

다낭성 신증 환자에서 병발되어 조직검사로 증명된 루푸스 신염

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Case of Biopsy Proven Lupus Nephritis in Autosomal Dominant Polycystic Kidney Disease

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Introduction: It is not easy to decide to perform renal biopsy in patients with autosomal dominant polycystic kidney disease (ADPKD). Therefore, precise diagnosis is often an issue of concern when ADPKD becomes complicated by another kidney disorder during the long clinical course of the disease. Herein, we report biopsy proven case of lupus nephritis in a patient with ADPKD.

Case: A 48-year-old female patient visited our hospital for generalized edema and arthralgia 8 years ago. She was diagnosed with ADPKD 3 years earlier. She had the evidence of acute glomerulonephritis such as nephrotic range proteinuria, microscopic hematuria and azotemia at admission. Her arthritis, low complement levels, high anti-ds DNA antibody titer, and positive FANA test were consistent with systemic lupus erythematosus (SLE). CT imaging revealed multiple cysts in liver and both kidneys, compatible with ADPKD. As her renal function declined rapidly, we performed laparoscopic kidney biopsy that confirmed WHO class IV lupus nephritis. She had been treated with steroid and cyclophosphamide for 4 years, which improved her renal function. It had been stable for 6 years, but began to decrease gradually from 2 years ago. Additional steroid pulse could not change the renal function deterioration and her serologic tests showed negative FANA, normal range of complements and anti-ds DNA antibody. With no evidence of lupus reactivation, her renal function decline seemed to be the progression of ADPKD. She reached end-stage renal disease and start hemodialysis in 8 years since diagnosed.

Conclusion: This case supports that renal biopsy is needed in ADPKD patients with nephrotic range proteinuria for accurate diagnosis and proper management.

Key Words: 다낭성 신증, 루푸스 신염, 신증후군
ADPKD, lupus nephritis, nephrotic syndrome