Renal impairment in a patient with IgG4-Related disease.

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Case

- 52 year old male
- Since 1997: malaise, abdominal tumor (mesenteric mass 5cm)
- Diagnostics : no explanation.
Diagnostic

- Elevated ESR and CRP, kidney function (creatinine 103 umol/L, eGFR 72 mL/min, urine protein: 0.25 g/l)

- CT abdomen:
Biopsy abdominal mass

IgG 20x

IgG4 20x
Diagnosis

- IgG4-related disease (IgG4-RD)
Diagnostic

- Elevated serum IgG4 (25 g/l)
Treatment

- prednisone 1mg/kg and azathioprine:
  - Decrease in inflammatory markers and serum IgG4
  - Decrease in abdominal mass (from 5cm to 3.6 cm)
While tapering prednisone, patient developed nephrotic syndrome and acute renal insufficiency:

- Creatinine: 150 umol/l
- eGFR: 46 mL/min
- Urine total protein: 8.28 g/l
- Serum albumin: 2.2gr/dl
- 1+ erythrocytes
Table 1: Common manifestations of IgG4-RD by organ system

<table>
<thead>
<tr>
<th>Organ System</th>
<th>Manifestations</th>
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<tbody>
<tr>
<td>Gastrointestinal</td>
<td>Autoimmune pancreatitis, Sclerosing cholangitis, Sclerosing mesenteritis</td>
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<tr>
<td>Head and neck</td>
<td>Eosinophilic angiocentric fibrosis (puffy, fibroinflammatory lesions of orbits and upper respiratory tract), Orbital pseudotumor, Riedel’s thyroiditis, Mikulicz’s disease (enlargement of lacrimal, salivary, and parotid glands), Kuttner’s tumor (salivary gland enlargement)</td>
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<tr>
<td>Allergy/respiratory</td>
<td>Asthma, atopy, allergy, Tracheal stenosis, Chronic sinusitis, Pleural and pulmonary nodules, interstitial lung disease, Fibrosing mediastinitis</td>
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<tr>
<td>Systemic</td>
<td>Multifocal fibrosclerosis (orbits, thyroid, retroperitoneum, mediastinum)</td>
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<tr>
<td>Large vessels</td>
<td>Inflammatory aortic aneurysm, Periaortitis and periarteritis</td>
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<tr>
<td>Renal/retroperitoneum</td>
<td>Retroperitoneal fibrosis (RPF), Intrinsic renal disease (IgG4-related kidney disease): Tubulointerstitial nephritis (IgG4-TIN); often with hypocomplementemia, Membranous glomerulonephropathy (IgG4-MGN); anti-PLA2R negative, Renal pyelitis</td>
</tr>
<tr>
<td>Nervous system</td>
<td>Hypertrophic pachymeningitis, Hypophysitis, Peri-neural masses</td>
</tr>
<tr>
<td>Blood and bone marrow</td>
<td>Eosinophilia, Polyclonal hypergammaglobulinemia with elevation in IgG4 and other immunoglobulins</td>
</tr>
</tbody>
</table>

Renal biopsy: PAS+ 20x
Renal biopsy: Jones 20x
Renal biopsy: Congo Red 20x
Renal biopsy: amyloid A immunohistochemistry
Renal biopsy:

- Immunofluorescence: No specific staining
- Mass-Spec not possible due to insufficient amount of tissue left.
Renal biopsy: diagnosis

- Renal amyloidosis type AA
Additional tests

- Serum amyloid: 105 mg/l (normal < 4.0 mg/l)

- Retrospective evaluation of serum:

- Serum amyloid during first presentation: 350 mg/l
Conclusion

1. Longstanding, untreated, IgG4-RD

2. Renal amyloidosis type AA secondary to IgG4-RD

Treatment: rituximab and monthly methylprednisolone 1000mg
TO THE EDITOR: Here, we describe a patient with renal amyloid A (AA) amyloidosis that was apparently associated with IgG4-related disease. A 53-year-old man with long-standing malaise and fatigue had a slow-growing mesenteric mass (5 cm in diameter) that had been present 16 years earlier on the basis of a review of previous imaging studies. (The case had been included in a case series of IgG4-related disease.) In July 2015, the patient had received a diagnosis of mesenteric IgG4-related disease with involvement of surrounding lymph nodes, which had fulfilled international consensus criteria (Fig. 1A and 1B). The only blood abnormalities at that time had been

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THANK YOU!