

Focal segmental glomerulosclerosis in atypical polycystic kidney disease

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A 77-year-old man presented complaining of generalized edema for 1 week. He had hypertension and polycystic kidney disease (PKD), which was diagnosed 45 years ago. Blood and urine tests showed low serum albumin (2.3 g/dL) and severe proteinuria (6.38 g/day). Abdominal computed tomography revealed multiple cysts with thin wall calcification in the left kidney, and several cysts in the right kidney and the liver (Fig. 1). Ultrasonography-guided percutaneous renal biopsy for the right kidney revealed focal segmental glomerulosclerosis (Fig. 2). The patient was prescribed diuretics, an angiotensin receptor blocker, aspirin, statin, a β blocker, and an aldosterone antagonist. One month after starting the medicines, he showed improved symptoms and laboratory findings (se-

rum albumin, 3.2 g/dL; urine albumin excretion, 654.8 mg/g creatinine), which further improved after another 3 months (serum albumin, 3.8 g/dL; urine albumin excretion, 128.4 mg/g creatinine).

Nephrotic syndrome is a rare coincidence in atypical PKD. Percutaneous renal biopsy can be performed in selected patients.

Conflict of interest

No potential conflict of interest relevant to this article was reported.

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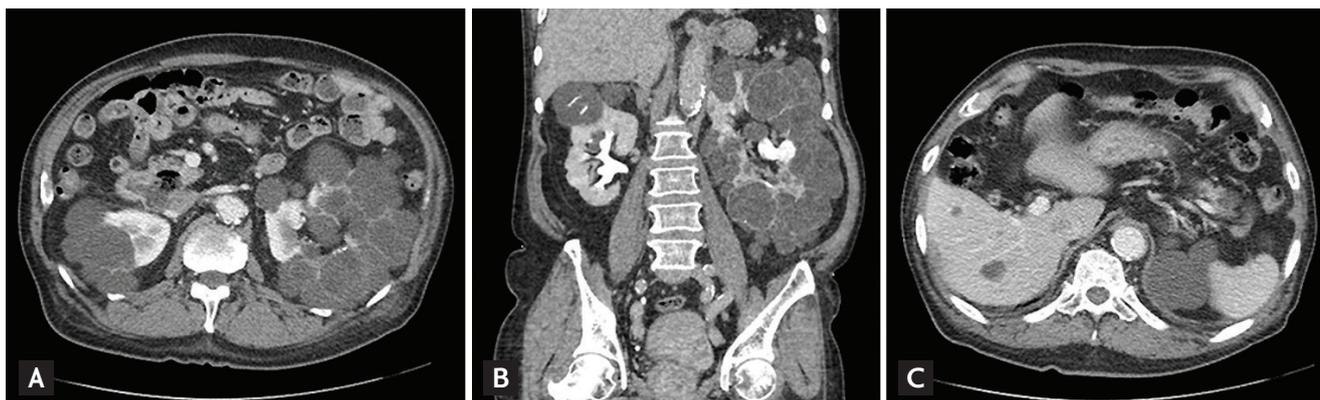


Figure 1. Abdominal computed tomography shows multiple cysts in the (A, B) left and right kidney and (C) several cysts in the liver.

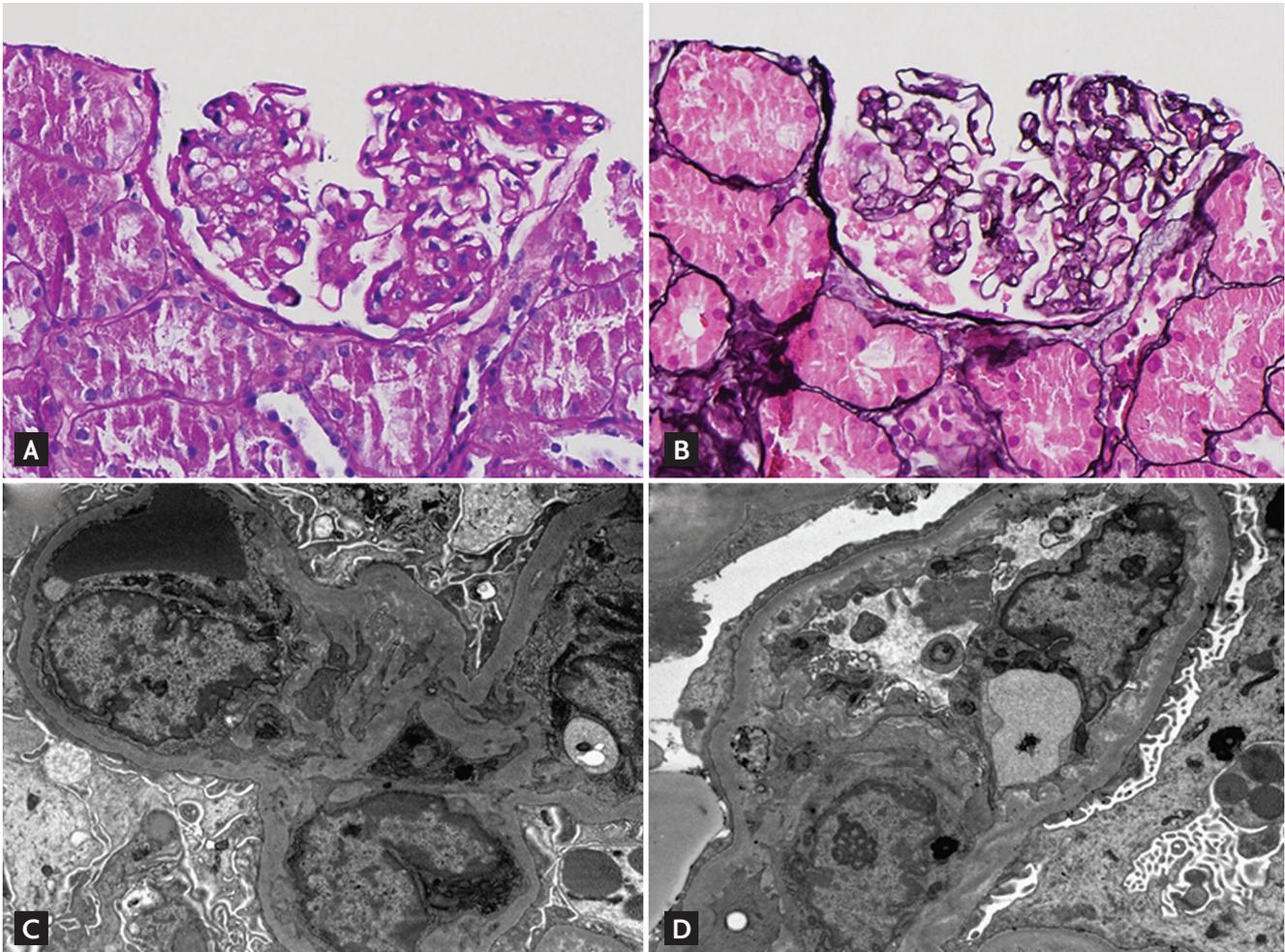


Figure 2. (A, B) Microscopic findings show endocapillary foam cells and extracapillary cytoplasmic hyaline droplets (A, periodic acid-Schiff stain, $\times 400$; B, Jones methenamine silver stain, $\times 400$). (C, D) Electron microscopy reveals swollen podocytes and diffuse effacement of the foot processes.