Concurrent renal and thyroid amyloidosis associated with early large vessel vasculitis

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Introduction: Amyloidosis is a disease caused by the deposition of amyloid fibrils. Amyloidosis can involve various organs, with the kidneys and heart being the most common, and its presentation varies depending on the organ involved. AA amyloidosis is often associated with systemic inflammatory disorders. There have been some reports of renal amyloidosis accompanying Takayasu arteritis. We report a case of concurrent renal and thyroid amyloidosis associated with early large vessel vasculitis.

Case: A 50-year-old woman admitted for acute elevation of serum creatinine and heavy proteinuria. She complained of fatigue, dyspnea on exercise, weight loss from 4 months ago. She had a history of recently diagnosed hypothyroidism with palpable goiter. At admission, serum creatinine was 3.99 mg/dL, C-reactive protein was 15.99 mg/dL, and spot urine protein-to-creatinine ratio was 8.187 g/g. Urine protein electrophoresis did not show any monoclonal peak. Kidney biopsy showed eosinophilic mesangial expansion with Congo-red-positive deposits (Fig. 1A, 1B), and randomly arranged fibrils under electron microscopy (Fig. 1C), compatible with AA amyloidosis. Thyroid biopsy also exhibited eosinophilic deposits and Congo-red-positive amyloid materials (Fig. 2A, 2B). Diffuse wall thickening from ascending aorta to both common carotid arteries was noted on chest computed tomography (Fig. 3). The patient was treated with prednisolone 1mg/kg/day, azathioprine 50mg per day, and levothyroxine. After 4 months of treatment, proteinuria was improved to 1.045 g/g of urine protein-to-creatinine ratio. After 10 months from diagnosis, the patient complained of orthopnea. Echocardiography revealed severe aortic regurgitation with post-inflammatory change of aortic valve. The patient's symptom was relieved with management for heart failure including angiotensin-receptor blocker, beta-blocker, and loop diuretics.

Conclusion: As AA amyloidosis involving multiple organs can be presented as an early manifestation of large vessel vasculitis, cautious evaluation and monitoring for additional manifestation is warranted.