CASE REPORT

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Posterior Reversible Vasogenic Cerebral Edema Syndrome: Rethinking Atypical PRES Presentations Without Encephalopathy

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Abstract

Posterior reversible encephalopathy syndrome (PRES) is an acute neurological disorder characterized by reversible subcortical vasogenic edema, predominantly affecting the posterior regions of the brain, particularly the parieto-occipital lobes. Common clinical manifestations include headaches, seizures, visual disturbances, and altered mental status, collectively referred to as encephalopathy. The pathophysiology of PRES is not entirely understood but is believed to involve endothelial dysfunction and impaired cerebral autoregulation, leading to hyperperfusion and subsequent vasogenic edema. While encephalopathy is considered a hallmark feature of PRES, the syndrome exhibits a broad spectrum of clinical presentations. Notably, cases without encephalopathy are underreported, potentially due to the variability in clinical manifestations and the absence of altered mental status, which may lead to misdiagnosis or delayed recognition. Such atypical presentations pose diagnostic challenges, emphasizing the necessity for heightened clinical suspicion and comprehensive evaluation. This report contributes to the existing literature by detailing a unique case of PRES without encephalopathy, underscoring the importance of considering PRES in patients presenting with visual disturbances and headaches, even in the absence of altered mental status. Early recognition and appropriate management are crucial, as timely intervention can lead to complete recovery, whereas delayed diagnosis may result in irreversible neurological deficits.

Keywords: Atypical PRES, encephalopathy, headache, hypertensive retinopathy, vasogenic edema

INTRODUCTION

Posterior reversible encephalopathy syndrome (PRES) is an acute neurotoxic condition characterized by reversible cortical-subcortical vasogenic edema, predominantly affecting the posterior regions of the brain, especially the parieto-occipital lobes.[1] The syndrome is frequently associated with severe hypertension, eclampsia, renal dysfunction, or immunosuppressive therapy.[2] Clinically, PRES commonly manifests with headaches, seizures, visual disturbances, and altered mental status, the latter often categorized under the umbrella of encephalopathy.[3] The pathophysiological mechanisms underlying PRES are not fully elucidated but are thought to involve endothelial dysfunction and impaired cerebral autoregulation, resulting in hyperperfusion and subsequent vasogenic edema.[4]

Although encephalopathy is traditionally regarded as a hallmark feature of PRES, the clinical spectrum is notably broad.[5] Atypical presentations lacking altered mental status are uncommon and, therefore, may be underrecognized or misdiagnosed. Such variability presents a significant diagnostic challenge, as the absence of classical neurological findings can delay appropriate intervention, increasing the risk of permanent neurological damage.[6] Prompt diagnosis is critical to initiate timely management strategies that can prevent irreversible neurological deficits.

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In this context, we present a rare and diagnostically challenging case of PRES without encephalopathy, where the patient presented with severe visual disturbances and headaches but maintained normal cognitive function throughout the disease course. This case underscores the importance of considering PRES in the differential diagnosis of patients with visual symptoms and hypertensive emergencies, even in the absence of altered mental status. Furthermore, it emphasizes the critical need for timely diagnosis and management to ensure favorable clinical outcomes.

CASE REPORT

A 42-year-old female without a known history of chronic illness presented with mild generalized headache and severe blurred vision in both eyes that began abruptly. She sought medical attention the following day at the ophthalmology clinic, where findings of papilledema and macular edema prompted referral to the neurology department for further evaluation. Upon admission to the neurology department, her blood pressure was critically elevated at 240/160 mmHg. Other vital signs were within normal limits. Neurological examination revealed no focal deficits, altered consciousness, or seizure activity. However, the patient exhibited significant visual impairment, with visual acuity reduced to counting fingers at 1 meter in both eyes. She was diagnosed by the ophthalmological examination with hypertensive retinopathy stage 4, characterized by retinal hemorrhages, hard exudates, cotton-wool spots, increased vascular tortuosity, and papilledema. Brain magnetic resonance imaging (MRI) revealed vasogenic edema predominantly involving the brainstem (midbrain and pons), cerebellum, and temporo-parieto-occipital lobes (Figure 1A). These findings effectively excluded differential diagnoses such as stroke, malignancy, central nervous system (CNS) demyelinating disorders, and leukoencephalopathy, while being consistent with a diagnosis of PRES. Further imaging, including magnetic resonance venography (MRV) and magnetic resonance arteriography (MRA), ruled out cerebral venous sinus thrombosis and reversible cerebral vasoconstriction syndrome (Figure 1B). Laboratory studies revealed no significant abnormalities, effectively excluding differential diagnoses such as hepatic encephalopathy, encephalitis, and CNS vasculitis. The patient initially received parenteral antihypertensive therapy with

