Brief reports

TRANSPLANT International

Transplant Int (1989) 2: 228-231

Fibromuscular dysplasia in donor kidneys - experience with three cases

R. Linder[1, *], H. Billing[2], A. Tibell[1], G. Tydén[1], and C.-G. Groth[1]

[1] Department of Transplantation Surgery and [2] Department of Radiology, Karolinska Institute, Huddinge Hospital, S-14186 Huddinge, Sweden

Abstract. In 1987, three patients received kidney grafts bearing medial fibroplasia at our hospital. Two of the grafts were from a cadaveric donor and one was from a living related donor kidney. The vascular affection was known before transplantation. Only one of the recipients developed stenosis and hypertension. With balloon catheter dilatation, the progressive stenosis of the renal graft artery could, however, be successfully corrected.

Key words: Fibromuscular dysplasia in kidney transplantation - Kidney transplantation, fibromuscular dysplasia - Arterial stenosis in kidney transplantation, fibromuscular dysplasia.

The first report about fibromuscular dysplasia of the renal artery leading to renovascular hypertension dates back 50 years and was presented by Leadbetter and Burkland [5]. It is known from angiography screening of potential kidney donors that the condition occurs with a frequency of 2%-3% [18, 19]. Still, to our knowledge, only one case of fibromuscular dysplasia in a renal allograft has ever been described [10].

Case reports

Case 1

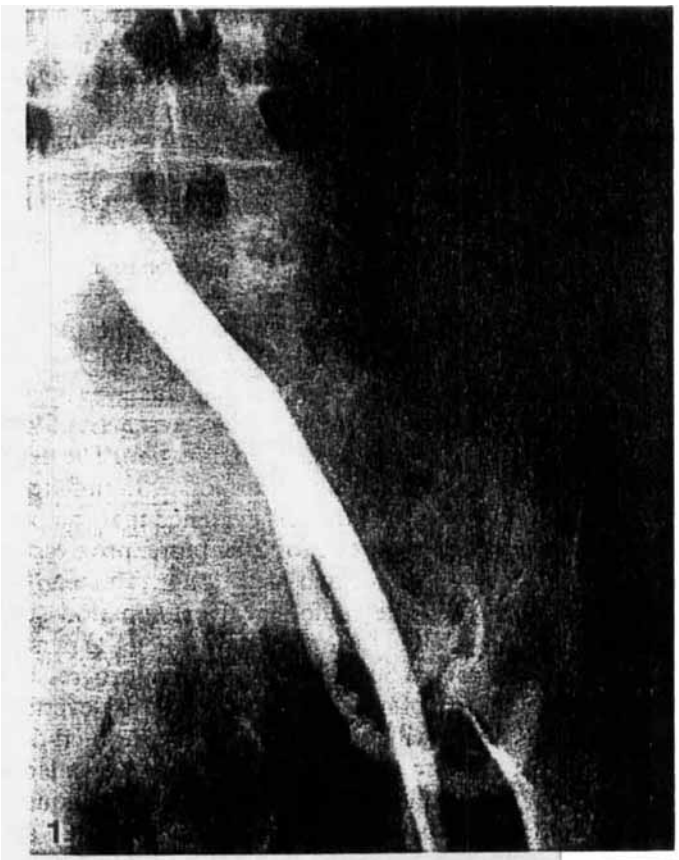
The recipient was a 30-year-old woman who had been suffering from type 1 diabetes for 20 years and had developed diabetic nephropathy. She was treated for hypertension with furosemide, metolazone, and prazosine. A combined pancreas-kidney transplantation was performed using a segmental pancreatic graft as described elsewhere [4]. The two grafts came from a cadaveric donor, a 53-year-old woman who died from subarachnoidal hemorrhage. She had had no history of hypertension or any other disease. During multiorgan procurement, moderate dysplastic changes restricted to the renal arteries were discovered. The renal artery was trimmed back to where it appeared normal. Histopathological examination of the resected specimen confirmed the suspected diagnosis of medial fibroplasia. An end-to-end anastomosis to the recipient's internal iliac artery and a venous end-to-side anastomosis to the common iliac vein were performed. Following transplantation the immunosuppressive treatment consisted of cyclosporin A, azathioprine, prednisolone, and antithymocyte globulin. Both grafts functioned immediately. The serum creatinine level dropped to within the normal range and exogenous insulin therapy was discontinued. Three rejection episodes had to be treated with steroids. Blood pressure came down gradually and the hypertension treatment was reduced stepwise and finally stopped.

