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LETTER TO THE EDITOR

Fibromuscular dysplasia of the right kidney in a woman who donated her left kidney

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Fibromuscular dysplasia (FMD) of the renal arteries develops at a frequency of up to 4% with a female predominance [1,2]. The disorder usually manifests with significant hypertension, which may be followed by progressive renal failure. Several risk factors for development of FMD have been identified; however, the exact underlying pathophysiology is still not fully understood [2]. End-stage renal failure (ESRF) associated with FMD is a known indication for renal transplantation (RT). However, with the advent of better diagnosis and therapy, nowadays, only few patients with FMD progress to ESRF. As a result of the shortage of available cadaveric allografts, living donation has evolved as a common source of renal allografts for RT [3]. With an increase in the number of living donations, donor safety has become a major concern. Although currently there is no clear evidence for an increase in ESRF because of donation of one kidney [4], an alarming number of reports show evidence for a significant perioperative and long-term risk for severe complications, particularly in some subpopulations [5–7]. FMD may only involve one kidney; however, it can also affect both kidneys. Several reports have focused on living donation of grafts from donors with FMD [8]. One approach is utilization of a diseased kidney, leaving the donor with an undiseased second kidney and at the same time treating the hypertension by the donor nephrectomy. Back-table, FMD is then surgically repaired and outcome of such kidneys in general has been reported to be good [9]. However, there still may be a risk for development of FMD in the remaining kidney. Parasuraman *et al.* [10] reported on rapid progression of native renal artery FMD following kidney donation, although at the time of donation, no abnormalities of the donor renal arteries had been detected. This donor was successfully managed by angioplasty; however, the case emphasizes the significant risk of live kidney donation. We would like to add another case to the literature, where the donor developed FMD 8 years after the donation.

Our female patient was 40 years old at the time of kidney donation. The recipient was her father who suffered from diabetic/hypertensive nephropathy. On preoperative evaluation, the woman had no signs of hypertension and her serum creatinine was 1.0 mg/dl. She was slightly obese with a body mass index of 32 kg/m^2 and was an active smoker. Preoperative contrast enhanced magnetic resonance angiography (MRA) showed bilateral normal kidneys and no signs of changes typical of FMD of the renal arteries (Fig. 1). She donated her left kidney, which was laparoscopically procured without any complications. Her postoperative course was uneventful and she was discharged on day 2 with a serum creatinine of 1.2 mg/dl and a BUN of 40 mg/dl. The transplanted kidney showed excellent initial function and the recipient also had an uneventful course and was discharged on day 5 with a serum creatinine of 1.5 mg/dl and a BUN of 55 mg/dl. His immunosuppression included induction with antithymocyte globulin, tacrolimus with trough levels of 6–10 ng/dl, mycophenolate-mofetil at a dose of 2 g/day and a steroid taper. Whereas the donor was well without any signs of hypertension or renal impairment, the recipient had an elevation in his serum creatinine together with development of hypertension after 3 years. He underwent MRA, which showed significant atherosclerotic changes in the iliac arteries; however, the graft artery and anastomosis

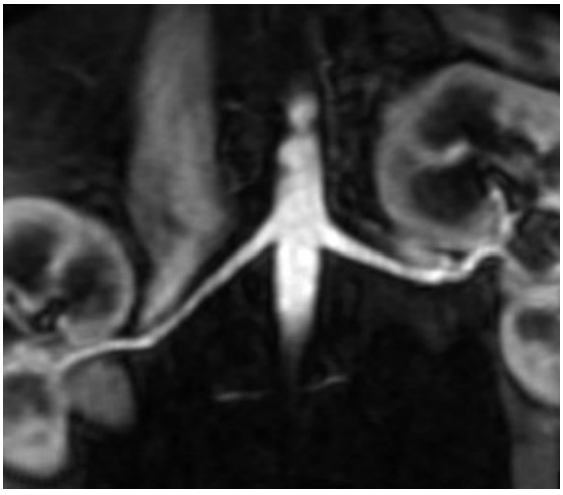

Figure 1 Preoperative donor evaluation: magnetic resonance angiography shows undiseased renal arteries.