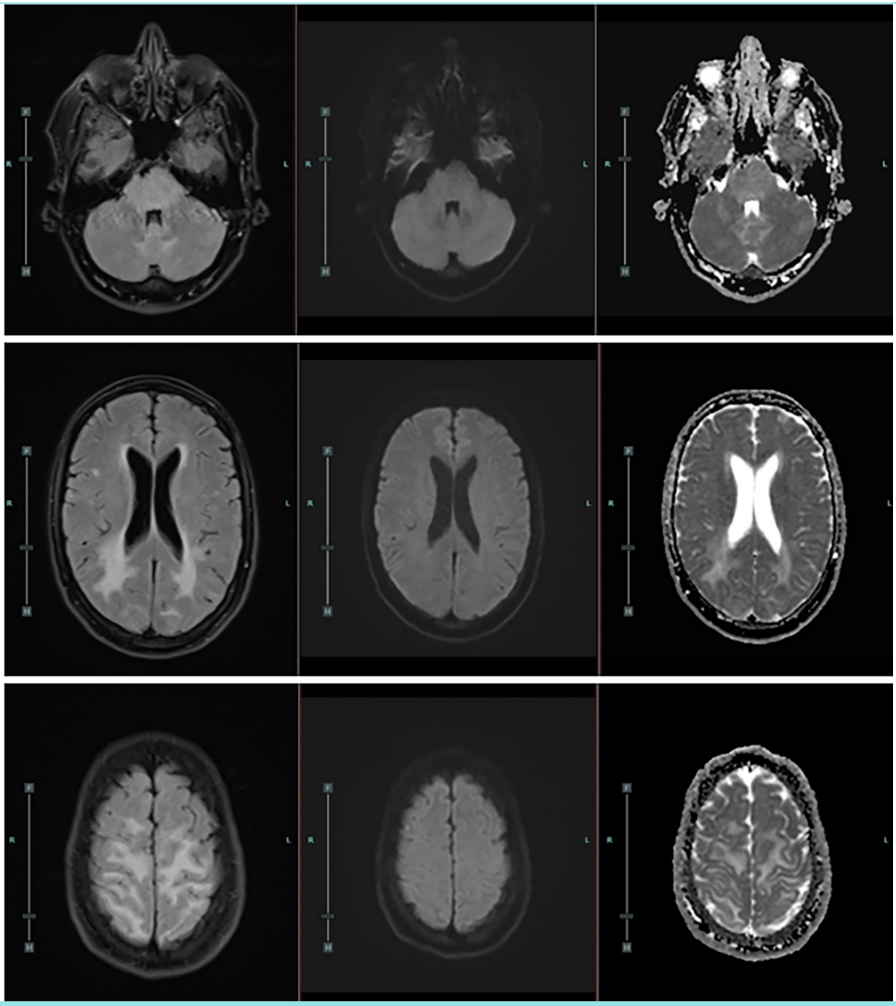

Figure 1A. The axial FLAIR, diffusion-weighted imaging (DWI), and apparent diffusion coefficient (ADC) sequences demonstrate vasogenic edema prominently involving the pons, cerebellum, and bilateral temporo-parieto-occipital lobes. The lesions appear hyperintense on FLAIR and DWI images, while corresponding ADC maps show no significant diffusion restriction, supporting the vasogenic rather than cytotoxic nature of the edema.

