

CASE REPORT

Association of autosomal dominant polycystic kidney disease, asymptomatic multiple giant coronary arteries aneurysms and abdominal aortic aneurysm: a case report

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Abstract

Autosomal dominant polycystic kidney disease (ADPKD) is a systemic disease characterized by the formation of multiple cysts in several organs. The formation of aneurysms accompanying this disease is being increasingly reported in the literature, and mutations in PKD-1 and PKD-2 are suspected in this etiology. Although the association between ADPKD and multiple coronary arteries aneurysms (CAA) was reported several times, we are presenting a case with the combination of ADPKD, multiple giant CAAs, abdominal aortic aneurysms and a suspected intracranial aneurysm, which has never been reported. The asymptomatic presentation of these multiple aneurysms might support the recommendations for further diagnostic investigations in these patients.

INTRODUCTION

Autosomal dominant polycystic kidney disease (ADPKD) is a multisystemic and progressive disorder characterized primarily by cyst formation in the kidneys, in addition to the extrarenal involvement of organs such as liver, pancreas and spleen [1]. Patients with ADPKD usually present with a variety of signs and

symptoms; however, the most common complaint is abdominal or flank pain. Hypertension is also one of the most common early signs of ADPKD and it occurs in 50–75% of the patients [2]. Mutations in one of two genes PKD1 and PKD2, which encode polycystin-1 and -2, respectively, play a pivotal role in this disorder [3]. These proteins are expressed in smooth muscle cells and

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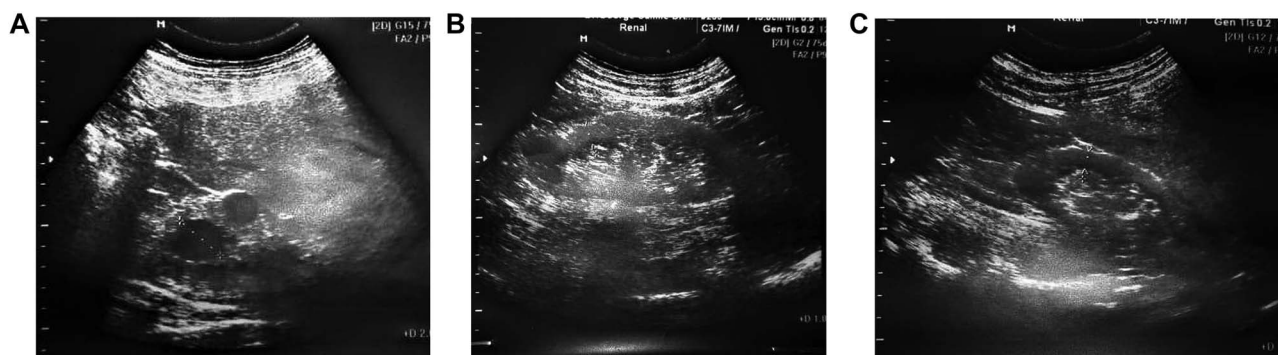


Figure 1: Renal echography showing multiple cysts in both kidneys.

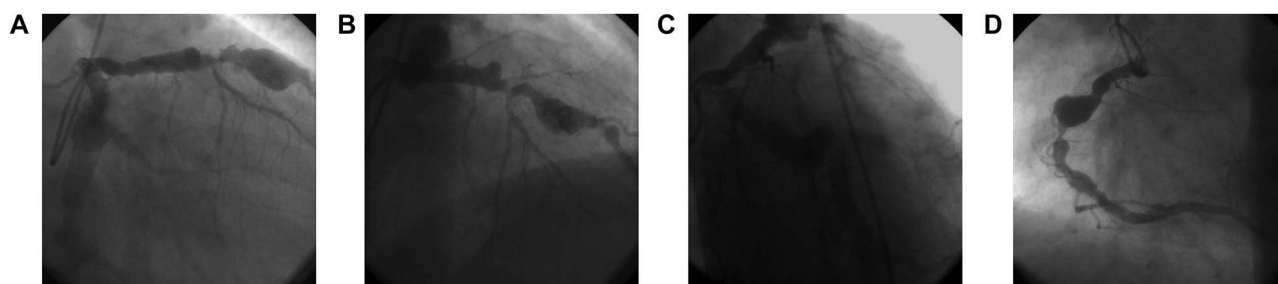


Figure 2: Coronary angiography of a patient with ADPKD showing multiple coronary aneurysms in: (A) the proximal LAD artery, the middle LAD and all of the circumflex artery. (B) The ostium, the proximal LAD and cystic one in the mid LAD. (C) The ostium, the proximal LAD and all of circumflex artery (spider view). (D) The first and the second segments of the right coronary artery.

myofibroblasts of the tunica media and in the endothelial layer of vessels, and when they are mutated, they lead to vascular wall weakness and the formation of aneurysms [4].

CASE REPORT

A 64-year-old male presented to our hospital complaining of polyuria, polydipsia, generalized edema, frequent nocturia and postvoid dribbling. His history included refractory hypertension for 7 years, though he was receiving multiple antihypertensive drugs, including Valsartan (320 mg), Atenolol (50 mg), amlodipine (10 mg), spironolactone (75 mg) and hydrochlorothiazide (75 mg). He also had a history of intracranial haemorrhage with complete recovery. His family history indicated a deceased cousin due to end-stage renal disease. On examination, he had elevated blood pressure (180/90), a distended but soft abdomen and a liver edge palpable 2 cm below the costal ribs.

