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A complicated case of renal artery stenosis

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Abstract

We present a boy with bilateral renal artery stenosis who presented with severe hypertension and haemorrhagic stroke. The diagnostic workup along with a complication of eventual surgical intervention are demonstrated.

MeSH: Hypertension, Renovascular, Renal Artery Obstruction

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Introduction

Renal artery stenosis due to fibromuscular dysplasia is a potentially fatal condition, and may result in end-stage renal failure. We present a boy who had bilateral renal artery stenosis with severe hypertension, who presented with haemorrhagic stroke. Diagnostic investigations are shown and a complication of surgical treatment is demonstrated.

Patient

Our patient presented at 13 years of age with sudden onset of slurred speech and right sided weakness. He had had left sided Bell's palsy four years prior and had been referred for physiotherapy by his general practitioner. Admission examination showed a blood pressure of 225/153 mmHg and right sided hemiplegia and he was treated with a labetalol infusion. Ophthalmological review showed signs of acute on chronic rise in blood pressure.

CT scan brain showed an intracerebral haemorrhage of 3 by 3 by 1.5 cm in the left basal ganglia and insula, without midline displacement and with normal ventricles (Figure1).

Echocardiography showed moderate left ventricular hypertrophy and no coarctation. Renal diethylenetriaminepentaacetate (DTPA) scan with captopril showed decreased uptake in the left kidney and no tracer was evident in the collecting systems of both kidneys at 30 minutes implying flow filtration pressure due to bilateral renal artery stenosis and baseline DTPA confirmed the diagnosis. Digital subtraction angiography (DSA) showed bilateral renal artery stenosis (Figure 2).

Figure 1 Computerized tomography of the brain showing haemorrhage in the left basal ganglia and insula

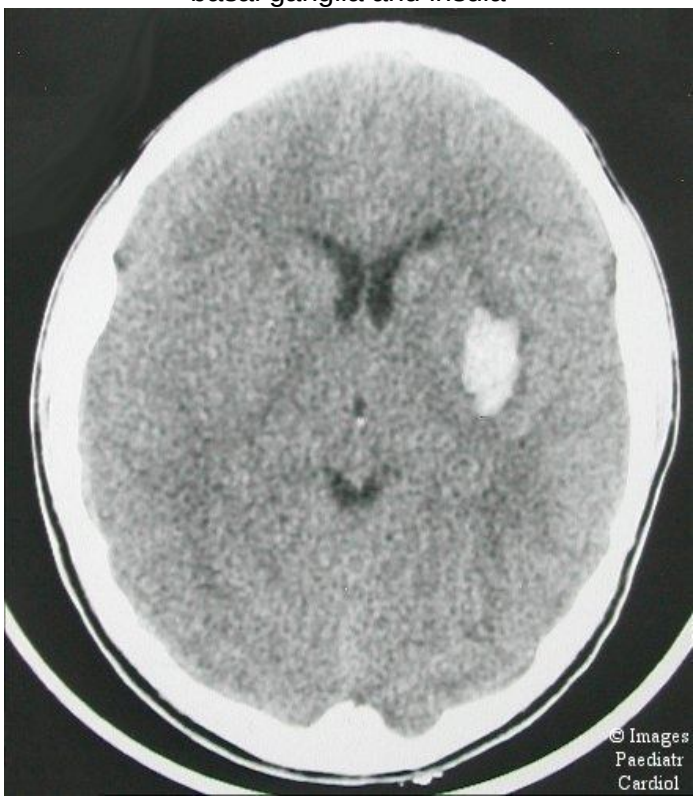
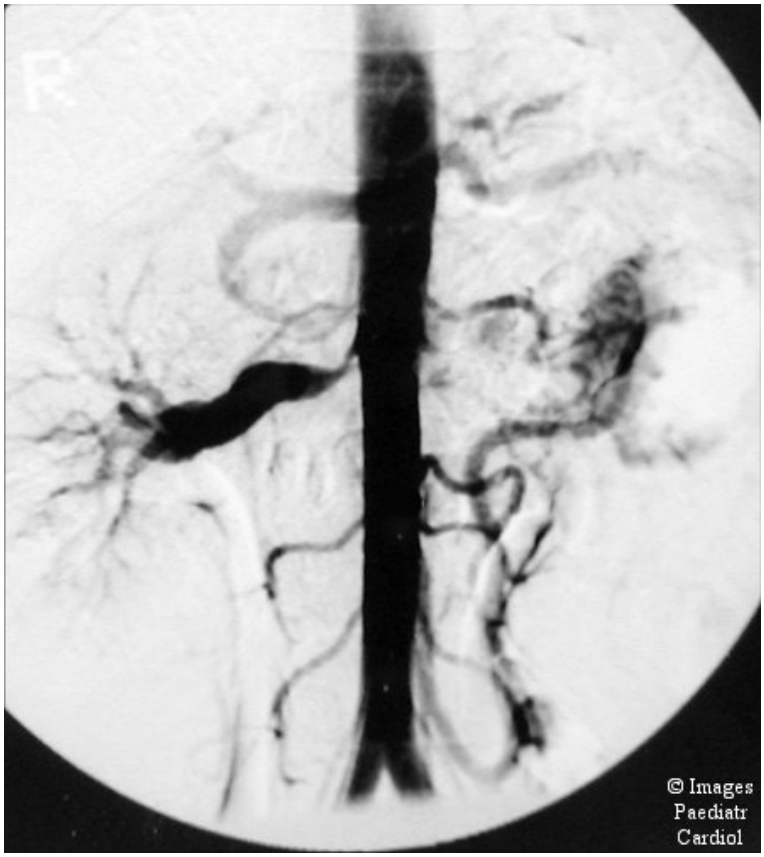


