Rescue from hemodialysis by late recanalization of renal artery occlusion

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ABSTRACT

We report on a patient with terminal renal insufficiency undergoing hemodialysis since four months. Imaging studies showed complete renal artery occlusion of a single kidney with collateral perfusion. Interventional recanalization of the renal artery was successful with a drop of serum creatinine from 1138 to 163 mol/l sparing the patient from further hemodialysis.

CASE REPORT

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We report on a 49-year-old female patient with a single kidney undergoing hemodialysis since four months due to terminal renal failure that occurred in the course of gastroenteritis. The left pelvic kidney had been removed six years earlier in the course of a caesarean section. Blood pressure was approximately 160/95 mmHg under double antihypertensive medication (Amlodipin and Carvedilol). Prior ultrasound guided biopsy of the right kidney was unobtrusive.

Initially, a CT-angiography was performed for assessment of renal perfusion and evaluation of potential renal artery recanalization, depicting total renal artery occlusion with collateral perfusion of the kidney (Figure 1 and 2). Interventional recanalization was considered as feasible and the patient underwent catheter angiography. Digital subtraction angiography (DSA), using a Terumo stiff wire (Terumo , Leuven, Belgium) and a Sidewinder I catheter, confirmed total renal artery occlusion and further depicted multiple segmental artery stenoses consistent with the diagnosis of fibromuscular dysplasia (FMD; Figure 3 and 4). Interventional recanalization of the right renal artery and segmental arteries by means of percutaneous transluminal angioplasty (PTA) and additional stent implantation (due to a focal dissection which occurred in the course of main artery dilatation) was successful. Overall, 200ml of i.a. contrast agent (Visipaque , GE Healthcare, München, Germany) and 30mg of Buscopan i.v. (Boehringer Ingelheim, Ingelheim, Germany) for reduction of bowel movement and improvement of image quality were administered. A drop of serum creatinine from 1138 to 163 mol/l (normal range 50-80 mol/l) within the first week after intervention could be appreciated and the patient was spared from further hemodialysis. Antihypertensive therapy was continued with a single medication (Captopril) and follow-up (five years) has been uneventful with no signs of recurrent disease. A life-long medication of 100mg salicylic acid has been recommended.

DISCUSSION

Fibromuscular dysplasia (FMD) is a non-inflammatory, non-atherosclerotic disease characterized by muscular hyperplasia in one or more layers [1]. FMD is a rare disease of unknown cause and accounting for less than 10% of renal artery stenosis. It most often affects the middle and distal portion of the main renal artery or its segment branches in contrast to atherosclerotic disease, which tends to be located at the origin or proximal portion of the renal artery [5].

Depending on the predominantly affected layer varying histologic types can be differentiated. Whereas medial

fibroplasia is the most common cause (75-80%), intimal and adventitial fibroplasia is less often (less than 10% and less than 1%, respectively). Although any artery can be affected, FMD most often occurs in the renal (60-75%; 35% bilateral) and carotid arteries [3]. FMD typically occurs in young women (less than 35 years). Symptoms may include arterial hypertension with a sudden onset of high blood pressure or an epigastric bruit, transient ischemic attack (TIA) and stroke. Renal failure appears less often. FMD may be diagnosed incidentally or in the course of evaluation for hypertension [4]. Diagnosis can be made by duplex ultrasound, however catheter angiography still is the gold standard with characteristic findings such as focal stenosis or a "string of beads" appearance indicating multiple stenoses and aneurysms [5].

Treatment is indicated in patients with renal artery stenosis and hypertension or impairment of renal function. In asymptomatic patients watch and wait strategy is implemented. Treatment of choice in symptomatic patients is interventional recanalization by means of percutaneous transluminal angioplasty. Cure or improvement of renal function can be achieved in a high percentage of these patients. Primary stent implantation is recommended only in case of suboptimal PTA result or dissection. Surgery may be necessary in patients presenting with macroaneurysms or complex arterial disease [6, 7]. It has been demonstrated before that delayed revascularization of renal artery occlusion can be successful and is recommended for treatment of renovascular hypertension and acute renal failure [8]. Based on our case report we would like to emphasize on this, and encourage to attempt renal artery recanalization in symptomatic patients with FMD even in the later course of the disease as affected patients are typically young and as this condition usually has a good prognosis.

