CASE REPORT

Unusual case of refractory hypertension: late presentation of the mid-aortic syndrome

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A 58 year old patient with refractory hypertension, chronic renal failure, and widespread arterial bruits is described. Investigations found hypoplasia of the major blood vessels, particularly the aorta, leading to low flow nephropathy.

A 58 year old female smoker was referred by her general practitioner for management of refractory hypertension. Her systolic blood pressure was persistently raised above 200 mm Hg. She did not give a history of claudication and the femoral pulses were palpable. Auscultation found a grade 3/6 aortic systolic murmur and widespread bruits in the carotid, renal, femoral, subclavian, and popliteal arteries. Preliminary biochemical investigations showed urea of 16 mmol/l and creatinine 283 mmol/l. Fasting cholesterol was 9.5 mmol/l. Creatinine clearance was 20 ml/h and a complete blood count showed normochromic normocytic anaemia. Ambulant 24 hour blood pressure monitoring indicated predominantly systolic hypertension with a mean systolic pressure of 170 mm Hg and mean diastolic pressure of 59 mm Hg. Carotid Doppler echocardiography showed only minor atheroma and a transthoracic echocardiogram showed left ventricular hypertrophy and aortic valve calcification with no significant gradient. Renal ultrasound showed that both the kidneys were small and hyperechoic with loss of corticomedullary differentiation suggesting chronic parenchymal renal disease.

At this point we were strongly suspicious of renovascular disease in view of widespread bruits, hypercholesterolaemia,

Figure 1 Magnetic resonance imaging of the aorta showing the hypoplastic section at the level of the kidneys.

Figure 2 Magnetic resonance imaging of the brachial vessels showing severe hypoplasia and extensive collateral vessel formation.

and smoking history. An isotope renogram showed bilateral reduction in the uptake but delayed perfusion of the left kidney. A renal angiogram conclusively showed no evidence of renal artery stenosis. The diagnosis was still not clear and magnetic resonance angiography of the aorta including the renal arteries was done. In the thorax the aortic arch had an unusually high configuration with a small lumen and was right sided. Magnetic resonance imaging of the heart and thorax showed major dilatation of the left heart with concentric hypertrophy. The subclavian, axillary, and brachial vessels were small. The abdominal aorta tapered down to a diameter of 6–7 mm at the supposed level of the renal artery (fig 1). The aorta also had an unusual position in that it was directly anterior to the midpoint of the vertebral bodies rather than to the left. Computed tomography of the abdomen with contrast again confirmed these features. The left renal artery was clearly visualised. The right renal artery was not visualised well but the right kidney was perfused normally. Below the renal arteries the abdominal aorta was small along with small common iliac and femoral arteries (fig 2). A renal biopsy was done to ascertain whether the renal damage was secondary to hypertension or caused by primary glomerulonephritis. Focal and segmental glomerulosclerosis was confirmed and focal fusion of foot processes was seen (a phenomenon secondary to low flow in this patient rather than primary glomerular pathology).

DISCUSSION

The renal impairment in this patient was secondary to low flow from aortic hypoplasia rather than renal artery stenosis,

hypertensive nephropathy, or primary glomerular disease. Clearly the aorta was hypoplastic and this, rather than luminal narrowing from atherosclerosis, was responsible for the low flow renal parenchymal damage leading to refractory secondary hypertension. The widespread bruits resulted from the hypoplastic brachial, axillary, and subclavian vessels.

Our patient appears to have a variant of mid-aortic syndrome with serious sequelae from renal damage. Quain first described hypoplasia of the distal aorta in 1847. However, the condition usually occurs early in life¹ (average age 9.5 years² and 19.5 years in two studies) rather than in the sixth decade as in our patient. For late presentations an atherosclerotic aetiology³ has been suggested but clearly this was not true in our patient. The principal clinical features are refractory hypertension, congestive cardiac failure, and arterial insufficiency of the lower extremities,³ which is often well tolerated. The distal thoracic and abdominal aorta is involved, often along with involvement of the visceral and renal arteries. The pathogenesis is controversial with arguments for congenital⁴ and inflammatory causes. It is postulated that decreased aortic flow during fetal development causes underdevelopment of the tunica media. Histopathological comparison of tissue from normal aortic arch and hypoplastic aorta has shown a paucity of cellular and supportive connective tissue constituents in this disease.⁵ This leads to diminished dimensions of the affected segment. There is a high incidence of single bifurcating lumbar arteries at the fourth and fifth lumbar vertebrae supporting the hypothesis that aortic hypoplasia may result from embryonic overfusion of dorsal aortas.⁶

Surgical treatment of this condition has been attempted in the younger subgroup. A variety of autogenous and prosthetic techniques have been described as one stage revascularisation procedures.⁷ Long term follow ups after such procedures have shown normal growth and development, preservation of renal function, and relief of myocardial insufficiency.⁷ The severity

of hypertension can be reduced in the vast majority, as shown in one study.² Percutaneous transluminal angioplasty has been used for localised stenosis of the abdominal aorta, but was found to be less reliable in relieving claudication than aortoiliac endarterectomy.⁸ Surgical procedures, whatever the method, are associated with low intraoperative mortality especially in the younger subgroup. Clearly a more diffuse involvement as in this patient militates against successful surgical treatment. Our patient required dialysis and is now on a renal failure regimen.

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