AA-amyloidosis in a kidney biopsy on top of pauci-immune crescentic glomerulonephritis

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In March 2011, a 56-year-old woman presented with fever of unknown origin accompanied with symmetric arthralgias and sinusitis. Her medical history was notable for diabetes mellitus Type 2, hypertension, smoking and multiple episodes of sinusitis. Admission blood chemistry tests revealed a leucocyte count of $14.3 \times 10^9/L$, a C-reactive protein (CRP) level of $4.4 \text{ mg/dL}$ ($<0.5 \text{ mg/dL}$), a creatinine level of $266.6 \text{ µmol/L}$ and a blood urea nitrogen (BUN) level of $47 \text{ mg/dL}$. Urinalysis demonstrated gross proteinuria with a total protein/creatinine ratio of $6.4 \text{ g/g}$ and nephritic sediment. Anti-nuclear, anti-GBM and MPO-ANCA antibodies were negative, Complement factor 3 and 4 in the normal range but PR3-ANCA antibodies were clearly detectable. Of note, computed tomography of her head and thorax displayed signs of pansinusitis but no pulmonary changes. A kidney biopsy was performed which revealed pauci-immune crescentic glomerulonephritis with crescent formation in 9 of 14 glomeruli. Most interestingly, glomeruli demonstrated Periodic Acid Schiff (PAS)-negative mesangial and perihilar deposits (Figure 1), which were positive for congo red (Figure 2, upper left insert). Immunohistochemistry revealed positivity for amyloid-P and amyloid-A (Figure 2, lower right insert). Electron microscopy confirmed fibrillary deposits consistent with amyloid (Figure 2). Amyloid was also present in arteriolar walls in the kidney. Since all other causes of AA-amyloidosis were excluded, the most probable underlying cause for AA-amyloidosis in our patient is long lasting granulomatosis with polyangiitis (Wegener’s). Since our patient suffered from multiple episodes of sinusitis in her medical history, it might be speculated that granulomatosis with polyangiitis (Wegener’s) was long-lasting, allowing the development of AA-amyloidosis.

Treatment of the patient was started with corticosteroids and intravenous cyclophosphamide. Because of a further decrease of kidney function requiring haemodialysis treatment, plasmapheresis was performed. After daily intensive plasmapheresis treatment for >2 weeks PR3-ANCAs became negative and fever as well as arthralgias resolved, but the patient’s kidney function has not recovered as yet.

Conflict of interest statement. None declared.

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Fig. 2. Typical fibrillary structure of amyloid deposits on transmission electron microscopy (uranyl acetate and lead citrate). Upper left inset: positive congo red staining of amyloid deposits. Lower right inset: positive immunohistochemistry for Amyloid A.