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Abdominal Aortic Dissection in a Patient With Autosomal Dominant Polycystic Kidney Disease After Starting Peritoneal Dialysis☆

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ABSTRACT

Autosomal dominant polycystic kidney disease (ADPKD), one of the most common genetic disorders, is caused by mutations in the PKD1 or PKD2 gene. ADPKD primarily affects the kidneys, causing the development of multiple bilateral cysts that are characteristic of this condition. Besides renal abnormalities, other manifestations of ADPKD include hepatic, pancreatic, and splenic cysts, intracranial aneurysms, aortic aneurysms, and mitral valve prolapse. Reports of ADPKD-associated abdominal aortic dissections are not rare, but there have been no reports of an ADPKD patient developing intestinal obstruction and abdominal aortic dissection after initiating peritoneal dialysis. Herein, we present one such case.

Introduction

Autosomal dominant polycystic kidney disease (ADPKD) is one of the most common life-threatening single-gene diseases. In America, ADPKD is the fourth leading cause of end-stage (ESRD). Currently, there is no treatment that can stop ADPKD from forming cysts in the kidneys, and the condition cannot be cured. In this case, a 41-year-old ADPKD patient accompanied by chronic renal failure (CRF) whose only therapy solution is dialysis. The aortic dissection finally appeared after initiating peritoneal dialysis, which finally caused a sudden death.

Case presentation

A 41-year-old Chinese man presented to the emergency department with complaints of nausea and vomiting for 10 days. Associated symptoms were edema, foam in the urine, and fatigue. His medical history included autosomal dominant polycystic kidney disease (ADPKD), accompanied by chronic renal failure since an age of 40 years.

On physical examination, the patient had a normal habitus, blood pressure of 159/92 mm Hg, pulse rate of 82/min, and respiratory rate of 20/min. His abdomen was flat, soft, and without tenderness, and both lower extremities showed mild pitting edema.

The values of relevant laboratory parameters were as follows: troponin, I 0.047 ng/mL; blood nitrogen, 226.9 mg/dL; serum creatinine, 12.62 mg/dL; carbon dioxide-combining power of plasma, 12.7 mmol/L; serum calcium, 2.64 mEq/L; serum sodium, 109 mEq/L; and hemoglobin, 10.7 g/dL. Urinalysis showed 2+ protein and 2-4 red blood cells. An electrocardiogram showed normal sinus rhythm and a complete right bundle branch block with T wave inversion in leads V4 through V6. A chest radiograph was normal (Fig. 1A).

The patient was admitted to the nephrology department on September 11, 2012, and emergency hemodialysis was administered for his severe electrolyte disorder, metabolic acidosis, and toxic uremia. After the treatment, the patient's nausea and

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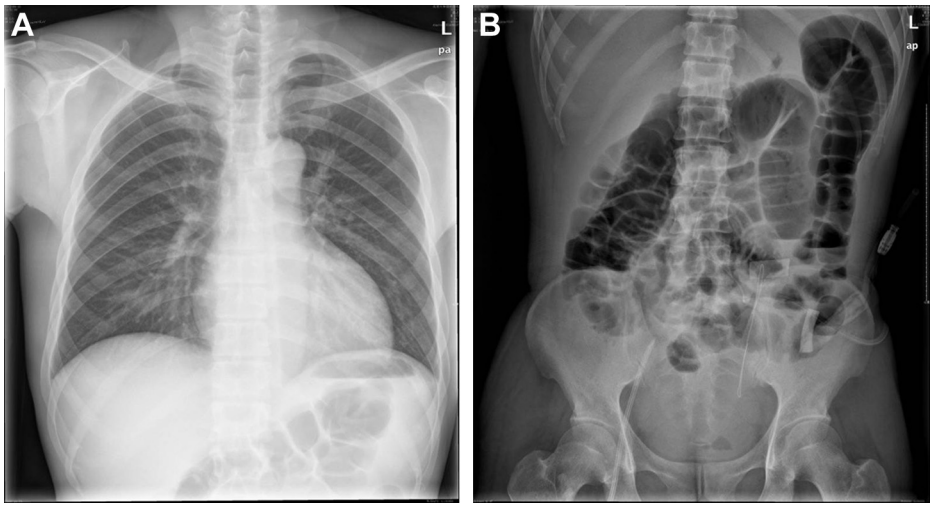