Four-and-a-half months after transplantation, blood pressure increased from 130/85 mm Hg to 220/115 mm Hg within 2 weeks despite the reintroduction of medical treatment. Renal graft angiography showed a significant stenosis of the renal artery distal to the anastomosis compatible with fibromuscular dysplasia (Fig. 1). A percutaneous transluminal balloon catheter dilatation was performed and the immediate result is shown in Fig. 2. Blood pressure improved again, reaching 150/80 mm Hg within 3 months. Medical treatment, with the exception of diuretics, was discontinued at this time because of side effects. Renal angiography and clinical examination 12 months after transplantation showed no recurrence of stenosis (Fig. 3) and a stable clinical condition of the patient with a serum creatinine level of 154 μmol/l and blood pressure of about 125/80 mm Hg under low-dose treatment with diuretics. Pancreatic graft function deteriorated 6 months after transplantation but graft angiography showed normal status of the graft artery and of the anastomosis (Fig. 4).

Case 2

This recipient was a 36-year-old woman with polycystic kidney disease. She suffered from severe hypertension that required quadruple therapy of metoprolol, nifedipine, clonidine, and captopril. This patient received the other kidney from the cadaveric donor described under case 1. Again, the renal artery was trimmed

* *Present address and address for offprint requests:* Klinik für Allgemeinchirurgie, Justus-Liebig-Universität, Klinikstrasse 29, D-6300 Giessen, FRG



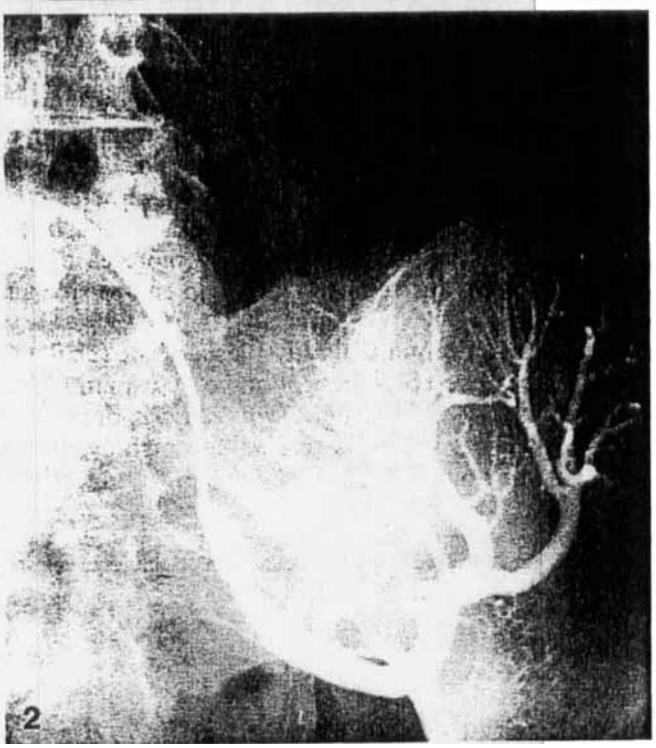
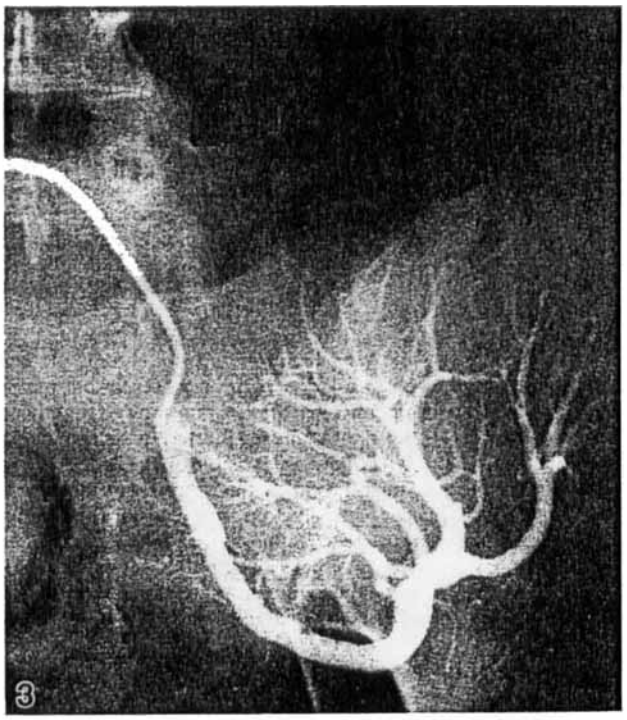


