KSN 2016 Abstract Submission

Clinical Nephrology

KSN2016ABS-1188

Spectrum of renal manifestations in pediatric common variable immunodeficiency

Heeyeon Cho*, Yeonhee Lee

Background: Common variable immunodeficiency (CVID) is a primary immunodeficiency characterized by deficiency of antibody production with unknown etiology. The clinical manifestations include recurrent bacterial infections associated with decreased serum immunoglobulin levels, and widespread involvement of other organ systems. Renal involvement in CVID has been known to be rare, and present with renomegaly, renal granuloma, and renal insufficiency. Additionally, the patients are exposed to the prolonged use of intravenous immunoglobulin, the frequent use of antifungal or antibiotics, and the genitourinary tract infection.

Methods: We describe 3 CVID patients with various renal manifestations.

Results: Case 1: She visited the emergency department because of seizure, and was diagnosed as having hepatosplenomegaly, cytomegalovirus infection, and chronic kidney disease (CKD) stage 4 at the age of 12 years. There was no evidence of glomerulonephritis, and the findings of ultrasonography were compatible with CKD. During follow-up, she was diagnosed as having CVID, and treated with intravenous immunoglobulin. She progressed to end-stage renal disease and peritoneal dialysis was started at the age of 14 years. She was supposed to have renal involvement of CVID. Case 2: She suffered from Langerhans cell histiocytosis, ulcerative colitis, recurrent pneumonia, acute pyelonephritis, sialadenitis, and thrombocytopenia from the age of 3 months, and was diagnosed as having CVID and CKD at the age of 24 years. The laboratory findings showed CKD stage 3 and tubulopathy requiring the supplementation of potassium. The initial ultrasonography showed multiple nodular lesions in both kidneys and small right kidney. During follow-up, she received the immunoglobulin and oral corticosteroids. The nodular lesions disappeared and size-discrepancy persisted. Renal function was stationary and renal biopsy was not performed. There is a possibility of renal granuloma and scarring. Case 3: She was diagnosed with CVID at the age of 5 years, and treated with intravenous immunoglobulin. During follow-up, she suffered from recurrent pneumonia and esophageal varix associated with portal hypertension. At the age of 17 years, she complained of polyuria and weight loss, and the laboratory findings were compatible with intestinal lymphangiectasia and tubulopathy requiring the supplementation of potassium, phosphorus, and magnesium. The ultrasonography showed normal size and echogenicity of kidneys. The glomerular function is normal and there is no albuminuria.

Conclusion: In the cases of CVID with multiple organs involvement, the clinicians should concern for the renal insufficiency associated with renal involvement, and the regular evaluation including tubular function, image work-up for kidney, and renal biopsy for the patients with suspicious granuloma is necessary.

Keywords: common variable immunodeficiency, renal insufficiency