THE KOREAN SOCIETY OF NEPHROLOGY
APCN & KSN 2024
June 13(Thu) - 16(Sun), 2024 Coex, Seoul, Korea
Promoting Sustainable Kidney Health: The Asia-Pacific and Beyond

Abstract Submission No.: A-0926

Clinical features and outcomes of Immunoglobulin G4-related kidney disease and Immunoglobulin G4-related retroperitoneal fibrosis in Korea

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Objectives : Immunoglobulin G4-related disease (IgG4-RD) is a newly recognized disease. Kidney and retroperitoneum are known to be one of the organs frequently involved. This research aimed to demonstrate the clinical features and outcomes of IgG4-related kidney disease (IgG4-RKD) and IgG4-related retroperitoneal fibrosis (IgG4-RPF).
Methods : Patients who satisfied the comprehensive diagnostic criteria on IgG4-RD from January 2009 to July 2020 at three medical institutions were included. They were classified into three groups as RKD, RPF, and Others group. The differences in symptoms, laboratory, histological and radiological findings, treatment, and final outcomes among three groups were evaluated.
Results : The number of newly diagnosed patients increased every year. Of total 94 patients, 13 (13.8 %) and 22 (23.4 %) patients were classified into the RKD and RPF group, respectively. The mean age at diagnosis in the RKD (62.2 ± 10.8 years) and the RPF groups (60.1 ± 15.2 years) was older than that in the Others group (51.1 ± 13.3 year) (P = 0.004). Eosinophilia was more common in the RKD group (38.5%) compared to the RPF (9.1%) or the Others groups (8.5%) (P =0.011). Serum IgG levels were no differences among groups. In the RKD group, the histological findings of all renal tissue were tubulointerstitial nephritis. In logistic regressions analysis, hypocomplementemia and renal function at the time of diagnosis were associated with renal involvement (Odds ratio 0.946 and 14.044, respectively). Older age, male and higher serum IgG4 levels were associated with retroperitoneal involvement (Odds ratio 1.054, 6.114 and 1.001, respectively). Glucocorticoids therapy was the most common treatment with no differences among groups. Final response rate was similar among groups.
Conclusions : Kidney and retroperitoneum are the major organs of IgG4-RD and they exhibit clinical features that distinguish them from others. Establishment universal consensus for the various subsets of IgG4-RD will be needed in the future.

Table 1.jpg

Table 1. Comparison of clinical features among three groups

Variables	RKD (n=13)	RPF (n=22)	Others (n=59)	*P*-value
Age at diagnosis (years), mean ± SD	62.2 ± 10.8*	60.1 ± 15.2*	51.1 ± 13.3	***0.004***
Sex, male, n (%)	10 (76.9)	19 (86.4)*	29 (49.2)	***0.001***
Follow-up duration (months), mean ± SD	42.6 ± 35.3	40.9 ± 25.3	37.1 ± 30.9	0.797
Comorbid diseases, n (%)				
DM	2 (15.4)	5 (22.7)	8 (13.6)	0.333
HTN	7 (53.8)	6 (27.3)	11 (18.6)	0.213
Autoimmune disease	0 (0.0)	1 (4.5)	2 (3.4)	0.900
Malignancy	2 (15.4)	0 (0.0)	3 (5.1)	0.575
Number of organ involvement, n (%)				0.795
Single	4 (30.8)	9 (40.9)	25 (42.4)	
Multiple (≥2 organs involved)	9 (69.2)	13 (59.1)	34 (57.6)	
Presence of symptoms, n (%)	9 (69.2)*	16 (72.7)*	54 (91.5)	***0.021***
Serum creatinine at diagnosis (mg/dL), mean ± SD	1.82 ± 1.49*	1.58 ± 1.46*	0.98 ± 1.38	***< 0.001***
CKD-EPI eGFR at diagnosis (ml/min/1.73m^2), mean ± SD	56.8 ± 33.6*	70.6 ± 33.4*	94.3 ± 23.7	***0.001***
Urine protein-to-creatinine ratio at diagnosis, mean ± SD	0.53 ± 0.46	0.67 ± 1.01	0.25 ± 0.38	0.108
Eosinophilia (> 500/ μL), n (%)	5 (38.5)	2 (9.1) †	5 (8.5) †	***0.011***
Elevated ESR (> 20 mm/hr), n (%)	9 (69.2)	12 (54.5)	29 (49.2)	0.152
Elevated CRP (> 0.47 mg/dL), n (%)	8 (61.5)	10 (45.5)	24 (40.7)	0.371
Hypocomplementemia, n (%)	4 (30.8)*	2 (9.1)	4 (6.8)	***0.003***
Serum IgG4 levels (mg/dL), median (IQR)	362 (122.3, 849.8)	429 (167.4, 1197.4)	334 (118.9, 983.5)	0.703
Elevated serum IgG4 (≥ 135 mg/dL), n (%)	9 (69.2)	14 (63.6)	39 (66.1)	0.485
Autoimmune Ab, n (%)	3 (23.1)	2 (9.1)	13 (22.0)	0.225
Biopsy done, n (%)	13 (100)	17 (77.3)	44 (74.6)	0.526
Disease grading, n (%)				
Definite	9 (69.2)	7 (31.8)	28 (47.5)	0.368
Probable	4 (30.8)	5 (22.7)	14 (23.7)	0.988
Possible	0 (0.0)	10 (45.5)†	17 (28.8)†	***0.016***

Table 1.jpg

Table 2. Logistic regression for associated factors with renal and retroperitoneal involvement

Logistic regression for associated factors with renal involvement

Variable	Univariate OR (95% CI)	*P*-value	Multivariate OR (95% CI)	*P*-value
Age at diagnosis (years)	0.965 (0.846, 1.101)	0.965		
Sex, male	2.292 (0.586, 8.966)	0.233		
Presence of symptoms	0.315 (0.070, 1.420)	0.133		
CKD-EPI eGFR at diagnosis (ml/min/1.73m^2)	0.970 (0.951, 0.989)	***0.002***	0.946 (0.913, 0.980)	***0.002***
Eosinophilia (> 500/ μL)	6.607 (1.696, 25.739)	***0.007***	0.402 (0.016, 10.205)	0.581
Hypocomplementemia	12.444 (2.234, 69.321)	***0.004***	14.044 (1.380, 142.945)	***0.026***
Serum IgG4 levels (mg/dL)	1.000 (0.997, 1.003)	0.458		

Logistic regression for associated factors with retroperitoneal involvement

Variable	Univariate OR (95% CI)	*P*-value	Multivariate OR (95% CI)	*P*-value
Age at diagnosis (years)	1.050 (1.010, 1.092)	***0.014***	1.054 (1.002, 1.109)	***0.044***
Sex, male	4.541 (1.410, 14.619)	***0.011***	6.114 (1.405, 26.613)	***0.016***
Presence of symptoms	1.508 (0.298, 7.636)	0.620		
CKD-EPI eGFR at diagnosis (ml/min/1.73m^2)	0.982 (0.967, 0.997)	***0.021***	0.984 (0.956, 1.012)	0.252
Eosinophilia (> 500/ μL)	0.513 (0.104, 2.523)	0.412		
Hypocomplementemia	1.378 (0.314, 6.049)	0.671		
Serum IgG4 levels (mg/dL)	1.001 (1.000, 1.002)	***0.015***	1.001 (1.000, 1.001)	***0.031***