Case Series of ANCA Vasculitis as Prognostic Marker for Severity or Relapse of Glomerulonephritis.

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Case 1 involves a 63 year old female with microscopic polyangitis, pulmonary-renal syndrome. She was managed conservatively with rituximab. The primary author and lead investigator do not have any have conflicts of interest to declare or financial disclosures.

Table 1. Case Series of ANCA vasculitis associated glomerulonephritis

<table>
<thead>
<tr>
<th>Case #, sex, age, diagnosis</th>
<th>Renal function at remission</th>
<th>Renal function at relapse</th>
<th>Special labs</th>
<th>Treatment course</th>
</tr>
</thead>
<tbody>
<tr>
<td>Case 1: Female 63 year old, Microscopic Polyangitis, Pulmonary-renal syndrome</td>
<td>Creatinine 0.90 mg/dL, eGFR 30/min</td>
<td>Creatinine 2.60 mg/dL, eGFR 19/min</td>
<td>P-ANCA titer 1:80 (positive)</td>
<td>• methylprednisolone 250 mg</td>
</tr>
<tr>
<td>Case 2: Female 60 year old, 2007, C-ANCA, anti PR3 positive, FSGS (biopsy)</td>
<td>Creatinine 1.8 mg/dL, eGFR 31/min</td>
<td>Creatinine 3.90 mg/dL, eGFR 11/min</td>
<td>Remission: ESR 8 to 16 mm/hr</td>
<td>• anti-MPO 80 U/mL (ref. -20)</td>
</tr>
<tr>
<td>Case 3: Female 78 year old, anti-PO, ANCA, glomerulonephritis</td>
<td>Creatinine 1.37 mg/dL, eGFR 37/min</td>
<td>Creatinine 1.81 mg/dL, eGFR 27/min</td>
<td>Remission: ESR 14 mm/hr</td>
<td>• Rituximab 375 mg/m²</td>
</tr>
</tbody>
</table>

Case Presentations

Case 1 involves a 63 year old female with microscopic polyangitis who developed pulmonary-renal syndrome requiring intubation, plasmapheresis and immunosuppression. Table 1 shows anti-MPO titer elevated to 80 U/ml with proteinuria and acute kidney injury.

Case 2 involves a 60 year old female with CKD stage 3 and focal segmental glomerulosclerosis in remission who relapsed and developed pauci-immune crescentic glomerulo-nephritis. In case 2, anti-PR3 titer was elevated (>100 U/mL) and associated with a 20-fold increase in the Protein/creatinine ratio of 2343 mg/dL. She eventually required peritoneal dialysis.

Case 3 involves a 78 year old female with CKD stage 3 and anti-PO vasculitis in remission but developed AKI with a 6 fold increase in anti-MPO titer and 10 fold increase in proteinuria. She was managed conservatively with rituximab.

Table 1 shows anti-MPO titer elevated to 80 U/ml with proteinuria and requiring intubation, polyangitis

Table

<table>
<thead>
<tr>
<th>Abbreviations:</th>
<th>Creatinine (mg/dL), eGFR (ml/min)</th>
<th>Prot/Crt ratio</th>
<th>Remission: ESR 8 to 16 mm/hr</th>
<th>• Rituximab 375 mg/m²</th>
<th>• cyclophosphamide</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>ref. (reference), eGFR (estimated glomerular filtration Rate), ESR (erythrocyte sedimentation rate), C-ANCA (cytoplasmic anti-neutrophilic cytoplasmic antibodies), P-ANCA (peri-nuclear anti-neutrophilic cytoplasmic antibodies), MPO (Myeloperoxidase), PR3 (Proteinase-3), PIGC (Pauci immune crescentic glomerulonephritis), FSGS (Focal Segmental glomerulosclerosis), Crt (Creatinine), Prot (Protein), Prot/Crt ratio ref. &lt; 160</td>
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</table>

Conclusion

- Although measuring ANCA to predict disease activity remains controversial, this case series demonstrates association between ANCA titer and severity of renal disease. ANCA titers may provide prognostic value that can be useful when discussing therapy options with the renal patient.

- Understanding the pathophysiology of disease activity and remission to help define better biomarkers to positively affect adverse events and patient outcomes.

References


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