Possible Involvement of the Natural Immune Systems in the Pathogenesis of Kawasaki Disease

Department of Pediatric Cardiology & Nephrology, Kyoto Prefectural University of Medicine Graduate School of Medical Science, Kyoto, JAPAN.

Introduction: Kawasaki disease (KD) is a pediatric vasculitis, which sometimes involves aneurysm as a sequel of coronary arteritis. Although its etiology remains to be elucidated, several preceding studies have indicated that altered immune responses could be involved in the pathogenesis of the disease. By employing an animal model for KD, we aimed at shedding light on the pathological mechanisms underlying the disease, from the viewpoint of natural immunity.

Materials, Method and Results: To induce KD, DBA/2 mice were administered Candida albicans water-soluble fraction (CAWS). Biochemical and immunohistochemical analyses revealed (1) increased levels of self-reactive immunoglobulin M (IgM)-type antibodies, (2) extravascular deposition of IgM around the aorta, (3) decreased levels of mannose binding protein-C (MBL-C) in plasma, and (4) deposition of MBL-C in the aortic intima, all suggesting that the natural immune system could be involved in early-stage CAWS-induced vasculitis. IgM and MBL-C have been known to activate the complement classical and lectin pathways, respectively. Interestingly, preliminary results showed that inhibition of the lectin pathway partially suppresses CAWS-induced vasculitis.

Conclusion: Taken together, these findings indicate that the pathogenesis of KD could be associated with dysregulation of natural immune systems, including the lectin pathway. This and other immune pathways may be possible therapeutic targets for KD.