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The difference of activation pattern of complement system between pediatric and adult lupus nephritis

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Objectives: Lupus nephritis (LN) has an etiopathogenesis caused by activation of the complement system. The purpose of this study is to investigate the differences and clinical implication of the activation pattern of complement system between pediatric and adult LN patients.

Methods: We retrospectively reviewed medical records of 14 pediatric and 26 adult patients whose tissue specimens were stored among patients diagnosed with LN through renal biopsy. The activation of complement system was evaluated by performing IHC for C4d (a component of the lectin pathway), C1q (a component of the classical pathway), and IF for C3 (a component of the alternative pathway) in renal tissue.

Results: The study enrolled 14 pediatric and 26 adult patients, and the proportion of female was significantly higher in both groups. The average age at diagnosis of pediatric patients was 11.7 years, and the average age of adult patients was 37.3 years. Except for age and C3 level, the baseline clinical characteristics of pediatric and adult patients were similar. Age-adjusted mean C3 value were significantly lower in pediatric patients, 33.0mg/dL in pediatric patients and 50.8 mg/dL in adult patients. As a result of complement staining of kidney tissue, the C3 and C1q positivity rate in pediatric/adult patients were 92.9/76.9% and 85.7/80.8%, respectively and there was no significant difference. However, the C4d positivity was 35.7% in pediatric patients and 76.9% in adult patients, which was significantly higher in adult than in pediatric patients. The C4d/C1q(+/+) group had poor prognosis than C4d/C1q(+/-) group. (82% vs 33%)

Conclusions: Pediatric LN patients had significantly lower C4d activation compared to adult LN patients and the co-positivity for C4d and C1q can be considered as a poor prognostic factor for LN patients. Therefore, we conclude that the activation of lectin pathway plays an important role in deciding the age difference and prognosis in LN.