The RPGN group has more severe clinical and biological features. The BVAS was 20 ± 4 vs 14 ± 7 in non-RPGN (P < 0.001). Mean serum creatinine was 436 ± 282 µmol/L vs 137 ± 75 (P < 0.001). In the SPRF group, mean serum creatinine was 167 ± 64 µmol/L. Patients presented few extrarenal symptoms (BVAS 15.0 ± 5.6) and had almost constantly anti-MPO Ab. On the other hand, in the NRF group, anti-PR3 specificity was noted in 40% of cases and lung/ENT involvement was frequent (BVAS 17.4 ± 4.9), although 50% of patients were already receiving an immunosuppressive therapy at the time of the biopsy. Although there was no significant difference in the therapeutic scheme, prognosis was statistically different between the three groups. At 12-months, CKD stage 4 or 5 was present in 14 (31%) patients of the RPGN group, in 2 (11%) of the SPRF group, and in 0% of the NRF group (P = 0.03).

Conclusion. In our cohort, 45% of the patients with renal AAV presented with a non-RPGN. Some of them showed slowly progressive renal failure, with few extra-renal symptoms, making the diagnosis more difficult. Patients already treated for vasculitis may present mild renal symptoms but significant pathological findings. The prognosis of patients with non-RPGN is excellent if renal vasculitis is detected and treated early.

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P127 Abnormal glycosylation of serum IgG from patients with ANCA-associated systemic vasculitis: Relation to disease activity

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Introduction. Modification of the serum IgG glycoform profile has been reported as a factor in the pathogenesis in several inflammatory autoimmune diseases including ANCA disease [1,2]. The subclass of N-linked oligosaccharides deficient in terminal sialic acid and galactose (IgG-G0) is significantly increased in the serum of patients with PR3 ANCA disease and correlates with disease activity [3]. Here we investigate whether there is a similar pattern of abnormal IgG glycosylation in the serum of MPO-ANCA patients.

Methods. IgGs were isolated from serum samples from 29 patients with MPO-ANCA, 21 patients with PR3-ANCA, and 30 healthy donors. Isolated IgGs were digested with trypsin and the released glycopeptides were identified and quantified by LC-ESI-QTOF.

Results. As previously shown [3], IgG-G0 levels of PR3-ANCA patients did correlate with disease activity. In contrast, IgG-G0 levels of MPO-ANCA patients were elevated both at the time of active disease and during disease remission and therefore did not correlate with disease activity.

Discussion. This study both confirms the findings of previous investigations documenting aberrant IgG glycan modification in PR3-ANCA disease [2,3] and provides new information regarding aberrant IgG and disease parameters in MPO-ANCA disease. The lack of correlation between IgG glycoform levels and disease activity in MPO-ANCA disease raises the possibility that the humoral immune dysregulation driving the production of aberrantly glycosylated IgG may substantially predate the clinical manifestations of the disease.

Conclusion. Significant differences exist between MPO- and PR3-ANCA diseases regarding the association of aberrantly glycosylated IgG levels with disease activity. It is conceivable that these differences may contribute to significant clinical differences in the disease course, severity, and relapse rate observed between the two diseases.

References

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Miscellaneous

P128 Renal histopathological classification of ANCA associated glomerulonephritis: A validation study of 92 patients

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Introduction. A novel histopathological classification was proposed recently for ANCA associated glomerulonephritis: focal, crescentic, mixed and sclerotic.

Objectives. Our aim was to perform a validation study and to determine the prognostic value of the classification for predicting renal outcome.

Methods. Ninety-two renal biopsies with ANCA associated glomerulonephritis were classified retrospectively. eGFR during follow-up was corrected for baseline eGFR. Renal survival was assessed using the Kaplan-Meier estimator, comparing the histological classes t. Age, gender, ANCA subtype, diagnosis, baseline eGFR and histological category were included as covariates in the Cox regression model.

Results. Median age was 61.9 years, 53.3% patients were male and 61.9% had microscopic polyangiitis (MPA) vs 38% granulomatosis with polyangiitis (GPA). Mean follow-up was 61.8 months. Twenty-six percent reached end stage renal disease (ESRD) within a mean time of 25.3 months. 23% died within a mean time of 45.3 months. 26.1% were classified as focal, 19.6% crescentic, 14.1% sclerotic and 40.2% mixed. Renal survival at 1 year was a 100% in the focal class, 86% in mixed group, 72% in crescentic and 61% in the sclerotic class (P = 0.002). The sclerotic class displayed the lowest eGFR at 5 years (P < 0.05) and increased mortality (38%). The crescentic group had the lowest baseline eGFR, but reasonable long-term renal recovery (Supplementary data).

Discussion. Similar to previous studies the focal group did best, and the sclerotic group worst in terms of ESRD. In contrast to previous results our crescentic group did no better and somewhat worse than the mixed group in terms of ESRD, which we associate with a lower baseline GFR. However, those crescentic patients without ESRD had better recovery than the mixed group.

Conclusion. Renal histology and baseline renal function were a better predictor of renal outcome than baseline eGFR alone. Whether this...