Fig. 1. Renal graft angiography of the recipient with recurrence of hypertension 4.5 months after transplantation. Extensive stenosis beyond the anastomosis secondary to progressive fibromuscular dysplasia

Fig. 2. Renal graft angiography immediately after balloon catheter dilatation



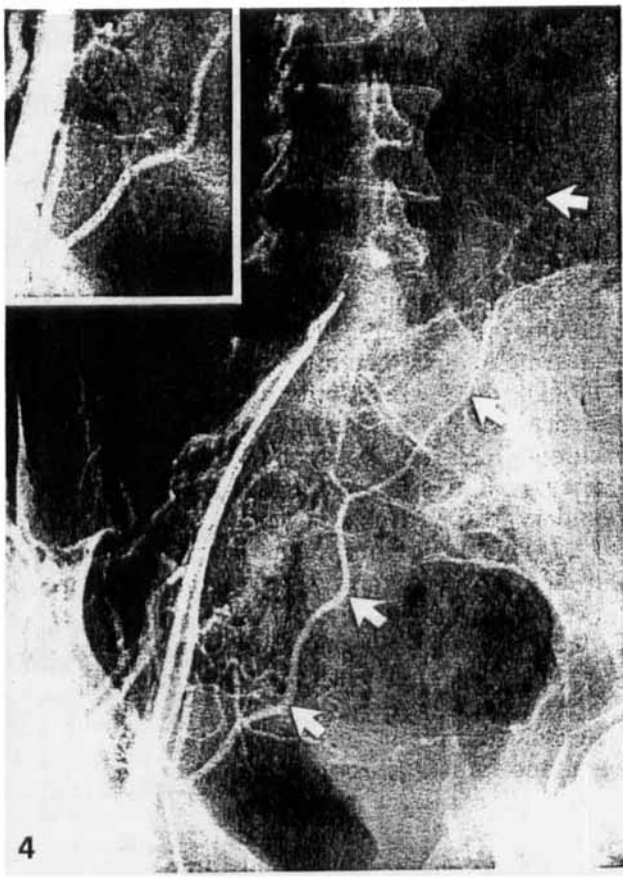


Fig. 3. Renal graft angiography 6 months after balloon catheter dilatation demonstrating unchanged therapeutic success

Fig. 4. Angiography of the segmental pancreatic graft showing the splenic artery *(arrows)* without any signs of fibromuscular dysplasia. *Inset:* normal appearance also at the site of the anastomosis

back and the vascular anastomoses were performed as described above. After transplantation the recipient was given cyclosporin A, azathioprine, and prednisolone. Hemodialysis was necessary for 3 weeks until the renal graft started to function. Four rejection episodes could be managed with steroids and OKT3 treatment. Serum creatinine levels dropped slowly and hypertension treatment could be reduced. Renal angiography 12 months after transplantation showed slight dysplastic changes of the renal graft artery but no stenosis.

