Abdominal Aortic Aneurysm and Polycystic Kidneys

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Dear Sir,

The association of autosomal dominant polycystic kidney disease (ADPKD), also known as adult polycystic kidney disease, with various cardiovascular disorders is well known. The diagnosis of abdominal aortic aneurysm (AAA) associated with ADPKD has been reported in only a few cases. A personal case of successful aneurysmectomy is reported and incidence and treatment of AAA in the ADPKD patient population are discussed.

A 48-year-old Caucasian male, with known ADPKD, arterial hypertension, and chronic renal failure since 1979, was admitted in March 1991. During the last year he complained of progressive abdominal pain, essentially in the left flank, irradiating to both legs. At physical examination, the abdomen was tender and distended by two very large polycystic kidneys palpable as far as the midline. Peripheral arterial pulses were normal. The systolic blood pressure was 170 mm Hg upon admission. Other physical findings were irrelevant. Serum creatinine was 3.7 mg/dl (320 μ mol/l). Computed tomography of the abdomen showed voluminous polycystic kidneys, liver cysts, and a 5 cm wide fusiform infrarenal aortic aneurysm, however, without signs of leakage. There was no evidence of other cardiovascular abnormalities. The pain was attributed to the cystic kidneys, and the patient was transferred to the Department of Vascular Surgery for further investigation and elective aneurysmectomy. During the operation, the kidneys were carefully retracted laterally as far as possible, giving limited access to the aneurysm and the aortic bifurcation. After opening of the aneurysmal sac, an aortobifemoral bypass with a bifurcated Dacron prosthesis was performed. The postoperative course was uneventful. Pedal pulses were normal, and serum creatinine remained at the preoperative level during 10 months. After this period, the renal function started slowly deteriorating. At the present time, 30 months after surgery, serum creatinine is 6.3 mg/dl (546 μ mol/l), but the patient is still not under dialysis treatment, and he remains in good general condition without vascular symptoms.

ADPKD is frequently associated with lesions in the cardiovascular system. In a combined retrospective and prospective study, the incidence was estimated to be 18% by clinical

investigation and 27% by necropsy [1]. Aneurysms of the cerebral arteries are the most frequent abnormality, reported in up to 41% of the ADPKD patients [2]. Other disorders include dilatation of the aortic root, bicuspid aortic valve, mitral valve prolapse, coarctation of the aorta, and an increased risk of dissection of the thoracic aorta. Very few cases of AAA in association with ADPKD have been reported in the literature. Although a large multicenter study has been recommended by several authors [3-7], this is still missing. So, the question of whether a higher incidence of AAA in ADPKD patients is present in comparison with a control population remains unanswered. As far as we know, 8 cases have been reported, including ours. Two cases have not been operated on [3, 4].

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Two cases of rupture are reported, both fatal, one despite emergency surgery [3, 5]. Four patients had elective aneurysmectomy. Three, including our case, were successful [6,7]. One patient died after aneurysmectomy combined with kidney transplantation and right nephrectomy [3]. Until the present day, there is no evidence in the literature that the incidence of AAA is significantly affected by polycystic kidney disease. However, the association of AAA with ADPKD has some particular features for diagnosis and treatment. Clinical diagnosis of the aneurysm is very difficult, since palpation of a pulsatile mass between large polycystic kidneys is not always possible. The arterial hypertension, which is frequently present in these patients [2], increases the risk of leakage and rupture. Polycystic kidney patients often present with abdominal pain [2], and for this reason the pain of a leaking aneurysm can be misinterpreted, causing a dangerous delay in diagnosis. It is, therefore, important to examine the diameter of the aorta during repeated ultrasound studies to which these patients are routinely submitted during follow-up of their disease. Once the diagnosis of aneurysmal dilatation has been made, elective aneurysmectomy is even more strongly recommended in ADPKD patients than in others. The technical difficulty of the exposure of the aorta is increased by the presence of voluminous kidneys, causing delay in cross-clamping during emergency laparotomy for rupture. This will obviously increase the risk of this already high mortality condition. Most ADPKD patients are excellent candidates for kidney transplantation. This should be kept in mind in the choice of the operative technique.

In summary, we conclude that, due to the difficulty of a clinical diagnosis in the presence of large kidneys, repeated controls of the diameter of the aorta during ultrasonography are strongly recommended. Due to the increased difficulty of aneurysmectomy in these patients, early elective surgery is mandatory, since no survival after rupture has been reported.

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