

CASE REPORTS

The right scan at the right time: reversible posterior leukoencephalopathy syndrome mimicking bilateral occipital lobe infarcts

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Abstract

Reversible Posterior Leukoencephalopathy Syndrome (RPLS) is a relatively recently characterised neurological syndrome, first described by Hinchey et al in 1996, with neuroimaging findings of reversible vasogenic subcortical oedema. The clinical presentation can vary, is often non-specific but can include headache, global encephalopathy, seizures and visual disturbances. In this article we present such a case in a 79 year old woman, followed by a discussion of the typical presentations, associations, pathomechanisms and neuroimaging findings.

Keywords: RPLS, elderly, PRES, stroke

A 79-year-old woman with a previous medical history of breast cancer, treated hypertension, toxic multinodular goitre and myeloma was admitted with acute epiglottitis requiring urgent sedation, endotracheal intubation and immediate transfer to the intensive care unit. Treatment included intravenous cefotaxime, flucloxacillin and dexamethasone, with blood cultures growing a β -haemolytic streptococcus. She developed hypertension and acute renal failure, with hypomagnesaemia (0.57 mmol/l) and hypokalaemia (2.8 mmol/l) taking 4 days to normalise. She was extubated and noted to be confused with a fluctuating conscious level and focal motor seizures which ceased following intravenous lorazepam. A contrast-enhanced cranial CT showed cerebral atrophy, while lumbar puncture excluded infection and subarachnoid haemorrhage. Despite treatment with antihypertensives, her blood pressure peaked on Day 7 (210/120 mmHg) with an ensuing flaccid tetraparesis. Urgent electroencephalogram demonstrated generalised excess of slow-wave activity only, while repeat cranial CT showed bilateral occipital pole and cerebellar hypoattenuation consistent with bilateral posterior cerebral artery (PCA) infarcts. She was transferred to a medical ward on Day 12 for supportive care. However, the following day she became spontaneously normotensive (despite not receiving antihypertensives for several days), alert and orientated with dramatic improvement in her limb weakness. Alternative diagnoses were not considered until cranial MRI

(Figure 1.) demonstrated non-haemorrhagic signal change involving the posterior part of each cerebral hemisphere mainly within the white matter. Diffusion-weighted (DWI) and fluid attenuation inversion recovery (FLAIR) images showed these changes as hyperintensities, with apparent diffusion coefficient (ADC) mapping indicating the absence of restricted water diffusion otherwise seen in acute ischaemia or infarction. These appearances were consistent with reversible posterior leukoencephalopathy syndrome (RPLS) and the patient continued to improve. After rehabilitation, she was discharged home independent on Day 25. Complete resolution of the white-matter changes on repeat MRI was demonstrated 2 months later.

Discussion

RPLS is characterised by headache, seizures, visual disturbances and altered mental functioning occurring in acutely ill hospitalised patients [1], particularly those with uncontrolled hypertension, renal failure or immunosuppression [2, 3].

The exact pathomechanisms of RPLS are postulated to be similar to that of hypertensive encephalopathy [4, 5] though hypertension is not always present [6]. Endothelial dysfunction secondary to drugs or increases in blood pressure/volume with resulting dysregulation of cerebral

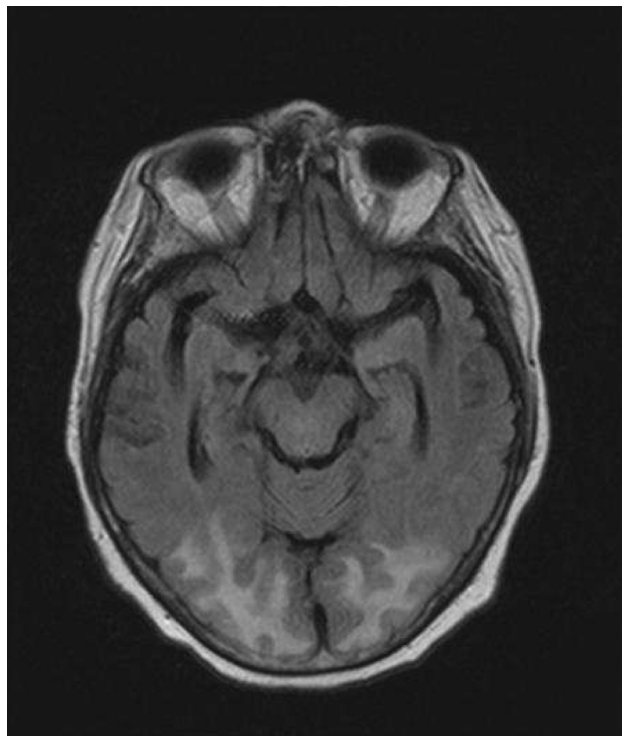


Figure 1. Cranial MRI (T2 FLAIR): bilaterally symmetrical, predominantly white matter hyperintensities in the occipital lobes.

blood flow is thought to cause dilatation of cerebral arterioles, breakdown of endothelial tight junctions and plasma leakage [1]. The vertebrobasilar territory is thought to be particularly vulnerable to disruption of autoregulatory mechanisms due to its relative paucity of sympathetic innervations, leading to over-perfusion and oedema [5] which is predominantly vasogenic and reversible, but may convert to cytotoxic in certain patients [7], especially if appropriate treatment is delayed. While anti-epileptic medications are essential in the acute treatment of seizures, most patients do not progress to chronic epilepsy [8].

Neuroimaging in cases of RPLS is classically associated with subcortical white-matter vasogenic oedema causing high-intensity signal principally in the posterior regions on T2-weighted MRI. DWI and ADC mapping are useful in differentiating vasogenic from cytotoxic oedema while distinction from bilateral PCA infarction, the 'top of the basilar' syndrome [9], is evident by the relative sparing of the cortical structures, with resolution of lesions on follow-up studies.

As the name RPLS suggests, reversibility of the neuroimaging lesions is an essential feature and though there remains much conjecture as to the timing of repeat brain

imaging, resolution probably occurs between 5 days and several weeks [8]. Lesions have been reported as progressing to irreversible leucomalacia [2, 8, 10] although these probably represented pre-existing ischaemic disease. Recognition of the syndrome is critical to ensure prompt intervention which includes removal of any potential triggers such as cytotoxics and aggressive management of hypertension and seizures.

While the majority of patients with RPLS are under 60 years of age, our patient is the oldest reported to date and her case highlights how appropriate neuroimaging is critical for the prompt recognition of this syndrome and its differentiation from stroke.

Conflicts of interest

There are no conflicts of interest to declare.

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