

Hypertension secondary to progressive vascular neurofibromatosis

A Kurien, P R John, D V Milford

Abstract

A 4 year old girl with neurofibromatosis type 1 (NF1) was referred for hypertension. An aortogram showed narrowing of the left main renal artery. An angiogram three and a half years later showed coarctation of the abdominal aorta. She underwent aortoplasty but the stenosis recurred. Vascular involvement in NF1 may be progressive and requires long term follow up.

(Arch Dis Child 1997;76:454-455)

Keywords: neurofibromatosis; coarctation; renal artery stenosis; hypertension

Vascular lesions are widely recognised to complicate neurofibromatosis type 1 (NF1) and include stenoses of the renal arteries, mesenteric artery, and aorta. We describe a child with hypertension complicating NF1 in whom renal artery stenosis was rapidly diagnosed but who later developed abdominal aortic coarctation. This case highlights the importance of measuring the blood pressure annually in children with NF1 and repeated meticulous catheter angiographic investigations in those who are hypertensive.

Case report

A 4 year old girl suffered a minor head injury while playing. Two hours later she developed progressive loss of function in the left arm and left leg and was admitted to the local hospital where a diagnosis of a right extradural haematoma was made. She was hypertensive and was noted to have approximately 20 café au lait spots over her abdomen, back, and arm and pigmentation in her right axilla. Her sister, brother, mother, and maternal grandmother also had café au lait spots.

On admission to this hospital she was well but her blood pressure was 147/100 in the left arm. Her left hemiparesis was resolving; no other abnormality was found. Renal function was normal, vanillylmandelic acid excretion was normal and echocardiography showed mild left ventricular hypertrophy and no structural abnormality. A renal ultrasound scan and dimercaptosuccinic acid scan showed two normal kidneys. Recumbent plasma renin samples were normal. A renal angiogram demonstrated a left renal artery narrowed at its origin from the abdominal aorta. The right renal, coeliac, and superior mesenteric arteries appeared normal.

Hypertension persisted despite treatment with lisinopril. Two weeks after discharge she was admitted to her local hospital with severe frontal headache, twitching of her right arm, and increasing drowsiness. She was transferred to our care. Blood pressure control was improved by captopril and labetalol and her neurological status returned to normal.

She was admitted 14 months after her original presentation because of poor blood pressure control and underwent balloon angioplasty of the left main renal artery stenosis. Her antihypertensive medication remained unchanged until repeat angiography 3.5 years after presentation that showed a 3 cm long stenosis of the abdominal aorta below the superior mesenteric artery at the level of the renal arteries. Lateral imaging (fig 1) demonstrated that the aorta was narrowed in the anterior-posterior diameter by 50% (normal aorta 5.9 mm, narrowed segment 2.7 mm). The main renal arteries appeared slightly irregular and their ostia were stenosed. After balloon aortoplasty, blood pressure was 103/31 above the stenosis and 95/48 below the stenosis. She remained stable for six months on a reduced dose of antihypertensives. Repeat angiography showed persisting coarctation and bilateral renal artery stenoses which were not corrected by further angioplasty.

Discussion

NF1 is inherited in an autosomal dominant manner and occurs with a frequency of approximately one in 3000 live births. It typically presents with café au lait spots, multiple neurofibromas, axillary freckling, and ocular Lisch nodules; the gene is located in the pericentric region of chromosome 17. NF2 presents with schwannomas of the acoustic nerve and the gene is localised to chromosome 22. Vascular abnormalities including renal, aortic, and mesenteric stenoses are a rare complication of NF1 and affect approximately 2%

Birmingham Children's Hospital,
Ladywood Middleway,
Birmingham B16 8ET:
Department of
Nephrology
A Kurien
D V Milford

Department of
Radiology
P R John

Correspondence to:
Dr Milford.

Accepted 29 January 1997

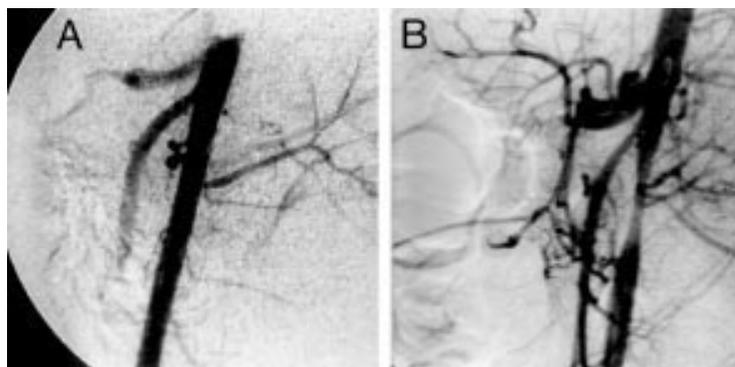


Figure 1 Oblique views of abdominal aorta showing no coarctation initially (A) but demonstrating marked anterior-posterior narrowing on repeat aortography (B).

of patients. Renal artery stenosis typically affects the origin of the vessel and may be associated with stenoses in other vessels. Case reports in the adult literature have also noted renal artery stenosis coexisting with coarctation of the abdominal¹ or thoracic aorta. Paediatric reports have similarly noted renal artery stenosis and abdominal and thoracic coarctation.^{2,3}

Greene *et al* described two histological appearances affecting vessels in neurofibromatosis, the first involving larger vessels (aorta, proximal renal arteries, carotid arteries) and the second mainly affecting smaller, more peripheral vessels.⁴ In the former the vessels are said to be surrounded by neurofibromatous or ganglioneuromatous tissue in the adventitia. There is intimal proliferation, thinning of the media, and fragmentation of the elastic tissue. In the latter there are no associated neural malformations and Greene *et al* speculated the findings were compatible with a dysplasia of the small vessels. Histologically there are multiple nodules in the affected vessels and ultrastructural examination demonstrates these are nodular aggregates of smooth muscle. However, these two appearances may coexist and in some instances the vascular abnormality may be widespread and involve arteries, arterioles, and veins in the kidneys.⁵

In their thorough review of the literature, Schurch *et al* noted that of 45 patients investigated, 13 had abdominal aortic coarctation and in 10 this occurred with renal artery stenosis which was either unilateral or bilateral and in some mesenteric or coeliac artery stenosis was

also noted.² Unlike fibromuscular dysplasia, which is usually isolated and involves the distal two thirds of the renal artery, the vascular involvement in neurofibromatosis may be proximal in 50% of cases and be associated with aortic stenosis.

Our case demonstrates the importance of annual reviews of children with NF1 and emphasises the need for blood pressure measurement. In addition, repeated screening for stenosis in the major branch vessels of the aorta in children with NF1 and renal artery stenosis is required, as in our case the aortic coarctation developed after the first angiogram. The nature of the vascular involvement in neurofibromatosis has tended to prompt suggestions that surgery is unlikely to resolve the problem, especially as the lesions may be diffuse. Angioplasty was of benefit in temporarily resolving the aortic stenosis but unlike previous reports⁶ the stenosis recurred.

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