Nephrectomy in Autosomal Dominant Polycystic Kidney Disease: A Patient with Exceptionally Large, Still Functioning Kidneys

Edwin M. Spithoven^a Niek F. Casteleijn^a Paul Berger^b
Roel Goldschmeding^c

^aDepartment of Nephrology, University Medical Center Groningen, University of Groningen, Groningen, and Departments of ^bSurgery and ^cPathology, University Medical Center Utrecht, Utrecht, The Netherlands

Key Words

Autosomal dominant polycystic kidney disease · Nephrectomy · Transplantation

Abstract

Autosomal dominant polycystic kidney disease (ADPKD) is the most common hereditary kidney disease. It is characterized by progressive cyst formation in both kidneys, often leading to end-stage kidney disease. Indications for surgical removal of an ADPKD kidney include intractable pain, hematuria, infection, or exceptional enlargement and small abdominal cavity hampering implantation of a donor kidney. We report the case of an extraordinarily large ADPKD kidney weighing 8.7 kg (19.3 lb) with a maximal length of 48 cm (19 inch), and with cysts filled with both clear and bloody fluid.

© 2014 S. Karger AG, Basel

Introduction

Autosomal dominant polycystic kidney disease (ADPKD) is characterized by progressive cyst formation in the kidneys leading to massive kidney enlargement. Over time, the kidney function deteriorates as functional parenchyma is replaced by growing cysts. Renal cysts may rupture leading to hematuria and pain. A pyogenic infection may also be located in these cysts. ADPKD patients with chronic pain related to the enlarged kidneys, recurrent hematuria or therapy-resistant cyst infection may benefit from nephrectomy. In addition, in some patients, the cystic kidneys are exceptionally enlarged leaving too little abdominal

Edwin M. Spithoven
Department of Nephrology, University Medical Center Groningen
University of Groningen, PO Box 30.001
NL–9700 RB Groningen (The Netherlands)
E-Mail e.m.spithoven@umcg.nl

space to allow for kidney transplantation. Here, we report nephrectomy of an extremely enlarged ADPKD kidney prior to transplantation in a 42-year-old man.

Case Report

A 42-year-old male patient with ADPKD visited our outpatient clinic while preparing for renal replacement therapy. His medical history included hypertension and myocardial infarction, complicated with aorta dissection, classified as a Stanford type A. He had experienced multiple periods with hematuria and kidney pain, most likely caused by recurrent cyst bleedings. He inherited ADPKD from his paternal side; his father started renal replacement therapy at 42 years of age and died at 46 years due to complications during a second kidney transplantation. His 35-year-old sister also has ADPKD with a 24-hour urine creatinine clearance of 137 ml/min and a total kidney volume of 1,974 ml. Physical examination of the patient revealed a man weighing 99 kg (218 lb) and measuring 189 cm (6 feet 2.4 inch), with a body mass index of 27.7. His waist and hip circumferences were 100 and 102 cm, respectively. His residual 24-hour urine creatinine clearance had decreased to 8 ml/min, and his serum creatinine was 812 μ mol (9.2 mg/dl). Since a potential living unrelated donor allograft was found for the patient, he was evaluated to undergo kidney transplantation. Magnetic resonance imaging showed exceptionally enlarged kidneys in a small abdominal space (total kidney volume is 10,280 ml, left kidney 5,628 ml and right kidney 4,652 ml) (fig. 1). Of note, the total kidney volume was measured on T2-weighted coronal images using the commercially available software Analyze Direct 8.0 (Analyze Direct, Inc., Overland Park, Kans., USA). Intra- and interreviewer coefficients of variation were 2.4 and 3.1%, respectively [1]. In addition, several small cysts were found in the liver. A left-sided nephrectomy was performed to allow for kidney transplantation. Postoperatively, no complications were reported and he was discharged from the hospital in a good condition 7 days later. The removed kidney weighed 8.7 kg (19.3 lb) and had a maximal length of 48 cm (19 inch) (fig. 2). Subsequent dissection of the removed kidney (online suppl. video 1; for all online suppl. material, see www.karger.com/doi/10.1159/000363378) revealed almost complete replacement of cortical as well as medullary kidney tissue by numerous cysts. Some of these cysts were filled with clear pre-urine, others with hemorrhagic fluid and again others with a chocolate-like substance. No pathological signs of (pre-)malignancy were found. After the nephrectomy, the patient was on temporary hemodialysis until he received a living unrelated renal allograft 3 months later. One week after the transplantation, he experienced an episode of vascular allograft rejection type IIA, which was successfully treated with antithymocyte globulin and plasmapheresis. Five months after the transplantation, he was doing well with mildly reduced but stable kidney function [creatinine level 124 μ mol/l (1.44 mg/dl)].

Discussion

ADPKD is the most common hereditary kidney disease. Most affected subjects have progressive renal function decline and need renal replacement therapy between their 40th and 70th year of age [2]. Nephrectomy before transplantation happens relatively often in ADPKD patients. However, this case is extraordinary because of the massive kidney enlargement. The maximal length of the removed kidney was 48 cm (19 inch). The weight of the removed kidney was 8.7 kg (19.3 lb) and its volume was 5,925 ml. This indicates that the

volumetric mass density (mass per unit volume) of the removed kidney was much higher than water. This may be due to the fibrotic deposition, multiple hemorrhages and clots. To our knowledge, only one case reported a patient with ADPKD that had kidneys with higher total weight than the present case [3].

Transection of the kidney revealed the presence of clear and cloudy fluid, to chocolate-brown semisolid contents in the cysts, consistent with the variable aspects of the cysts in the magnetic resonance image (fig. 2a). The dark contents of some cysts probably reflect passed bleeding events that may have been associated with a sudden increase of tension on the kidney capsule, a cause for pain and discomfort reported by the patient, although symptomatic episodes probably underestimate the true frequency of cyst hemorrhage.

In the removed kidney, most renal parenchyma in both the cortex and the medulla had been replaced completely by cysts and extensive scar tissue. Since, in general, both kidneys are usually affected to a similar degree by ADPKD, it seems remarkable that the patient's preoperative creatinine clearance was still 8 ml/min.

Disclose Statement

The authors have no conflicts of interest.

References

- 1 Meijer E, Rook M, Tent H, et al: Early renal abnormalities in autosomal dominant polycystic kidney disease. *Clin J Am Soc Nephrol* 2010;5:1091–1098.
- 2 Grantham JJ: Clinical practice. Autosomal dominant polycystic kidney disease. *N Engl J Med* 2008;359:1477–1485.
- 3 Ekser B, Rigotti P: Images in clinical medicine. Autosomal dominant polycystic kidney disease. *N Engl J Med* 2010;363:71.

Fig. 1. Magnetic resonance image showing the patient's voluminous bilaterally enlarged polycystic kidneys (total kidney volume is 10,280 ml). Several cysts are present in the liver.

Spithoven et al.: Nephrectomy in Autosomal Dominant Polycystic Kidney Disease: A Patient with Exceptionally Large, Still Functioning Kidneys

Fig. 2. **a** A resection specimen of the left polycystic kidney. **b** A histological sample after transection showing that both the cortex and medulla have been replaced completely by cysts and extensive scar tissue.