Giant aneurysms: a gender-specific complication of Kawasaki disease?

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Background: Kawasaki disease (KD) is a pediatric vasculitis of unknown origin. Its main complication is the development of coronary artery aneurysms (CAA) with giant CAA at the end of the spectrum.

Methods: In this cohort study, we evaluated the association between patient characteristics and the development of giant CAA based on z-scores. Multivariable, multinomial logistic regression analysis was used to identify variables associated with giant CAA.

Results: A total of 301 KD patients, comprising of 216 patients without enlargement, 45 with small-sized, 19 with medium-sized, and 21 with giant CAA with all echocardiographies at our center were retrospectively included. Remarkably, 95% of patients with giant CAA were boys. In addition to ‘no/late intravenous immunoglobulin (IVIG) treatment’, ‘male gender’ (OR 15.56, 95% CI 1.86-130.07), ‘age <1 year’ (OR 8.06, 95% CI 2.56-25.35), and ‘IVIG re-treatment (6.38, 95% CI 1.86-21.88)’ were significantly associated with an increased risk of giant CAA, with patients without enlargement as reference. Compared to patients medium-sized CAA, ‘IVIG re-treatment’ was significantly associated with giant CAA. The majority of giant CAA continued to increase in size during the first 40 days.

Conclusions: We identified risk factors associated with an increased risk of giant CAA. The difference in variables between the giant CAA group and the other CAA subgroups suggests a separation between patients with the treatment-resistant giant CAA and the other IVIG-responsive patients, in which gender may be factored as a most relevant genetic trait. The increase in size during the first 2 months indicates the need for repeated echocardiography.