TEACHING POINT

Fibromuscular dysplasia (FMD) is a rare non-inflammatory, non-atherosclerotic disease characterized by muscular hyperplasia in one or more layers and accounting for less than 10% of renal artery stenosis. Characteristic imaging findings include focal stenosis or a "string of beads" appearance indicating multiple stenoses and aneurysms. Renal artery recanalization in symptomatic patients with FMD by means of percutaneous transluminal angioplasty should be attempted even in the later course of the disease as affected patients are typically young and as this condition usually has a good prognosis.

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FIGURES

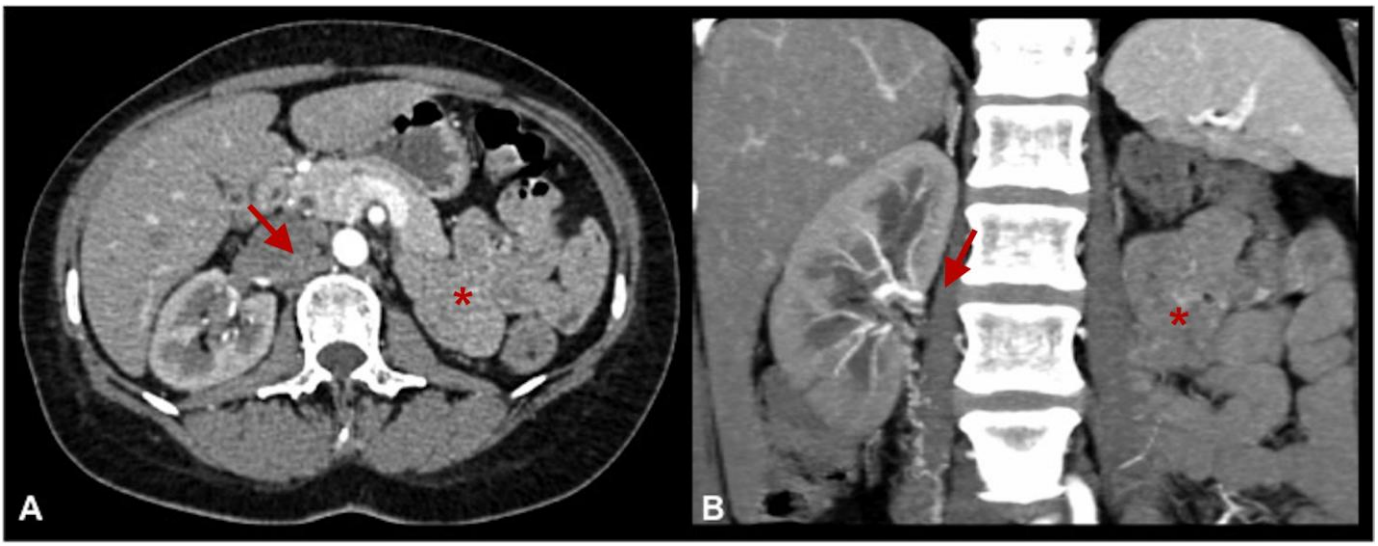

Figure 1: 49-year-old female patient with diagnosis of FMD. CTA (arterial phase; 120 kV, 1247 mAs) after injection of 120ml Ultravist (Bayer HealthCare, Leverkusen): On axial (a) and coronal (b) reformations no right renal artery is seen in terms of total vessel occlusion (red arrows). Parenchymal contrast is reduced with proper differentiation of renal medulla and cortex. Status post nephrectomy on the left side (red asterisk).

