Case 8

A case of freguent relapsing nephrotic syndrome combined with IgA nephropathy that showed the diffuse global sclerosis at second renal biopsy

Joo Hoon Lee[1], Yong Mee Cho[2], Young Seo Park[1].

Department of Pediatrics, Asan Medical Center, University of Ulsan College of Medicine, Seoul, Korea[1]

Department of Pathology, Asan Medical Center, University of Ulsan College of Medicine, Seoul, Korea[2]

[Background]

Children with frequent relapsing nephrotic syndrome were treated initially with cyclophosphamide. Cyclosporine was usually used in the frequent relapser after cyclophosphamide therapy. Cyclosporine-induced remission is not long-lasting and most patients relapse within a few months following cessation of treatment. Thus, cyclosporine may have to be administered for long periods of time, exposing to the risks of nephrotoxicity. Renal biopsy is indicated to detect the cyclosporine nephrotoxicity after long-term treatment of cyclosporine. IgA nephropathy shows variable rate of progression to chronic renal failure. A high proportion of glomeruli showing sclerosis, crescents, or capsular adhesion, the presence of severe tubulointerstitial changes on renal biopsy are associated with a poor outcome. We report a case of frequent relapsing nephrotic syndrome combined with IgA nephropathy that had about 50% of diffuse global glomerular sclerosis on second renal biopsy without progression to renal failure.

[Clinical Case]

An 8-year-old boy was admitted for follow-up renal biopsy of frequent relapsing nephrotic syndrome combined with IgA nephropathy. Five years ago, renal biopsy was performed due to steroid resistant nephrotic syndrome and revealed mild mesangial expansion with mesangial electron dense deposits, one globally sclerotic glomerulus and diffuse mesangial deposit of IgA. Cyclosporine was administered and it was effective. He had been managed with cyclosporine for 1 year. He was relapsed after cessation of cyclosporine, and he changed to be steroid dependent. Cyclophosphamide had been administered for 3 months. Cyclosporine has been administered again because relapse was frequent after cyclophosphamide therapy since 4 years ago. Renal biopsy was performed to investigate cyclosporine nephrotoxicity at 1 year ago. Renal biopsy revealed chronic advanced glomerulopathy with diffuse global glomerular

sclerosis (24/46), focal fibrocellular crescent formation (5/46), and severe chronic tubulointerstitial changes and mesangial IgA deposit. But he had no symptom or any specific physical finding and CBC, chemistry and urinalysis were all normal. Even though renal biopsy findings were serious, we continued cyclosporine therapy without any additional treatment because his clinical findings and other laboratory findings were normal.

[Method]

One year later, we performed third renal biopsy at contralateral right kidney.

[Results]

In contrary to previous findings, only focal global glomerular sclerosis (1 glomeruli over total 29 glomeruli) with mild chronic tubulointerstitial changes without crescent was found. We performed Tc-99m DMSA SPECT to find out if there is focal lesion on left kidney. Cortical defects were found in lower pole of both kidneys which would be the location of biopsy.

[Conclusion]

We concluded that second biopsy findings might be a manifestation of a complication of previous biopsy such as trauma or transient arteriovenous fistula. In general, renal biopsy findings are associated with prognosis of nephrotic syndrome and IgA nephropathy. We should be cautious in the interpretation of second renal biopsy findings with diffuse global sclerosis localized in renal subcapsular area in cluster in patients with glomerulonephritis with good clinical features.