Glomerulocystic Kidney in a Patient Affected with Progressive Systemic Sclerosis

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Dear Sir,

We were very much interested in the report by Oh et al. [1] concerning a glomerulocystic kidney (GCK) in an adult patient. GCK is mainly observed in infants and young children, and congenital malformations are very often present. Therefore, maldevelopment is accepted as a cause of GCK. We report herein a case of GCK which has developed in a 60-year-old male patient affected with progressive systemic sclerosis. We performed a left kidney needle biopsy in this patient when he was 56 years old, because of proteinuria accompanied by microscopic hematuria. We found mesangial proliferative glomerulonephritis but no GCK in the renal cortex. The kidney was removed 4 years after the percutaneous biopsy, because of persistent gross hematuria due to infarction of the kidney: we found GCK disease. Figures 1 and 2 show that the cysts were egg-shaped, 3–4 times larger than a normal glomerulus in diameter and consisted of dilated Bowman's spaces and collapsed glomerular tufts (fig. 1) with immuno-fluorescent filaments of IgG and albumin in the urinary space (fig. 2). This appearance is not compatible with ischemic glomerular lesions [2].

It is not clear which mechanism(s) are responsible for the cystic dilatation of the glomerulus and by which mechanism(s) glomerular plasma filtration can be maintained in the collapsed tuft, causing the presence of preurine in the glomerular cysts.

The progressive dilatation of Bowman's space might indicate an alteration in the collagen component of Bow-

Fig. 1. Scattered glomerular cysts in the renal cortex. PAS. $\times 120$.

man's capsule with a concomitant slow increase in the intracapsular glomerular pressure; the hydrostatic filtration pressure might be maintained, together with a slowed deflow of preurine at the glomerular urinary pole.

Although it is difficult to determine the origin of the large GCK, the case we discuss shows that GCK may not only be a congenital but also an acquired lesion.

In progressive systemic sclerosis, renal involvement consists of arterial intimal thickening and interstitial fibrosis with development of severe hypertension [3]; such features were present in

our patient. A generalized disorder of connective tissue with fibrosis and degenerative changes in the kidney might be a cause of GCK disease.

174

Sessa/Giordano/Meroni/Battini/Torri-Tarelli/Volpi

Fig. 2. Immunofluorescence microscopy. Filaments of IgG in the urinary space. $\times 250$.

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