A case of unilateral nephrectomy performed for autosomal dominant polycystic kidney disease with marked unilateral enlargement

Shiho Makabe¹ · Hiroshi Kataoka^{1,2} · Tsunenori Kondo³ · Kazunari Tanabe⁴ · Ken Tsuchiya⁵ · Kosaku Nitta¹ · Toshio Mochizuki^{1,2}

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Abstract

Autosomal dominant polycystic kidney disease (ADPKD) is characterized by the occurrence of multiple cysts that increase the size of both kidneys, progressively reducing kidney function. Usually the cysts occur bilaterally, and there is no difference in the degree of cyst enlargement between the left and right. Here, we report a case of ADPKD in which kidney size increased markedly on the left side and was accompanied by severe abdominal distension and discomfort. Renal dynamic scintigraphy revealed a severe reduction in function of the left kidney compared with the right. Open left nephrectomy was performed. No change in renal function was observed postoperatively [preoperative estimated glomerular filtration rate (eGFR): 57.6 mL/min/1.73 m², 3-month postoperative eGFR: 56.4 mL/min/1.73 m²], and the abdominal symptoms subsided. When one kidney is markedly larger than the other, the cause and status of the laterality should be evaluated by using renal dynamic scintigraphy in addition to other examinations such as computed tomography or magnetic resonance imaging. Unilateral nephrectomy should be considered as a potential treatment.

Keywords Autosomal dominant polycystic kidney disease · ADPKD · Unilateral nephrectomy · Renal dynamic scintigraphy

Introduction

Autosomal dominant polycystic kidney disease (ADPKD) is the most common hereditary kidney disease; it is characterized by multiple renal cysts on both sides, increasing kidney size, and progressive decline in renal function [1]. *PKD1* and *PKD2* have been identified as the causal genes. Cysts develop when one of these genes does not function in renal tubular cells. Usually the cysts occur bilaterally, and there is no difference in the degree of cyst enlargement between the left and right. Here, we report a case of unilateral nephrectomy performed for ADPKD with markedly enlargement on one side.

Case report

The patient was a 28-year-old female with a chief complaint of left abdominal distention and discomfort. Her father had begun dialysis from age 50 years because of ADPKD. Gross hematuria had been observed in the woman in 2004, at age 19 years, and PKD was diagnosed. Her first visit to our

✉ Toshio Mochizuki
mtoshi@twmu.ac.jp

¹ Department of Medicine, Kidney Center, Tokyo Women's Medical University, 8-1 Kawada-cho, Shinjuku-ku, Tokyo 162-8666, Japan

² Clinical Research Division for Polycystic Kidney Disease, Department of Medicine, Kidney Center, Tokyo Women's Medical University, 8-1 Kawada-cho, Shinjuku-ku, Tokyo 162-8666, Japan

³ Department of Urology, Tokyo Women's Medical University Medical Center East, 2-1-10 Nishiogu, Arakawa-ku, Tokyo 116-8567, Japan

⁴ Department of Urology, Kidney Center, Tokyo Women's Medical University, 8-1 Kawada-cho, Shinjuku-ku, Tokyo 162-8666, Japan

⁵ Department of Blood Purification, Kidney Center, Tokyo Women's Medical University, 8-1 Kawada-cho, Shinjuku-ku, Tokyo 162-8666, Japan

hospital was in December 2008, at age 23 years. Renal function was normal (blood urea nitrogen 11.6 mg/dL, serum creatinine 0.75 mg/dL, estimated glomerular filtration rate (eGFR) 79.9 mL/min/1.73 m²), and her urinalysis findings were also normal. Plain computed tomography (CT) in December 2008 revealed a right kidney volume (RKV) of 361 cm³, left kidney volume (LKV) 2152 cm³, total kidney volume (TKV) 2513 cm³, and height-adjusted TKV (htTKV) 1621 cm³ (Fig. 1). All kidney volumes were measured using the 3D workstation of Ziostation2 Version 2.4.2.3.

Gradual abdominal distention and increased distress caused by the left kidney enlargement began in about October 2009. Pain when in the upright position began to accompany the abdominal distention in 2012. Magnetic resonance imaging (MRI) in June 2012 yielded the following findings: RKV 472 cm³, LKV 2707 cm³, TKV 3178 cm³, and htTKV 2050 cm³. The abdominal circumference had increased by 10 cm during the 5 years since 2008.

