CASO CLÍNICO/CASE REPORT

A Reversible Event Leading to Lifestyle Reversion: About a Case of PRES

Um Evento Reversível e uma Reversão do Estilo de Vida: Sobre um Caso de PRES

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Abstract

The posterior reversible encephalopathy syndrome (PRES) is a heterogeneous clinical-radiological characterized by a combination of encephalopathy, altered mental status, epileptic seizures, visual disturbances, headache, and focal neurologic signs.

Other neurologic deficits, such as ataxia, may occur. PRES can be found in a non-posterior distribution, mainly in watershed areas, including the frontal, inferior temporal, cerebellar, and brainstem regions.

We present the case of a middle age male presenting with headache, dizziness, ataxia, and hypertensive emergency. PRES was suspected, and the diagnosis was confirmed by brain magnetic resonance imaging. Computerized tomography showed bilateral hypodensities of the cerebellar white matter with associated narrowing of the cortical sulci and basal cisterns.

This case represents a rare presentation of PRES with unusual clinical and topographical involvement, such as brainstem and cerebellum.

Resumo

A síndrome da encefalopatia reversível posterior (PRES) é um síndrome clínico-radiológico heterogéneo caracterizado por uma combinação de encefalopatia, estado mental alterado, convulsões epilépticas, perturbações visuais, dores de cabeça e sinais neurológicos focais.

Outros défices neurológicos, como a ataxia, podem ocorrer. PRES pode ser encontrado numa distribuição não-posterior, principalmente em áreas de bacia hidrográfica, incluindo as regiões frontal, temporal inferior, cerebelar, e do tronco cerebral.

Apresentamos o caso de um homem de meia idade que se apresenta com dores de cabeça, tonturas, ataxia, e emergência hipertensiva. Suspeitou-se de PRES, e o diagnóstico foi confirmado por ressonância magnética cerebral. A tomografia computorizada mostrou hipodensibilidades bilaterais da matéria branca cerebelar com estreitamento associado dos sulcos corticais e cisternas basais.

Este caso representa uma apresentação rara de PRES com um envolvimento clínico e topográfico invulgar, como o tronco cerebral e o cerebelo.

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Introduction

Posterior reversible encephalopathy syndrome (PRES) is an increasingly recognized clinical-radiological disorder. We expose the case of a middle age male presenting with headache, dizziness, ataxia, and hypertensive emergency. The clinical presentation is variable and unusual symptoms such as ataxia, dizziness, or even focal deficits may occur.[1,2] As the name suggests, PRES typically affects posterior regions. However, other topographies are involved, such as watershed areas, cerebellar, and brainstem regions. The brainstem and basal ganglia involvement is considered uncommon, representing less than 5% of the cases described in the literature.[2,3] Arterial hypertension and blood pressure fluctuations are present in a significant proportion of patients at the time of neurologic deficits onset.[1] We describe a case of PRES associated with a hypertensive crisis with unusual symptomatology and neuroimaging. We briefly discuss some aspects of this heterogeneous syndrome of unclear aetiopathogenesis for which there are still no established diagnostic criteria.

Case Report

We present the case of a middle age male smoker (35 packs/year) with primary arterial hypertension diagnosed three years earlier with established target-organ damage (nephrosclerosis, cardiomyopathy, and retinopathy). There was no family history of hypertension. He lost to follow-up in Nephrology and Cardiology consultations, admitting to erratic therapeutic compliance.

He presented in the emergency department with occipital headache, pulsatile, without prodromes, aura, photo or phonophobia, postural or exertional worsening, accompanied by blurred vision, dizziness, and ataxia for the last five days, with progressive worsening. There was no history of fever or other focal neurological symptoms, including seizures. He was admitted to the emergency room for a hypertensive emergency (250/170 mmHg in the left upper limb; 245/165 mmHg in the right upper limb). He was conscious, cooperative, and oriented, with no changes in the cranial nerves, language deficits, or other cognitive impaired. He had no change in pain sensitivity, asynergy, dysmetria in the finger-nose and heel-knee tests or other changes in the coordination tests; motor strength was preserved with live, symmetrical osteo-tendinous reflexes and flexed cutaneous-plantar response.

The biochemical blood tests showed an acute renal injury with plasma creatinine of 2.4 mg/dL and blood urea nitrogen (BUN) of 89 mg/dL when in the last years, the patient had values of plasma creatinine between 1.5 and 1.8 mg/dL and BUN between 56-64 mg/dL. The remaining analytical study, as well as the electrocardiogram, were regular.

A cranial computed tomography (CT) scan was performed, and it showed signs of ischemic leukoencephalopathy and bilateral cerebellar white matter hypodensity associated with fading of the cortical sulci and base cisterns hemorrhagic lesions were ruled out (**Fig. 1**).

