P-87

말기신부전으로 발현한 성인에서의 특발성 사구체낭성 신질환

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Sporadic Glomerulocystic Kidney Disease Manifested by End Stage Renal Disease in an Adult

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Glomerulocystic kidney disease (GCKD) is a rare type of renal cystic disease characterized histologically by cystic dilation of Bowman's space and collapse of the glomerular tuft. This disease is often reported in infants and young children with heredity and extrarenal anomalies. However, some sporadic cases were presented in adults. We additionally report sporadic GCKD in 28 years adult Korean man manifested with end stage renal disease (ESRD). He was transferred to our emergency room due to overt azotemia and persistent vomiting. One month before visit, He had felt general weakness and fatigue. We could not catch his extra renal abnormality and he had no family history who presented with ESRD. His last serum creatinine (Cr) level available at 3 years ago in our hospital was 1.2 mg/dL. Initial blood urea nitrogen and serum Cr was 151 mg/dL and 12.1 mg/dL respectively. No hydronephrosis was detected and kidney size was normal on Ultrasonography. Kidney biopsy revealed marked dilated and empty Bowman's spaces and atrophy of glomerular tuft. He is undergoing regular hemodialysis through left radio-cephalic fistula. Although GCKD is extremely rare cause of ESRD, we consider this and do renal biopsy when we confront unknown etiology of ESRD accompanied with normal size kidney.

Key Words: 사구체낭성 신질환, 특발성, 말기신부전
Glemerulocystic kidney disease, Sporadic, End stage renal dise