FLAIR: Fluid-attenuated inversion recovery

nimodipine (1 mcg/kg/hour) under close monitoring, followed by oral amlodipine 10 mg once daily for maintenance. Electroencephalography was performed and revealed normal findings. Levetiracetam (500 mg, twice daily) was initiated as a prophylactic measure, considering the high-risk of seizures associated with the observed vasogenic edema. No seizures occurred during the hospital stay. Echocardiography revealed mild left ventricular hypertrophy with an ejection fraction of 60%. Renal artery Doppler ultrasonography excluded renal artery stenosis. With antihypertensive therapy, the patient experienced gradual relief from headaches and a progressive improvement in visual acuity. By discharge on day four, visual acuity had improved to 20/25 with a reported 80% subjective recovery. At one-month follow-up, complete resolution of visual deficits and normalization of retinal findings were observed (Figure 2). On follow-up MRI, the vasogenic edema observed in prior imaging was no longer evident, indicating radiological resolution (Figure 3). Levetiracetam tapering was initiated in light of sustained clinical and radiological recovery. Written informed consent was obtained from the patient for publication of this report and accompanying images.

DISCUSSION

PRES is commonly characterized by the presence of encephalopathy, a key clinical feature that manifests in up to 94% of patients. This spectrum ranges from mild confusion and cognitive deficits to stupor and, in severe cases, coma.[7] The presented case of PRES is notable for its absence of encephalopathy, a finding that deviates from the classical clinical profile commonly described in the literature.[8] Such presentations are underreported and pose significant diagnostic challenges, as the absence of altered mental status may lead to misdiagnosis or delayed

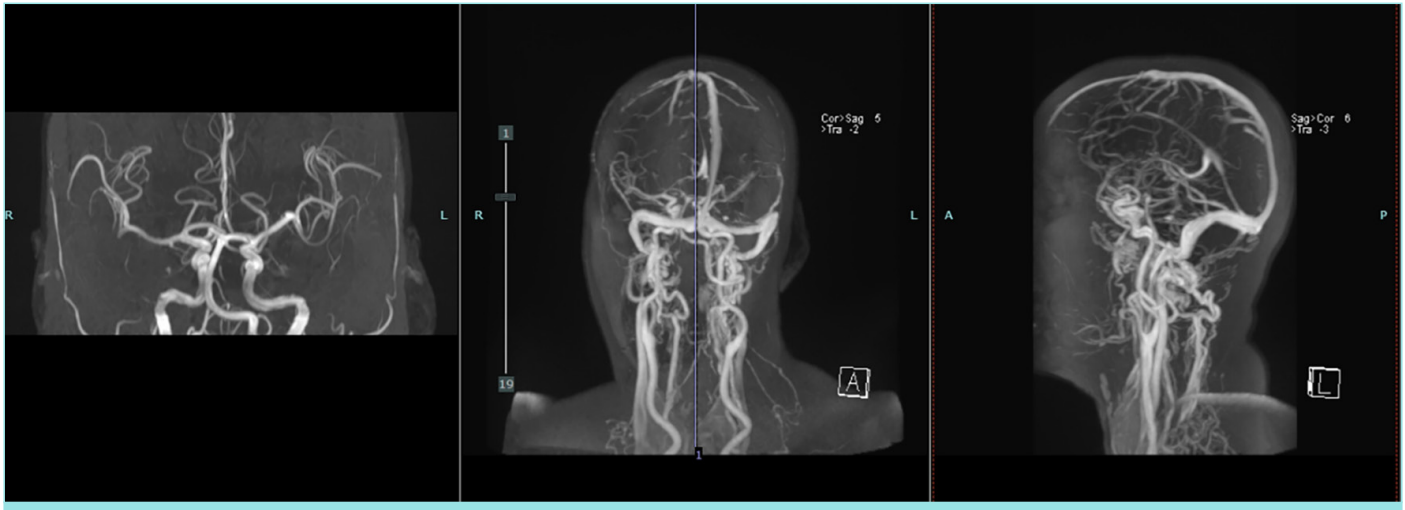

Figure 1B. The study involves displaying vascular imaging studies, including magnetic resonance arteriography (MRA) in the first image and magnetic resonance venography (MRV) in the subsequent two images. The MRA shows normal intracranial arterial structures without evidence of segmental narrowing or beading. Similarly, the MRV sequences demonstrate normal venous flow within the major dural sinuses.