Figure 1. **(A)** Chest radiograph showing normal findings (9/11/2012). **(B)** Upright abdominal radiograph (9/21/2012) showing marked gaseous abdominal distension and short air fluid levels. The end of the catheter was at lumbar level 4.

vomiting were alleviated. On September 20, he received a peritoneal catheterization operation. Unfortunately, the next day, he experienced catheter occlusion, and his chief complaints were constipation and abdominal discomfort. An upright abdominal radiograph demonstrated partial intestinal obstruction and migration of the catheter, the end of which was at lumbar level 4 (Fig. 1B). Although multiple position changes and an enema were attempted, the occlusion remained. Peritoneal dialysis (PD) was therefore halted and temporarily changed to hemodialysis. After fasting and an anti-infective therapy, an abdominal plain-film review indicated that his partial intestinal obstruction was cured. On September 25, the patient presented with a sudden-onset excruciating back pain and sweating, with no radiating pain and shortness of breath. He remained hemodynamically stable, but his blood pressure rose to 180/90 mm Hg. The patient's electrocardiogram showed no change from before the treatment and cardiac enzymes, measured for the purpose of excluding an acute coronary syndrome diagnosis, and were found to be within the reference range. However, his D-dimer level was 8 times higher than normal, raising the possibility of a pulmonary embolism. Although under analgesia, the patient continued to complain of pain. Finally, the echocardiographic evaluation was completed, revealing aortic dissection, left ventricular hypertrophy, and slight aortic and pericardial regurgitations. Immediately, computed tomographic angiography of the abdomen was performed, which showed an aortic dissection extending from the descending aorta distal to the left subclavian artery to the iliac arteries bilaterally with a patent false lumen (Figs. 2, 3). The patient was diagnosed with a DeBakey type IIIB aortic dissection. Surgical intervention was recommended, and he was evaluated for endovascular stent-graft repair. Unfortunately, the patient did not have an uneventful postoperative course. His blood pressure was uncontrolled, and his condition deteriorated 2 days after surgery.

Discussion

ADPKD is one of the most common serious hereditary illnesses, found in 1 of 400-1 of 1000 individuals. Renal replacement therapy is at least as successful in ADPKD patients as it is in non-ADPKD patients, and extrarenal complications are not frequent.[1] PD is widely recognized as a feasible treatment option for most ADPKD patients with end-stage renal disease, whose overall survival rate and risk of peritonitis are reportedly similar to those of nondiabetic

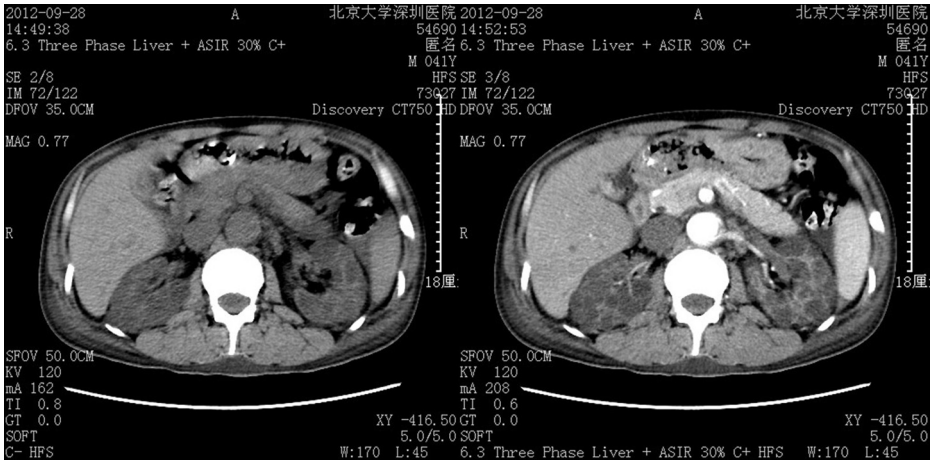

Figure 2. Upright abdominal radiograph (9/21/2012) showing marked gaseous abdominal distension and short air fluid levels. The end of the catheter was at lumbar level 4.

