

Aortic Dissection Associated with Autosomal Dominant Polycystic Kidney Disease: A Case Report

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ABSTRACT

A 78-year-old man who had been diagnosed with autosomal dominant polycystic kidney disease (ADPKD) and hypertension presented with chest pain. His family history was positive for ADPKD. Chest computed tomography (CT) revealed a type A aortic dissection with thrombotic occlusion of a false lumen and an ulcer-like projection in the ascending aorta, an aneurysm of the ascending aorta, and pericardial effusion. Abdominal CT showed multiple renal and hepatic cysts. At surgery, aortic dissection with thrombotic occlusion of the false lumen and an intimal tear in the distal ascending aorta were observed. Hemiarch replacement including the intimal tear was performed. The patient is doing well without requiring dialysis and without recurrence of aortic dissection or aneurysm under strict antihypertensive therapy 3 years after the operation. Pathological examination of aortic wall specimens revealed no degenerative abnormality. ADPKD should be kept in mind as one of the causative disorders of aortic dissection.

INTRODUCTION

Autosomal dominant polycystic kidney disease (ADPKD) is caused by mutations in polycystic kidney disease-1 (PKD-1) or PKD-2 gene, and is characterized by progressive cyst formation in the kidney leading to end-stage renal disease. In addition to this main feature, extra-renal manifestations such as hepatic cysts, colon diverticula, and cardiovascular abnormalities are well recognized in ADPKD [Torres 2007]. Among the cardiovascular abnormalities, mitral valve prolapse, enlarged aortic root and annulus, cerebral aneurysm, coronary aneurysm, and myocardial hypertrophy are common morbidities [Leier 1984]. However, a small number of thoracic aortic dissections associated with ADPKD have been previously reported [Silverio 2015; Sung 2017]. We report a case of type A acute aortic dissection in a patient with ADPKD, and emphasize the importance of strict control of blood pressure and regular follow-up for the cardiovascular system.

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CASE REPORT

A 78-year-old man who was under treatment for hypertension presented with chest pain. The patient had been diagnosed with ADPKD and hypertension at the age of 65, and ascending aortic aneurysm at the age of 71. His family history was positive for ADPKD in one of his older brothers, however, it was negative for an older sister and his two children. The presence of ADPKD in his parents was unknown. His blood pressure was 114/86 mmHg and pulse rate was 67/min. No cardiac murmur was audible. Multiple large, round masses were palpable on the abdomen. Creatinine concentration was 3.19 mg/dL, and estimated glomerular filtration rate was calculated as 15.6 mL/min/m². A chest X-ray showed mild cardiomegaly and a widened upper mediastinum. An ECG revealed left ventricular hypertrophy without ischemic change in ST segments. Transthoracic echocardiography showed a normal-sized left ventricle, dilated ascending aorta, and moderate pericardial effusion. Mitral valve prolapse was not observed. Computed tomography (CT) demonstrated a type A acute aortic dissection with thrombotic occlusion of a false lumen and an ulcer-like projection, an aneurysm (5 cm) of the ascending aorta, and moderate pericardial effusion (Figure 1). Abdominal CT showed multiple cysts in the kidneys, the liver, and the spleen (Figure 2). At surgery, the pericardium was opened through a median sternotomy. Moderate bloody effusion was present in the pericardial cavity. Cardiopulmonary bypass was established between the right axillary artery and the right atrium, and myocardial protection was achieved with antegrade and retrograde cold blood cardioplegia. In addition to moderate hypothermic perfusion, perfusion pressure of 70–80 mmHg and perfusion volume of 2.2–2.4 L/min/m² were maintained for renal protection during cardiopulmonary bypass. When the ascending aorta was incised, the aortic dissection with the occluded false lumen extended from the ascending aorta to the arch, and an intimal tear was found in the distal ascending aorta. Hemiarch replacement including the intimal tear was performed during moderate hypothermic circulatory arrest with selective cerebral perfusion at the rectal temperature of 26°C. Duration of circulatory arrest was 68 min. The peak creatinine concentration was 4.34 mg/dL on postoperative day 3, thereafter, the patient recovered without requiring renal replacement therapy. Pathologic examination of aortic wall specimens showed mild atherosclerotic changes, but degenerative abnormalities such as cystic medial necrosis were not found. The patient is

