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Clinical characteristics of childhood onset immune complex-mediated MPGN and complement-mediated C3 glomerulopathy

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Objectives: As pathophysiologic understanding of membranoproliferative glomerulonephritis (MPGN) advanced to introduce new classification, we aimed to compare the clinical characteristics between childhood onset immune complex-MPGN (IC-MPGN) and C3-glomerulopathy (C3G).

Methods: We retrospectively reviewed the medical records of 21 patients diagnosed as MPGN from a single center. Biopsy findings were reviewed for re-classification, and clinical characteristics were analyzed.

Results: Electron microscopy(EM) based classification comprised of 12 MPGN type 1, 6 MPGN type 3, and 3 MPGN type 2(Dense deposit disease(DDD)), and immunofluorescent(IF) based new criteria classified these patients into 11 IC-MPGN, and 11 C3G(8 C3GN, 3 DDD). Mean age of onset was 12.9±2.3 years for IC-MPGN, 11.2±1.6 for C3G.(p=0.054) Abnormal urinalysis from school checkup was the most common presentation in both entities(60% in IC-MPGN, 72.7% in C3G). Initial estimated glomerular filtration rate(eGFR, ml/min/m^2) and serum albumin(mg/dl) showed no significant difference between two groups.(129.8 vs 126.8, p=0.877 and 2.7 vs 3.1, p=0.342, respectively) At initial presentation, serum C3 level was decreased in 40% of the IC-MPGN patients(mean 60.3±45.7) while all C3G patients showed decreased C3 level(mean 20.6±13.8) at presentation.(p=0.024) Among 7 out of 11 C3G patients whose follow up complement level was available, 5 patients showed normalization of C3 level during median 2.83 (range 0.33-7.33) years; 4 of them subsequently reached remission with serum albumin normalization and proteinuria resolution. Full remission was achieved in 20% of IC-MPGN, and 45.5%(C3GN 4/8, and DDD 1/3) of C3G patients.(p=0.361) 30.0% of IC-MPGN patients progressed to end stage renal disease(ESRD) in median 11.1(range 5.3-11.5) years, while 18.2%(2/9 from C3GN, and none from DDD) of C3G patients progressed to ESRD in median 13.9(range 9.5-18.2) years.(p=0.635)

Conclusions: Childhood onset IC-MPGN and C3G shows no significant difference in the clinical characteristics except for the incidence of decreased C3 level. Normalization of C3 level was associated with loss of disease activity in C3G.

Table 1. comparison between IC-MPGN and C3G

	IC-MPGN (n = 10)	C3GN (n= 8)	DDD (n= 3)	*p*
Age of onset (Mean±SD), yrs	12.9±2.3	11.2±1.6		0.045
Initial presentation				
Urinalysis abnormality from school (n)	7	6	2	-
Generalized edema (n)	2	2	0	-
Gross hematuria (n)	1	0	1	-
Crescent formation in renal biopsy (n)	1	2	1	-
Initial eGFR (ml/min/min^2), (mean±SD)	129.8±45.2	126.8±42.0		0.877
Initial serum albumin level (mg/dl)	2.7±0.9	3.1±0.9		0.342
Change in C3 level				
Initial C3 level (mean±SD) (mg/dl)	60.3±45.8	20.6±13.8		0.024
Initial decreased C3 level (n)	4	8	3	-
Normalization of C3 level (n)	1	5	0	-
Persistent C3 level decrease at last f/u (n)	0	1	1	-
Follow up data unavailable (n)	3	2	2	-
Renal outcome				
Complete remission (n)	2	4	1	0.361
ESRD (n)	3	2	0	0.635
CKD stage 3-4 (n)	1	1	0	-

IC-MPGN; immune complex mediated membranoproliferative GN, C3GN; C3 glomerulonephritis, DDD; dense deposit disease, n; number, yrs; years, SD; standard deviation, ESRD; end stage renal disease, CKD; chronic kidney disease