Posterior reversible encephalopathy syndrome in (low renin) essential hypertension

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Physicians still encounter hypertensive crises, which merit rapid diagnosis and treatment. Posterior reversible encephalopathy syndrome (PRES) is usually associated with acute changes in blood pressure in patients with complex medical problems. We present the case of a patient with PRES as a presenting feature of essential hypertension.

CASE REPORT

A 43-year-old woman presented with a two-month history of intermittent throbbing, mid-frontal headache lasting from hours to days at a time. On admission the headache had worsened and was associated with nausea and photophobia. In casualty she had two partial seizures, with left arm abduction and facial twitching, followed by secondary generalization. She was not known to be hypertensive and there was no other past medical history. On examination she was drowsy, apyrexial and hypertensive, with a blood pressure of 180/100 mmHg. Fundoscopy was normal with no hypertensive changes. Apart from mild unsteadiness there were no focal neurological signs.

The working diagnosis was of encephalitis and she was initially treated with phenytoin and acyclovir. The persistent hypertension was treated with 4 mg slow-release doxazosin. Contrast-enhanced head computed tomography (CT) was normal. Cerebrospinal fluid (CSF) was of normal pressure, acellular and sterile with normal glucose and protein. Herpes simplex virus and varicella-zoster virus DNA and enterovirus RNA were absent. CSF oligoclonal bands were negative. An electroencephalogram (EEG) was encephalopathic. There was a microcytic anaemia (Hb 10.8 g/dL; Hb E trait), a leukocytosis of $12.5 \times 10^9/\text{L}$ and a thrombocytosis of $609 \times 10^9/\text{L}$. Urea and electrolytes were normal. There were no casts, cells or protein in the urine. Thyroid function tests, calcium and magnesium were normal and an autoantibody profile was negative.

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(a)

(b)

Figure 1 (a) MRI T₂ FLAIR brain scan showing high signal within the occipitoparietal deep white matter, also involving the grey matter and spreading anteriorly. (b) MRI T₂ FLAIR brain scan showing complete resolution at six months

Magnetic resonance imaging (MRI) of the brain showed patent venous sinuses with appearances typical of PRES (Figure 1a).¹ The hypertension was controlled with a combination of doxazosin and amlodipine (in order to facilitate investigation of the renin–angiotensin–aldosterone axis). She was treated with oral agents, as her blood pressure was not initially felt to be significantly elevated. With improvement of the hypertension her symptoms and MRI appearance (Figure 1b) resolved. Subsequently, blood pressure control remained excellent and doxazosin was discontinued. Further assessment revealed she had suppressed plasma renin activity with normal aldosterone levels and no evidence of adrenal pathology on triple phase CT scanning. Exhaustive investigation failed to disclose any secondary causes of hypertension.

DISCUSSION

PRES, which is frequently associated with hypertension, usually occurs in patients with complex medical problems. Early diagnosis with prompt treatment of hypertension

often results in gradual resolution of the clinical and radiographic findings.¹ Common precipitants, singly or in combination, include acute elevations in blood pressure (e.g. hypertensive encephalopathy and eclampsia), uraemia and treatment with immunosuppressive drugs (e.g. cyclosporin A).^{1,2} Headache, altered alertness, seizures, vomiting and visual disturbances often occur. Seizures may be early, often of focal onset with secondary generalization. Limb weakness and ataxia may develop.¹ Occipitoparietal cortical and subcortical vasogenic oedema, typically seen on MRI T₂ FLAIR, is characteristic of PRES.^{1,3} The calcarine and paramedian regions of the occipital lobes are usually spared, important in differentiating the imaging appearances from basilar artery occlusion.¹

PRES is more likely to occur in previously normotensive individuals who develop acute hypertension.^{2,4,5} A sudden rise rather than absolute blood pressure may be the major determinant of the subsequent pathology. Propensity for encephalopathy, with an associated cerebral hyperperfusion syndrome and vasogenic oedema, occurs once the mean arterial blood pressure has exceeded the upper limit of cerebral autoregulation—less than 120 mmHg in previously normotensive individuals. Individuals with chronic hypertension are less prone to develop this disorder, as their upper limit of autoregulation is reset at higher levels of mean arterial pressure (up to 150 mmHg).⁶ Disturbed autoregulation, particularly of the posterior circulation, which is known to be more sparsely innervated with sympathetic fibres, may account for the characteristic radiographic features of posterior cerebral oedema.⁷

Hypertensive emergencies are still encountered by physicians. Delaying treatment may result in progression with cerebral infarction and/or haemorrhage and long-term

neurological disability.⁸ It is important to be aware that diagnosis of hypertensive emergencies should not be solely based upon absolute blood pressure values and that a sudden rise in blood pressure can be responsible for acute end-organ damage in previously normotensive individuals.⁶

Competing interests None declared.

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