Renal dynamic scintigraphy (with ^{99m}Tc-MAG3; technetium-99m mercaptoacetyltriglycine-3) was performed in January 2013, when the patient was 28 years of age. MAG3 capture was 15.4% in the left kidney and was markedly reduced compared with the right (84.6%). Because the effective renal plasma flow (ERPF) in the left kidney was 46.7 mL/min/m², compared with 256.2 mL/min/m² in the right kidney, and the function of the left kidney was evaluated as severely reduced (renogram index $L/R = 0.1882$) (Fig. 2), we predicted that the decrease in renal function that would result from excision of the left kidney would be minor. Abdominal contrast-enhanced CT was performed in August 2013 and revealed that the left kidney was markedly larger than the right. In the patient's lower abdomen, the left kidney extended beyond the midline of the body to the right, and this had led to distressing symptoms (Fig. 3). Because of the patient's distress due to the abdominal distention, unilateral left nephrectomy was chosen, and the patient was admitted to the urology department of our hospital in

Fig. 2 Renographic curves from renal imaging using ^{99m}Tc-MAG3. MAG3 capture and effective renal plasma flow were markedly lower in the left kidney than in the right, and the function of the left kidney was evaluated as severely reduced. KCPM kilo-counts per minute

October 2013. MRI in September 2013 showed the following: RKV 657 cm³, LKV 3444 cm³, TKV 4101 cm³, and htTKV 2646 cm³. The annual rate of growth was: LKV 21.8%/year, RKV 31.4%/year, and TKV 23.2%/year (Fig. 4).

On admission, blood urea nitrogen was 15.4 mg/dL, serum creatinine was 0.96 mg/dL, eGFR was 57.6 mL/min/1.73 m², and other blood analysis and urinalysis were within normal limits. Open left unilateral nephrectomy was performed in the urology department. The excised kidney weighed 2600 g. Pathology examination revealed multiple cysts of various sizes in the kidney parenchyma and glomerular reduction and renal tubule expansion in the remaining parenchyma. On the basis of these findings, polycystic kidney disease (PKD) was diagnosed. The postoperative course was uncomplicated, and the patient was discharged on postoperative day 5. No further deterioration in her renal

Fig. 1 Plain computed tomography (CT) in December 2008 revealed a right kidney volume (RKV) of 361 cm³, left kidney volume (LKV) 2152 cm³. LKV was approximately six times RKV

Fig. 3 a–c Results of abdominal contrast-enhanced computed tomography in August 2013. The left kidney was markedly larger than the right. The cortex of the right kidney was enhanced by contrast medium, but the cortex of the left kidney was barely enhanced

function was observed after left nephrectomy (preoperative eGFR: 57.6 mL/min/1.73 m², 3-month postoperative eGFR: 56.4 mL/min/1.73 m²) (Fig. 5). RKV was 794 cm³ in September 2015, and the annual rate of growth in RKV was 10.4%/year, which was not faster than that before unilateral nephrectomy.

Discussion

Here, we report a case of ADPKD with marked enlargement of the left kidney, accompanied by severe abdominal distension and distress. Renal dynamic scintigraphy by renogram revealed severely reduced function of the left kidney compared with the right. Open left nephrectomy was performed. No further deterioration in renal function was observed 3 months postoperatively, and the patient's abdominal symptoms subsided. The reasons for performing the unilateral nephrectomy in this case were: (1) the substantial difference in size between the left and right kidneys; (2) the severe decrease in function of the left kidney according to MAG3

scintigraphy; and (3) the patient's request for treatment, even by nephrectomy.

Renal transcatheter arterial embolization (TAE) also can reduce kidney volume, and many ADPKD patients undergoing hemodialysis have received TAE treatment. Suwabe et al. reported renal volume reduction rate by TAE treatment ranged from 3.9 to 84.8% [2]. She strongly complained to remove the left kidney completely because of distress. Furthermore, unilateral renal TAE often cause the other kidney bleeding. In this case, right renal function should be protected. Taken together, we decided to choose nephrectomy not renal TAE.

Usually, there is no marked difference in size between the left and right kidneys in ADPKD. However, in our patient, LKV was approximately six times RKV at the initial examination in 2008 (Fig. 1); therefore, we consider this case to be unique. Irazabal et al. [3] radiologically classified ADPKD patients as typical (Class 1) or atypical (Class 2). Class 2 accounted for only 8.8% of patients and was divided into Class 2A and Class 2B. Class 2A included unilateral, segmental, asymmetric, or bilateral atypical presentations, and Class 2B included acquired polycystic kidney with unilateral

Fig. 4 Changes in kidney volume. Magnetic resonance imaging (MRI) examination in June 2012 showed right kidney volume (RKV) 472 cm³, left kidney volume (LKV) 2707 cm³, and total kidney volume (TKV) 3178 cm³. MRI examination in September 2013 showed RKV 657 cm³, LKV 3444 cm³, and TKV 4101 cm³. The rate of change was 21.8%/year for LKV, 31.4%/year for RKV, and 23.2%/year for TKV

or bilateral atrophy. Our patient's imaging findings could not be classified into any of these.