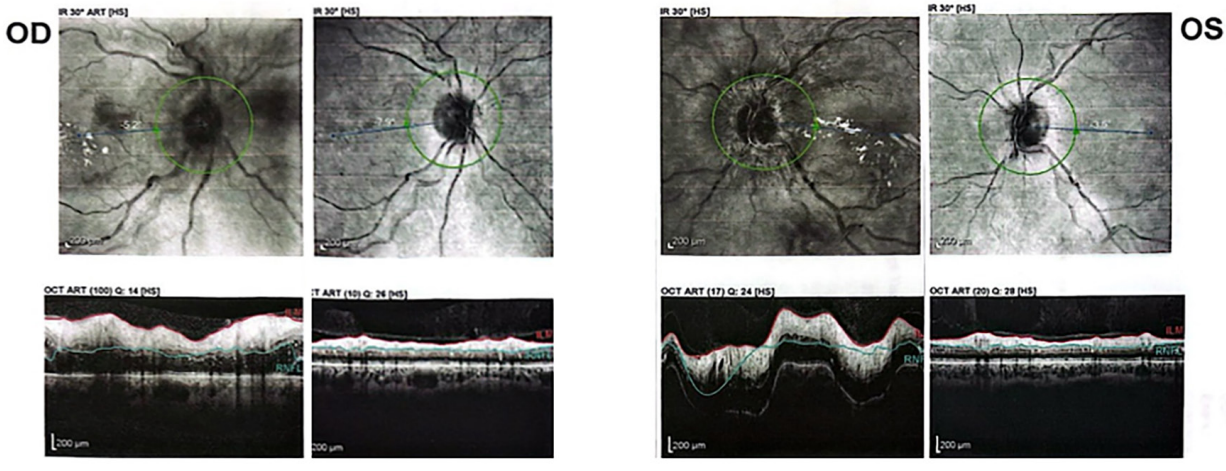

Figure 2. Comparing pre- and post-treatment fundus photographs and optical coherence tomography (OCT) of the retinal nerve fiber layer, (RNFL), images of both eyes involves an analysis of these data. Initial fundus findings include retinal hemorrhages, hard exudates, cotton-wool spots, vascular tortuosity, and papilledema, while OCT shows diffuse RNFL thickening consistent with optic disc edema. Post-treatment images demonstrate marked radiological and clinical improvement.

