Case Report
Giant Symptomatic Polycystic Kidneys

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Abstract

Background: Autosomal Dominant Polycystic Kidney Disease (ADPKD) is one of the most common inherited kidney diseases and is the 4th commonest reason for dialysis. ADPKD is usually diagnosed in the 4th decade of life and is caused by mutations in genes PKD1 (80%) and PKD2 (20%). In the late stages ADPKD is characterised by fibrosis and inflammatory damage, which leads to complications and rapid progressive deterioration of the kidney. Methodology: Through an educational radiological imaging, a clinical case of a patient with giant polycystic kidneys who experienced an infectious complication prompting kidney removal was described. Results: Fifty nine years old woman with end-stage chronic kidney disease due to ADPKD was admitted presenting with left flank pain, fever, tachycardia, haematuria and dyspnea. Blood examination showed leukocytosis (46700 cells $\mu$L$^{-1}$), elevation of inflammatory indexes and raised serum creatinine (8.9 mg dL$^{-1}$). An abdominal CT scan showed kidneys 3 time bigger than normal (~30 cm each) and super-infected cysts. Severe sepsis with bacteremia from extended spectrum $\beta$-lactamase producing E. coli was diagnosed. The patient was managed with percutaneous drainage of the larger abscess (removing 1.2 L of pus) and intravenous meropenem. A bilateral nephrectomy was planned in order to prevent further infections and to reduce dyspnea due to diaphragm compression. Conclusion: Patients with ADPKD usually suffer from complications such as recurrent infections of the urinary tract, cyst haemorrhage and symptoms related to mass and nephrolithiasis. Although, treatments which prevent progression to the end-stage renal disease are not available, early detection and treatment of hypertension can prevent kidney damage. Total kidney volume may predict the future need for nephrectomy but may be particularly warranted if complications arise e.g., abdominal symptoms, recurrent infections, haemorrhage and cystic lesions suspected for neoplasm.

Key words: ADPKD, computed tomography, E. coli, end-stage, hypertension

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Data Availability: All relevant data are within the paper and its supporting information files.
INTRODUCTION

ADPKD is one of the most common inherited kidney diseases and is the 4th commonest reason for renal replacement therapy. Herein, a case of a patient with complicated giant polycystic kidneys was described.

CASE STUDY

Fifty nine years old woman with hypertension and stage-V (end-stage) chronic kidney disease was admitted to the infectious diseases ward presenting with left flank pain, fever, tachycardia, haematuria and dyspnea. She had been diagnosed with Autosomal Dominant Polycystic Kidney Disease (ADPKD) 5 years previously. On admission blood examination showed marked leukocytosis (46700 cells µL⁻¹), elevation of inflammatory indexes and raised serum creatinine (8.9 mg dl⁻¹). A Computed Tomography (CT) scan of the abdomen showed kidneys 3 time bigger than normal (right kidney 16×28 cm, left kidney 16×33 cm) (Fig. 1). In addition multiple abscesses, air-filled cystic formations and thickening of the peri-renal fascia were observed likely due to super-infection of the kidney cysts. Severe sepsis was diagnosed and blood cultures flagged positive with extended spectrum β-lactamase producing E. coli. The patient was managed with percutaneous drainage of the larger abscess (removing 1.2 L of pus) and intravenous meropenem after which clinical improvement was noted. A bilateral nephrectomy was planned in order to prevent further infections and to reduce dyspnea due to diaphragm compression.

ADPKD is usually diagnosed in the 4th decade of life. The disease is caused by mutations in genes PKD1 (80-85%) and PKD2 (15-20%)¹. There are clear genotype-phenotype correlations for the onset of end-stage renal disease and mutations of PKD2 have the better outcome. The most important prognostic factors are genotype, age, sex, estimated glomerular filtration rate (eGFR) and total kidney volume²³.

CONCLUSION

Total kidney volume may be measured by magnetic resonance imaging or CT and if eGFR is preserved is an early and accurate index of cystic burden and can predict grow rate. Patients with ADPKD may have normal renal function for many years because of compensatory hyperfiltration of healthy nephrons. In the late stages ADPKD is characterised by fibrosis and inflammatory damage, which leads to rapid progressive deterioration of the kidney. Patients with ADPKD usually suffer from complications, such as recurrent infections of the urinary tract, cyst haemorrhage and symptoms related to mass and nephrolithiasis.

Although, treatments which prevent progression to the end-stage renal disease are not available, early detection and treatment of hypertension can prevent kidney damage. Total kidney volume may predict the future need for nephrectomy but may be particularly warranted if complications arise e.g., abdominal symptoms, recurrent infections and haemorrhage and cystic lesions suspected for neoplasm.

REFERENCES


Fig. 1: A Computed Tomography (CT) scan of abdomen showed Autosomal Dominant Polycystic Kidney Disease (ADPKD) affected kidney (left)