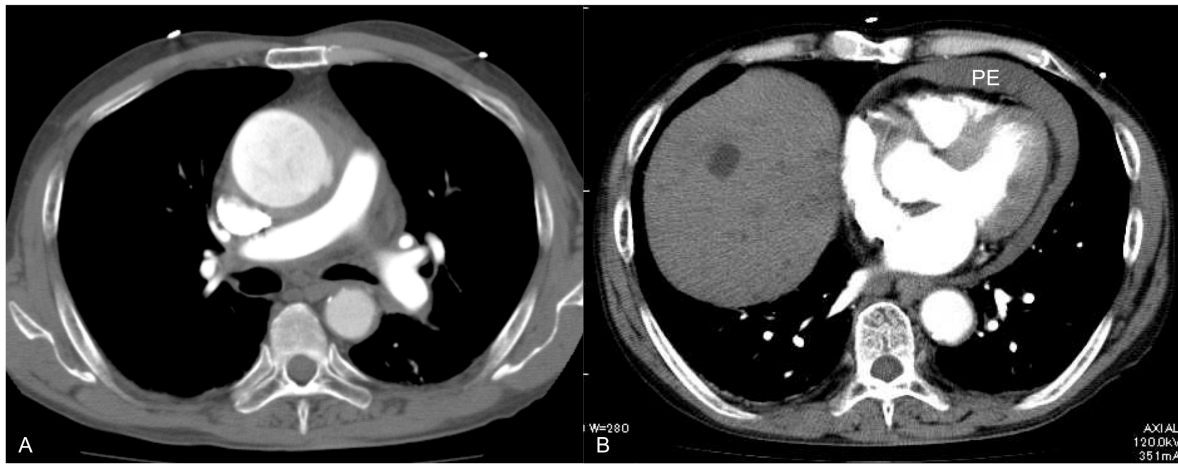


Figure 1. Chest CTs showing the type A aortic dissection with thrombotic occlusion of the false lumen and an ulcer-like projection, the ascending aortic aneurysm (A), and moderate pericardial effusion (B).

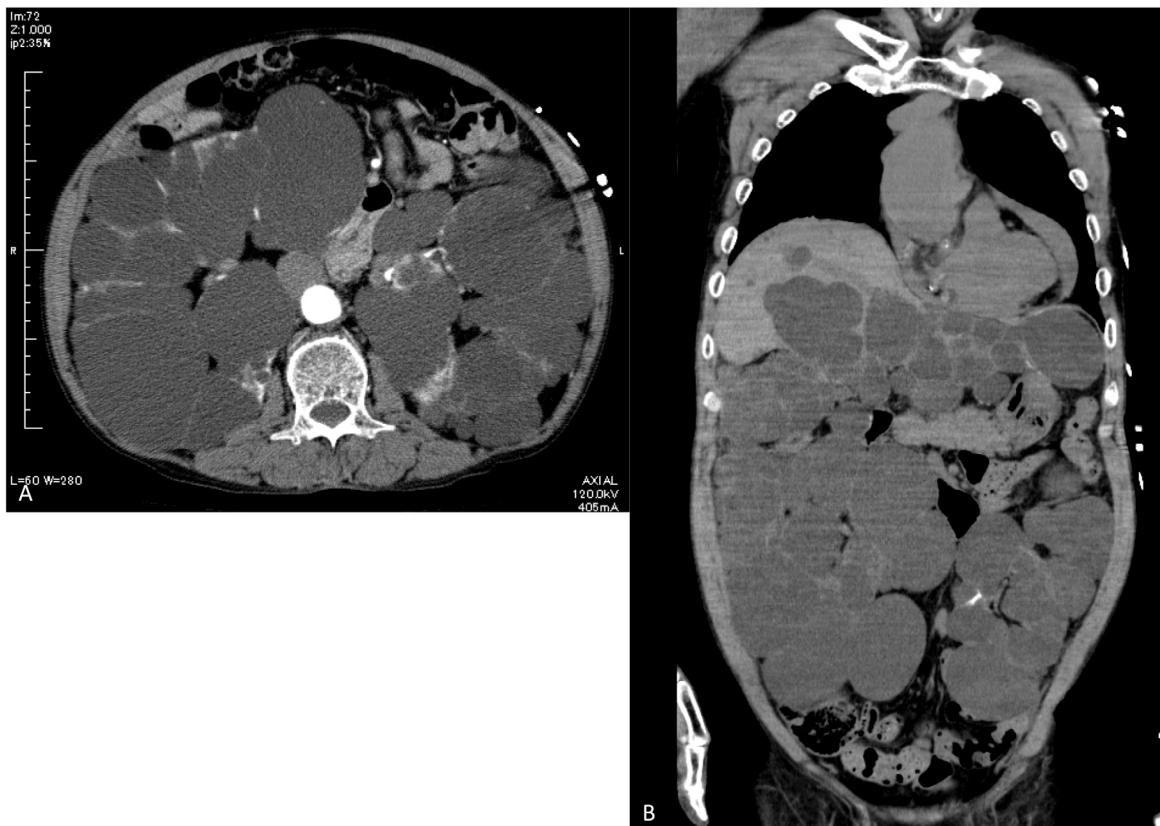


Figure 2. Abdominal CTs showing multiple cysts in the kidney, liver, and spleen.

doing well without requiring dialysis and without recurrence of aortic dissection under strict antihypertensive therapy with an angiotensin-II receptor blocker and other antihypertensive

drugs 3 years after the operation, although renal impairment is gradually getting worse, with the latest creatinine concentration at 5.74 mg/dL.