Figure 2 Digital subtraction aortogram showing poor filling of both renal arteries, worse on the left



Catheterisation of the renal arteries showed a filling defect in both kidneys which were supplied by collateral arteries probably via the cortical artery (Figures 3-6).

Figure 3 Digital subtraction angiography - cannulation of left renal artery showing a large filling defect.

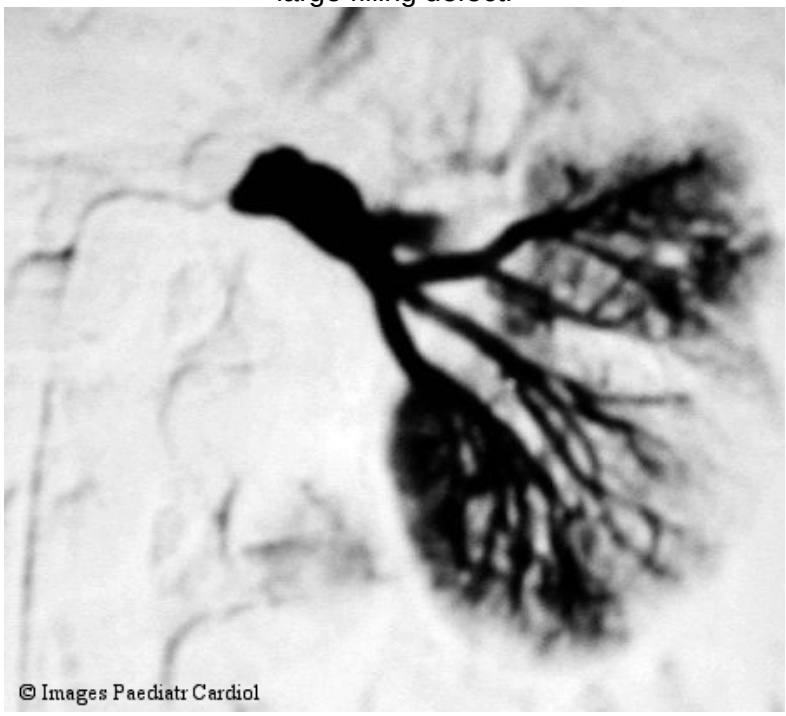


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Figure 4 Digital subtraction angiography - filling defect in left kidney supplied by a collateral.

