SHORT REPORT

Polycythaemia and hypertension caused by renal artery stenosis

M G Coulthard, W H Lamb

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A girl with failure to thrive and a haemoglobin of 140 g/l at 1.3 years died from a brain haemorrhage 2.5 years later. Renal artery stenosis had caused severe, chronic hypertension and increased erythropoietin secretion (haemoglobin 182 g/l). Blood pressure should be measured in all unwell children, including those failing to thrive.

A 1 year old girl whose height was on the 25th centile suffered a notable weight loss from the 50th to the 2nd centile after an acute diarrhoeal illness. However, her weight remained low, and she was investigated for failure to thrive at 1.3 years. Routine physical examination was unremarkable, but did not include measurement of her blood pressure. Urinalysis revealed only + proteinuria. Her haemoglobin was raised at 140 g/l but red cell morphology, white cell and platelet counts, and concentrations of vitamin B₁₂, folate, and ferritin were all normal, and this result was not pursued further. The following investigations were also all normal: plasma creatinine (45 µmol/l), urea, electrolytes, calcium and phosphate, liver function tests, immunoglobulins, endomysial antibodies, sweat sodium concentration, stool culture, and examination of stool for fat droplets.

Her growth subsequently improved, and by 3.8 years she was a developmentally normal child on the 25th centiles for both height and weight. She then became suddenly unwell, with confusion, slurred speech, and reduced tone, and rapidly became comatose. She had a systolic blood pressure of 260 mm Hg, and gross papilloedema. Her haemoglobin was very high at 182 g/l in the absence of dehydration, and in the presence of an erythropoietin concentration of 19 mU/ml (polycythaemia in the absence of chronic hypoxia would normally cause suppression of the erythropoietin to below the detection limit of 5 mU/ml). On admission, her plasma creatinine concentration was normal at 43 µmol/l, and urinalysis revealed blood +++ and protein ++. She promptly developed acute anuric renal failure, and was transferred to the regional paediatric nephrology and intensive care unit for management of her hypertension, acute renal failure, and encephalopathy. A renal tract ultrasound was unremarkable. A computerised tomographic scan of her head was normal, but she became decerebrate and died within 27 hours of the onset of her first symptoms.

Postmortem examination revealed the cause of death to be an acute brain stem haemorrhage which had ruptured from the medulla into the floor of the fourth ventricle. The left kidney was smaller than the right (weights 36 and 50 g respectively), and its artery was tightly stenosed throughout most of its length as a result of fibromuscular dysplasia. The left kidney was her only organ that was histologically normal, all the rest showing arteriosclerotic changes consistent with chronic exposure to hypertension (fig 1). The right kidney also showed early glomerular collapse with periglomerular fibro-

Figure 1 Cerebral arteriole showing marked hyalinosis.

sis, and areas of tubular atrophy with interstitial fibrosis. Her heart showed massive left ventricular hypertrophy, and weighed 135 g (expected weight 70 g); her aorta had prominent fatty streaks.

DISCUSSION

It is virtually certain that this girl was hypertensive as a result of her left renal artery stenosis when she presented with failure to thrive and a raised haemoglobin at 1.3 years. The outcome would have been very different if this had been identified. Her blood pressure could have been reduced medically, and her renin drive then controlled by balloon dilatation or surgery to the stenosed renal artery, or by left nephrectomy.

Failure to thrive is a well recognised feature of hypertension in childhood. However, as a tertiary referral unit we have never seen hypertension diagnosed as a result of screening for failure to thrive, though we have seen many children with severe hypertensive symptoms in whom weight loss had also been a major concern, often for many months. We strongly support the view that blood pressure measurement should be a routine part of the examination of any unwell child, including those presenting with failure to thrive.¹

Polycythaemia is a rare complication of renal artery stenosis in adults.^{2–4} It occurs because erythropoietin as well as renin may be released inappropriately from the affected kidney. However, the increase in erythropoietin release as a result of a fall in renal blood flow is only moderate compared to its release in response to anaemia.⁵ This is the first case we are aware of that has been described in childhood. Severe polycythaemia itself could lead to cerebrovascular sequelae because of an increased thrombosis risk secondary to hyperviscosity, but in this case our patient died of a brain stem bleed which is likely to have been a direct consequence of hypertension.

In summary, because the symptoms of hypertension are so varied and often non-specific, and the potential hazards of delay so severe, it is justified to measure the blood pressure in

all children who have an unexplained illness.¹ This includes measuring it routinely in children with failure to thrive. Though hypertension is relatively uncommon, early identification which allows the sequelae of uncontrolled hypertension to be avoided is of great value. Finally, renal artery stenosis should always be considered in a child with significant and unexplained polycythaemia, especially in the presence of hypertension.

Authors' affiliations

M G Coulthard, Department of Paediatric Nephrology, Royal Victoria Infirmary, Queen Victoria Road, Newcastle upon Tyne NE1 4LP, UK
W H Lamb, Bishop Auckland General Hospital, Bishop Auckland, County Durham DL14 6AD, UK

Correspondence to: Dr M G Coulthard, Department of Paediatric

Nephrology, Royal Victoria Infirmary, Queen Victoria Road, Newcastle upon Tyne NE1 4LP, UK; malcolm.coulthard@ncl.ac.uk

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