Glomerulocystic Kidney in a Patient with Nephrotic Syndrome

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Dear Sir,

Glomerulocystic kidney (GCK), which is characterized by cystic dilatation of Bowman's capsule, has been described in infants and children. However, recently GCK has been reported in an adult patient with renal failure [1] and in a patient with progressive systemic sclerosis [2].

Thus now, GCK is regarded not to be confined to the pediatric population.

We describe herein a case of GCK, observed in a 69-year-old male patient. He was referred to our hospital because of nephrotic syndrome. There was no previous history of renal disease. The BUN was 19 mg/dl, serum creatinine 1.7 mg/dl, albumin 2.8 g/dl. A urinalysis revealed 4+ proteinuria without occult blood. Bence-Jones protein in the urine was negative. Creatinine clearance was 40 ml/min/1.73 m². Computed tomography showed one cyst of 1.5–2.0 cm in diameter in the left kidney. There was no evidence of urinary tract obstruction. Cystic lesions were not seen in other organs. A right kidney needle biopsy was done. Figure 1 shows the glomerular cysts that are exclusively localized near the cortical surface. These cysts were egg-shaped, and slightly larger than a normal glomerulus in diameter. Epithelial cells on Bowman's capsule were flattened and proteinaceous material was seen within the space. Most of the glomerular tufts were obviously collapsed. There was remarkable interstitial fibrosis in the cortex, but not in the medulla. No cystic changes involving the tubules were found. Non-cystic glomeruli showed mild mesangial proliferation associated with expansion of mesangial matrix. Immunohistochemistry of the bioptic specimen showed linear staining of IgA, IgG and IgM on the endothelial surface of the noncystic as well as cystic glomeruli (fig. 2). Of particular interest, the tubular epithelium was diffusely positive for IgG and dispersely for C3 (fig. 3).

The pathogenesis of GCK and the mechanism leading to the formation of cysts remain unclear.

Though urinary tract obstruction may underlie the development of GCK

Fig. 1. Scattered glomerular cysts in the renal cortex. × 50. PAS.

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Fig. 2. Endothelial staining of IgG by immunohistochemistry in the noncystic glomerulus. x 100.
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matosus. Some members of her family showed GCK-like cortical cysts, suggesting autosomal dominant inheritance.

In the present case, it is difficult to explain the mechanism of development of GCK. However, the immunohistochemical findings may suggest the participation of an immunological insult in the build-up of GCK.

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Fig. 3. Dispersed C3 deposition in the tubular epithelium. × 50.

in infants [3], the documented cases of adult GCK [1, 2], including our case, were not associated with such lesions. In a similar vein, the absence of other anomalies that are usually present in infants strongly speaks against the congenital origin in adult GCK. More recently, Carsen et al. [4] reported an adult GCK in a 20-year-old woman with nephrotic syndrome due to systemic lupus erythe-

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