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학교소변검사에서 발견되어 5년간 임상 및 신생검 소견을 추적 관찰한 C3 신염 증례

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A Case of C3 Glomerulonephritis Diagnosed From School Urine Screening: Follow-up Clinical and Biopsy Findings Over a 5-year Observation Period

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C3 glomerulonephiritis (C3GN) is a recently-described entity, defined as strong C3 deposition but without any immunoglobulins, C1q, or C4 on immunofluorescence (IF) microscopy. Proliferation either can or cannot be found on light microscopy (LM), and electron dense deposits in mesangium and/or subendothelium can be seen on electron microscopy (EM). However, treatment planning, prognosis, morphologic evolution are still unknown. We present a case of a 10-year old girl who visited for microscopic hematuria and proteinuria on school urine screening. She was asymptomatic with normal renal function. Persistent C3 hypocomplementemia led her to a kidney biopsy. First biopsy showed membranoproliferative glomerulonephritis (MPGN) pattern with only C3 deposition on IF. She was under supportive therapy including angiotensin-converting enzyme (ACE) inhibitor for over two years. During the period, she did not have symptoms nor proteinuria. But microscopic hematuria and C3 hypocomplementemia persisted. After 33 months of treatment, the patient quit medication on her own. Over one year later, she revisited clinic with microscopic hematuria and normal renal function at the age of 15. She denied of any symptoms during the entire period. Kidney function was within normal range, without decline, and C3 hypocomplementemia persisted. A second kidney biopsy was taken for nephrotic-range proteinuria (53.37 mg/m^2/hr) which showed MPGN pattern on LM, but only C3 deposition on IF as same as those of first biopsy. Pathologic changes include decrease in glomerular neutrophil infiltration, increase in mesangial cell proliferation, electron dense deposits, capillary wall doubling, tubular atrophy, and interstitial fibrosis. The intensity of C3 deposition increased, and more positive areas were found. Under the new entity, she was diagnosed as C3GN. Currently, she is treated with ACE inhibitor with addition of angiotensin-receptor blocker for persistent proteinuria, with no change in microscopic hematuria. The reclassification clarifies our patient's stable renal function status better than conventional MPGN which would present with worse outcome. This is the first case report of C3GN with a follow-up biopsy. We were able to once again identify the usefulness of school urine screening.

Key Words: C3신병증, C3신염, 대체보체경로
C3 glomerulonephritis, Alternative complement pathway