DISCUSSION

ADPKD, the most common inherited kidney disease, is a systemic disease resulting from mutations in either PKD-1 or PKD-2 gene. Mutations of these two genes induce systemic abnormalities of collagen and extracellular matrix. Clinically, along with progressive cyst formation of the kidney and other organs, patients with ADPKD are at an increased risk for a variety of cardiovascular abnormalities, such as aneurysms, dissections, and cardiac valve diseases [Leier 1984]. Nowadays, these cardiovascular complications are the most common cause of death in ADPKD [Silverio 2015]. Genetically, PKD-1 gene encodes the polycystin-1 protein, and PKD-2 gene encodes polycystin-2, and these two proteins are expressed in vascular smooth muscle cells and endothelial cells of large arteries such as the aorta and intracranial arteries [Griffin 1997]. Accordingly, mutations of these genes may induce a structural abnormality of the aortic or arterial wall which leads to development of coexisting cardiovascular abnormalities and renal cysts.

Generally, aortic aneurysms are estimated to occur in 1% to 10% of ADPKD patients [Hassane 2007]. Thoracic aortic dissection was seven times more common in patients with ADPKD than in the general population in an autopsy series [Leier 1984], whereas only a small number of thoracic aortic dissections have been clinically reported in ADPKD patients [Silverio 2015; Sung 2017]. However, recent clinical studies have clarified a significantly higher frequency of aortic dissection in ADPKD patients than in the general population. Sung et al demonstrated that occurrence of aortic events (aortic dissection and/or aneurysm) in ADPKD patients was significantly more frequent than in the general population, and showed that the ADPKD patients had an 8-fold greater risk for development of aortic dissection than non-ADPKD patients [Sung 2017]. Furthermore, they reported that patients with coexisting ADPKD and hypertension had a very high risk for development of aortic events. Silverio et al reviewed clinical features of 27 ADPKD patients who developed aortic dissection, and indicated that the ADPKD patients with aortic dissection were significantly younger than the patients from the International Registry of Acute Aortic Dissection, and hypertension was markedly prevalent in ADPKD patients [Silverio 2015]. As a result, they suggest that aortic dissection occurs more frequently and at an earlier age in ADPKD patients due to a combination of structural abnormalities of the arterial wall and hypertension. Hypertension is found in approximately 80% of patients with ADPKD [Sung 2017]. Therefore, strict control of blood pressure is especially important for suppressing aggravation of renal impairment and decreasing catastrophic vascular complications because it is not possible to prevent progression of the aortic wall damage caused genetically by a decrease or altered pattern of PKD expression in the aorta [Hassane 2007; Muller 2002]. The current guidelines recommend antihypertensive therapies to control blood pressure at less than or equal to 130/80 mmHg [Leier 1984]. Helal et al showed that more aggressive blood pressure control and renin-angiotensin-aldosterone system inhibition were associated with better preservation of renal function,

later onset of end-stage renal disease, and better survival [Helal 2013]. We used an angiotensin-II receptor blocker postoperatively as one antihypertensive drug to strictly maintain the recommended blood pressure.

In our patient, the aortic aneurysm and dissection occurred 6 years and 13 years after the diagnosis of ADPKD, respectively. Sung et al found that the incidence of aortic events abruptly increased after a time interval of 8 years (a mean period of 4 years) from diagnosis of ADPKD [Sung 2017]. These results suggest that even asymptomatic patients with ADPKD should be regularly followed up with image techniques such as echocardiography or CT after diagnosis of ADPKD, particularly after an interval of 3 years from the diagnosis, to enable early detection of vascular complications; and once patients with ADPKD complain of chest or abdominal discomfort, aortic dissection or rupture of an aortic aneurysm should always be kept in mind as the differential diagnosis. Generally, repair of the ascending aorta or aortic root is recommended when aortic diameter is 5.5 cm or greater in patients without risk factors; however, replacing the aorta should be considered for patients with ADPKD and hypertension when the diameter is 5 cm or larger, similar to patients with other connective tissue disorders such as Marfan syndrome.

In conclusion, we report a case of acute aortic dissection associated with ADPKD, and emphasize the importance of strict control of blood pressure and regular follow-up of the cardiovascular system. ADPKD should be kept in mind as one of the causative disorders of aortic dissection.

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