An IgA nephropathy with elevated IgG4 in Eosinophilic granulomatosis with polyangiitis (EGPA)

경상대학교 의과대학 내과학교실

*김성제, Mingyo Kim

Introduction: There have been several studies showing that elevated IgG4 levels are commonly observed in eosinophilic granulomatosis with polyangiitis (EGPA), as known as Churg-Strauss syndrome (CSS). Up to 50% of patients with IgG4-RD have allergic diseases such as bronchial asthma or chronic sinusitis. The characteristic renal manifestations of EGPA and IgG4-related disease (IgG4-RD) are Pauci-immune necrotizing glomerulonephritis and tubulointerstitial nephritis, respectively. However, the occurrence of IgA nephropathy in patients with EGPA is extremely rare. Here, we present a case of IgA nephropathy with elevated IgG4 in an EGPA patient. **Case presentation:** A 47-year old male who has a history of asthma presented persistent nasal congestion and petechia in the lower extremities. Laboratory data demonstrated leukocytosis (17.880/mm^3) with eosinophilia (10,066/mm^3) of 56.3%, a positive finding of perinuclear anti-neutrophil cytoplasmic antibodies (P-ANCA), elevated IgG (1642.9mg/dl) and IgG4 (718mg/dl), increased IgG4/IgG ratio (0.43), and proteinuria. Computed tomography showed swelling of paranasal sinuses and multifocal centrilobular nodules and ground-glass opacities in both lungs. Skin biopsy of lower extremities revealed lymphocytic infiltrations with eosinophilia and a positive finding of IgG4 immunohistochemical stain in the perivascular area. Although the clinical manifestation of the patient would be met the EPGA criteria, we performed a kidney biopsy to rule out the IgG4-RD. The results of the kidney biopsy showed interstitial and perivascular eosinophilic infiltrations with IgA nephropathy and a negative finding of IgG4 immunohistochemical stain.(Figure.A,Figure.B). **Conclusion:** This report remarks that renal manifestation of EGPA would be expressed as IgA nephropathy. It should be considered a pathophysiologic link between EPGA and IgA nephropathy.

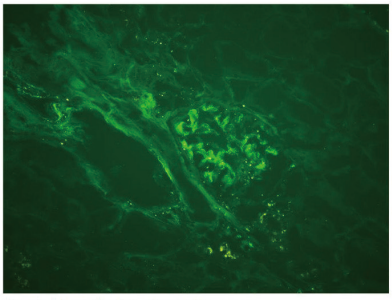

Figure A. IgA was identified at Immunofluorescence.

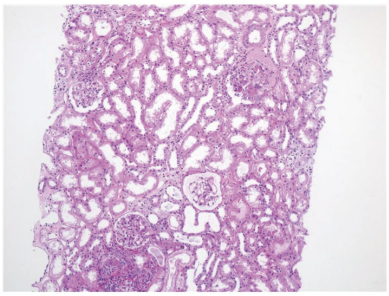

Figure B.IgA was identified at Immunohistochemistry.