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C3 glomerulonephritis in Children

JI HYUN KIM, Myung Hyun Cho, IL-Soo Ha, Hae Il Cheong, Hee Gyung Kang
Department of Pediatrics-Nephrology, Seoul National University Hospital, Korea, Republic of

Objectives:

C3 glomerulonephritis (C3GN) is marked by subendothelial and/or mesangial electron dense deposits, which displays isolated glomerular C3 deposition without concomitant staining for Ig. Since clinical manifestations and long-term outcome of C3GN have not been clarified yet, especially those in children, we reviewed our cases of pediatric C3GN.

Methods:

Among more than two thousand biopsy cases performed in our center since Jan 2004 (for 15 years), ten cases (M:F 5:5) were compatible to C3GN. Their medical records were retrospectively reviewed.

Results:

Pediatric C3GN patients at our center presented at their median age of 9.2 years with asymptomatic urinary abnormality (n=6), acute glomerulonephritis (n=3) or nephrotic syndrome (n=1). Hematuria was noted in nine (6 microscopic and 3 gross hematuria). Initial renal function was normal range in all. Initial serum C3 levels were low in all the cases but one. Pathologic diagnoses were C3GN for three, but those who were presented before 2015 had been initially diagnosed as PIGN (n=3) or MPGN (n=4); One patient with pathologic diagnosis of PIGN had follow-up biopsy in 20.2 months and finally diagnosed C3GN. All the patients were managed with ACE inhibitor or ARB, and eight were treated with additional steroid and other immunosuppressants of CNi or MMF. One patient with poor compliance progressed to ESRD in 5 years, despite therapeutic trial of eculizumab. The other 9 patients had normal renal function at last visit; Last serum C3 levels were normal in 4 and persistently low in the rest.

Conclusions:

Pediatric cases of C3GN showed diverse presentation, and some of the were initially mis-diagnosed as PIGN. Interestingly their outcome was fair, although our number of subjects was too small to draw any conclusion. For better understanding of this entity, further study on regulatory system of ACP is required, as well as other markers which can predict clinical course of C3GN.