CASE REPORT

Posterior reversible encephalopathy syndrome: A case of unusual diffusion-weighted MR images

Encéphalopathie postérieure réversible : un cas d’aspect inhabituel en IRM de diffusion

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Summary

Posterior reversible encephalopathy (PRES) represents an uncommon entity related to multiple pathologies, the most common of which is hypertensive crisis. PRES is classically characterized as symmetrical parieto-occipital edema, but may affect other areas of the brain. Diffusion-weighted magnetic resonance imaging (DWI) is important for differentiating between vasogenic and cytotoxic edema. We present here the case of a 43-year-old woman, known to suffer from arterial hypertension and severe renal failure, who developed PRES with restricted apparent diffusion coefficients (ADC) in various cerebral areas, suggesting irreversible tissue damage. Nevertheless, follow-up cranial MRI revealed complete remission, indicating that restricted diffusion does not always lead to cell death in this pathology. The underlying pathophysiological mechanism is not well understood. Such reversibility of diffusion anomalies has already been reported with transient ischemia, vasospasm after subarachnoid hemorrhage and epilepsy but, to our knowledge, never before in PRES.

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MOTS CLÉS

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Introduction

Posterior reversible encephalopathy syndrome (PRES) refers to a complex syndrome typically characterized by headache, altered mental functioning, visual disturbance and seizure. The most common causes of PRES are hypertensive encephalopathy, uremic encephalopathy, cyclosporine A neurotoxicity and preeclampsia or eclampsia [1]. Neuroimaging features in PRES include symmetrical diffuse or focal hyperintensity on T2-weighted or fluid-attenuated inversion recovery (FLAIR) images, predominantly in the supratentorial white matter and especially in the posterior circulation territories [2]. Previous studies [1,3–5] have shown that vasogenic edema accounts for the changes observed in PRES. Diffusion-weighted MR imaging (DWI) has proved reliable in distinguishing vasogenic edema in PRES from cytotoxic edema in the setting of cerebral ischemia [4,5]. The notion that high DWI signal intensity with low apparent diffusion coefficient (ADC) values in PRES is due to cytotoxic edema and, therefore, heralds the progression to infarction has been previously recognized in a few reported series [3].

We present here the case of a 43-year-old woman with posterior encephalopathy and signs of cytotoxic edema on DWI. However, although irreversible tissue damage was the expected outcome, we observed normalization on magnetic resonance imaging (MRI), indicating that restricted diffusion on ADC mapping does not always lead to irreversible tissue damage.

Case report

A 43-year-old woman, known to suffer from diabetes mellitus, arterial hypertension and severe renal failure, was found in a coma (Glasgow coma scale: 7) with no focal deficit, and high arterial blood pressure (170/100 mmHg), requiring intubation. Laboratory tests showed elevations in blood glucose, blood creatinine (645 mmol/L) and urea (1.18 mmol/L). Her sodium and potassium levels were 130 mmol/L and 5.7 mmol/L, respectively. Ophthalmological examination showed bilateral papillary edema and severe hypertensive retinopathy.

The cerebral MRI performed on admission showed extensive, bilateral, hyperintense signals on fluid-attenuated inversion recovery (FLAIR) sequences in the subcortical white matter of the parieto-occipital and frontal regions, thalami, corpus callosum, cerebellar hemispheres andpons (Figs. 1A, 1B and 2A). DWI showed increased signal intensity in the parietal and frontal lobes, and corpus callosum (Fig. 2B). ADC values from the right and left subcortical parietal lobes were reduced by approximately 45% (525.10^{-6} mm²/s) and 65% (351.10^{-6} mm²/s), respectively, compared with those from normal white matter (791.10^{-6} mm²/s) (Fig. 2C).

On the basis of these findings, the diagnosis of hypertensive encephalopathy with regions of cerebral cytotoxic edema was made. After four days of treatment in the intensive care unit, which included treatment with insulin, hemodialysis and intravenous antihypertensive medication, the patient was extubated and her loss of consciousness rapidly resolved.

