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Case Report

Posterior reversible encephalopathy syndrome with isolated infratentorial involvement: A case report

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ARTICLE INFO

Article history:

Received 6 January 2019

Revised 10 February 2019

Accepted 11 February 2019

Available online 1 March 2019

Keywords:

Posterior reversible encephalopathy syndrome

Spinal cord involvement

Infratentorial structures

Hypertensive emergency

ABSTRACT

Posterior reversible encephalopathy syndrome (PRES) is a clinical and radiological entity of acute neurological symptoms associated with characteristic MRI finding. Vasogenic edema in the white matter of parieto-occipital regions is the classical MRI findings. Spinal cord involvement in PRES is extremely rare and frequently underrecognized condition. Recently, a variant-type PRES with isolated involvement of infratentorial structures is getting more attention. Herein, we present a case of hypertensive emergency and associated radiological features of PRES with isolated involvement of the brain stem, cerebellum, and spinal cord.

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Introduction

Posterior reversible encephalopathy syndrome (PRES), first described in 1996 [1], is a radiological and clinical diagnosis made in patients who present with acute neurological symptoms and radiological magnetic resonance imaging (MRI) findings of increased white matter vasogenic edema typically in the bilateral parieto-occipital regions [2]. Clinically, PRES could variably present with a headache, altered mental status, vision changes, and possible seizure; and has frequently been reported in cases of hypertensive encephalopathy, chronic obstructive pulmonary disease, autoimmune diseases,

preeclampsia/eclampsia, and cytotoxic drugs [3–5]. Although the pathophysiological basis of PRES is still not fully understood, the most likely explanation is disruption of the cerebral vascular auto-regulation and endothelial dysfunction leading to vasogenic edema. The most common radiological manifestation of PRES is increased signal intensity in T2-fluid-attenuated inversion recovery (FLAIR) [6]. With antihypertensive and supportive treatments, the outcome of the clinical and radiological findings of PRES is a complete resolution in most of the cases.

PRES with spinal cord involvement is an extremely rare entity and has recently been distinguished from classical PRES [7]. There are only a handful of cases in the literature that

Competing Interests: The authors have declared that no competing interests exist.

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<https://doi.org/10.1016/j.radcr.2019.02.009>

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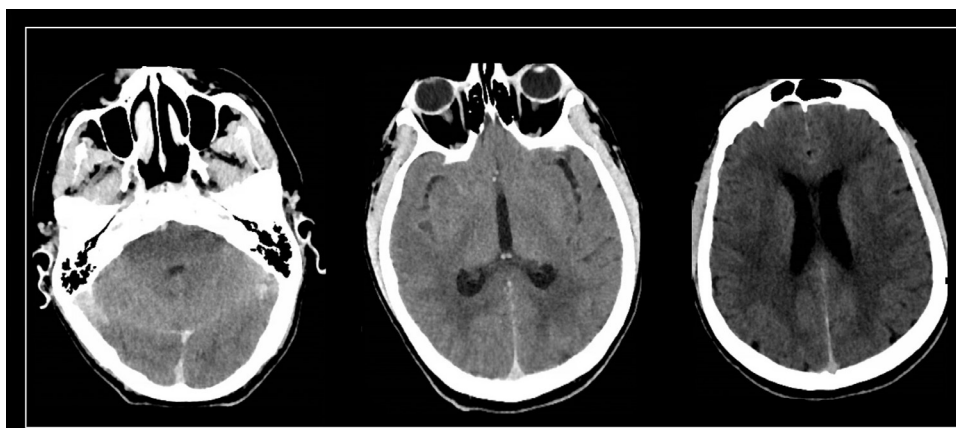


Fig. 1 – Computed tomography (CT) scan of the patient at the time of the presentation showing no evidence of intracranial bleeding and radiological findings suggestive of hypertensive encephalopathy; diffuse hypodensity of the white matter.

report isolated symptomatic and asymptomatic brain stem and spinal cord involvement in PRES [7–10], and names like atypical and variant PRES have been introduced to describe such phenomenon [6]. Herein, we describe a case of posterior reversible encephalopathy with isolated involvement of infratentorial structures.

Case report

A 42-year-old male patient with poorly controlled hypertension, not taking antihypertensive medications and poor medical follow-up, presented with hypertensive emergency. The patient was referred to the Emergency Department from the out-patient clinic with a blood pressure of 250/130. In the Emergency Department, the patient was treated with Metoprolol, Furosemide, Aspirin, and Lorazepam. The initial computed tomography (CT) scan showed some periventricular edema, mildly enlarged ventricles, and effacement of basilar cistern suggesting hypertensive emergency (Fig. 1).