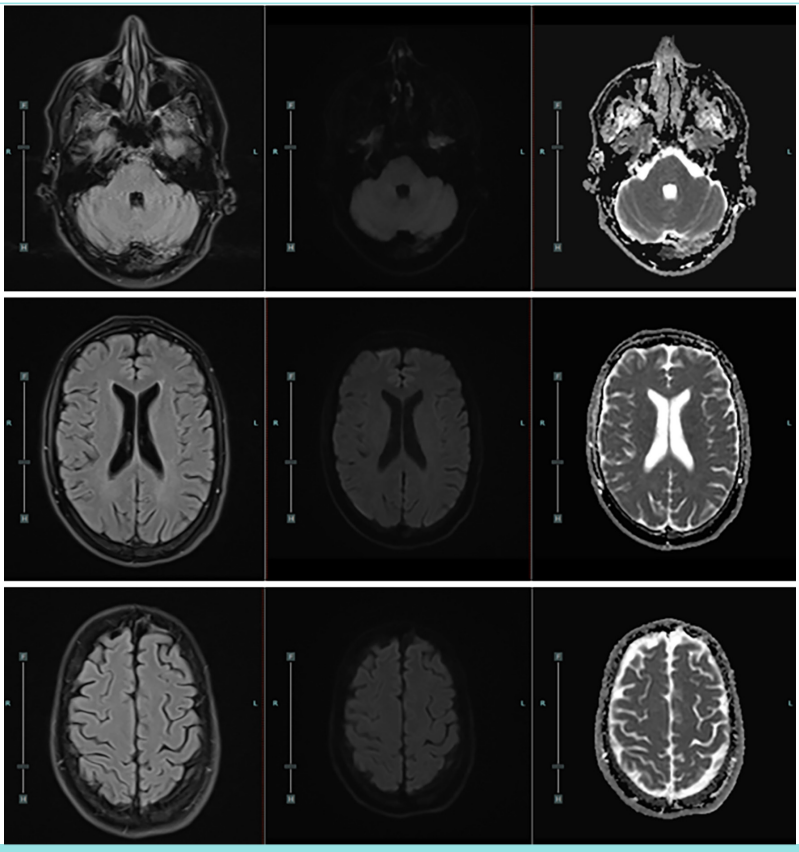

Figure 3. Demonstrating complete resolution of previously noted vasogenic edema in the brainstem, cerebellum, and temporo-parieto-occipital regions, indicating radiological improvement in follow-up MRI.

MRI: Magnetic resonance imaging

recognition.[9] Literature suggests that encephalopathy a hallmark feature of PRES and is present in the majority of cases. However, studies have highlighted that PRES is a spectrum disorder, and clinical manifestations may vary depending on the extent and location of cerebral involvement.[10] In particular, atypical presentations lacking encephalopathy are often overlooked, despite the presence of imaging findings consistent with vasogenic edema.[9] This case exemplifies such a diagnostic dilemma and emphasizes the necessity for heightened clinical vigilance when evaluating patients presenting with severe hypertension and visual disturbances.

The pathophysiology of PRES is believed to involve endothelial dysfunction and impaired cerebral autoregulation, resulting in hyperperfusion, blood-brain barrier disruption, and subsequent vasogenic edema.[2] The absence of encephalopathy in this case suggests possible individual variability in cerebral autoregulatory capacity or regional susceptibility to hypertensive injury. Sympathetic-mediated vasoconstriction tends to be more pronounced in the anterior circulation and relatively weaker in the posterior regions, potentially contributing to the predilection for vasogenic edema in posterior brain structures.[11] This mechanism could explain why the patient's clinical presentation was dominated by visual symptoms without altered mental status. However, further research is warranted to elucidate the mechanisms that may confer protection against altered mental status in certain patients.

The co-occurrence of hypertensive retinopathy with PRES further substantiates the concept of shared vascular pathology between the brain and retina. Both conditions are rooted in endothelial dysfunction and impaired autoregulatory mechanisms triggered by acute hypertensive insults. Retinal findings, such as papilledema, hemorrhages, and exudates, closely parallel the vasogenic edema observed in PRES, reflecting a common pathophysiological mechanism.[12] This relationship highlights the systemic nature of hypertensive crises and positions retinal changes as potential markers of CNS involvement. Such insights emphasize the importance of interdisciplinary diagnostic approaches and suggest that retinal assessments could facilitate early identification of CNS pathology, particularly in resource-limited settings.

Given the predominance of vasogenic edema in imaging findings, the term "Posterior Reversible Vasogenic Cerebral Edema syndrome"(PRVCES) has been informally proposed to highlight the central imaging and pathophysiological feature of vasogenic edema in PRES, offering conceptual clarity particularly in atypical cases without encephalopathy where traditional clinical markers may be absent.[13] While this terminology may provide a more descriptive and pathophysiologically accurate framework, it has not yet been incorporated into international classification systems or clinical practice guidelines. Accordingly, its use should be regarded as a descriptive and explanatory tool rather than as an established diagnostic category. Moreover, "PRES" remains the widely accepted and internationally recognized term in both clinical and academic literature, encompassing the broad clinical spectrum of

the syndrome, including both typical and atypical presentations.[14,15] Therefore, although PRVCES may contribute to enhanced understanding of unusual cases, its broader adoption would require validation in larger case series and consensus among experts. Until such recognition is achieved, the term should be applied with caution.