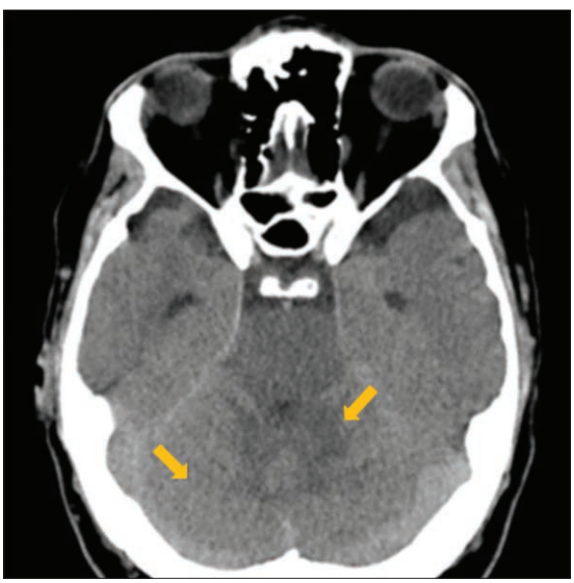

Figure 1. Areas of a diffuse white matter hypodensity at the cerebellum level, some space conflict, and blackout of cortical grooves.

The patient was admitted to a surveillance unit, requiring labetalol infusion (maximum of 45 mg/h) for maintaining systolic blood pressure below 140 mmHg and diastolic blood pressure below 90 mmHg, for the first three days. After testing for dysphagia, oral therapy with enalapril (20 mg 2id), chlorthalidone (50 mg 1id), carvedilol (25 mg 2id), nifedipine (30 mg 2id) and dinitrate (50 mg 2id) was progressively introduced.

He presented a total resolution of the headache, visual alterations, and progressive improvement of ataxia and dizziness with the normalization of the blood pressure. He was transferred to the general ward of Medicine, maintaining the previous good evolution.

In the face of hypertension, neurological imaging, and symptomatic improvement with blood pressure control, the hypothesis of PRES was placed, and cranial magnetic resonance imaging (MRI) was requested.

The MRI showed hyperintense lesions in T2-weighted

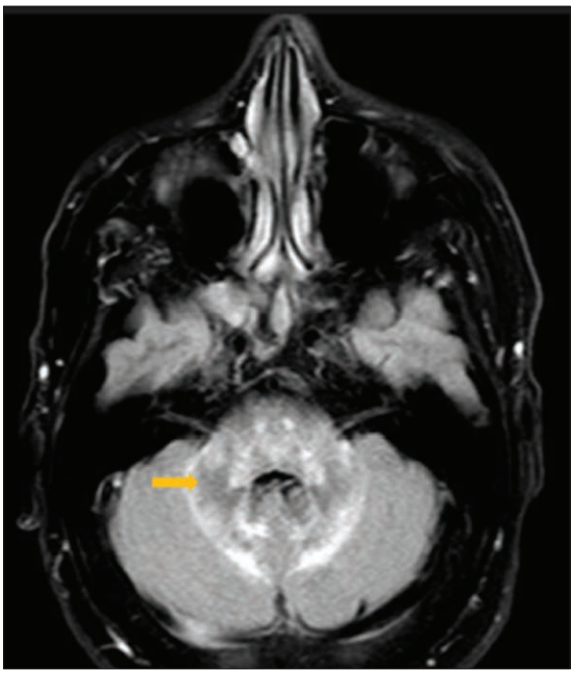

Figure 2. Brain magnetic resonance imaging: hyperintense lesions in T2/FLAIR at the cerebellum, medulla oblongata.

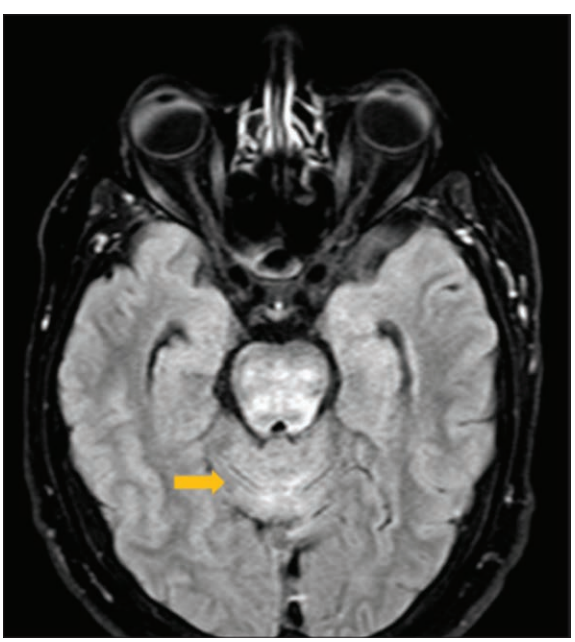

Figure 3. Brain magnetic resonance imaging: hyperintense lesions in T2/FLAIR at pons and vermis.

sequences without restricted diffusion at the cerebellum, oblong medulla, pons, and midbrain (**Figs. 2 and 3**).