The patient endorsed a 1-week history of morning bifrontal-occipital headache, associated with blurry vision, and some flashes of light and floaters over the past 4 days. Besides, the patient reported occasional neck stiffness, exertional dyspnea, and episodes of vomiting over the past 2 weeks before admission. He denied any orthopnea, leg swelling, agitation or aggressive behaviors, weakness, diplopia, numbness or tingling, back pain, skin rashes, fever, chills, or a tick bite recently. He acknowledged drinking 5–6 beers a day and smoking 1 pack of cigarette a day for the past 20 years. Neurological exam revealed significantly foggy mental status, but he was alert and oriented to person, place, and time. Small visual field defects in the left inferior temporal field and right inferior nasal field were also present. Color perception and visual acuity were intact—20/25 vision on the bedside vision test. The fundoscopic examination showed bilateral grade 4–5 papilledema and grade 4 bilateral hypertensive retinopathy. Otherwise, the neurological exam was unremarkable. His laboratory tests on admission were as the following: Creatinine, 2.32 mg/dL; troponin, 0.09 ng/mL; urine protein, 300 mg/dL;

magnesium, 2.3 mEq/L; and pro-BNP 21.38 pg/mL. Chest X-ray showed mild bilateral pleural effusion. Transesophageal echocardiography showed mild decreased left ventricular systolic performance with ejection fraction 48%. Renal ultrasound showed no evidence for renal artery stenosis. Cerebrospinal fluid and electroencephalogram examination were normal. Lyme disease antibodies and antinuclear antibody testing were negative. The erythrocyte sedimentation rate was within normal limits. Serum electrolytes and complete blood count were normal.

MRI showed numerous foci of confluent T2 hyperintensity involving most of the cerebellar hemispheres with additional confluent areas of the increased T2 signal in the brainstem involving most of the medulla, the pons and extending into the upper cervical spinal cord. These findings were suggestive of PRES. Moreover, extensive patchy and rounded foci of the increased T2 signal were seen in the deep, periventricular, and subcortical white matter; suggestive of chronic small vessel ischemic disease. There was also an increased T2 signal in the optic chiasm, prechiasmatic optic nerves, and the optic radiations. There was no diffusion restriction and no contrast enhancement (Fig. 2). Aquaporin-4 water channel auto-antibodies (also known as NMO-IgG; a biomarker for the diagnosis of neuromyelitis optica) were negative.

The patient was managed with clevidipine, antiepileptic medications, and other supportive measures. Within the next few days, blood pressure and clinical symptoms improved, and laboratory tests were completely normalized. A follow-up MRI, 60 days later, showed complete resolution of the brainstem, cerebellum and cervical spinal cord lesions (Fig. 3). Based on clinical and imaging findings, the diagnosis of PRES with isolated involvement of infratentorial structures was retrospectively made.

Discussion

This case demonstrates a rare presentation of PRES with isolated involvement of brainstem, cerebellum, and spinal cord. The case presentation was in the setting of hypertension