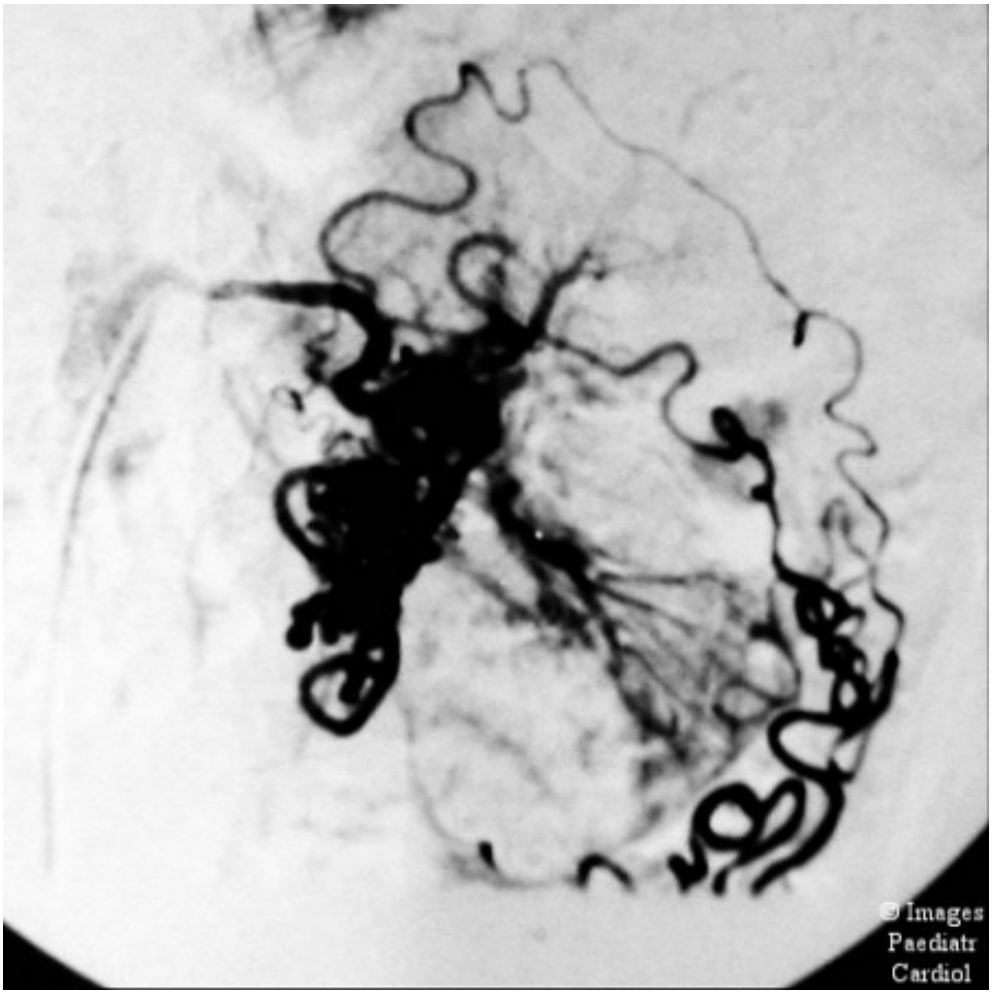


Figure 5 Digital subtraction angiogrpahy - cannulation of right renal artery also showing a large filling defect.

