NDT Plus (2011) 4: 73–74
doi: 10.1093/ndtplus/sfq189
Advance Access publication 2 November 2010

NDT PLUS
Nephrology Dialysis Transplantation

Images in Nephrology
(Section Editor: G. H. Neild)

Non-hereditary multiple renal cysts in unilateral kidney

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Keywords: adult; computed tomography; renal scintigraphy; unilateral renal cystic disease

A 52-year-old male, who noticed a sudden onset of general fatigue and appetite loss, visited our hospital. No other family member had a history of renal disease. He had been diagnosed with alcoholic hepatopathy 10 years before. His blood pressure was 112/72 mmHg. His right kidney was felt during palpation at the upper right abdominal area. Laboratory findings showed elevated levels of liver enzymes, but renal function was normal (serum creatinine 1.01 mg/dL). On urinalysis, occult blood and urine protein were negative. Abdominal computed tomography revealed multiple large cysts in the right kidney, a normal left kidney, and no evidence of further cystic disease in other organs (Figure 1). A diffuse low-density area in his liver was interpreted as fatty liver due to alcoholic hepatopathy. There was no evidence of urinary tract malformation. ^{99m}Tc-DTPA renal scintigraphy showed asymmetrical accumulation, and the right kidney was non-functioning on scintigraphy (Figure 2).

Unilateral renal cystic disease (URCD) is a rare and poorly understood condition, first described in 1979 [1]. Only 55 cases have been reported [2,3]. Except for its unilateral localization, it resembles autosomal dominant polycystic kidney disease (ADPKD) on gross and histological examination [4]. However, unlike ADPKD, URCD patients show neither a genetic background nor progressive deterioration in renal function. To diagnose URCD, characteristic CT findings are required in addition to genetic and clinical findings. URCD is characterized by cysts of varying sizes localizing in a diffusely enlarged kidney but not forming a distinct encapsulated mass, and the absence of intervening normal renal parenchyma between the cysts. In the present case, unilateral localization, a negative family history, no progression to chronic renal failure, and the absence of cysts or malformations in other organs were taken to confirm the diagnosis of URCD in our adult patient. In URCD, the contralateral unaffected kidney in adult patients may occasionally show a few simple cysts as has been documented [2], and the evolution of unilateral disease into bilateral disease has been reported [5]. Moreover, it is possible that development of complicated cysts due to rupture or infection of cysts, occurrence of malignancy, or growth of a kidney may appear. Therefore, our patient required long-term follow-up observation.

Conflict of interest statement. None declared.

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Received for publication: 9.9.10; Accepted in revised form: 13.10.10

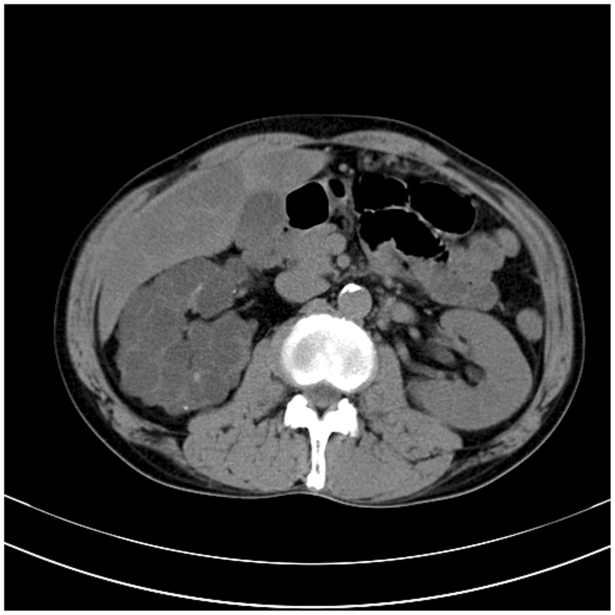

Fig. 1. An abdominal computed tomography scan demonstrated multicystic involvements of the right kidney, which measured 126 × 88 × 84 mm and extended into the pelvis. No further cystic involvement of the contralateral kidney or other abdominal organs was detected.

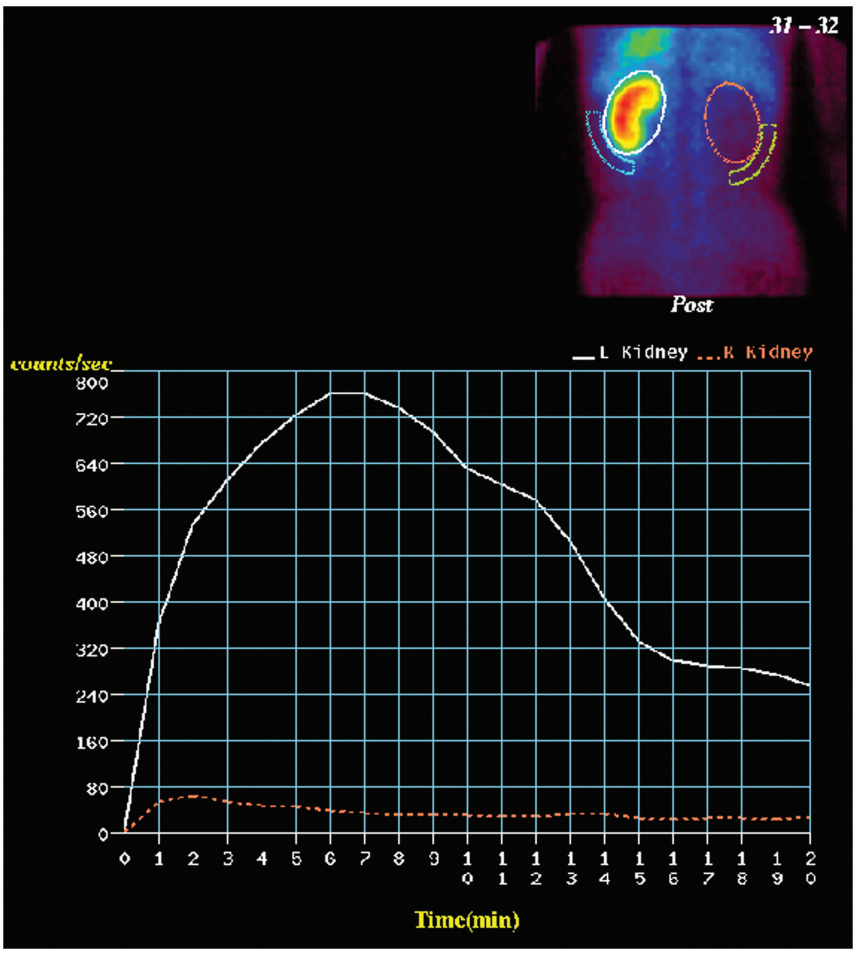

Fig. 2. ^{99m}Tc-DTPA renal scintigraphy showed asymmetrical accumulation, and the estimated glomerular filtration rates determined by scintillation count were decreased in the right kidney (right 5.2 mL/min; left 64.8 mL/min). The right kidney was non-functioning on scintigraphy.