

Letters to the Editor

Reversible posterior leukoencephalopathy syndrome: a recurrent and atypical case in hemodialysis[☆]

Leucoencefalopatía posterior reversible: un caso recurrente y atípico en hemodiálisis

To the Editor,

Reversible posterior leukoencephalopathy syndrome (RPLS) is a clinical and radiological syndrome described in 1996. Its pathogenesis is still unclear and there are two theories: (1) cerebral hyperperfusion and (2) severe vasospasm.¹⁻³ It is associated with malignant hypertension, eclampsia and other coexisting conditions in patients with chronic kidney disease (CKD) such as hypertension, vascular or autoimmune diseases, as well as immunosuppressants, erythropoietin and transplantation.²

A higher number of cases have been described in women. Clinical symptoms include seizures (93%), visual disturbances (67%), loss of consciousness (53%) and headache (47%). Between 7 and 13% present aphasia, hemiplegia, hemiparesis or mental impairment.¹

In 2010, a large case series (113 adults) was published, among which only 6% were recurrent⁴: from 2 weeks to 36 months after the initial episode.^{4,5}

Cranial MRI is the gold standard; it shows symmetrical parieto-occipital white matter abnormalities (cortical or subcortical edema), which are reversible after days or weeks of treatment.¹⁻⁵

Only a fast and adequate control of triggering events prevents irreversible complications. Sometimes antiepileptics, general anesthesia and mechanical ventilation must be administered; it has not yet been confirmed that corticosteroids reduce vascular edema.² There is no association between the severity of hypertension and the sequelae.⁵

In the differential diagnosis, multiple diseases should be considered² (Table 1).

We describe a case that we find interesting because of its unusual clinical presentation and subsequent recurrence:

Table 1 – Differential diagnosis of RPLS.

Vascular disease	Non-vascular disease
Cerebral venous sinus thrombosis	Infectious encephalitis
Intracranial hemorrhage	Autoimmune encephalitis
Posterior circulation stroke	Metabolic/toxic encephalopathy
Primary CNS vasculitis	

RPLS: reversible posterior leukoencephalopathy syndrome.

34-year-old male, on HD with CKD secondary to obstructive uropathy (verumontanum). His hypertension was treated with atenolol 50 mg/day. He presented to the emergency department for aphasia and agitation. Fever (38.8 °C), BP: 232/124 mmHg. No significant findings on physical examination. Blood count: normal leukocytes, slight neutrophilia and lymphopenia. Normal cranial CT and lumbar puncture. Negative blood and CSF cultures, and negative serology. Ocular fundus: hypertensive retinopathy grade III. He was hospitalized to Infectious Disease Service for suspected meningoencephalitis; empirical treatment was initiated with ampicillin, vancomycin, acyclovir, rifampicin and isoniazid.

It remained difficult to control the blood pressure (SBP: 150–200 mmHg, DBP: 90–120 mmHg); this was initially treated with labetalol via infusion pump, ACE inhibitors and calcium antagonists, but oral minoxidil 10 mg and atenolol 100 mg and adjustment of dry weight were also needed.

As a result of progressive reduction in the level of consciousness, reaching a score of 4 on the Glasgow scale, he was transferred to ICU. An MRI was ordered and the findings suggested RPLS: bilateral parieto-occipital subcortical white matter hyperintensity in T2 and FLAIR weighted sequences. Ten days later, the follow-up MRI was normal (Fig. 1).

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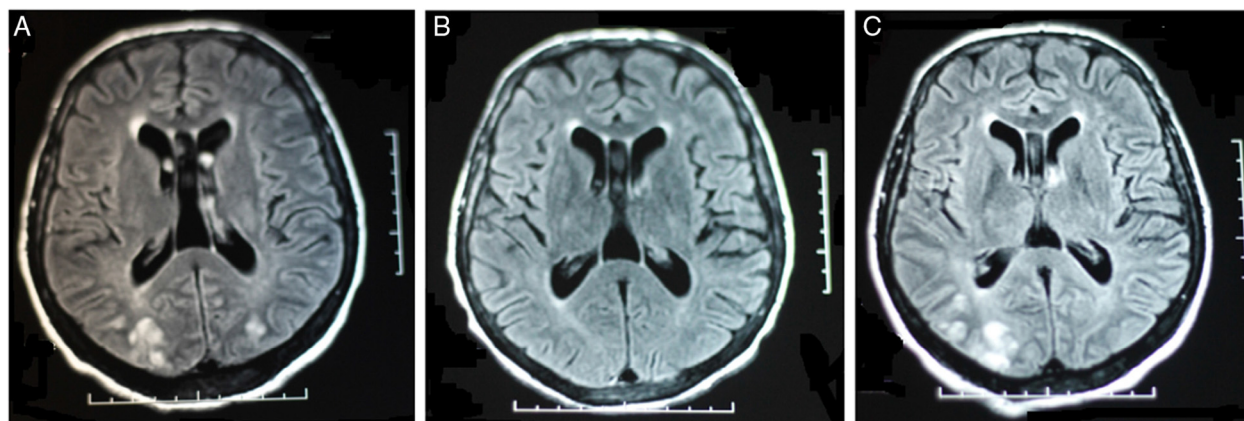


Fig. 1 – Cranial MRI findings: (A) bilateral parieto-occipital subcortical white matter hyperintensity; (B) follow-up MRI after 10 days of treatment, with complete resolution of lesions; (C) MRI confirming recurrent RPLS, 6 months after the first episode.

After 22 days in ICU, no neurological sequelae were present and he was transferred to Nephrology with a diagnosis of RPLS, probably hypertensive. Antihypertensive therapy was adjusted at discharge to maintain SBP at ≤ 150 mmHg.

Six months later he was readmitted for right hemiparesis and dysarthria secondary to hypertensive emergency (280/120 mmHg), with normal cranial CT. He was admitted to ICU with decreased level of consciousness, motor agitation and poor BP control. The MRI showed similar changes in the previous location, confirming recurrent RPLS in the context of a hypertensive emergency.

Pharmacological treatment was optimized with irbesartan 150 mg (0–0–1), carvedilol 25 mg (1/2–0–1/2), Carduran neo[®] 4 mg (1–0–1), and enalapril 20 mg (1/2–0–1/2).

This is a rare case because it is a male who began with fever and neurological disturbances in the context of hypertensive emergency, and had a recurrence after 6 months.

In the literature, hypertensive crises, CKD and sudden changes in pressure and volume in HD are related as triggers of disease recurrence.⁵ Early diagnosis through MRI and treatment of risk factors, in this case controlling the blood pressure, are essential for reversing the clinical picture and preventing recurrence. If it persists, it can be fatal or it may result in serious neuropsychological sequelae.

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