An unusual cause of fitting in a 20 year old girl

DJ Beckett, A Malins, ML Watson & CA Kelly

Abstract

Native renal artery stenosis resulting in hypertensive encephalopathy is exceptionally rare, with only 3 previous case reports in adults. We report such a case in a previously well 20 year old female.

MeSH keywords

Renovascular, hypertensive encephalopathy

Case History

A 20 year old girl with a past medical history only of migraines and mild asthma presented with a witnessed tonic-clonic seizure. She had been unwell for the previous two weeks with diarrhoea, vomiting and headaches. Her only medications were the oral contraceptive pill, which she had been taking for the past year, and co-codamol for the headache. She was a non-smoker.

On arrival at hospital she was noted to have a low grade pyrexia, with a tachycardia of 120 beats/min and blood pressure recorded on manual sphygmomanometer as 220/140 mmHg. Following the seizure she was disorientated to time and place, but neurological examination, including fundoscopy, was normal. ECG revealed a sinus tachycardia and chest x-ray was normal. Blood tests on admission revealed hypokalaemia (K^+ 2.5mmol/l) with normal renal function. Urinalysis revealed an inappropriate kaliuresis (K^+ 39mmol/l) but no proteinuria. Following two further seizures she underwent CT brain scan and lumbar puncture, which were entirely normal. She was transferred to the high dependency unit for close monitoring.

She became increasingly confused, agitated and restless requiring multiple doses of intravenous benzodiazepines. Despite this her blood pressure was persistently elevated up to 240/160. Further clinical examination did not demonstrate any radial-radial or radial-femoral pulse delay. A transthoracic echocardiogram was completely normal with no left ventricular hypertrophy. A provisional diagnosis was made of accelerated hypertension causing fitting as a result of hypertensive encephalopathy.

She was initially treated with intravenous labetalol, which resulted in gradual lowering of her blood pressure over the following 48 hours, with associated improvement in her mental status. At this point she was converted to oral atenolol (50mg) and amlodipine (10mg), along with spironolactone (100mg).

Further investigations included a normal low dose dexamethasone suppression test with undetectable levels of ACTH. Her 24 hour urinary normetadrenaline level was 4.2 μmol/24hr (normal 0.4-3.4), consistent with physiological stress but not suggestive of phaeochromocytoma. Plasma renin activity was substantially elevated at 29.8ng/ml/hr (normal 0.5-2.6) with high normal aldosterone 424pmol/l (normal 30-440).

Magnetic resonance imaging of her abdomen revealed normal adrenal glands, but a marked discrepancy in the size of her kidneys (right 12cms, left 8cms). Subsequent Magnetic resonance angiography (MRA) demonstrated a high-grade obstruction 1cm from the left renal ostium (Fig 1). Formal renal angiogram confirmed the presence of a left renal artery stump with no distal filling. Scanty collaterals were seen with no nephrogram except at the upper pole (Fig 2). Renal isotope scanning demonstrated that the left kidney contributed only 3% of total renal function.

After two weeks she was discharged home on atenolol 50mg and amlodipine 10mg. Renal artery stenting, normally the treatment of choice in such cases, had been deemed inappropriate due to complete occlusion of the affected vessel, and the non-functionality of the culprit kidney. She therefore underwent laparoscopic nephrectomy. Following this procedure, she was initially seen on a monthly out-patient basis and her blood pressure has been no higher than 120/80, off all anti-hypertensive medication. The oral contraceptive pill has been discontinued.

Retrospective examination of her medical records revealed that routine blood pressure checks had always been normal (110/60 and 120/80 one and five years previously). The lack of end-organ damage is further evidence of a rapid onset of hypertension in this case.

DJ Beckett
MRCP
High Dependency Unit,
Royal Infirmary of Edinburgh,
51 Little France Crescent,
Old Dalkeith Road,
Edinburgh,
EH16 4SA

A Malins
MRCP

ML Watson
FRCP

CA Kelly
FRCP

Correspondence:
Daniel Beckett
E-mail :
dbeckett@doctors.org.uk
25 Albert Road,
Falkirk,
FK1 5LS
Tel: 07958 403471

An unusual cause of fitting in a 20 year old girl

Pathological analysis revealed a histologically normal renal artery with a grossly atrophic kidney. Despite the negative histology and lack of typical findings on angiogram, the likeliest aetiology of renal artery stenosis in a female of this age remains fibromuscular dysplasia. The fact that the operation was undertaken laparascopically may have made identification of the affected segment of artery difficult, leading to its incomplete excision. Furthermore there was no clinical or laboratory evidence for a more widespread vasculitic process, and she had no risk factors for atherosclerotic disease. In view of this she will require annual MRA of her remaining renal artery, so that prophylactic stenting could be offered if necessary.

