A young woman with high blood pressure on haemodialysis: it is never too late to evaluate hypertension

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Introduction

Once a patient is on haemodialysis long-term renal replacement therapy is usually needed because of irreversible nephron loss. Severe kidney disease accompanied by high blood pressure is often due to extensive glomerular injury. However, some patients do come off haemodialysis, making a careful clinical work-up mandatory to rule out curable disorders. Stenosis of the renal artery should be included into the differential diagnosis, since it is a reversible cause of severe hypertension and renal dysfunction. This important fact is underlined by the present case. We observed a young female with severe arterial hypertension and renal failure requiring haemodialysis. Haemodialysis could be stopped, once a dysplastic renal artery had been surgically revised and renal function and blood pressure had returned to normal limits.

Case report

A 28-year-old woman from Yugoslavia was admitted to the university hospital in Basel for dyspnea and headaches. In Yugoslavia she had been suffering from severe renal failure of unknown aetiology for the past 9 months. An underlying glomerulonephritis had been suspected. On admission the patient was in severe distress with pulmonary oedema and arterial hypertension (170/130 mmHg). She had pitting oedema of the lower extremities. The serum creatinine was 861 μmol/l and urea 395 μmol/l. The urinary sediment showed microscopic haematuria and proteinuria (dip stick ++). The patient was immediately placed on haemodialysis and aggressive ultrafiltration. The further work-up included an abdominal ultrasound and CT scan revealing a small right kidney (length 5.5 cm) and an 11-cm-long left kidney. A subsequent angiogram of the left renal artery demonstrated an obstruction 3 cm distal to its origin (Figure 1). Collateral capsular and lower pole vessels were well developed indicating that the disease process was not of acute onset. Blood flow in the small right kidney was slow, however, arterial abnormalities and stenoses were not observed. The occlusion of the left renal artery was surgically repaired by insertion of a saphenous vein graft, which was followed by recovery of urine output within minutes. A follow-up renal angiogram 9 days post-surgery revealed patency of the vascular graft and good arterial perfusion of the renal parenchyma (Figure 2). Intraparenchymal arteries were unremarkable. However, one lower pole branch immediately distal to the anastomotic site demonstrated segmental ‘beading’, indicative of dysplasia (Figure 2). Within 2 weeks following surgery the serum creatinine had decreased to 89 μmol/l. The blood pressure ranged between 120 and 130 systolic and 80–90 diastolic. Urinalysis was normal except for mild proteinuria (dip stick +). Follow-up 4 months post-surgery confirmed stable renal function and normal blood pressure. The cause of the small right kidney remained undetermined; possibly, hypoplasia is the underlying condition. A segment of the left renal artery was resected and a needle biopsy of the left kidney was performed during surgery. The specimens were examined histologically.

Histopathology

Histological examination of the renal artery showed dysplastic changes. The arterial wall architecture was
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Fig. 1. Pre-operative angiogram of the left renal artery showing an occlusion 3 cm distal to the origin (arrowheads). Collateral vessels are well-developed.

Fig. 2. Post-operative angiogram demonstrating good arterial filling centrally and peripherally. Only one branch of the renal artery supplying the lower pole displays ‘beading’ along a short segment (arrowheads). This angiographic pattern is consistent with fibromuscular dysplasia. The small intraparenchymal ‘aneurysms’ (arrows) are a consequence of the renal biopsy.

Fig. 3. Fibromuscular dysplasia of the renal artery, perimedial type, with marked deposition of collagen in the outer vessel wall (large asterisk). The lamina elastica interna (arrows) is segmentally missing (arrowheads). The intima consists of dense fibrous tissue (small asterisk). Elastica van Gieson stain; 20× original magnification.

severely altered by deposition of fibrous tissue, focally ‘replacing’ the outer half of the arterial wall, in particular the outer media (Figure 3). Higher power magnification revealed that smooth muscle cells were not eradicated, but highly atrophic and ‘strangulated’ by collagen (Figure 4). Segmentally, connective tissue appeared to infiltrate from the outer portion of the media into the inner layers, sparing only small islands of muscle. In segments with better preserved media, rudimentary elastic lamellae were located amidst smooth muscle bundles separating the outer ‘longitudinal’ from the inner ‘circumferential’ portion (Figure 4). Structural changes were also found along the internal elastic lamina with partial fragmentation and loss (Figures 3 and 5). Connective tissue expanded the intima which showed features of fibroplasia along the outer aspect (alpha smooth muscle actin positive myofibroblasts) and an organized thrombus with iron deposition and neo-vascularization centrally (Figures 3 and 5). The radiographically observed vascular occlusion was due to thrombus formation. Arterial wall thinning or aneurysms were not present.

The renal biopsy contained seven normal glomeruli lacking any light microscopical or immunohistochemical evidence of glomerulonephritis. Tubules, arterioles and small arteries were unremarkable. The interstitium revealed only mild patchy inflammatory fibrosis and minute foci of non-specific mononuclear cell infiltrates. Arcuate type arteries were not sampled.

Comment

Excessive persisting hypertension despite aggressive ultrafiltration prompted an intensive search for under-
lying renal vascular lesions which proved rewarding in this young lady who presented with (sub) acute renal failure. She had unilateral renal artery dysplasia with occlusion and became dialysis dependent because the contralateral kidney was hypoplastic.

Renal artery dysplasia mostly affects patients in their second or third decades, is more common among women and usually involves only one side. Renal veins remain unchanged [1]. The mechanisms leading to structural changes in the arterial wall are undetermined. It has been speculated that mechanical injury due to increased mobility of the kidney and stretching of the renal artery may lead to fibrous remodelling of the vessel wall [2,3]. However, this hypothesis seems
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Fig. 4. (a) Complex changes in the arterial wall: dense collagen is deposited in the outer media (large asterisk). Connective tissue seems to infiltrate into the inner media (fat arrow) and accumulates along the lamina elastica interna (small asterisks), which is focally ruptured (thin arrow). Rudimentary elastic lamellae are located along the inner aspect of the thickened intima (large arrowheads) as well as between smooth muscle bundles of the media (small arrowheads). The lumen is nearly completely occluded by an organized thrombus. Elastica van Gieson stain; 25× original magnification. (b) Highly atrophic smooth muscle cells (brown) in the outer media are surrounded by abundant collagen. Only small bundles of muscle in the inner media are spared (arrows). Immunohistochemistry, incubation to detect alpha-smooth muscle actin; 125× original magnification.

Fig. 5. (a) Higher power view illustrating changes of the inner vascular wall: the lamina elastica interna (arrow) is segmentally fragmented and ruptured (arrowheads). Dense connective tissue, containing numerous alpha smooth muscle actin positive cells, is deposited on the medial and intimal side (small asterisks) of the elastica. A thrombus occludes the lumen (large asterisk). Elastica van Gieson stain; 60× original magnification. (b) Neo-vascularization (dark brown) marks the area of the organized thrombus (large asterisk). Clearly separated is the thickened intima (small asterisks; elastica interna marked by arrowheads). The thickened intima has features of fibroplasia. Same view as (a); immunohistochemistry, incubation to detect endothelial cells (CD34); 80× original magnification.

Teaching Point

1. The possibility of renovascular lesions should be considered in young patients with apparent end-stage renal failure and severe hypertension despite aggressive ultrafiltration.

2. Unilateral arterial dysplasia may cause renal failure if it induces hypertension and parenchymal damage in the contralateral kidney, or—as in this case—when the contralateral kidney is hypoplastic.

3. Three categories of dysplastic arteries can be distinguished, but morphological subtyping is sometimes arbitrary.

References


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