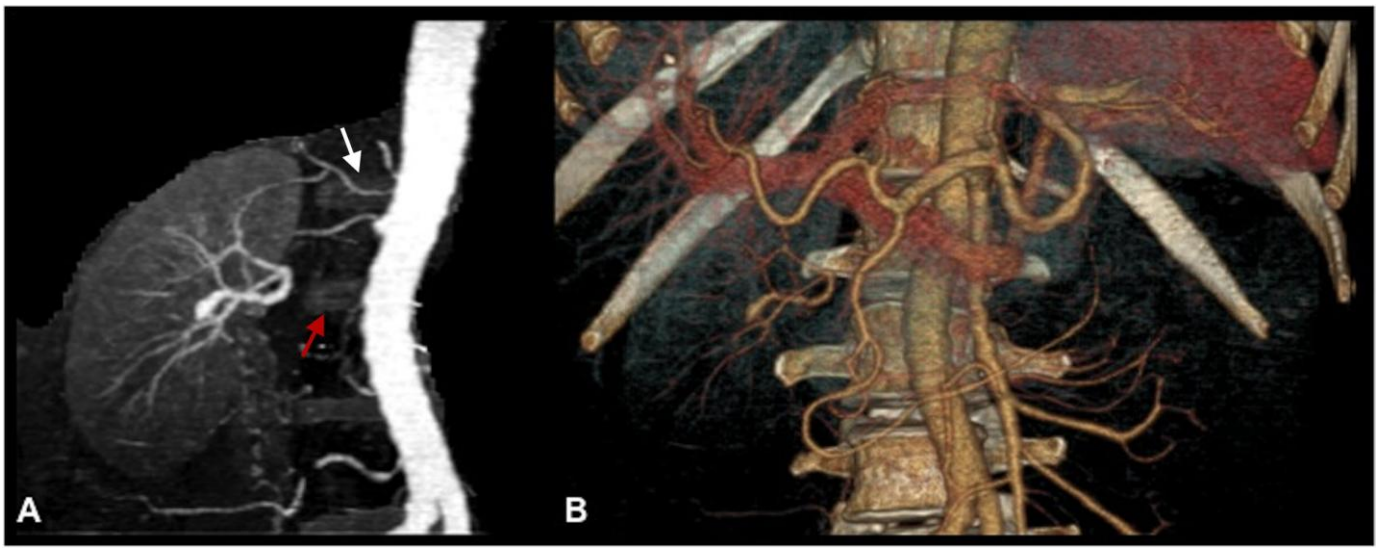

Figure 2: 49-year-old female patient with diagnosis of FMD. Coronal maximum intensity projection (MIP, a) and 3D volume rendered image (VR, b): Complete occlusion of the right renal artery (red arrow) with refilling of the segmental arteries via lumbar collaterals (white arrow) can be appreciated.

