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Case Report

Successful Endovascular Stent-Graft Repair for Complicated Type B Aortic Dissection Developed in a Patient with Polycystic Kidney Disease

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Polycystic kidney disease (PCKD) is rarely associated with aortic dissection (AD), which is a life-threatening disease. Although endovascular stent-graft repair tends to show better outcomes than conventional therapies in complicated type B AD (TBAD), successful endovascular intervention of TBAD with malperfusion in a patient with PCKD has not been reported. This case shows a 37-year-old male who had sudden onset of sharply stabbing epigastric pain with severe hypertension, who was diagnosed with TBAD and PCKD by a computed tomography and initially underwent medical treatment. Four days after discharge, he was rehospitalized with left leg pain and paresthesia due to left lower leg malperfusion. Thoracic endovascular stent-graft repair covering the primary tear site of dissection was performed successfully, leading to a decrease in false lumen and improvement of symptoms. We report the case of complicated TBAD in a patient with PCKD treated with endovascular stent-graft repair.

Key Words: Aortic diseases, Polycystic kidney diseases, Endovascular procedures

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INTRODUCTION

Aortic dissection (AD) is a life-threatening condition involving the aorta, occurring at an estimated incidence of 3-8 cases per 100,000 people [1]. Systemic hypertension is the most common predisposing factor and polycystic kidney disease (PCKD) is a rare but important risk factor for AD. Therapies in complicated type B AD (TBAD) in patients with PCKD is not different from the general population [2,3]. In uncomplicated TBAD, medical treatment has a much better survival rate of 89% at 1 month compared with surgical treatment [4]. However, conventional therapies such as medical and surgical treatment in complicated TBAD are disappointing. Now, endovascular stent-graft repair is considered as an alternative to conventional therapies [5].

CASE

A 37-year-old male presented to the emergency room with sudden onset of sharply stabbing epigastric pain never felt before. He had a history of hypertension with no medication. His vital signs were as follows: blood pressure, 220/150 mmHg; pulse rate, 87 beats/minute; respiratory rate, 18 breaths/minute; and body temperature, 36.6°C. Physical examination revealed a weak pulsation on the left femoral artery and mild tenderness in the epigastric area. An electrocardiogram showed left ventricular hypertrophy with normal sinus rhythm. Chest radiography showed small

amounts of bilateral pleural effusion without cardiomegaly. All blood chemistry profiles were within normal limits except mild renal dysfunction; they were as follows: creatinine 1.28 mg/dL (estimated glomerular filtration rate: 71.0 mL/min), blood urea nitrogen 19 mg/dL, random glucose 152 mg/dL, total cholesterol 160 mg/dL, and uric acid 7.9 mg/dL. In a computed tomography (CT) of the chest and abdomen with contrast, TBAD originating from the aortic arch just next to the left subclavian artery and terminating at both common and external iliac arteries was observed (Fig. 1A, B). The left common iliac artery and celiac trunk were almost obstructed by thrombus in the false lumen (Fig. 1C). Multiple variable-sized cysts in the liver and both kidneys were found, compatible with PCKD (Fig. 1D). The family history for autosomal dominant PCKD was not evident, because he had no siblings and his parents died when he was young. Echocardiogram showed left ventricular hypertrophy (septal wall thickness on diastole, 14.3 mm) and minimal amount of pericardial effusion with a left ventricular ejection fraction of 56%. The ankle-brachial index (ABI) was 0.41 at 2 days after admission and improved to 0.74 before discharge. Follow-up CT scan showed a similarly severe but shorter occlusion of the left common iliac artery compared with the previous CT scan (Fig. 2A, B). He had medical treatment with carvedilol 25 mg, nifedipine 66 mg, diltiazem 90 mg for twice-daily dosing and candesartan 16mg, minoxidil 5 mg for once-daily dosing. He was discharged 11 days after admission under stable conditions including a mild claudication.

Four days after discharge, he was hospitalized again for severe left leg pain and paresthesia while walking. Blood pressure was normal during hospitalization, but the left femoral artery pulse was not palpable. Emergent arteriography revealed complete obstruction of the left common iliac artery. There was normal perfusion of the celiac, superior mesenteric and renal arteries via the true