Case 3

This was a 19-year-old woman suffering from chronic glomerulonephritis. She received a kidney from her 50-year-old mother. During donor work-up, the renal arteriogram showed slightly narrowing changes of the right main renal artery with suspicion of medial fibrodysplasia. Following excision of the kidney, it was found that the proximal 1.5 cm of the artery bore most of the changes. This part was resected before anastomosing the kidney in the recipient using the same technique as mentioned in cases 1 and 2. As yet, no complications have occurred in the postoperative course. Histopathological examination of the resected artery showed moderate lesions of medial fibroplasia.

Discussion

Most, if not all, of our knowledge about fibromuscular dysplasia comes from patients showing more or less severe symptoms, mainly renovascular hypertension [1]. The histological categorization of the different variants of the lesion remains unsettled [15]. The most common site of occurrence is the renal artery, the second most common being the carotid artery and its branches. Fibrodysplasia can, however, affect almost all medium-sized branches of the aorta, including the celiac, superior mesenteric, inferior mesenteric, and iliac arteries [6, 11, 13-15, 21]. Although there are relatively few symptomatic instances of the disease in visceral arteries, signs of chronic intestinal arterial occlusion and forming of aneurysms of the hepatic and splenic arteries secondary to fibrodysplasia have been reported [6, 11, 12, 14, 15, 20]. Reports about the spontaneous course of this disease are rare. A long-term follow-up study in 91 hypertensive patients who had not been operated upon showed interesting results: progression occurred in only 2 of the 40 patients with medial fibroplasia, whereas in 7 of the 8 patients with perimedial fibroplasia, medial fibromuscular hyperplasia, and intimal fibroplasia, new disease developed or existing disease progressed [7]. Another series, consisting of 42 patients with different types of renal artery fibroplasia, showed all forms as being progressive with variable rates of progression. However, the study population was restriced to patients who had evidence of the development of new disease or of the progression of existing disease [2]. On the other hand, cases with symptomatic renal artery fibrodysplasia showing improvement or even reversal without surgical or balloon catheter correction have been reported [9]. Obviously, there is a distinct number of relatively mild or even asymptomatic cases, especially in the most common variation, i.e. medial fibroplasia. Thus, it may be that fibrodysplastic changes are discovered by chance in kidney donors and that a decision then has to be made for or against using the kidney.

Based on the available data concerning the nature of the disease, and taking into account the observations we made in our three patients, we suggest that living donors with bilateral changes, even of low degree, should be excluded from donation. The decision is more difficult in living donors with unilateral changes. If there is sufficient evidence that the person is not carrying one of the more progressive forms, he can be accepted as a donor. This can be presumed when the radiological examination suggests merely medial fibroplasia and when the person is between 45 and 50 years of age, when progression or development of new disease is unlikely. Nevertheless, this donor will remain at a slightly elevated risk. He must agree to this and this has to be considered for his later follow-up. Cadaveric donor kidneys should not be used when there is anatomically advanced fibroplasia. Thus, significant stenotic lesions that cannot be resected and aneurysms would preclude use of the kidney. Cadaveric donor kidneys with moderate or mild lesions but without apparent stenosis could be considered for transplantation since progression is usually slow and, if it were to occur, might be treated by balloon catheter dilatation [3, 8, 16, 17, 22]. If eventually indicated, balloon dilatation can also be performed more easily in an end-to-end anastomosis than in an end-to-side anastomosis.

Thus, fibromuscular dysplasia of the renal artery is a relative contraindication to transplantation of the kidney. Should significant stenosis or hypertension ensue, it can be treated successfully, as illustrated by our case 1.