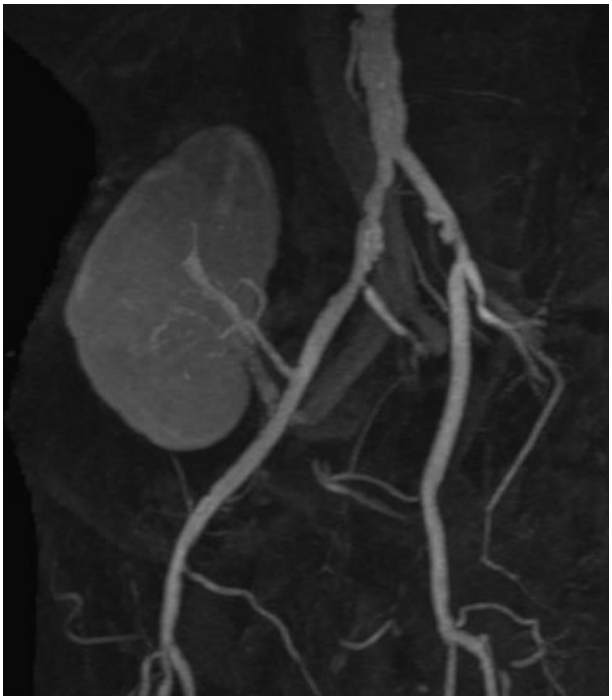

Figure 2 Magnetic resonance angiography of the recipient 3 years posttransplant: significant arteriosclerotic changes of both iliac arteries, but no signs of renal graft arterial disease or anastomotic changes.

showed no abnormalities (Fig. 2). Renal biopsy revealed signs of calcineurine inhibitor toxicity and accordingly, the TAC dose was reduced. Also, antihypertensive therapy was initiated. During further follow-up, the recipients' blood pressure was well and the serum creatinine was stable between 1.2 and 1.4 mg/dl. In fall 2008, the donor started to experience palpitations. Her blood pressure gradually increased to values of 160–190 over 90–110 mmHg. She was started on antihypertensive therapy including labetalol, hydralazine and propranolol. Also, her serum creatinine started to increase peaking at 1.6 mg/dl.

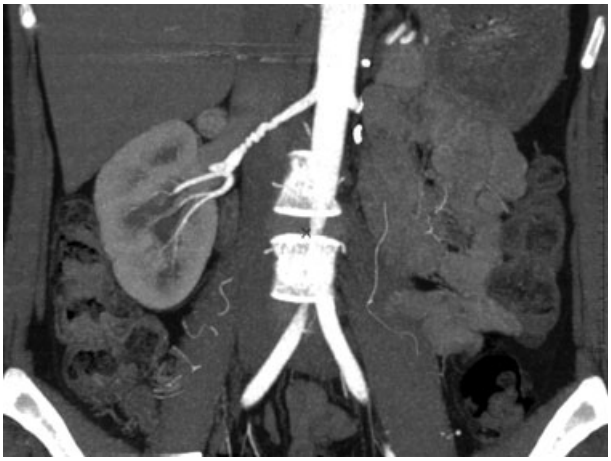

Figure 3 Computed tomography angiogram of recipient 9 years postdonation; clips on stump of left renal artery and vein of donated left kidney; typical string of beads appearance of the right renal artery indicative of fibromuscular dysplasia.

At this point, a Doppler ultrasound examination was performed, which revealed >90% stenosis in the mid right renal artery, a peak systolic velocity of >400 cm/s and a renal aortic ratio of >9; however, the exact pathology could not be identified. Therefore, computed tomography angiogram (CTA) was performed, which showed typical signs of FMD (Fig. 3). Subsequently, she underwent percutaneous transluminal angioplasty (PTA) (Fig. 4). Within few days after the procedure, her blood pressure normalized and serum creatinine values returned to 1.3 mg/dl. Both donor and recipient are under close observation with regard to progression/development of (de novo) FMD.

