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, antiPR3 specificity was noted in 40% of cases and lung/ENF 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 subfamily 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 donors 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 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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2. Addenbrooke’s hospital, Cambridge, United Kingdom

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
system could be useful to guide treatment, further prospective studies are needed.

Supplementary data associated with this article can be found on the website of La Presse Médicale (http://www.em-consulte.com/revue/lpm).

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P129

Aortitis evidenced by PET-CT 18FDG in the setting of ANCA-associated vasculitis: About four cases

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Introduction.– We have previously reported an atypical case of anti-MPO ANCA-associated vasculitis (AAV) presenting with aortitis evidenced by PET 18FDG [1]. The presumptive diagnosis was an overlap between granulomatosis with polyangiitis (GPA) and Takayasu’s arteritis and we discussed the significance of the occurrence of aortitis in GPA. Besides, a recent cluster analysis of 673 patients with AAV introduced the concept of different phenotypes of AAV, one of them being “cardiovascular AAV”, representing 9% of cases [2].

Methods.– We reviewed all PET-CT 18FDG performed in patients with AAV followed in our hospital in order to detect aortic and/or large vessels hypermetabolism. Hypermetabolic activity in other organs was also systematically reviewed. For each patient, we looked for the diagnostic specificity of ANCA, the presence of cardio-vascular clinical events and recorded the final diagnosis according to classification of AAV.

Results.– Thirty-three exams were performed in 14 patients, four of these patients (29%) presented aortic hypermetabolism. No patient experienced one of the cardiovascular events listed in the Birmingham Vasculitis Activity Score (version 3). Most of the patients presented atypical features of AAV (Supplementary data).

Conclusion.– This small series suggests that the occurrence of aortitis in the setting of AAV may be underestimated and deserves to be evaluated by the mean of PET-CT in larger cohorts. Although these imaging findings are not associated with obvious cardiovascular symptoms, we believe that they support the recent concept of “cardiovascular AAV”.

Supplementary data associated with this article can be found on the website of La Presse Médicale (http://www.em-consulte.com/revue/lpm).

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References


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P130

The informational needs of patients with ANCA-vasculitis – a multinational study

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Introduction.– ANCA-associated vasculitis (AAV) is a rare, complex, often relapsing, multisystem disease. Patients with AAV often seek information about their disease.

The aim of the study was to compare the informational needs of British, North American, and European patients.

Methods.– We developed a vasculitis informational needs questionnaire (VINQ). Patients rated the importance of various aspects of information using 33 questions relating to five domains (symptoms, investigations, treatment, physical, psychological) on a 5-point Likert scale (1 = unimportant, 5 = extremely important). The preferred mode of information delivery was also rated. The VINQ was distributed to members of Vasculitis UK (VUK) by regular mail, and to patient registrants of the Vasculitis Clinical Research Consortium (VCRC) via the internet.

Results.– There were 314 VUK 273 VCRC respondents. Demography is given in the table. Respondents rated information on diagnosis, prognosis, diagnostic tests, treatment, and side effects as extremely important (table I). Information on patient support groups and psychosocial care was less important. There was no difference in the ratings of needs based on group (VUK or VCRC), sex, age, disease duration, or disease subtype. The most preferred methods of providing information for both groups was by a doctor (with or without written material) or by the internet; educational courses and CD/DVD were the least preferred methods.

Table I

<table>
<thead>
<tr>
<th>Study Group Characteristics</th>
<th>All subjects</th>
<th>Vasculitis UK</th>
<th>VCRC</th>
</tr>
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<tbody>
<tr>
<td>Group Size</td>
<td>587</td>
<td>314</td>
<td>273</td>
</tr>
<tr>
<td>Age: median (range)</td>
<td>60 (51–67)</td>
<td>63 (52–70)</td>
<td>58 (49–54)</td>
</tr>
<tr>
<td>Women n (%)</td>
<td>300 (32)</td>
<td>116 (37)</td>
<td>184 (67)</td>
</tr>
<tr>
<td>GPA n (%)</td>
<td>448 (76)</td>
<td>255 (61)</td>
<td>193 (71)</td>
</tr>
<tr>
<td>MPA n (%)</td>
<td>34 (6)</td>
<td>13 (4)</td>
<td>21 (8)</td>
</tr>
<tr>
<td>EGPA n (%)</td>
<td>105 (16)</td>
<td>46 (15)</td>
<td>59 (22)</td>
</tr>
</tbody>
</table>

Ratings of Informational Elements

<table>
<thead>
<tr>
<th></th>
<th>All subjects</th>
<th>Vasculitis UK</th>
<th>VCRC</th>
</tr>
</thead>
<tbody>
<tr>
<td>Diagnosis (ie, prognosis)</td>
<td>4.5</td>
<td>4.3</td>
<td>4.5</td>
</tr>
<tr>
<td>Investigations (type + results)</td>
<td>4.5</td>
<td>4.5</td>
<td>4.5</td>
</tr>
<tr>
<td>Treatments (inc side effects)</td>
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<td>4.5</td>
<td>4.6</td>
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<tr>
<td>Lifestyle</td>
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<td>3.7</td>
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<tr>
<td>Patient support groups</td>
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<td>Psychological care</td>
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