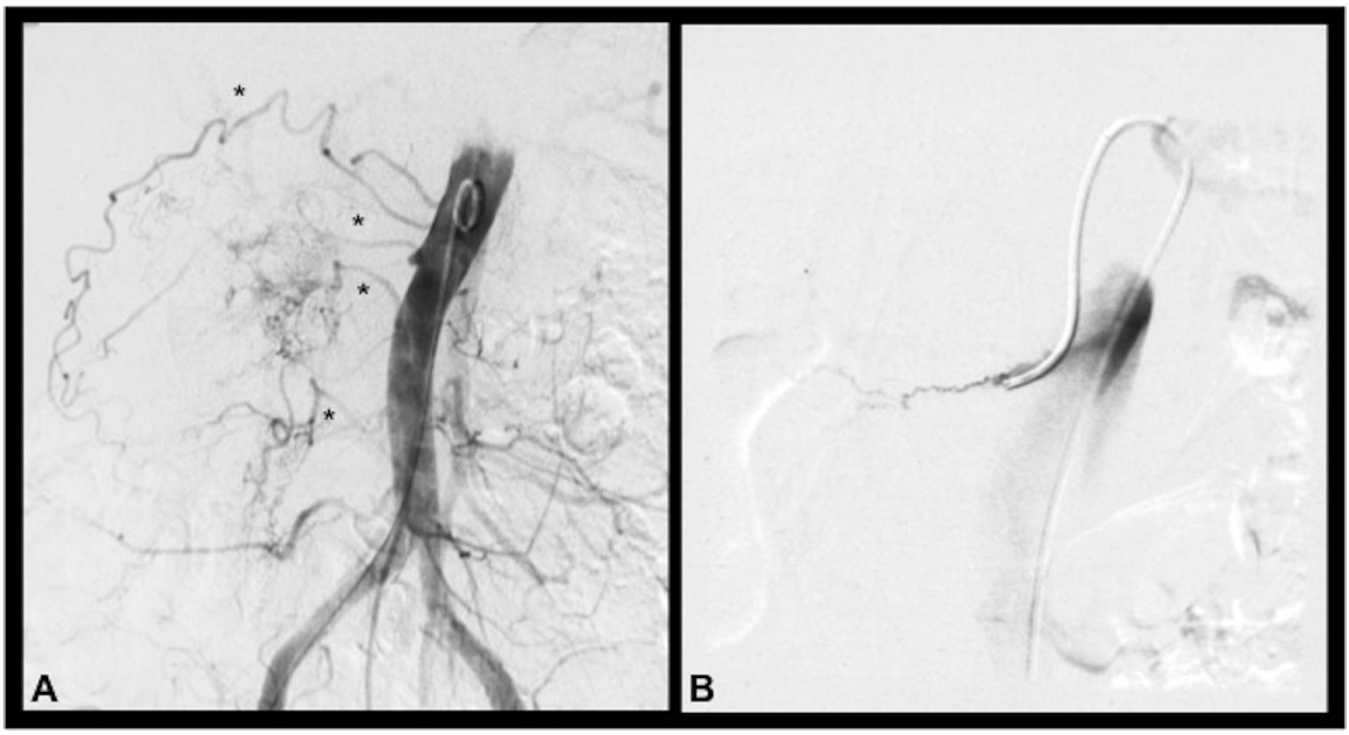

Figure 3: 49-year-old female patient with diagnosis of FMD. Digital subtraction angiography (DSA): Pigtail catheter placed in the aorta at the level above the renal arteries and demonstration of complete occlusion of the right renal artery. The right kidney is supplied by strong lumbar collaterals (*). Selective probing of the right renal artery using a Terumo stiff wire (Terumo , Leuven, Belgium) and a sidewinder II catheter, no parenchymal contrastation is seen (b).

