Background: Although hypocomplementemia has been frequently reported in IgG4-related kidney disease (IgG4-RKD), few studies have investigated differences in the clinicopathological features of IgG4-RKD with and without hypocomplementemia in a relatively small cohort [1, 2].

Objectives: To compare the clinicopathological features of Japanese patients with and without hypocomplementemia in IgG4-RKD.

Methods: We retrospectively examined the clinicopathological features of 60 patients with definitively diagnosed IgG4-RKD, collected from the institutions associated with the Japan IgG4-RKD working group between December 2010 and May 2019, with reference to the presence of hypocomplementemia.

Results: Among the patients included, 42 (70%) had hypocomplementemia. In the latter group, serum levels of IgG and non-IgG4 IgG, calculated as total IgG minus IgG4, were significantly higher (mean IgG level, 3832 vs 2626 mg/ml, p =0.005, mean non-IgG4 IgG level, 2775 vs 1827 mg/ml, p =0.000). Renal function at diagnosis tended to be lower (mean eGFR level, 3832 vs 2626 mg/ml, p =0.000). There were no significant inter-group differences in the levels of serum IgG4 and IgG, or in the number of extra-renal involved organs. Renal pathology specimens were obtained from 53 patients, 70% of whom had hypocomplementemia. In the hypocomplementemia group, light microscopy demonstrated a significantly broader extent of interstitial inflammatory cell infiltration (p=0.035), and immunofluorescence revealed a higher frequency of IgG or complement deposition on the renal tubular basement membrane (p=0.048). C1q deposition on the TBM was evident only in the hypocomplementemia group. There was no significant inter-group difference in the presence of storiform fibrosis, the degree of interstitial fibrosis, the number of infiltrating IgG4-positive cells, or the frequency of membranous glomerulonephritis.

Conclusion: Hypocomplementemia in IgG4-RKD is associated with elevated levels of IgG subclasses other than IgG4, and may be related to progression of renal inflammation.

References:

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Figure 1. Retention rate of Certolizumab pegol in bio-naive and bio-experienced patients with non-infectious uveitis. Abbreviations: DRT, Drug Retention Time; LTFU, lost to follow-up; DRR, Drug Retention Rate; PAR, Patients at Risk; AE, adverse event.

Figure 2. Drug Retention Rate of Certolizumab pegol in bio-naive and bio-experienced patients with non-infectious uveitis. Abbreviations: DRT, Drug Retention Time; LTFU, lost to follow-up; DRR, Drug Retention Rate; PAR, Patients at Risk; AE, adverse event.

Conclusion: CZP in NIU showed an excellent retention rate at 24 months in bio-naive patients. However, it was more than halved when CZP was started as a ≥2nd biologic. Discontinuation of CZP in bio-experienced patients was due to lack or loss of efficacy in 22% and to adverse events in an additional 26% of patients.

References:

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