

Incomplete Kawasaki disease, is it time for modified criteria?

Hanna B. (1), Tadros M. (2)

(1) Cairo University, Cairo, Egypt; (2) ELGalaa Military Hospital for Women and Children, Cairo, Egypt

Introduction: The diagnosis of Kawasaki disease (KD) in the absence of the classical criteria is a big challenge especially that features are non-specific. The terms "atypical" and "incomplete" have been used to describe such cases and guides the administration of intravenous immunoglobulins (IVIG).

Patients and Methods: We analyzed the data of 4 patients who were diagnosed with KD not fulfilling criteria for incomplete KD from 02/2014 to 05/2015.

Results: The first was a 4 months-old boy presenting with generalized mucopapular rash, purulent conjunctivitis, lip fissuring and 1 day of fever. His left coronary system was ectatic initially, that progressed to aneurysmal dilatation of the circumflex artery (CX). Symptoms responded to IVIG after an initial episode of severe anaphylaxis. He continues to have CA aneurysm and is on anticoagulation. Second was a 5 year-old boy with a 20-day history of fever without any other manifestations, his left anterior descending artery (LAD) had a fusiform dilatation at its midportion. Fever resolved following IVIG administration and CA dilatation regressed in 2 months.

Third was a 4 year-old boy presenting with a 7-day history of fever associated with nonpurulent conjunctivitis, follicular tonsillitis, with no other criteria of KD. Both coronary systems were dilated. Responded to IVIG and his coronary dilatation regressed within 1 month.

Fourth is a 2 year-old boy presented with 5 days of fever and generalized lymphadenopathy, hepatomegaly, and marked elevation of transaminases. Pericardial effusion was discovered on work-up, and on follow-up the LAD became ectatic and finger-peeling appeared. Fever responded to IVIG, effusion resolved in 3 days and coronary dilatation regressed in 2 weeks.

Conclusion: We report 4 cases diagnosed with KD despite insufficient criteria: typical fever being absent in one, fever was the only manifestation in another, a third with only 2 criteria in addition to fever and a fourth with generalized lymphadenopathy as the predominant feature besides fever. All of which had CA dilatation and responded clinically to IVIG, with the persistence of large coronary aneurysm in one case. We suggest that the criteria for KD be modified to a scoring system including major and minor criteria.