Follow-up MRI was performed one week after the initial abnormal presentations—at which time, the patient’s condition was clinically normal. MRI findings were normal (Fig. 2D) except for a small, residual hyperintense signal on FLAIR images of the left thalamus and corpus callosum. ADC maps and ADC values were nearly normal. The clinical improvement allowed her to be transferred to a nephrology medical unit on day 10 for treatment of the persistent renal failure.

Discussion

This case of PRES was characterized by its unusual DWI findings. The patient presented with hypertensive encephalopathy with acute uremia. The pathophysiology of hypertensive encephalopathy has been studied extensively, and two main mechanisms have been suggested [1]. One hypothesis is that cerebral vasospasm results in ischemia and the subsequent development of T2 hyperintensity. Alternatively, it has been suggested that there is a temporary failure of the autoregulatory capabilities of the cerebral vessels, leading to hyperperfusion, breakdown of the blood–brain barrier and, as a consequence, vasogenic edema [3].

The predominant involvement of posterior cerebral regions is a recognized hallmark of PRES [2]. It has been proposed to be the result of regional differences in the distribution of intracranial adrenergic receptors [6]. Vessels of the posterior circulation have a sparse sympathetic innervation and are, therefore, poorly equipped to initiate protective vasoconstriction in response to a sudden increase in arterial blood pressure, resulting in disruption of the blood–brain barrier and passive extravasation of fluid into the interstitium.

However, despite the emphasis on posterior supratentorial lesions, the associated involvement of the brainstem,
A, B. A 43-year-old woman with arterial hypertension and severe renal failure is admitted in a comatose state: axial fluid-attenuated inversion recovery (FLAIR) imaging shows bilateral, symmetrical hyperintense signals in the cerebellar hemispheres, pons, and subcortical white matter of the parietal, occipital and frontal lobes, thalami and corpus callosum.

Uremic encephalopathy is due to an elevated level of blood urea nitrogen (BUN) that directly results from renal failure. Neurological deficit from simple uremia is not a frequent occurrence, and the pathophysiology of this entity is not well known. It has been suggested that a breakdown of the blood–brain barrier due to endothelial injury (as in the kidneys) leads to an increased concentration of “uremic toxins” in the brain parenchyma and associated neurological abnormalities [7]. In our patient, as both acute uremia and acute hypertension were present, we cannot confirm which condition induced the neurological and morphological changes, but can only suppose that both may be relevant.

In our patient, reversible restricted water diffusability was seen on DWI early after the clinical symptoms, together with related FLAIR imaging abnormalities. Areas affected by PRES showed ADC reductions in the order of 45–65% in the parietal regions. Many studies have shown that increased ADC values indicate vasogenic edema, while decreased ADC values indicate cytotoxic edema that inevitably induces cell death [2–5]. However, to our knowledge, no other study has reported a patient with ADC reduction and a favorable clinical outcome in PRES— with no evidence of parenchymal...
This reversibility of diffusion anomalies has previously been reported during acute transient cerebral ischemia [8] and vasospasm after subarachnoid hemorrhage [9], which also casts some doubt on previous assumptions that lesions visible on DWI are areas of cytotoxic edema that inevitably lead to cell death. In addition, the literature contains anecdotal reports of transient cerebral ischemia in which abnormalities observed on DWI sequences have proved to be reversible. In an animal model, Li et al. [10] showed that ADC loss could be reversed if occlusion lasted less than 60 minutes. Beyond that time, however, lesions were irreversible [11]. Using an ischemia/anoxia model, Harris et al. [12] showed that the ADC has to fall by 10–25% before anoxic depolarization is triggered—in other words, before proton pump failure occurs. ADC and DWI abnormalities have, furthermore, been observed during epilepsy concomitantly with reversible cell membrane and fluid changes [13]. However, Hasegawa et al. [14] suggested that there might be an ADC threshold above which lesions are reversible.

The pathophysiology of these reversible abnormalities observed on DWI, and especially in the presence of transient ischemia, is not clear. It may be that DWI abnormalities are associated with proton pump dysfunction and cerebral blood flow anomalies [9], but this hypothesis has yet to be confirmed.

In conclusion, we infer from our present case that MRI is an efficient means of early diagnosis and follow-up in PRES, and that a decrease in ADC values does not always correlate with nonreversibility of such lesions, as is often described in the literature for this pathology.

References