Abdominal ultrasonography revealed abdominal aortic aneurysm (AAA) in addition to multiple cysts in both kidneys (Fig. 1). These findings were consistent with the ones in Multi-Slice Computerized Tomography (MSCT), in which three variable-sized cystic lesions were detected in both kidneys with a diameter of 32, 38 and 47 mm. The MSCT also showed the AAA that measured 3.6 cm, in addition to prostate hyperplasia. Furthermore, thorax MSCT images showed multiple aneurysms larger than 22 mm arising from the left anterior descending (LAD) artery, the left circumflex and the right coronary artery. On echocardiography, there was an eccentric left ventricle hypertrophy with an ejection fraction (EF) of 60%, while the electrocardiogram (ECG) uncovered a prominent left axis deviation. Magnetic Resonance Imaging of the brain showed an increased size of the cerebral ventricles in addition to multiple hypertensive lesions of 2–24 mm on Fluid-Attenuated Inversion

Recovery and T2-Weighted Image. Laboratory tests revealed high levels of serum creatinine (6 mg/dl), urea (105 mg/dl), haemoglobin (9.1 g/dl) and platelets count ($187 \times 10^3/\text{dl}$). Twenty-four hours urinalysis showed an additional massive proteinuria of 4200 mg/dl, in addition to a trace of red blood cells without cellular casts in microscopic examination. The patient underwent a coronary catheterization in order to prepare him for renal transplantation, which clearly revealed ecstatic coronary arteries with multiple aneurysmal dilations in the three main coronary arteries (Fig. 2). Currently, the case is managed with haemodialysis sessions twice a week.

DISCUSSION

Herein, we report a case of an association between ADPKD and coronary arteries aneurysms (CAA). To our best knowledge, this rare association is only described 16 times in the literature [5]. The diagnosis of ADPKD in our patient was based on the presence of multiple bilateral kidney cysts and extrarenal manifestations (Ravine's Criteria), after ruling out other possible pathologies [6]. Moreover, the clinical course of the disease, the patient's age and the normal values of inflammatory markers make Kawasaki disease in this patient less likely. On the other hand, patients with ADPKD have a high prevalence of intracranial aneurysms between 9 and 12% [7], which suggests that the haemorrhagic stroke in our patient was probably underlined by a cerebral aneurysm. Further study with magnetic resonance angiography is recommended. Neves *et al.* deduced in their systematic review that 6 out of 23 ADPKD patients (40%) have multiple coronary aneurysms as in our case [5]. Although these multiple involvements of coronary arteries are more frequently reported in ADPKD patients, aneurysms in the other vessels are still quite a rare association in the general population [5].

The most common associated vascular aneurysm with CAA is an AAA [8]. The association between multiple giant coronary aneurysms, which is quite rare, and other aneurysms including the AAA and the suspected cerebral aneurysm makes the presentation of our case extremely rare [8], because the combination between these three pathologies in the context of ADPKD was never reported before.

Furthermore, the giant cardiac aneurysms were diagnosed accidentally in our patient who did not have any cardiac symptoms, and this is consistent with the findings of the aforementioned systematic review, which showed that 6 out of the 13 patients with coronary artery aneurysms were asymptomatic (21%). This should draw attention towards the significant asymptomatic cardiac manifestations in ADPKD, which might prevent grave prognosis [5].

Although there are many recommendations regarding the isolated AAA and CAA, still such recommendation in the setting of ADPKD are unavailable. Boyer *et al.* recommended the surgical revascularization when a CAA involves the main stem of the left coronary artery, multivessel coronary artery disease is identified, a giant CAA is present or when other cardiac condition mandate surgery [9]. Moreover, the National Institute for Health and Care Excellence showed that all men aged 66 or more should be screened if they have not already been screened or they have one of the mentioned risk factors including hypertension. Additionally, they consider aneurysm repair for people with an unruptured AAA in case of symptomatic aneurysm, asymptomatic and larger than 4.0 cm with a growth by more than 1 cm in 1 year, or asymptomatic measuring ≥ 5.5 cm [10].

Rare associations of vessels aneurysms might happen in patients with ADPKD, including the coexistence of AAA, giant multiple coronary aneurysms and intracranial aneurysms. Hypertension also represents a common manifestation of this entity and could be refractory as in our patient. Therefore, it is recommended to investigate for secondary causes in patients with refractory hypertension as early as possible and to screen for asymptomatic aneurysms in cases of ADPKD, in order to prevent serious complications.

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ABBREVIATIONS

Autosomal polycystic kidney disease (ADPKD), coronary artery aneurysm (CAA), abdominal aortic aneurysm (AAA), multi-slice computerized tomography (MSCT), magnetic resonance angiography (MRA).

CONFLICT OF INTEREST STATEMENT

The authors declare that they have no competing interests.

FUNDING

No funding was required.

ETHICAL APPROVAL

Not required.

CONSENT

Written informed consent was obtained from the patient for the purpose of the publication of this case report and all accompanying images.

GUARANTOR

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