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Case Report

Endovascular Management of Renal Artery Pseudoaneurysm in Autosomal Dominant Polycystic Kidney Disease: A Case Report

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Autosomal dominant polycystic kidney disease (ADPKD) is one of the most common hereditary kidney diseases. In addition to renal involvement, vascular complications including intracranial arterial, aortic aneurysms and dissections are common in these patients. We report the case of a 35-year-old male patient with ADPKD who presented with hematuria and was diagnosed with two intrarenal arterial pseudoaneurysms. Endovascular embolization using coils was performed to resolve these symptoms. Vascular complications are often encountered in patients with ADPKD; hence, sufficient clinical suspicion and timely diagnosis can help manage the disease. The most common causes of hematuria in ADPKD patients are cyst hemorrhage or infection; however, vascular aneurysms should also be considered a possibility.

Key Words: Autosomal dominant polycystic kidney, Embolization, Pseudoaneurysm

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INTRODUCTION

Autosomal dominant polycystic kidney disease (ADPKD) is the most common genetic disorder leading to renal failure. Mutations in the polycystic kidney disease 1 (PKD1) or polycystic kidney disease 2 (PKD2) genes, or both are responsible for the development of multiple cysts in both kidneys, eventually replacing renal parenchyma and resulting in renal failure [1]. As a systemic disorder, ADPKD also affects other organs, such as the liver and pancreas along with the genital and central nervous systems. They can be both cystic and non-cystic. Acute renal symptoms often results from cyst growth, leading to intracystic hemorrhage, infection, rupture, or development of nephrolithiasis.

Vascular involvement is one of the most common extrarenal manifestations of ADPKD, resulting in serious complications. Intracranial aneurysms are well-documented in patients with ADPKD [2,3]. However, extracranial visceral artery involvement is rare, with only a few cases reported in the literature, affecting the renal [4], renal and hepatic [5], and esophageal arteries with the development of Takayasu arteritis [6]. One case reported the use of percutaneous thrombin injection for the embolization of a renal pseudoaneurysm [4]. Here, we present a case in which endovascular coiling was used for the embolization of the renal pseudoaneurysm.

Ethical clearance was waived for this single patient case report. Adequate permission and consent were obtained from patient.

CASE

A 35-year-old male presented to the emergency department with left flank pain and gross hematuria. The patient was diagnosed with ADPKD in 2019 and had been on hemodialysis support for the last 4 years, with a usual urine

output of approximately 1-1.5 L/24 hours. The event was spontaneous with no preceding trauma, and the patient was at home during the episode. The patient had previously been admitted to our institution for gross right pleural effusion and acute flank pain due to renal cyst hemorrhage. There was no family history of similar diseases.

On examination, fullness and tenderness were noted in the left flank. The patient was afebrile with a blood pressure of 140/90 mmHg and a heart rate of 110 bpm. Laboratory investigations revealed a hemoglobin level of 7.1 gm/dL at presentation, which dropped to 6.0 gm/dL in 6 hours. The serum creatinine was 3.63 mg/dL. The patient received intravenous fluids, broad-spectrum antibiotics, and multiple blood transfusions due to a significant drop in hemoglobin levels.

Ultrasonography revealed multiple cysts of varying sizes in bilateral kidneys, with a large heteroechoic collection at the upper pole of the left kidney. The patient had previously shown evidences of cystic hemorrhages, which were spontaneous in nature and unrelated to injury. Given the persistent decline in the hemoglobin level despite aggressive conservative management, a renal pseudoaneurysm was suspected. Computed tomography (CT) scans revealed enlarged kidneys with multiple cysts, some of which appeared hyperdense. Multiple cysts were also observed in the liver (Fig. 1A). A large hematoma was observed at the upper pole of the left kidney (Fig. 1B). In the arterial phase, two contrast-filled outpouchings were seen arising from segmental branches of the upper and interpolar arteries on the left side (Fig. 1C).

The patient underwent digital subtraction angiography via a transfemoral approach. The left renal artery was cannulated using a renal double-curve catheter. Post-cannulation angiography confirmed two contrast-filled outpouchings arising from the segmental branches of the upper and interpolar arteries (Fig. 2A). Superselective cannulation was performed with a Progreat 2.7 Fr Microcatheter (Terumo), and these branches were embolized using pushable microcoils (VortX 18, Boston Scientific) (Fig. 2A-E). A total of two microcoils (2 mm in diameter, 3 cm in length) were used. Post-embolization angiography revealed non-opacification of the pseudoaneurysms (Fig. 2F). Following the procedure, hematuria resolved, and no further hemoglobin drop was observed. The patient was discharged when in stable condition after two days. We hypothesized that the occurrence of the pseudoaneurysm was secondary to a previous episode of cyst infection and hemorrhage.

The patient remains under follow-up and is undergoing regular hemodialysis, with adequate urine output. Ultrasonographic monitoring showed significant reduction in the size of the hematoma over 2 months post-procedure. No further episodes of hematuria or cyst infections have been observed.