Unilateral polycystic kidney disease has been reported as a different disease from ADPKD. In 1989, Levine and Huntrakoon [4] named them unilateral renal cystic disease (URCD) in adults, and it has been known by a variety of different names, such as localized cystic disease of the kidney

(LCDK) [5]. Although some cases of URCD/LCDK have been reported, the mechanism of unilateral cystogenesis has not been elucidated [6, 7]. Because multiple cysts are present on only one side in URCD/LCDK, no progressive deterioration in renal function is observed. Generally, the differential diagnosis between URCD/LCDK and ADPKD is not difficult, because in URCD/LCDK patients there is no genetic background and no cysts are present in the unaffected kidney or in other intra-abdominal organs. However, careful follow-up in young patients with URCD/LCDK is important because of the possibility of asynchronous ADPKD or malignancy. Our patient had a family history of ADPKD, and multiple cysts were observed in both kidneys, thereby clearly differentiating her condition from URCD/LCDK.

Substantial differences in size between the left and right kidneys in ADPKD have been reported before. Poster et al. [8] reported three cases of unilateral ADPKD among 182 ADPKD cases. A *PKD1* gene mutation was observed in all three cases, but two cases were considered to have been caused by contralateral renal agenesis, and the third by severe contralateral renal hypoplasia. Xu et al. [9] reported two cases of unilateral ADPKD caused by ectopic unilateral multicystic dysplastic kidney (MCDK) among 605 ADPKD cases; a *PKD1* gene mutation was observed in both cases. These are cases of unilateral ADPKD in which one kidney was non-functional because of aplasia, hypoplasia, or MCDK, and the size of the other kidney therefore increased. In contrast, in our patient, severe reduction in function of the enlarged left kidney was observed; therefore, we considered our patient's case to be different from these others.

Generally, among the causes of difference in size between the left and right kidneys, prerenal disorders such as abnormal blood flow, renal disorders such as glomerular disease, or postrenal disorders such as lower urinary tract problems

Fig. 5 Changes in estimated glomerular filtration rate (eGFR) level, demonstrating no obvious deterioration in renal function after left nephrectomy

should be considered. Before nephrectomy in our patient, image of the left urinary tract was of poor resolution in both contrast-enhanced CT and MAG3 scintigraphy. These findings suggested the possibility of blockage of the lower urinary tract on the left side. Interestingly, Galarreta et al. [10] indicated that there is a common mechanism of progressive renal disorder in unilateral ureteral obstruction (UUO) and PKD. When a cyst reaches a certain size in PKD, it separates from the renal tubule and becomes an atubular glomerulus on the nephron. In UUO, formation of an atubular glomerulus is induced as a result of oxidative stress and extensive proximal tubular impairment. It is possible that, in our patient, ADPKD was accompanied by UUO in the left kidney, accelerating disease progression.

MAG3 scintigraphy is a nuclear scanning method that measures ERPF on the basis of the renal uptake and clearance of ^{99m}Tc -MAG3; it is a noninvasive way to evaluate renal blood flow, differential renal function, urinary tract patency, and kidney shape. This imaging modality allows left and right renal function to be evaluated separately. In our patient, radioisotope uptake in the left kidney was so severely reduced that it could not be recorded. Reduced uptake in the right kidney was observed in the upper region, but both uptake and ERPF were more than five times greater in the right kidney than in the left kidney, and a marked difference between left and right renal function was observed (Fig. 2). MAG3 scintigraphy in our patient was performed in a phase when a marked difference in size between left and right was observed, but MAG3 scintigraphy and urography should preferably be performed in the early disease phase to determine the cause of the differences between left and right and to consider any association with ureteral obstruction.

In conclusion, the current case of ADPKD showed marked enlargement of the left kidney accompanied by severe abdominal distention. Renal dynamic scintigraphy revealed severely reduced function of the left kidney, and open left nephrectomy was performed. When one kidney is substantially larger than the other, the cause and status of the laterality should be evaluated by using renal dynamic scintigraphy, in addition to other examinations such as CT or MRI. Unilateral nephrectomy should be considered as a potential treatment. Although there was no marked deterioration in right kidney function in the patient 3 months after surgery, close follow-up is needed to observe the long-term results.

Compliance with ethical standards

Conflict of interest Toshio Mochizuki and Ken Tsuchiya received travel fees and honoraria for lectures from Otsuka Pharmaceutical Co.

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Ethical approval All procedures performed in the patient study were in accordance with the 1964 Helsinki Declaration and its later amendments, or with comparable ethical standards.

Informed consent Informed consent was obtained from the patient.

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