Kawasaki Disease: A Condition of Many Guises

Dear Editor,

Kawasaki disease (KD) is a systemic vasculitis that primarily affects children younger than 5 years of age. Its incidence rate varies from 3.4 per 100,000 children <5 years of age in Thailand[1] to 218.6 per 100,000 children <5 years of age in Japan.[2] Though there are well defined criteria for the diagnosis of KD,[3,4] its presentation can be varied[5] and the diagnosis of KD remains a challenge in febrile children who do not fulfill the diagnostic criteria but have several findings compatible with those of KD.

Case Report

The patient is a 5 months old male infant who presented with a 3-day history of fever (highest temperature reached was 38.4°C) associated with mild running nose, red and cracked lips, bilateral non-suppurative conjunctivitis and generalised rash with involvement of the trunk and peripheral limbs predominantly. Physical examination of the patient on admission revealed a playful afebrile infant with mild bilateral non-suppurative injected conjunctiva. There were bilateral small palpable cervical lymph nodes measuring less than 0.5 cm in greatest diameter. There were, however, no features of red lips or strawberry tongue, rash, erythema (in contrast to what had been highlighted in the history), oedema of the hands and feet or erythema/induration at Bacillus Calmette-Guerin (BCG) inoculation site, to suggest the presence of KD. Cardiovascular and abdominal examinations in the infant were unremarkable.

Investigations performed included a full blood count (FBC) which showed total white cell count of 16.11 x 10^9/L, with absolute neutrophil count of 6.27 x 10^9/L and absolute lymphocyte count of 7.13 x 10^9/L, with occasional reactive lymphocytes seen. Haemoglobin level was 10.2 g/dL, and platelet count was 484 x 10^9/L. C-reactive protein (CRP) level was 26 mg/L. Liver function test performed did not reveal any elevated transaminases or hypoalbuminaemia (serum albumin 38 g/L) and urine formed elements did not reveal any pyuria. Respiratory virus immunofluorescence, including adenovirus, was negative.

The infant was initially treated symptomatically with oral paracetamol with good effect. The infant remained afebrile for the subsequent 28 hours during the hospital stay before developing a temperature of 37.7°C (day 5 of illness). The patient was also noted to develop red and cracked lips and a macular rash over the right cheek subsequently on day 6 of illness. A decision was hence made to repeat FBC which revealed a total white cell count of 20.29 x 10^9/L, with absolute neutrophil count of 8.74 x 10^9/L and absolute lymphocyte count of 8.51 x 10^9/L. Haemoglobin level was 10.4 g/dL and platelet count was 621 x 10^9/L (platelet clumps were seen suggesting that the true platelet count was higher). Repeat CRP was also mildly increased to 34 mg/L. A 2-dimensional echocardiography was thus performed for the patient in light of these new findings, evaluating for possible incomplete KD. The scan showed a mildly ectatic right coronary artery (RCA) with a measurement of 2.2 mm (Z-score + 2.7). A diagnosis of incomplete KD was hence established in view of presence of fever of more than 5 days, rash and red lips, and further supported by CRP of 34 mg/L and echocardiographic finding of ectatic RCA with Z-score of + 2.7.[6]

The patient was thus given intravenous immunoglobulin (IVIG) of 2g/kg and commenced on high-dose oral aspirin of 100 mg/kg/day. The infant became afebrile whilst on IVIG infusion and remained so for more than 48 hours after completion of IVIG. His lips were no longer red and there was resolution of rash. Repeat FBC 48 hours post-IVIG showed a decrease in total white cell count and CRP to 10.91 x 10^9/L and 7 mg/L, respectively.

The above case report illustrates the treachery involved in the diagnosis of incomplete KD and the unusual progression of clinical signs and symptoms, including a playful, not irritable infant, good response of fever to paracetamol and patient becoming afebrile for more than 24 hours before the redevelopment of fever and other symptoms associated with KD.

Incomplete KD is more common in young infants[7] and may be made in cases with fewer classical diagnostic criteria but with several fitting clinical, laboratory or echocardiographic findings after exclusion of other causes of febrile illness. The diagnosis of incomplete KD should be entertained in children—especially young infants—with unexplained fever of ≥5 days associated with 3 or less of the clinical criteria but with compatible laboratory and/or echocardiographic features.[6] Infants ≤6 months old with fever ≥5 days and evidence of systemic inflammation

with no obvious explanation may also benefit from echocardiography which will contribute to clinching the diagnosis of KD.

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