Images in Nephrology

(Section Editor: G. H. Neild)



Generalized aneurysmal disease in association with autosomal dominant polycystic disease

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Keywords: ADPKD; coronary aneurysms; femoral aneurysms

A 78-year-old man with end-stage kidney disease secondary to autosomal dominant polycystic disease (ADPKD) on haemodialysis, presented to our institution following a myocardial infarction. He was known to have cystic involvement of his liver and spleen, significant mitral regurgitation secondary to mitral valve prolapse, and a distant history of abdominal aortic aneurysm repair. There was no family history of ADPKD or history of cerebral aneurysms.

Coronary angiogram revealed widespread large coronary artery aneurysms affecting both branches of the left and right coronary artery, as well as significant atherosclerotic disease (Figure 1). During work up for coronary artery bypass surgery and mitral valve replacement, multiple other sites of aneurysmal disease were identified, including a 5 cm thoracic aortic aneurysm and bilateral femoral artery aneurysms (Figure 2).

Whilst waiting for cardiac surgery the patient developed septic shock secondary to an infected renal cyst, requiring prolonged intensive care admission and extended rehabilitation. Given his age and frailty a decision was made

to manage his widespread aneurysmal disease conservatively with regular radiographic surveillance.

Previous case series have suggested a high incidence of coronary artery aneurysms in association with ADPKD. Hadimeri et al. [1] observed coronary aneurysms in 4 out of 30 (13%) ADPKD patients. This is markedly higher than the incidence observed in the general population, of ~1.5% of patients studied by coronary angiography or autopsy [2].

There is less evidence regarding the link between ADPKD and aortic, thoracic and femoral aneurysms. While Bailey et al. [3] studied the association between aortic aneurysms and ADPKD, the study was unable to conclusively link the two conditions. Evidence linking thoracic or femoral aneurysms with ADPKD appears predominantly limited to case studies [4].

Given that the gene responsible for ADPKD has been linked with abnormalities of vascular smooth muscle, myofibroblasts, extracellular matrix and collagen defects [5], it seems reasonable for there to be widespread vascular abnormalities

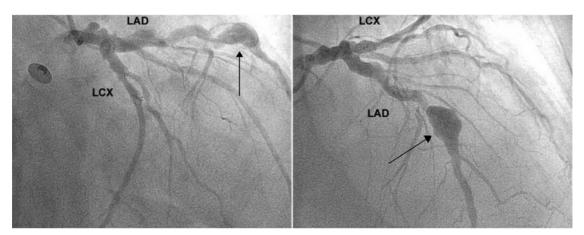


Fig. 1. Aneurysmal involvement of the mid to distal left anterior descending coronary artery (black arrow).

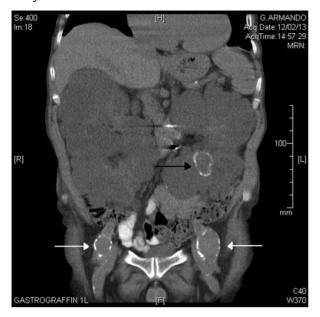


Fig. 2. Bilateral femoral artery aneurysms (white arrows) together with large polycystic kidneys. Note incidental finding of calcified cyst in left polycystic kidney (black arrow).

in this patient population, although it seems to be seldom described.

In summary, we present a rare case of a patient with ADPKD and associated coronary artery aneurysms as well as aorto/iliac aneurysms.

Conflict of interest statement. The results presented in this paper have not been published previously in whole or part.

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Received for publication: 30.4.14; Accepted in revised form: 13.5.14