KSN 2020
FULLY VIRTUAL MEETING

**Abstract Type : Poster**
**Abstract Submission No. : PO-1170**

# Nephrotic syndrome in a boy with methylmalonic acidemia

**JIYEON SONG**, Seong Heon Kim
Department of Pediatrics-Nephrology, Pusan National University Yangsan Hospital, Korea, Republic of

**Case Study:**

Background

Methylmalonic acidemias (MMA) are a group of metabolic disorders of diverse etiology characterized by impaired conversion of methylmalonyl CoA into succinyl-CoA. There has been a paucity of nephropathological studies of MMA-associated kidney disease, and the published renal biopsies all show a predominantly tubulointerstitial process that is not reportedly distinctive. We report a case of nephrotic syndrome in a patient with MMA.

Case

A 30 month old boy visited our clinic for fever, diarrhea, and decreased oral intake for 3days. He diagnosed with MMA. His vital signs was blood pressure of 92/60 mmHg, heart rate of 120/bpm, respiratory rate of 22/min, body temperature of 38.1 °C, and oxygen saturation of 96%. The initial laboratory test and urinalysis are as follows: Hb 13.6 g/dL, platelet 364K, Na+ 131 mmol/L, K+ 4.4 mmol/L, BUN 9.1 mg/dL, Creatinine 0.21 mg/dL, Albumin <1.5 g/dL, cholesterol 296 mg/dL, Urine protein 4+, RBC 0-2 /HPF, urine protein/creatinine ratio > 3.0. We planned the renal biopsy because MMA also can cause renal problem and differential diagnosis from minimal change nephrotic syndrome is necessary. In pathologic findings, there were mild and focal changes of tubular atrophy, fibrosis, and subsequent electron microscopy revealed diffuse effacement of foot process referring minimal change disease (MCD).
After biopsy, we started oral steroid (2mg/kg) for treatment of NS, complete remission was achieved after 4 weeks, and steroid was tapered and stopped. He relapsed about 3 months after discontinuation of medication, restarted oral steroid, and steroid was tapered and discontinued after steroid therapy because he had remission. During follow-up, he currently maintained complete remission without proteinuria.

Conclusion

This case is the first report that MMA patient was diagnosed by primary NS. However, the association is not clear yet.