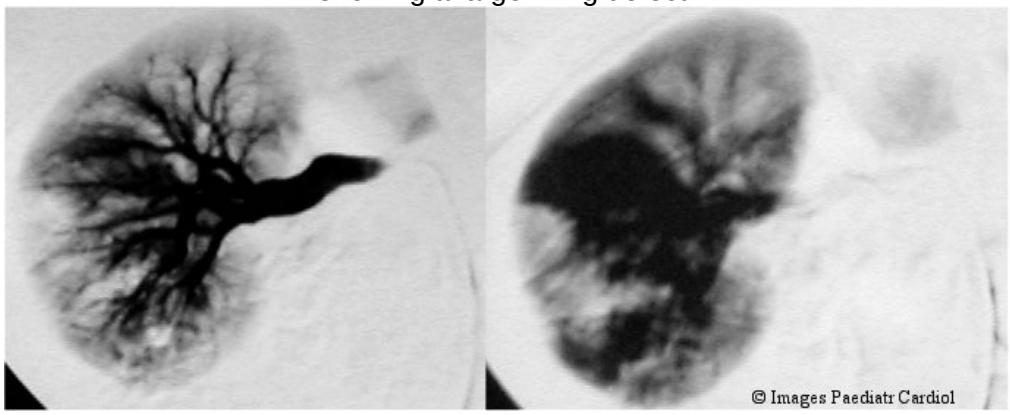
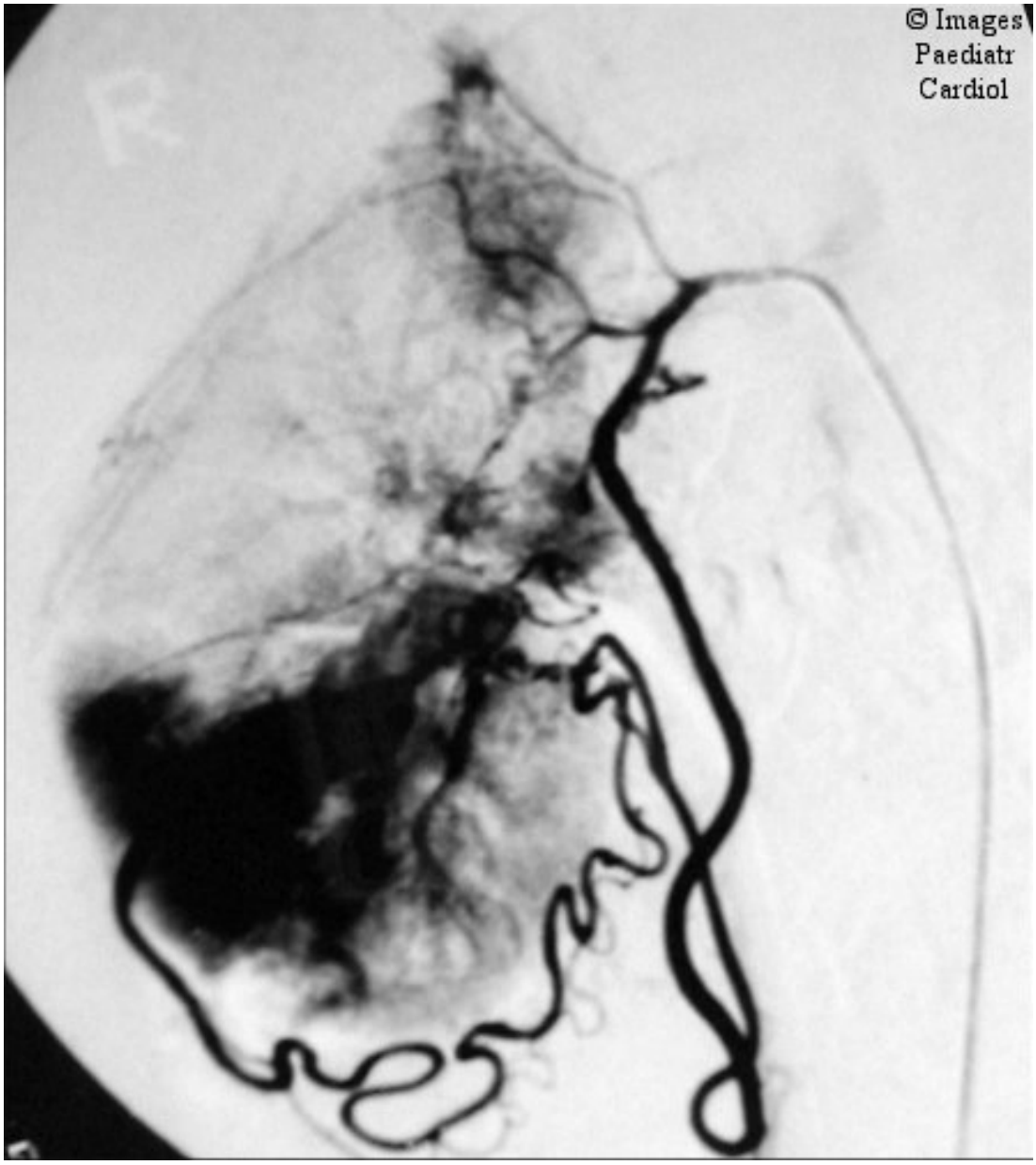


Figure 6 Digital subtraction angiography - filling defect in right kidney also supplied by a collateral.



Blood pressure was controlled at 150/60 mmHg with a combination of nifedipine, atenolol and hydrallazine. He was transferred to a tertiary referral centre two weeks later and balloon dilatation of the left renal artery stenosis was attempted. Seven months later, a left aortorenal Goretex bypass graft was performed, at the same tertiary centre following which, he was commenced on aspirin. However, the patient's blood pressure once again began to rise. He was reinvestigated and shown to have recurrent bilateral renal artery stenosis. He was once again transferred to a tertiary centre, two years after the first operation for further surgery. This consisted of a bifurcated Dacron trousers graft from the anterior surface of the aorta just below the inferior mesenteric artery to both renal arteries distal to the stenoses. Superior mesenteric artery occlusion was noted, but this was not causing any problems, and this was thought to be due to a large collateral from the splenic artery to the inferior mesenteric artery.