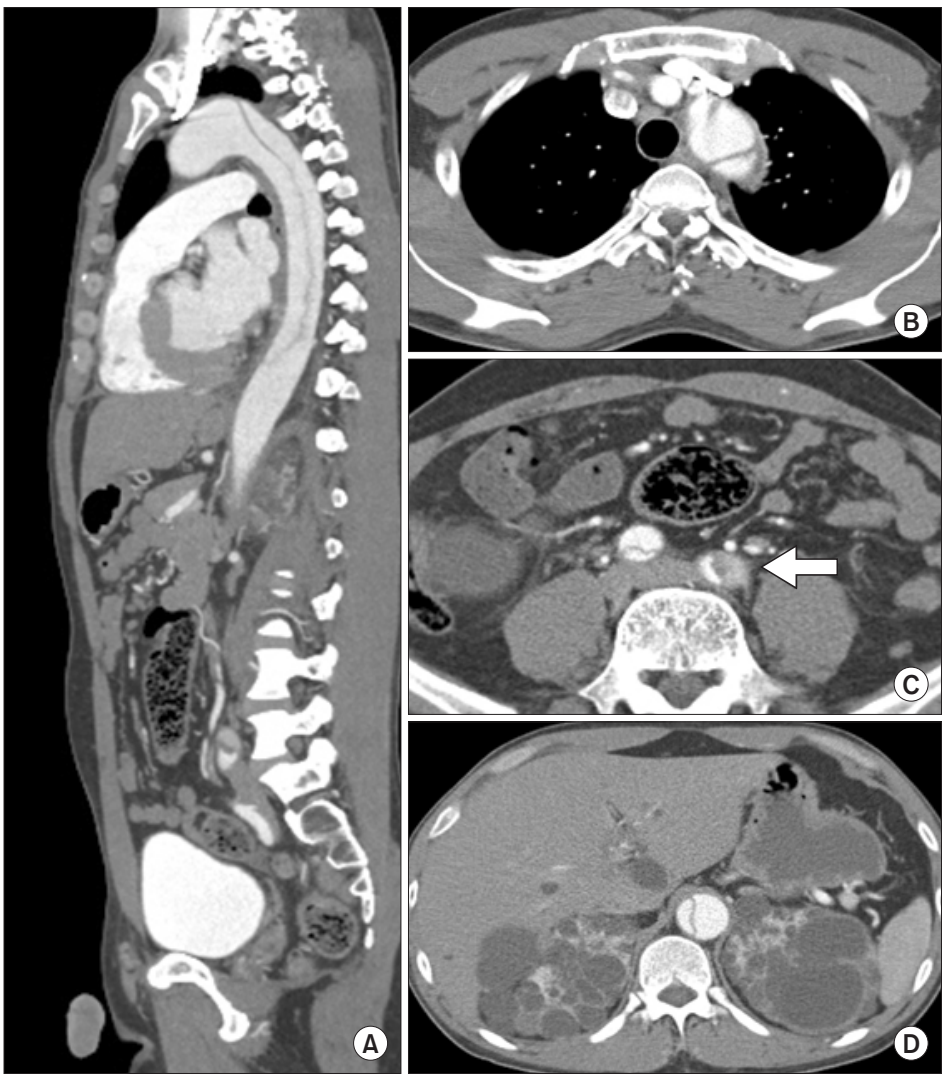

Fig. 1. Computed tomography of the chest and abdomen. (A) Dissection of the aortic arch distal to the left subclavian artery, sagittal view and (B) transverse view. (C) Left common iliac artery is almost obstructed by the false lumen with thrombus (arrow). (D) Multiple variable-sized cystic lesions in the liver and both kidneys.

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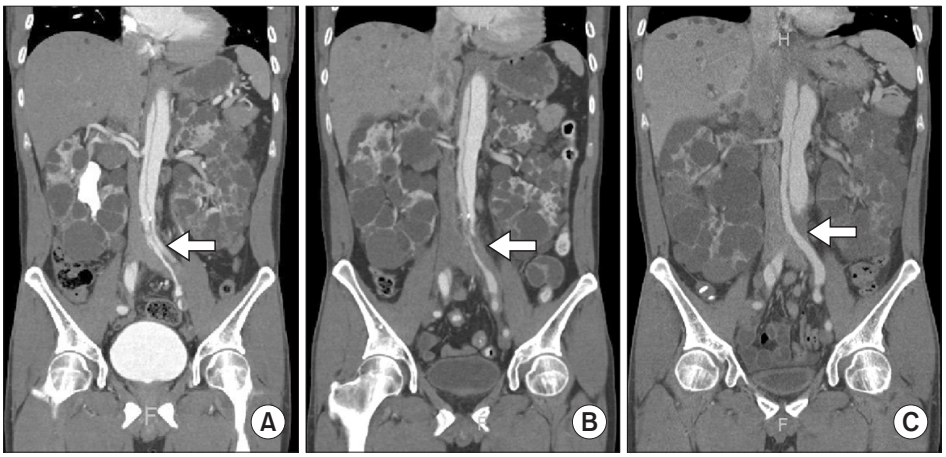

Fig. 2. Computed tomographies (CT) of the abdomen. (A) Severe long segment occlusion of the left common iliac artery on admission (arrow). (B) Similar severe but shorter segment occlusion of the left common iliac artery compared with the first CT 5 days later (arrow). (C) Resolved occlusion of the left common iliac artery 2 years after thoracic endovascular stent-graft repair (arrow).