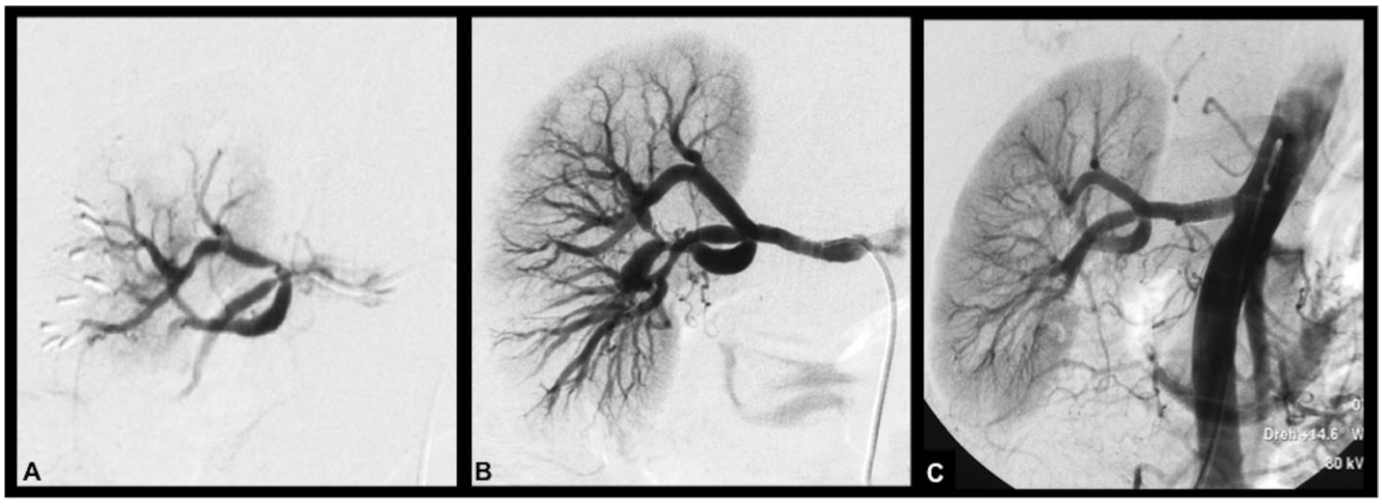

Figure 4: 49-year-old female patient with diagnosis of FMD. Placement of a cobra catheter further distal in the right renal artery depicts in addition multifocal stenoses of the segmental renal arteries (a). Step by step dilation of the main renal artery up to 7mm (using a 7mm Wanda balloon catheter) and segmental arteries using a 4mm Wanda balloon catheter and control series in between (b). Final control after additional placement of a Herkulink stent (7mm) in the proximal portion of the right renal artery because of an intima flap (not shown). No residual stenosis and homogeneous parenchymal contrastation can be appreciated (c).

Etiology	Cause unknown; non-inflammatory, non-atherosclerotic
Incidence	Rare (accounts for less then 10% of renal artery stenoses)
Gender ratio	W > m (3:1)
Age predilection	<35 years (most patients)
Risk factors	Not known; can be associated with fibromuscular dysplasia of other aortic branches or carotid arteries
Treatment	Watch and wait in asymptomatic patients; percutaneous transluminal angioplasty (PTA)
Prognosis	Usually good
Findings on imaging	Focal stenosis or "string of beads" appearance indicating multiple stenoses and aneurysms

Table 1. Summary table for fibromuscular dysplasia (FMD)

	US	**CT**	**MRI**	**Angiography**
FMD	Visible ridges, thickened artery wall, stenosis; color Doppler: damped appearance of arterial Doppler waveform, loss of early systolic peak	Discrepant renal sizes; evidence of prior renal infarct or global ischemia; CTA: multifocal stenoses	T1w: Atrophic kidney, corticomedullary thinning; T1w contrast enhanced: prolongation of cortical nephrographic phase and persistent corticomedullary differentiation; MRA: similar findings as CTA	Focal stenosis or "string of beads" appearance; most often affects the middle and distal portion of the main renal artery or its segment branches; uni- or bilateral
Atherosclerotic stenosis	Visualization of renal artery stenosis; damped appearance of arterial Doppler waveform, loss of early systolic peak; decrease in renal length over time	Visualization of atherosclerotic plaque; CTA: stenosis most often located in ostium or proximal 2cm of renal artery	T1w: decrease in renal size over time; T1w contrast enhanced: prolongation of cortical nephrographic phase and persistent corticomedullary differentiation MRA: Visualization of renal artery stenosis;	Tends to be located at the origin or proximal portion of the renal artery; eccentric / concentric stenosis; uni- or bilateral

Table 2. Differential diagnosis table for fibromuscular dysplasia (FMD)

ABBREVIATIONS

CTA = computed tomography angiography
DSA = digital subtraction angiography
FMD = fibromuscular dysplasia
MIP = maximum intensity projection
PTA = percutaneous transluminal angioplasty
TIA = transitory ischemic attack
VR = volume rendered

KEYWORDS

Fibromuscular dysplasia; renal artery occlusion; percutaneous transluminal angioplasty (PTA); string of beads