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Successful cyclosporin a therapy for diffuse mesangial sclerosis associated with WT1 mutations

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Wilms' tumor suppressor gene 1 (WT1) mutations are found in Denys-Drash syndrome, Frasier syndrome and isolated diffuse mesangial sclerosis; these mutations lead to the occurrence of diffuse mesangial sclerosis (DMS) and focal segmental glomerulosclerosis. Nephrotic syndrome (NS) caused due to DMS is unresponsive to drug therapy and is characterized by rapid progression to end-stage renal disease. Here, we report a case of a 3 years and 5 months old girl with NS caused due to DMS who responded favorably to cyclosporin A (CsA) and angiotensin-I converting enzyme inhibitor (ACE-I). The light microscopic findings of the renal biopsy before CsA therapy revealed the early stage of DMS, which showed small glomerulus with diffuse mesangial matrix increase and mesangial hypercellularity and hyperplastic podocytes. However, prominent epithelial proliferation was not found in the specimen. CsA therapy induced a dose-dependent decrease in her urinary protein/creatinine ratio and resulted in partial remission of NS and maintenance of normal renal function for over 3 years. The second biopsy at 3 years old revealed the improvement on the light microscopic findings. CsA may be effective for DMS with WT1 mutations, if therapy is started before creatinine levels increase and in the early stage of DMS. In children with WT1 mutation, CsA therapy may prevent prompt progression to end-stage renal disease.

Biography

Koji Nagatani has completed his graduation from Hamamatsu University, School of Medicine, Shizuoka, Japan in 1998 and belonged to Ehime University Graduate School of Medicine, Department of Pediatrics. He is a Member of The Japanese Society for Pediatric Nephrology, Japanese Society of Nephrology, International Pediatric Nephrology Association and International Society of Nephrology. He is the Director of Department of Pediatrics, Uwajima City Hospital, Japan

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