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A case of multi-organ involvement in IgG4-related disease with superimposed IgA nephropathy

JI Wook Choi, Sun Moon Kim
Department of Internal Medicine-Nephrology, Chungbuk National University Hospital, Korea, Republic of

Case Study: IgG4-related disease (IgG4-RD) is a systemic disease associated with high serum IgG4 levels. It has various clinical manifestations such as salivary gland enlargement, autoimmune pancreatitis, and interstitial nephritis. There have been few studies about the relationship between IgG4-RD and IgAN. Here, we present a case of IgG4-RD superimposed with IgAN.
A 50-year old man was admitted for progressive renal dysfunction. Four years and seven months before admission, serum creatinine and urinalysis were normal. Six months before admission, both parotid glands were swollen. At admission, serum creatinine was 2.6 mg/dL, and spot urine protein-to-creatinine ratio was 0.6 g/g. After adimission, chest CT scan showed interstitial pneumonitis in the both lower lobes and abdominal CT exhibited multiple intraabdominal lymph nodes enlargements and swollen pancreas. Serum amylase levels were increased. Serum IgG levels were increased, specifically IgG4 levels increased to 5370 mg/dL. Renal biopsy revealed histologic findings compatible with IgG4-RD including marked tubular attenuation with diffuse interstitial infiltration and increased IgG4 positive cells (30-45/HPF). Further, it also demonstrated glomerular mesangial IgA staining compatible with IgAN. With high dose steroid treatment, serum creatinine was decreased. This case suggests a possible link between IgG4-RD and IgAN.