This case further underscores the critical role of neuroimaging in the diagnostic process, particularly in patients presenting with limited symptoms and signs. MRI findings of vasogenic edema involving the brainstem, cerebellum, and posterior cortical regions were pivotal in establishing the diagnosis of PRVCES. The integration of MRV and MRA facilitated comprehensive vascular assessment, effectively ruling out cerebral venous sinus thrombosis and reversible cerebral vasoconstriction syndrome-both important differential diagnoses in patients presenting with severe hypertension and neurological symptoms. The early use of these advanced imaging modalities not only ensured diagnostic accuracy but also guided therapeutic decisions. Timely imaging likely contributed to the favorable clinical outcome observed, while delayed or inadequate imaging in atypical presentations may risk diagnostic uncertainty, mismanagement, and irreversible neurological complications. The emphasis on vasogenic edema as the key diagnostic feature in PRVCES further underscores the importance of prioritizing imaging in cases with atypical presentations, where encephalopathy is absent.

Antiepileptic drugs (AEDs) play a crucial role in the management of patients with PRES, particularly those who experience seizures during the disease course. In cases with clinical seizure activity, AEDs are essential to stabilize the patient and prevent recurrent episodes. However, the use of AEDs in PRES patients without seizures remains controversial, and is often guided by the extent of vasogenic edema and the perceived seizure risk.[16] Prophylactic treatment, as seen in the presented case, is typically individualized and aims to mitigate potential complications in high-risk scenarios. Gradual discontinuation of AEDs is an essential consideration in the management process and should be based on a comprehensive assessment of both clinical and radiological findings. In this case, tapering was planned following evidence of radiological resolution and clinical stability. This approach minimizes unnecessary medication exposure while safeguarding against potential seizure recurrence. However, the optimal timing for AED withdrawal in PRES remains undefined and warrants further investigation.

CONCLUSION

PRES can present without encephalopathy, as demonstrated in this patient with hypertensive retinopathy and severe hypertension. Clinicians must maintain a high index of suspicion for PRVCES in atypical presentations of PRES, particularly in the absence of encephalopathy, to avoid diagnostic delays and mismanagement. This report contributes to the growing recognition of the diverse clinical spectrum of PRES and emphasizes the critical need for further research into its pathophysiological mechanisms, diagnostic strategies, and management approaches to optimize patient outcomes.

MAIN POINTS

- Posterior reversible encephalopathy syndrome (PRES) may present without altered mental status; the presence of isolated visual disturbances in the setting of a hypertensive emergency should raise clinical suspicion for this diagnosis, even in the absence of encephalopathy.
- The coexistence of advanced hypertensive retinopathy and radiologically confirmed vasogenic edema underscores the shared pathophysiological basis between the retinal and cerebral microvasculature, highlighting the diagnostic value of ophthalmological findings in neurovascular syndromes such as PRES.
- This case reinforces the indispensable role of advanced neuroimaging in the early identification of atypical PRES presentations and advocates for a broader diagnostic paradigm that accounts for regional cerebral vulnerability and interindividual variability in autoregulatory responses.

ETHICS

Informed Consent: Written informed consent was obtained from the patient for publication of this report and accompanying images.

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Footnotes

Authors Contributions

Surgical and Medical Practices: Ö.Ö., Ş.I., Concept: Ö.Ö., Design: Ş.I., Data Collection and/or Processing: Ö.Ö., Analysis and/ or Interpretation: Ö.Ö., Ş.I., Literature Search: Ş.I., Writing: Ö.Ö.

DISCLOSURES

Conflict of Interest: No conflict of interest was declared by the authors.

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