Case #2 A 9 years old girl of heavy proteinuria with chronic kidney disease by school urine screening. What is your differential diagnosis?

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Case presentation:

A 9 years old girl visited to out patient clinic due to heavy proteinuria in school urine screening. Her body weight was 35kg, body length was 125cm, blood pressure was 120/72mmHg. Physical findings were almost normal. Hemogram showed Hb 12 gm%, WBC:9240/cumm, platelete: 220000/cumm. Serum albumin was 4.2 mg/dl, BUN: 34 mg/dl,Cr: 2.1 mg/dl. 24hours urine protein was 2250mg. Renal sonogram showed only increasing ecchogenicity of renal parenchyma. She received short course of methylprednisolone pulse therapy and in vain. Then she received renal biopsy.

Answer. oligomeganephronia

Pathological findings:

(1)microscopically, the section of renal biopsy contains two completely obsolescent and another 5 non-obolescent but moderately enlarged glomeli revealing significant increased in glomeular size and component cells with increased capillary lumina mesangial cellularity, two loci of focal segmental glomerulosclerosis, and focal thickening of focal thickening of Bowmen's capsule. The nember of glomeruli was decreared $(0.91/\text{mm}^2)$ and diameter $(410~\mu\text{ m})$ of glomeruli was increased. The tubulointerstitial compartment shows evident tubular luminal dilatation and/or enlargement, frequent hyaline cast formation evident, foamy change of promximal tubular epithelium, patchy tubular atrophy (up to 20% in area), focal interstitial fibrosis.

- (2) The PAS and CSM stains delinetate foci of focal segmental glomerulsclerosis, significant mesangial hypercellulaerity, and enlarged glomerular with hypercellulaerity without increase in mesangial matrix.
- (3) Electron microscopic study: There are no electron dense deposit found. The glomerular basement membrane (GBM) show no remarkable change, no lamination or splitting change of the lamina densa present. There are partial foot processes effacement of the podocytes seen.
- (4) Immunofluorescence stain was negative finding.

According to the above features, secondary focal segmental glomerulosclerosis (2/5) in the background of oligomeganephroniac hypoplasia (oligomeganephronia) with patchy tubular atrophy (20% in area) and mild interstitial fibrosis is firstly considered.

Discussion:

The pathological features, consisting of a reduced number of enlarged glomeruli, were diagnostic of oligomeganephronia.