This episode was revolutionary in the patient's life, previously non-compliant with pharmacological and non-pharmacological therapy, having adopted a new lifestyle. At the hospital discharge from the episode described, he was discharged with five antihypertensive drugs. At the last consultation assessment, he had excellent blood pressure control with only three drugs and was familiar with the importance of a regular diet and physical activity.

The patient followed in our internal medicine consultation, agrees with the publication of this case.

Discussion

PRES arises in the context of vascular dysfunction and consequent vasogenic edema. The cause is attributed to two competing theories: tension alterations with mean blood pressure above the limit of autoregulation (known as "vasogenic theory") and endothelial dysfunction resulting from systemic toxicity, known as "endothelial theory."

"Vasogenic theory" argues that rapidly developing hypertension with failure of cerebral autoregulation cause the breakdown of the blood-brain barrier and secondary vasogenic edema.[2] The relative lack of sympathetic innervation of the posterior circulation is the likely mechanism for the preferential involvement of this part of the brain from PRES. This hypothesis supports that adequate tension control allows for clinical improvement and radiological reversal.

In "the endothelial theory," there is an endothelial dysfunction due to the cytotoxic effects of a systemic condition (infection, sepsis, neoplasm, drugs) triggering an imbalance between vasoconstrictor and vasodilator mediators in favor of the former, which promotes hypoperfusion.

Arterial hypertension and blood pressure fluctuations are considered prevalent in the development of PRES. Patients with uncontrolled hypertension, as in the case described, are particularly susceptible to hypertensive crises resulting in hypertensive encephalopathy and PRES.[1]

In this case, despite the narrower differential diagnosis in a hypertensive emergency, the clinical and imaging pattern was atypical, making the PRES diagnosis particularly challenging. Although not specific, four symptoms are considered typical. They are frequently reported: encephalopathy and seizures are the two most common symptoms, followed by visual disturbances and headache,[2-8] present in the described case. Ataxia and dizziness associated with PRES occur in a minority of cases, 3.6% and 6.8%, respectively, and are considered uncommon manifestations. These symptoms occur due to the involvement of atypical regions, such as the cerebellum and brainstem, in this patient.[9,3]

Brain MRI, especially the T2-weighted and fluid-at-

tenuated inversion recovery (FLAIR) sequences, is the most sensible way to show the bilateral, subcortical, and symmetrical vasogenic edema, which is the characteristic imaging pattern from PRES.[2,3]

There are three characteristic patterns at PRES – posterior, frontal, and diffuse.[1,5] Occasionally, the edema may have a central-variant pattern that affects the brainstem, basal ganglia, posterior limb of the internal capsule, periventricular regions, and cerebellum.[2] The affection of these regions is relatively uncommon

Our suspicion of PRES based on clinical symptoms, hypertensive crisis, and bilateral hypodensities of the cerebellar white matter on CT was confirmed by MRI. **Figs. 1 and 2** show a hyper signal on long TR sequences in the cerebellum (including vermis), bulb, pons, and mesencephalon, consistent with PRES but in regions considered unusual. Recent data show involvement of each area in only 22.3% and 8.6% of cases, respectively.[3] Affectation of the pons and midbrain is particularly rare. The reversibility of vasogenic edema is a characteristic hallmark of imaging findings in PRES.

Due to the observed evolution, with a continuous and complete resolution of the symptoms concomitant with tension control, and expected evolution for a PRES, we did not perform MRI control. We admit, however, it is a limitation. Complete reversibility of the lesions should be observed at MRI. Imaging reversibility corresponds with clinical improvement, occurring over days to weeks.[1,5]

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Our patient had the expected evolution with the stabilization of the blood pressure profile, and the symptoms progressively improved until he was completely asymptomatic.

Conclusion

With this case, we intend to draw attention to a rare entity like PRES, where clinical suspicion is essential to establish the diagnosis. Timely diagnosis and institution of adequate therapy have a significant impact on the prognosis.

We highlight the importance of timely suspicion in atypical PRES responsible for unique presentations that may delay diagnosis.

Further investigations are needed to understand the pathophysiology of PRES better. A standardized algorithm that incorporates clinical and laboratory markers, imaging features and a risk factor for PRES should be sought in future studies. ■

Contributorship Statement / Declaração de Contribuição

SSP: manuscript elaboration and final approval.
CCA: manuscript elaboration and final approval.
HM: manuscript elaboration and final approval.
RA: manuscript review and final approval.

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