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Clinical significance of C4d deposition in pediatric HSP nephritis

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Objectives: HSPN (Henoch-Schönlein Purpura Nephritis) occurs in 30-50% of pediatric HSP patients. Major pathologic findings include mesangial proliferation and IgA deposition as in IgA nephropathy. Recently, the role of complement system in IgA nephropathy has been reported. The purpose of this study was to investigate the clinical significance of C4d in pediatric HSPN.

Methods:

We have retrospectively reviewed pathologic findings and medical records of 35 children and 12 adults with HSPN diagnosed at the Department of Pediatrics and Internal Medicine, Kyungpook National University Hospital from 2006 to 2018. And C4d staining was performed to investigate the correlation between pathologic findings and clinical features.

Results:

The mean age of the 35 children with HSPN was 9.2 ± 3.9 years old and the male to female ratio was 18:17. Among them, 20 patients (57.1%) were positive for C4d. In the HSPN classification according to ISKDC, 5 (14.3%) were class I, 19 (54.3%) were II, 8 (22.9%) were IIIA and 3 (8.6%) were IIIB. The positive rate of C4d according to class was 40% (2/5) in class 1, 58% (11/19) in class II, 63% (5/8) in class IIIA and 67% (2/3) in class IIIB. The higher the ISKDC classification, the higher the C4d positive rate. The positive rate of C4d according to age was 57.1% in 35 pediatric HSPN patients, while the positive rate was 41.7% in 12 adult patients. The positive rate of C4d in children less than 10 years of age were significantly higher than those who were 10-18 years old (68.0% vs 30.0%).

Conclusions: The positive rate of C4d in pediatric HSPN showed a tendency to increase with the younger the age, severe mesangium proliferation and crescent formation. This suggests that activation of the complement system plays an important role in the pathogenesis of HSPN and further studies are needed.