

## KSN 2017 Abstract

KSN-17-P268

### Focal segmental glomerulosclerosis associated with polycythemia progressed to chronic renal failure in 16-year-old boy with severe obesity

Kyung-mi JANG, \*Young-hoon PARK

Pediatric nephrology, Youngnam University Hospital, Korea, South

**Introduction:** Focal segmental glomerulosclerosis (FSGS) is divided into primary and secondary FSGS, depending on etiology. Secondary FSGS may be the result of hypertrophy and hyperfiltration developing to compensate for various causes such as polycythemia. There are few cases of FSGS secondary to polycythemia event in adult. To our knowledge, there is no case report yet in children. Now, we report this extremely rare case of FSGS secondary to polycythemia despite CKD in 16-year-old boy with severe obesity.

**Case:** Healthy looking obese boy (BMI 32.7 kg/m<sup>2</sup>) was referred to our hospital for the evaluation of proteinuria and hypertension (147/88 mmHg) on school urinary screening test. Laboratory investigations on admission were as follows: serum total protein 6.8 g/dL, serum albumin 4.3 g/dL; BUN 17.9 mg/dL, serum creatinine 2.48 mg/dL; uric acid 10.0 mg/dL; serum total cholesterol 250 mg/dL, LDL 107 mg/dL, triglycerides 522 mg/dL, HDL 36 mg/dL; aspartate aminotransferase 48 U/L, alanine aminotransferase 71 U/L. The 24-hour urine study showed heavy proteinuria (127.5 mg/m<sup>2</sup>/hr) and decreased GFR (43.8 mL/min/1.73 m<sup>2</sup>). Serum anti-neutrophil cytoplasmic antibodies were negative. Full blood count revealed hemoglobin 18.9 g/dl, hematocrit 56%, red blood cells 6.48 x 10<sup>6</sup>/mm<sup>3</sup>, WBC 5,980/mm<sup>3</sup> with a normal differential count, platelet 211,000/mm<sup>3</sup> and erythropoietin level of 17 mIU/mL (reference range, 5–20). Polycythemia was noted instead of anemia despite CKD. Bone marrow aspiration and biopsy showed normal nucleated hematopoietic cells distribution. There were no symptoms of cardiac, cerebral or pulmonary abnormalities. A renal biopsy was performed. Light microscopy showed FSGS with glomerular enlargement. No immune accumulation was determined on IF staining. Ultrastructural examination revealed diffuse loss of foot process without electron-dense deposits. Negative findings of gene studies (JAK 2, MPL, CALR mutation) for polycythemia vera (PV) suggested that there is no relevance to the PV. While we are trying to search for cause of polycythemia, his father was founded to have polycythemia, proteinuria and microscopic hematuria on the recent health examination. Patient and his parents' genetic test for PFCP (primary familial and congenital polycythemia) is pending. We thought that his obesity also affect the progress of the FSGS secondary to polycythemia to CKD.

## **KSN 2017 Abstract**

The patient was treated by repeated phlebotomies and ACE inhibitor, and HMG-CoA reductase inhibitor with dietary management and physical exercise. Aspirin was added for his high viscosity. After 6 months of treatment, BP, cholesterol, aspartate aminotransferase, alanine aminotransferase became normal. However, his LDL, triglyceride and HDL did not recover normal reference range. The amount of 24-hour urine protein (88 mg/m<sup>2</sup>/hr) and BMI (29.8 kg/m<sup>2</sup>) were partially controlled. Meanwhile, GFR (50 mL/min/1.73 m<sup>2</sup>) did not deteriorate any longer.

**Conclusion:** FSGS secondary to polycythemia discovered by school urine screening test was progressed to CKD in a patient with severe obesity. Phlebotomy for lowering the hematocrit, and ACEI, HMG-CoA reductase inhibitor, reducing body weight appeared clinically beneficial to the course of renal disease in FSGS secondary to polycythemia. Long-term observation on phlebotomy and drug treatment effects on progressing CKD in FSGS secondary to polycythemia will be required.

**Keywords :**