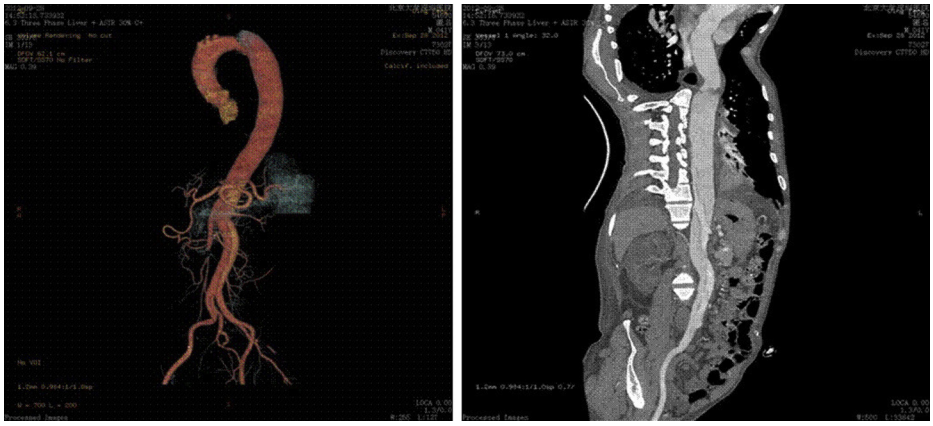

Figure 3. Abdominal computed tomographic images (9/28/2012) showing the uncountable kidney cysts, which appeared as rounded structures with near-water attenuation (HU = 0). The walls were very thin and regular, often not imperceptible.

PD patients.[2] There have been no reports in the literature on ADPKD patients with intestinal obstruction or abdominal aortic dissection after starting PD. In this article, we present one such case.

According to the available literature, intestinal obstruction is an unusual complication of ADPKD. Goncalves et al[3] reported one case of a 70-year-old man undergoing chronic hemodialysis for polycystic kidney disease, who had a partial intestinal obstruction caused by huge cysts that filled most of the abdominal cavity, leading to compression of the surrounding organs. This was relieved by a unilateral right nephrectomy. As for our patient, we inferred that his renal cysts were not as large as those in the previous case; therefore, the same complication was not encountered until he received the PD. The surrounding organs were compressed when 1-2 L of dialysis solution reduced the size of the abdominal cavity. This may be supported by the fact that the obstruction was easily resolved after PD was discontinued. However, more cases should be observed to identify the underlying mechanism.

Worse still, the patient was troubled not only by an intestinal obstruction but also by an abdominal aortic dissection since receiving PD. Aortic dissections are uncommon, yet they are highly lethal. If untreated, an aortic dissection can be fatal within the first 24-48 hours. Several risk factors are associated with aortic dissections, including high blood pressure (hypertension), genetic disorders affecting the blood vessel wall, atherosclerosis, cocaine use, and trauma. Although not recognized as a risk factor for life-threatening cardiovascular outcomes such as Marfan syndrome, the prevalence of aortic aneurysms in ADPKD patients varies from 1% to 10%.[4] Therefore, all physicians should pay attention to this potential complication. Standing hypertension, hemodialysis, and old age are closely related to the discovery of aortic aneurysms and dissections in ADPKD.[5] However, cases of aortic dissection occurring after PD are rarely reported. Whether an association exists between aortic dissection and PD remains unknown. We considered that PD might have just been an indirect factor, whereas hypertension could be the factor directly responsible. Because the patient developed intestinal obstruction 2 days before, his blood pressure could not be controlled during this period because of his continual complaints of constipation and abdominal discomfort. Unfortunately, the combination of ADPKD and uncontrolled hypertension resulted in an aortic dissection. Although surgical intervention was undertaken immediately, the patient did not survive. Sudden death occurred 2 days after the operation. We considered that perhaps he had a second dissection. However, the cause of death could not be ascertained because the patient's family refused an autopsy.

Conclusion

There have been no reports of an ADPKD patient developing intestinal obstruction and abdominal aortic dissection after initiating peritoneal dialysis. Despite the enormous effort putting into the diagnosis and treatment, the combination of ADPKD and uncontrolled hypertension finally resulted in an aortic dissection which caused a sudden death. We are showing this case in order to give medical staffs some alerts that same cases should be avoided in the treatment of ADPKD.

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