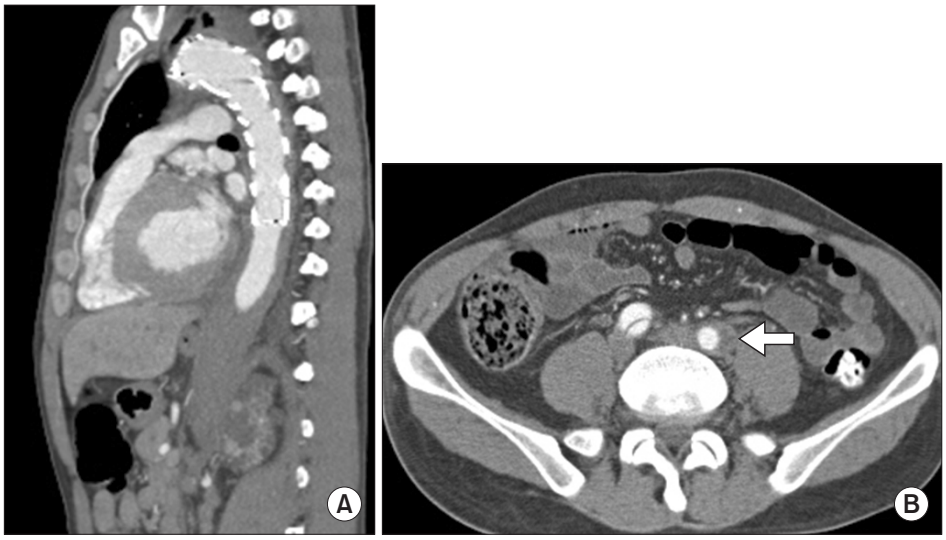

Fig. 3. Post-procedure computed tomography of the chest and abdomen. (A) Thoracic endovascular stent-graft repair. (B) Improvement of the left common iliac artery obstruction and false lumen thrombosis (arrow).

lumen. After being diagnosed as left lower leg malperfusion (LLM), thoracic endovascular stent-graft repair (TEVAR) was performed immediately via femoral cut-down approach under general anesthesia. The proximal edge of the dissected aorta had a diameter of 33 mm. The selected graft was 34×150 mm sized Valiant Thoracic Stent Graft (Medtronic Vascular, Santa Rosa, CA, USA). Post-CT scan of the aorta with contrast after the procedure showed improvement of flow of the left common iliac artery (Fig. 3). Pain and paresthesia of the left leg was relieved dramatically after the procedure. Since then, he was free of any symptoms. Two years later, follow-up CT confirmed no signs of malperfusion (Fig. 2C).

DISCUSSION

AD has been associated with diverse risk factors including atherosclerosis, connective tissue disorders, trauma, iatrogenic injuries, pregnancy, and especially hypertension [6]. Thoracic AD was seven times more common in patients with PCKD than in the general population in autopsy series [7]. Uncontrolled hypertension and impaired polycystin expression related to vascular wall integrity may increase the risk for developing an AD in patients with PCKD [3]. Complications such as unrelenting pain, progressive aortic dilatation, malperfusion syndromes or imminent rupture occur in 20%-45% of TBAD and LLM occurs in 19%-48% of complicated TBAD. Medical treatment in TBAD patients with life-threatening complications is disappointing but mild LLM can be improved with strict control of blood pressure and heart rate [8]. In our case, a mainly static obstruction of the left common iliac artery by thrombus from the false lumen and weak pulsation on physical examination with low ABI results were found, therefore interventional treatment was more favorable than medical treatment. We should have performed the intervention for LLM based on the follow-up CT scan before the first discharge, but we informed the patient about the possibility of an urgent

intervention.

Conventional surgical treatment demonstrates a high postoperative complication and mortality rate. Although the benefit of endovascular procedure over surgical treatment has not been defined properly due to lack of evidence, 30-day mortality/morbidity and postoperative complications after endovascular procedure tend to be better than surgical treatment. The concept of endovascular stent-graft repair is coverage of the primary tear site, leading to complete thrombosis and obliteration of the false lumen. It may also lead to true lumen expansion and recovery of LLM. The goal of therapy is to restore distal perfusion and correct end-organ ischemia [5,8]. Our report underlines a case of complicated TBAD requiring TEVAR with careful interpretation of follow-up CT scan.

Medical treatment with strict blood pressure and heart rate control are recommended in TBAD with PCKD. However, in case of complicated TBAD, careful observation and timely optimal intervention may improve outcomes.

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