained by any other infectious etiology. Even though his echocardiography was normal, he was accepted as having incomplete KD without fever, with IVIG treatment being introduced with high-dose aspirin.

This is the first case report of KD without fever from Turkey. Based on the previous reports, we wanted to attract attention to the paucity of symptoms in young infants with KD. Fever, which is a constant feature, may even be absent in some of the patients, and clinicians should bring to mind KD when they are faced with the other features and increased inflammatory markers in young children.

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References

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