Discussion

Hypertension is well recognised as a secondary phenomenon following seizures, and is the result of a surge of adrenergic hormones. However the existence of an aetiological link between hypertension and adult onset seizures is a subject of some controversy. One study[1] described an increased incidence of hypertension in adult patients presenting with a first seizure, compared with control subjects, and postulated sub-clinical cerebrovascular disease as a mechanism, but this has since been refuted.[2,3]

Accelerated hypertension as a cause for seizure is very rare. In a series of 247 patients presenting to hospital with a first seizure, none were attributable to accelerated hypertension.[4] However in patients with hypertensive encephalopathy, seizures are a common feature, occurring in up to 25% of cases.[5]

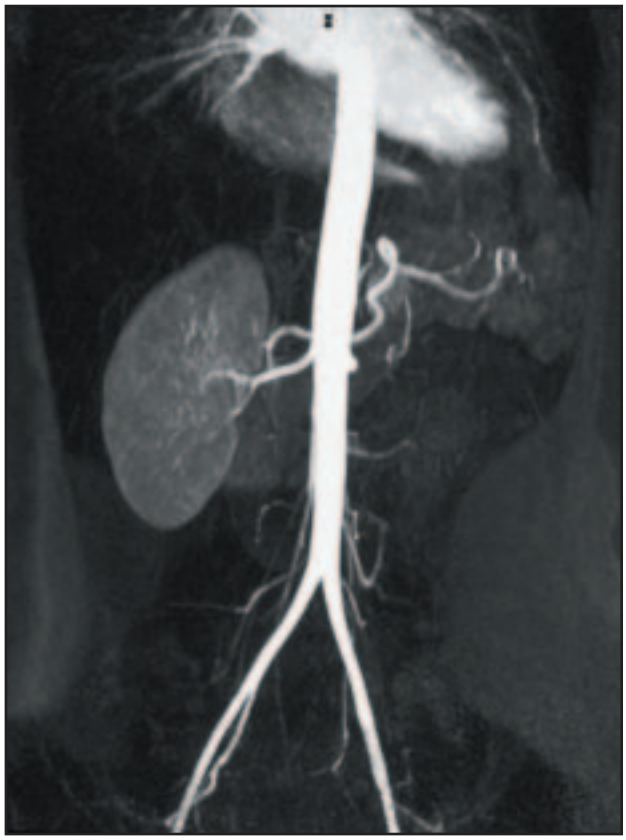

Figure 1. Magnetic Resonance Angiogram demonstrating a high grade obstruction 1cm from the left renal ostium

Renal artery stenosis causing hypertensive encephalopathy is well recognised in renal transplant recipients[6] and in children.[7] However, there are only three previous case reports of native renal artery stenosis causing hypertensive encephalopathy in adults.[8,9] In one of these cases the clinical picture was complicated further by a small sub-arachnoid haemorrhage.[9] All the previous cases were described in young females (16, 19 and 27 years of age). In two of the previous cases, as in this case, the patients were taking the oral contraceptive pill, and both were noted to have normal blood pressure recordings in the six months prior to admission. All of these cases were found to be due to fibromuscular dysplasia, and were treated with either renal artery stenting, or anti-hypertensive medication.

Fibromuscular dysplasia accounts for less than 10% of cases of renal artery stenosis,[10] the vast majority being due to atherosclerotic disease. Other causes include Takayasu's arteritis, polyarteritis nodosa and solitary renal myofibromatosis. Fibromuscular dysplasia is a non-inflammatory, non-atherosclerotic angiopathy that affects medium-sized arteries predominantly in young women of childbearing age. Renal involvement occurs in 60-75% and is well recognised to cause refractory hypertension. The second commonest site affected is the cerebral vasculature (25-30% of patients), and other affected areas may include the viscera, limbs and coronary arteries. In about a quarter of cases, disease is found in more than one arterial region.

The aetiology of Fibromuscular dysplasia is not well understood. There are several reports of familial occurrences, suggesting a genetic basis. The greater prevalence in women suggests hormonal factors may also be important. One study suggested that smoking may be important but found no link with use of the oral contraceptive pill.[11]

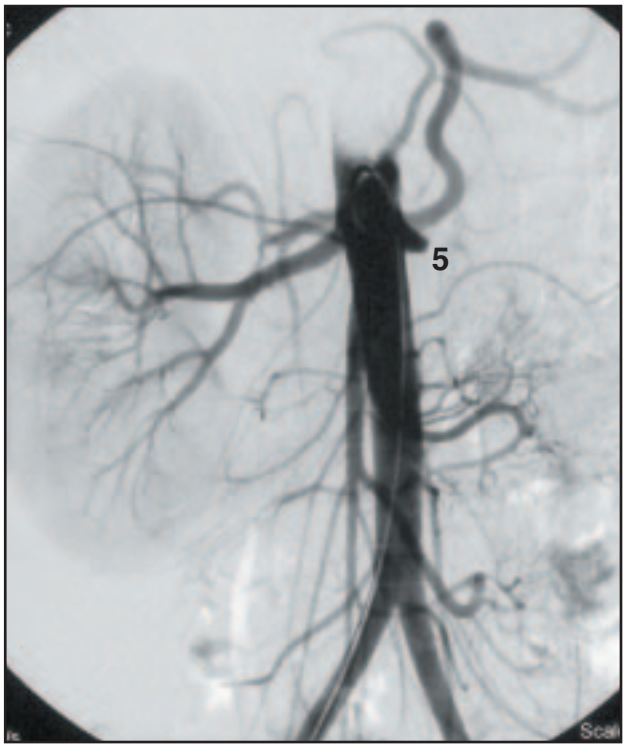

Figure 2. Renal angiogram showing left renal stump (5)

The gold standard treatment remains open surgical revascularisation. However this is associated with considerable morbidity and mortality. Renal artery angioplasty was first carried out in 1978, and stenting followed in the early 1990s. A recent trial of percutaneous therapy specifically for fibromuscular dysplasia demonstrated a 95% primary patency rate with improved or cured hypertension in 79% of patients overall.[12]

In conclusion, hypertension is a well recognised consequence of seizures, but in exceptional cases may be the causative factor. Although rare, renal artery stenosis should be considered as a cause of hypertensive encephalopathy. The four cases reported to date describe rapid onset hypertension in young females, three of whom were taking the oral contraceptive pill, although a causative link remains to be proven.

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