Acknowledgement. The authors gratefully acknowledge the support they received from the Deutsche Forschungsgemeinschaft, D-5300 Bonn 2.

References

1. Foster JH, Oates JA, Rhamy RK, et al. (1969) Hypertension and fibromuscular dysplasia of the renal arteries. Surgery 65: 517
2. Goncharenko V, Gerlock AJ, Shaff MI, Hollifield JW (1981) Progression of renal artery fibromuscular dysplasia in 42 patients as seen on angiography. Radiology 139: 45-51

3. Grossmann RA, Dafoe DC, Shoenfeld RB, et al. (1982) Percutaneous transluminal angioplasty treatment of renal transplant artery stenosis. Transplantation 34: 339-343
4. Groth CG, Tydén G (1988) Segmental pancreatic transplantation with enteric exocrine drainage. In: Groth CG (ed) Pancreatic transplantation. Saunders, Philadelphia, pp 99-112
5. Leadbetter WF, Burkland CE (1938) Hypertension in unilateral renal disease. J Urol 39: 611-626
6. Marston A, Clarke JMF, Garcia Carcia J, Miller AL (1985) Intestinal function and intestinal blood supply: a 20 year surgical study. Gut 26: 656-666
7. Meaney TF, Dustan HP, McCormack LJ (1968) Natural history of renal artery disease. Radiology 91: 881-887
8. Mollenkopf F, Matas A, Veith FJ, et al. (1983) Percutaneous transluminal angioplasty for transplant renal artery stenosis. Transplant Proc 15: 1089-1091
9. Nemcek AA, Holmburg CE (1986) Reversible renal fibromuscular dysplasia. Am J Radiol 147: 737-738
10. Nghiem DD, Schulak JA, Bonsib SM, Ercolani L, Corry RJ (1984) Fibromuscular dysplasia: an unusual case of hypertension in the transplant recipient. Transplant Proc 17: 555-558
11. Palubinskas AJ, Wylie EJ (1961) Roentgen diagnosis of fibromuscular hyperplasia of the renal arteries. Radiology 76: 634
12. Patchefsky AJ, Paplanus SH (1967) Fibromuscular hyperplasia and dissecting aneurysm of the hepatic artery. Arch Pathol 83: 141
13. Perry MO (1974) Fibromuscular dysplasia. Surg Gynecol Obstet 139: 97-104
14. Ripley HR, Levin SM (1966) Abdominal angina associated with fibromuscular hyperplasia of the celiac and superior mesenteric arteries. Angiology 17: 297
15. Rose AG (1983) Diseases of medium-sized arteries including hypertension. In: Silver MD (ed) Cardiovascular pathology, vol 2. Churchill Livingstone, New York, pp 739-796
16. Sniderman KW, Sprayregen S, Sos TA, et al. (1980) Percutaneous transluminal dilatation in renal transplant arterial stenosis. Transplantation 30: 440-444
17. Sos TA, Pickering TG, Souheil S, et al. (1984) The current role of renal angioplasty in the treatment of renovascular hypertension. Urol Clin North Am 11: 503-513
18. Spanos PK, Simmons RL, Kjellstrand CM, et al. (1974) Screening potential related transplant donors for renal disease. Lancet I: 645-649
19. Spring DB, Salvatierra O, Palubinskas AJ, et al. (1979) Results and significance of angiography in potential kidney donors. Radiology 133: 45-47
20. Stanley JC, Fry WJ (1974) Pathogenesis and clinical significance of splenic artery aneurysms. Surgery 76: 898-909
21. Stanley JC, Gewertz BL, Bove EL, Sottiurai V, Fry WJ (1975) Arterial fibrodysplasia. Arch Surg 110: 561-566
22. Tellis VA, Matas AJ, Veith FJ (1988) Vascular complications of transplantation. In: Cerilli CJ (ed) Organ transplantation and replacement. Lippincott, Philadelphia, pp 423-432