Our case emphasizes that kidney donation may be associated with a significant risk for long-term complications and donors should be thoroughly evaluated for potential secondary complications. Blood pressure monitoring and annual determination of serum creatinine and

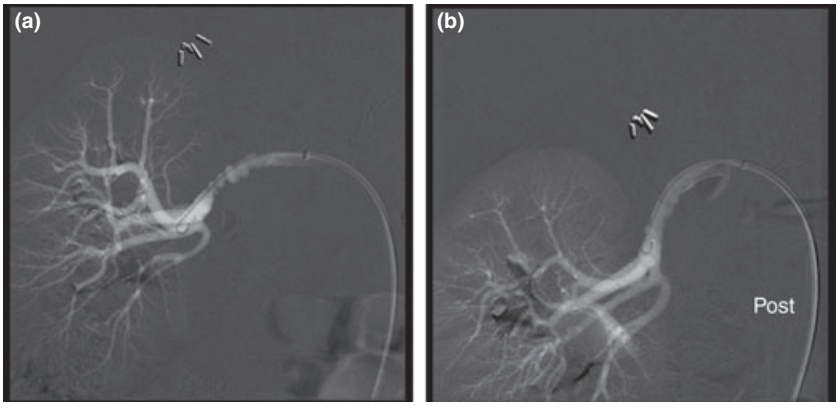

Figure 4 Angiography: (a) significant fibromuscular dysplasia (FMD) of right renal artery prior to angioplasty. (b) Completion angiogram following percutaneous transluminal angioplasty shows near complete resolution of FMD.

potentially, proteinuria may detect many disorders at an early stage and allow intervention before development of severe damage to the remaining kidney [11]. Ultrasound examination of the remnant kidney at fixed intervals may be an appropriate study, particularly in living donors with potential risk factors such as obesity, advanced age or nikotin abuse. During preoperative evaluation, FMD may be overlooked when only using CTA and MRA. However, as in our case, FMD could well have developed years after donation and pre donation, even angiography may have been without pathological findings. Although the diagnostic accuracy of cross-sectional vascular imaging has never been validated in a large trial for FMD, it is likely that a significant number of especially 'subtle' cases evade detection. Also, the diagnostic performance of CTA and MRA for detection of renal artery stenosis depends critically on the availability of state of the art scanners and local expertise. A large multicenter trial assessing CTA and MRA for the detection of atherosclerotic renal artery stenosis on older equipment used for evaluation of our donor demonstrated sensitivities of 64% and 62% and specificities of 92% and 84% for CTA and MRA, respectively [12]. In addition, only moderate interobserver agreement was found for both modalities. The latest generation imaging systems undoubtedly are far superior to older systems and it is questionable if routine catheter angiography is necessary for all potential donors and capable of detecting early stages of this particular disease. It has been reported that kidney donation was rejected when during pre-operative evaluation using CTA, ultrasound and MRA, FMD was detected particularly in cases of bilateral involvement. With regard to FMD, digital subtraction angiography (DSA) has the best predictive value, however, is no longer considered gold standard for donor evaluation [13–15]. Andreoni *et al.* [16] questioned this approach showing that a significant number of patients would have donated a kidney based on CTA/MRA and thus would have been left with a diseased second kidney, while DSA was able to demonstrate FMD. While older publications mainly focused on the recipient outcome if kidneys from patients with FMD were utilized, equal attention is required for the donors as the disorder may develop many years after kidney donation such as in our case. Our patient was obese and kept smoking, which may have contributed to development of FMD. Despite several attempts to convince her that these factors may put her at risk for a disease of her remnant kidney, she was not compliant and unable to change her eating or smoking habits. Donors must be advised already before donation that obesity, DM, hypertension and smoking may pose significant risks to their remaining kidney and appropriate risk factor modification is indicated. In the reported case, severe arterial hypertension led to performance of Doppler ultrasound and subsequently CTA, which established the diagnosis of FMD. PTA was successfully applied and within few days after the intervention, blood pressure and serum creatinine normalized. As a result of the known tendency of FMD to recur after successful PTA, long-term follow-up including Doppler ultrasound is required for our patient [17].

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