His creatinine slowly rose from 86umol/l to 216umol/l four years after the second operation. In March 2003, he presented with a short history of diarrhoea and vomiting

followed by progressive acute left iliac fossa pain radiating to the back with tenderness to percussion. Blood pressure had climbed to 180/105 mmHg. Urinalysis showed microscopic haematuria and proteinuria. DTPA showed an unperfused left kidney. DSA showed a normal right kidney but the left portion of the Goretex graft and left kidney were not visualised at all.

He was once again urgently transferred to a tertiary centre and a catheter was placed in the left portion of the Goretex graft, clot aspirated and tissue plasminogen activator (tPA) infused. Check angiography showed multiple filling defects within the main hilar vessels of the left kidney implying partial revascularisation. He was continued on heparin and a magnetic resonance angiogram showed a normal right kidney with good flow, and partial recanalisation of the left side of the Goretex graft and renal artery. However, only about half of the kidney demonstrated evidence of function, probably as a result of ischaemic damage. Further investigations failed to reveal a prothrombotic disorder. He continues on a combination of aspirin and warfarin, with normalisation of creatinine levels.

Discussion

Renovascular hypertension is the most common cause of secondary hypertension, and the commonest cause of renovascular hypertension is renal artery stenosis. Reduced kidney perfusion causes renal release of renin, triggering the angiotensin system, resulting in hypertension.

Renal artery stenosis is also increasingly recognised as an important cause of chronic renal insufficiency and end stage renal disease. Unilateral renal artery stenosis causes ischaemic nephropathy on the affected side and uncontrolled hypertension leads to hypertensive nephrosclerosis on the nonaffected side, therefore both uni- and bilateral renal artery stenosis may lead to progressive renal failure.

Renal artery stenosis may be caused by athersclerosis (90% of cases), or by congenital fibromuscular dysplasia due to thickening of the renal arterial walls (10% of cases). It occurs most commonly in young adults, particularly women aged 20 to 40 years, and rarely affects African Americans or Asians. In the older population, artherosclerosis is by far the most common aetiology. Fibromuscular dysplasia may be associated with William's syndrome, Alagille syndrome, neurofibromatosis type 1 and cutis laxa. Abdominal bruits are highly specific for this condition.

Investigations should include renal function tests including 24 hour urine collection to measure the degree of proteinuria, and urinalysis and microscopy to exclude glomerulonephritis. Serologic tests for systemic lupus erythematosus or other causes of vasculitis, and measurement of peripheral renin activity are also recommended.

Ultrasound is useful insofar as it may detect a difference in size between a normal and an ischaemic kidney. Doppler ultrasound of the renal arteries is also helpful. Radionuclide scanning particularly following a single dose of captopril is more useful especially in patients with normal renal function. Spiral CT angiography avoids arterial catheterisation and produces accurate images of renal artery anatomy but requires iodinated contrast material that may further impair renal function. Magnetic resonance angiography allows direct visualisation of the renal arteries without the use of iodinated contrast material and provides measurement of absolute blood flow rate, glomerular filtration rate and renal perfusion. Conventional arteriography remains the criterion standard for the confirmation and identification of renal artery occlusion.

Treatment involves medical control of hypertension and relief of stenosis by percutaneous transluminal balloon angioplasty (catheter), with or without stenting, or revascularization surgery.

Balloon angioplasty alone has a high restenosis rate. The recent use of stents to maintain lumen patency has dramatically increased both short and long term success rates for this modality of treatment. Resistant cases may need surgical angioplasty, which also has significant failure rates, and the site of the obstruction may have to be bypassed by a prosthetic tube, connecting the aorta to the renal artery distal to the stenosis. Bypass procedures include aortorenal, hepatorenal, splenorenal and ileorenal conduits constructed with autologous saphenous veins, autologous arteries or prosthetic material. Nephrectomy may occasionally be recommended for a unilaterally atrophied kidney.

Medical control of the hypertension involves the use of angiotensin-converting enzyme inhibitors, and angiotensin II antagonists in patients who are intolerant to the first class of drugs.

In view of the natural history of renovascular disease, patients require serial determinations of serum creatinine levels, adequate blood pressure control and serum potassium levels. Duplex ultrasound if available, allows regular radiologic progression follow-up.

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References

1. Khosla S. Renal artery stenosis: a review of therapeutic options. Minerva Cardioangiol. 2005;53:79–91.[PubMed: 15788982]