During the follow-up, the patient underwent magnetic resonance imaging of the brain with time-of-flight angiography (a non-contrast study was selected due to the patient's renal profile). The study revealed no evidence of intracranial or extracranial aneurysms or vascular disorders.

DISCUSSION

Vascular complications of ADPKD are well documented in the literature. Perrone et al. [2] described the association between cardiovascular involvement and ADPKD, and hypothesized that genetic mutations in PKD1 and PKD2 predispose patients to the development of aneurysms and

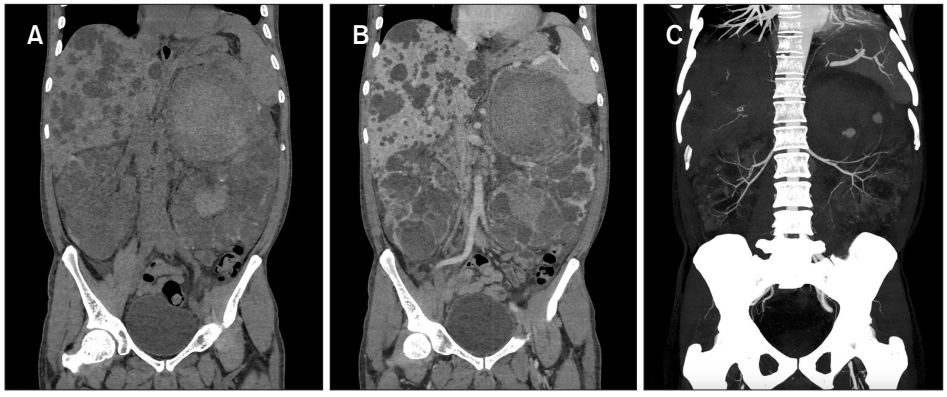

Fig. 1. (A) Non-contrast enhanced coronal reformatted section showing enlargement of bilateral kidneys with multiple heterogeneous cysts within the parenchyma. A hyperdense collection is seen at the upper pole of the left kidney suggestive of hematoma. (B) Contrast enhancement phase showing the presence of multiple cysts in bilateral kidneys as well as liver. (C) Maximum intensity projection reconstruction of the arterial phase showed two contrast-filled out-pouching arising from segmental branches of the left renal artery.

