nation (ECMO). Symptoms resolved during treatment with prednisolone and cyclophosphamide. Six-months-mortality was 15%, however deaths were not directly related to the lung involvement (one patient due to bowel ischemia, one patient due to multi-organ failure).

Discussion. – Among patients with ARDS routine screening for ANCA can rapidly establish the diagnosis of AAV, whereas biopsy is often difficult to obtain. ARDS was more commonly associated with PR3-ANCA positivity. Early mortality was not related to pulmonary involvement. Overall outcome of patients with AAV and ARDS was favourable, even in patients requiring ECMO therapy.

Conclusion. – Outcome of patients with AAV and ARDS is favourable. Early mortality was not related to pulmonary involvement. Among patients with ARDS routine screening for ANCA can rapidly establish the diagnosis of AAV.

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Focal necrotizing and crescentic glomerulonephritis in patients with normal serum creatinine


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Introduction. – Focal necrotizing (FNGN) and crescentic glomerulonephritis (CGN) are common renal manifestations of systemic vasculitis. They usually present as rapidly progressive glomerulonephritis and have a poor prognosis if untreated. These pathological findings, however, are not always accompanied by abnormalities of renal function. We aimed to establish the frequency and outcomes of patients presenting with FNGN/CGN and normal serum creatinine at our centre.

Patients. – We conducted a retrospective review (1995–2011) of all adult patients who presented with native renal biopsy proven FNGN/CGN and normal serum creatinine (< 120 micromol/L).

Results. – Thirty-eight patients were identified, median age 57 years (range 17–78), 29% male. Biopsies showed median 14 glomeruli (4–33), with 32% (4–100%) of glomeruli affected by necrosis/crescents. All patients received immunosuppression in accordance with local protocols. Median duration of follow-up was 45 months (2–120) months, average 39.5 months. Clinical features and outcomes are summarised in Table I

Table I

<table>
<thead>
<tr>
<th>Diagnosis</th>
<th>Number of cases (%)</th>
<th>% affected glomeruli</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pauci-immune GN</td>
<td>23 (74%)</td>
<td>32% (4–100%)</td>
</tr>
<tr>
<td>Lupus Nephritis</td>
<td>7 (18%)</td>
<td>17% (4–50%)</td>
</tr>
<tr>
<td>Anti-GBM disease</td>
<td>2 (5%)</td>
<td>36% (26–47%)</td>
</tr>
<tr>
<td>IgA Nephropathy</td>
<td>1 (3%)</td>
<td>50%</td>
</tr>
</tbody>
</table>

Biochemistry

<table>
<thead>
<tr>
<th>Test</th>
<th>At biopsy</th>
<th>At 1 year</th>
<th>At last follow-up</th>
</tr>
</thead>
<tbody>
<tr>
<td>Creatinine (µmol/L)</td>
<td>84.00 (52–115)</td>
<td>82 (58–145)</td>
<td>77.0 (57–107)</td>
</tr>
<tr>
<td>Albumin (g/dL)</td>
<td>29.0 (10–40)</td>
<td>38 (30–46)</td>
<td>37.0 (22–45)</td>
</tr>
<tr>
<td>uPCR (mg/mmol)</td>
<td>71.2 (0–681)</td>
<td>23 (0–272)</td>
<td>15.0 (0–238)</td>
</tr>
</tbody>
</table>

1 Results expressed as median (range). Censored for ESRF/death.

The vast majority of patients had good outcomes at 1 year and at last follow-up. The majority of patients had extra-renal manifestations of vasculitis or autoimmune rheumatic disease. Two patients progressed to ESRF (both secondary to lupus nephritis, at 21 & 29 months) and four patients died during follow-up (at 2, 12, 96 & 122 months).

Discussion. – FNGN/CGN may occasionally present in patients with normal serum creatinine. This occurs most commonly in patients with pauci-immune GN secondary to ANCA-associated vasculitis. Abnormal urinary findings in association with extra-renal manifestations of disease may alert clinicians to this diagnosis. Confirmation of organ-threatening involvement on renal biopsy may significantly influence treatment decisions.

Conclusion. – Low threshold of clinical suspicion for FNGN/CGN, prompt biopsy and early initiation of treatment may prevent irreversible kidney damage and improve long-term outcomes in these patients.

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P168

Clinical and pathological study on 34 patients with primary ANCA-associated systemic vasculitis with renal immune complex deposition

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Introduction. – To analyze the clinical and pathological characteristics of Chinese patients with primary antineutrophil cytoplasmic autoantibody (ANCA)-associated systemic vasculitis (AASV) with renal immune complex deposition.

Patients. – Thirty-four patients diagnosed with ANCA-associated systemic vasculitis in Shanghai Ruijin Hospital with renal immune complex deposition were enrolled in this study. Their clinical and pathological data were collected and studied, and compared with other 76 AASV patients having classic pauci-immune glomerulonephritis.

Results. – Of the 34 patients, 27 patients were microscopic polyangiitis (MPA), six patients were Wegener’s granulomatosis (WG) and one patient was Churg-Strauss syndrome (CSS). The mean age was 56.4 ± 16.4 years with a male/female ratio of 1:1.27 (19/15). Kidney and lung were the most common organs involved, taking 100% (34) and 76.5% (26) respectively. 79.4% (27) of patients had impaired renal function, with an average serum creatine of 390.3 ± 284.9 (median 352.46–1067) µmol/L. C3 (82.4%) and IgM (50%) were the most common immune complex deposits observed in kidney, mostly located in mesangial areas and capillary loops. During the follow-up [median 39 (1–120) months, average 39.5 ± 31.7 months], all following a therapy of daily oral OCS combined with immunosuppressants, six (17.7%) patients died and 11 (32.4%) finally progressed to end-stage renal disease (ESRD). Compared with patients having classical pauci-immune glomerulonephritis, patients with renal immune complex deposition had more significant proteinuria, with a higher prevalence of nephrotic syndrome and hypocomplementaemia, and also a higher risk for progressing to ESRD.

Conclusion. – Our study found that AASV patients with renal immune complex deposition had significantly more proteinuria, more hypocomplementaemia and a higher risk for progressing to ESRD than those having classic pauci-immune glomerulonephritis, and that might indicate a worse renal prognosis.

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