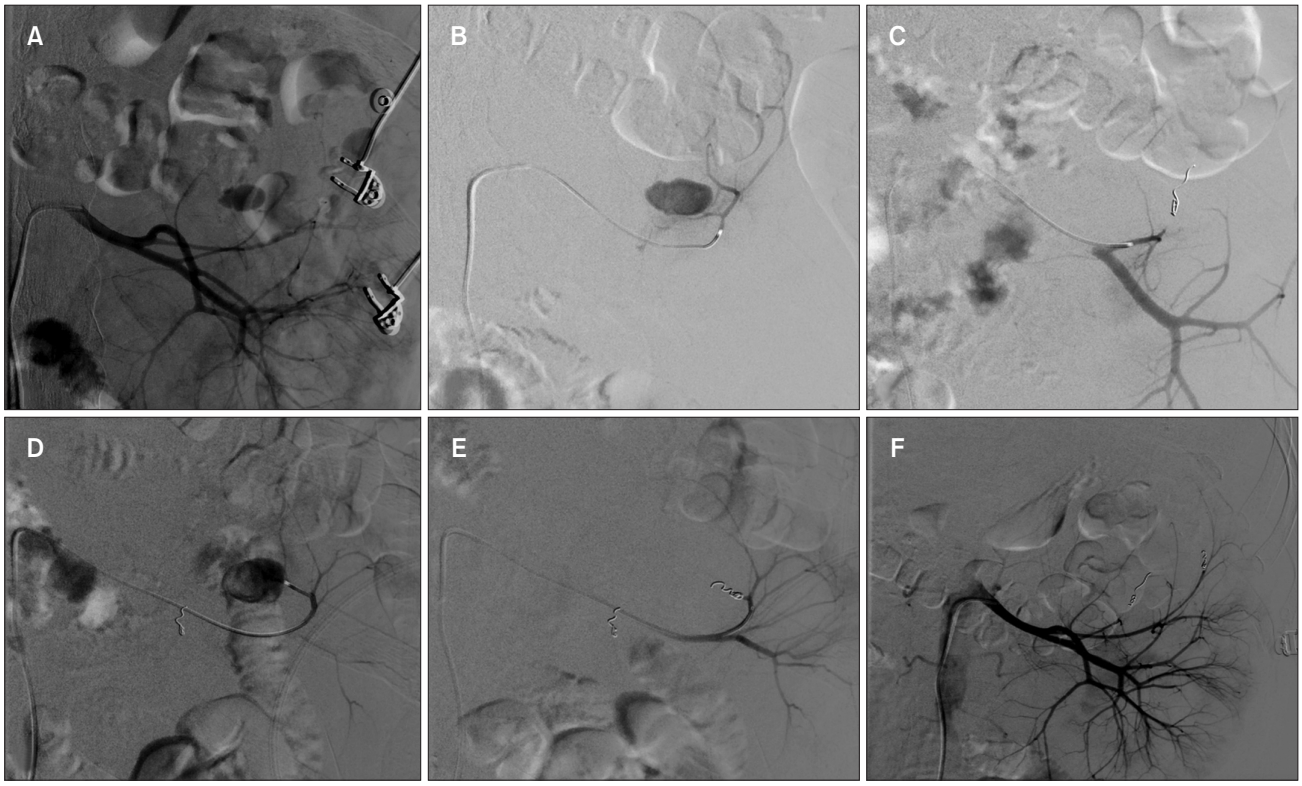

Fig. 2. Digital subtraction angiography images showing (A) guide catheter angiogram through the left renal artery showing two contrast-filled out-pouching arising from the segmental arteries suggestive of pseudoaneurysm. Selective catheterization and coil embolization of middle polar (B and C) segmental artery pseudoaneurysm and upper polar (D and E) artery pseudoaneurysm. The final guide catheter angiogram through the left renal artery (F) showed non-opacification of the pseudoaneurysms.

dissections. These genetic links have also been observed in patients with a family history of intracranial aneurysms associated with ADPKD [7,8]. Koska-Ścigała et al. [3] conducted a review of vascular involvement in ADPKD, highlighting cerebral artery aneurysms, cervical encephalic artery dissection, aortic aneurysm and dissection, and intracranial arterial dolichoectasia as common manifestations.

Involvement of the extracranial visceral artery is exceedingly rare in ADPKD. A literature review revealed three cases of extracranial vascular involvement. The earliest case of an intrarenal pseudoaneurysm in a patient with ADPKD was reported by Benjaminov and Atri [4], in which a 58-year-old male on hemodialysis presented with a pseudoaneurysm after a fall on his right lateral chest wall and flank. The patient was treated with ultrasound-guided percutaneous injection of bovine thrombin. Another case, reported by Kumar et al. [5], described aortic, hepatic, and renal arterial aneurysms in a 55-year-old female with ADPKD. Kim et al. [6] reported a 47-year-old female with an esophageal artery pseudoaneurysm and esophageal hematoma, successfully treated with endovascular embolization. In our case, we opted for endovascular coil embolization of an intrarenal pseudoaneurysm.

Renal artery aneurysms and pseudoaneurysms are among the most common visceral aneurysms, constituting 22%–25% of cases [9]. The most common causes include iatrogenic factors such as percutaneous biopsy, trauma, and infection; vascular dysplasia such as Marfan syndrome, Ehlers-Danlos syndrome, and atherosclerosis; and small- and medium-sized vessel vasculitis such as Takayasu arteritis, fibromuscular dysplasia, and polyarteritis nodosa.

Regarding acute intracystic hemorrhage in patients with ADPKD, patients on dialysis, total kidney volume, young age, hypertension, and increased renal artery luminal volume are factors that increase the risk of intracystic hemorrhage [10]. Bello-Reuss et al. [11] suggested an association between vascular malformations, such as aneurysms, and cysts in patients with ADPKD due to excessive angiogenesis and stretching of fragile vessels along the cyst walls.

Recently, endovascular techniques have become the mainstay in the management of renal artery aneurysms and pseudoaneurysms. Angiographically, aneurysms and pseudoaneurysms arising from segmental and small accessory arteries are classified as type III [12]. Superselective embo-

lization of the segmental branch is the preferred treatment for type III renal aneurysms and pseudoaneurysms. It can achieved by liquid embolic agents such as N-butyl cyanoacrylate, Onyx (ethylene vinyl alcohol copolymer), or coils deployed via microcatheters [13].

Transarterial embolization (TAE), along with nephrectomy, is an underdeveloped method for reducing kidney size and managing cyst-related complications in patients with ADPKD [14]. These volume reduction therapies are primarily performed before renal transplantation to create space in the pelvis for the graft, and are mostly recommended in patients with complete renal function loss [14,15]. TAE can also be considered in cases of recurrent cyst infections and hemorrhages. The patient in our case had moderate renal failure, with a urine output of 1.5 L/day. Loss of residual renal function, with complete loss of glomerular filtration is a serious complication of TAE that contributes to higher mortality rates [15]. In addition, severe complications are reported in patients with non-selective TAE such as post-embolization syndrome [14]. Hence, to preserve the residual function and reduce the complications after embolization, we opted for superselective embolization in our case.

In summary, patients with ADPKD presenting with hematuria should be evaluated for vascular complications, which, although rare, can be effectively managed using an endovascular approach, depending on lesion morphology.

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CONFLICTS OF INTEREST

The authors have nothing to disclose.

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