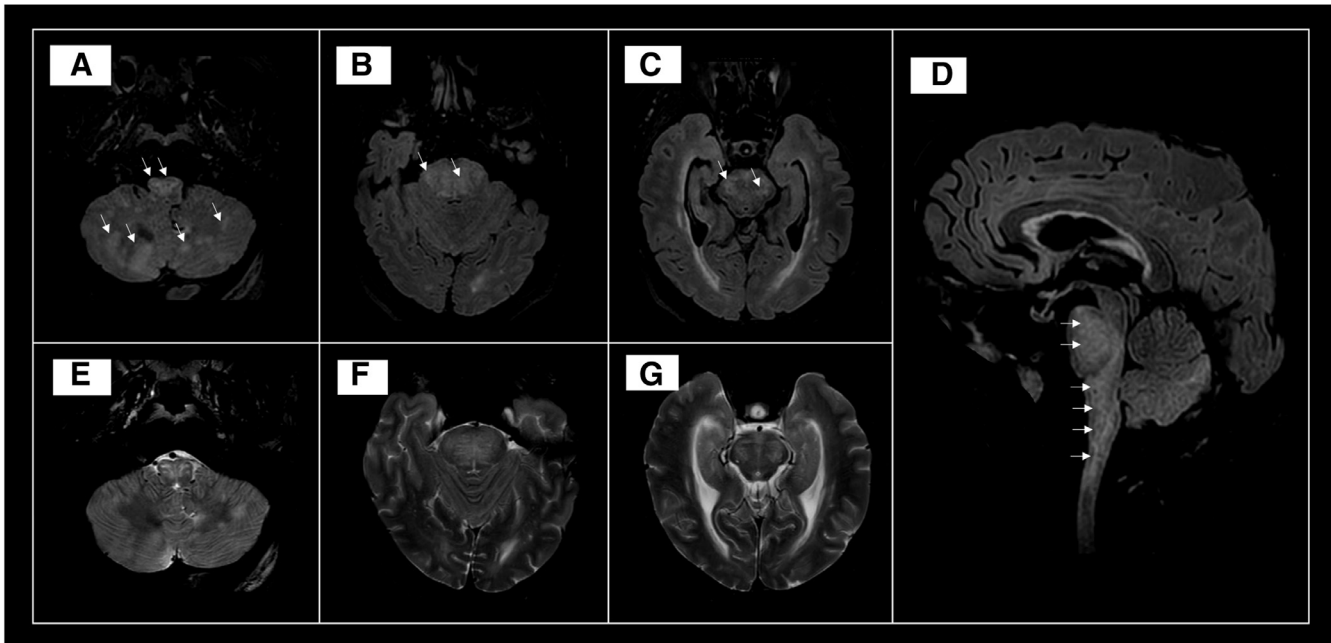


Fig. 2 – (A, B, C) Axial MRI fluid-attenuated inversion recovery (FLAIR), and (E, F, G) MRI T2-weighted MRI showing hyperintense lesions in the cerebellum, and brain stem without the typical parieto-occipital and temporal lesions. (D) A sagittal view of FLAIR images showing longitudinal hyperintense lesions involving the brainstem, cerebellum, and upper spinal cord (arrows indicate the abnormalities in each photo).

emergency with a complete reversal of the clinical and radiological findings after blood pressure normalization. Recently, isolated involvement of infratentorial structures in PRES has caught more attention. One case report and review of the literature done by Ou et al. have investigated various clinical and radiological features of isolated-infratentorial PRES variant compared to classical PRES [11]. The authors concluded that this variant of PRES is more common in males (70%), with a mean blood pressure of 216/135, and clear medical history of hypertension (46%), and renal impairment (51%). In our case, blood pressure at presentation was 250/130 and associated renal impairment was also present.

The exact pathophysiological mechanism underlying the development of PRES is still controversial. One theory suggests that when the elevated blood pressure exceeds the cerebrovascular auto-regulatory limits, vasodilation and disruption of the blood-brain barrier occurs leading to vasogenic edema and the characteristic MRI findings. On the other hand, the endothelial theory is based on the assumption that the initial trigger of the vasogenic edema occurs by endothelial cell injury, followed with the excessive release of cytokines and vasoactive molecules, increased vascular permeability, and interstitial edema [12]. Several papers have also highlighted the importance of the potential role of the lower density of sympathetic innervations in the posterior vertebrobasilar circulation in the higher susceptibility of the posterior regions of the brain to vasogenic edema [1,13].

The differential diagnosis of the brain stem, cerebellar and spinal cord lesions could include acute disseminated encephalomyelitis (ADEM), neuromyelitis optica (NMO), severe metabolic, toxic or electrolyte disturbance, multiple

sclerosis, and other demyelinating conditions. Although NMO can present with extensive longitudinal spinal cord lesions, similar to the ones seen in our patient, the clinical presentation was not consistent with NMO given the patient presentation in the settings of hypertensive emergency, and absence of signs of optic neuritis. Besides, aquaporin-4 water channel auto-antibodies (a diagnostic biomarker for NMO) were negative. Consistent with our initial diagnosis of PRES, a 60-day later follow-up MRI scan showed complete resolution of the brainstem, cerebellum, and cervical spinal cord lesions. In general, the MRI findings in PRES include T1 signal hypo-intensity, with variable enhancement, and T2 signal hyper-intensity. The diffusion-weighted imaging (DWI) are usually normal. The most distinguishing feature of PRES is the complete resolution of the imaging changes after management of the initial precipitating event. In NMO, the spinal cord involvement is usually more extensive (also known as longitudinally extensive spinal cord lesion) and go across more than 3 vertebral segments. Moreover, in NMO, the follow-up scans could show persistent T1 signal hypo-intensity and tissue atrophy due to inflammation [14]. On the other hand, lesions associated with ADEM are more common in children and typically preceded by a recent vaccination or viral infection. Besides, ADEM lesions are usually located in the periventricular-subcortical regions, with T2 signal hyper-intensity and, often, demonstrate ring enhancement (open ring sign) and peripherally restricted diffusion in DWI [15]. Collectively, the white matter lesions observed in our patient are mostly characteristic of PRES and retrospectively confirmed by the complete resolution in the follow-up scan. The subcortical white matter lesions observed in our patient could indicate

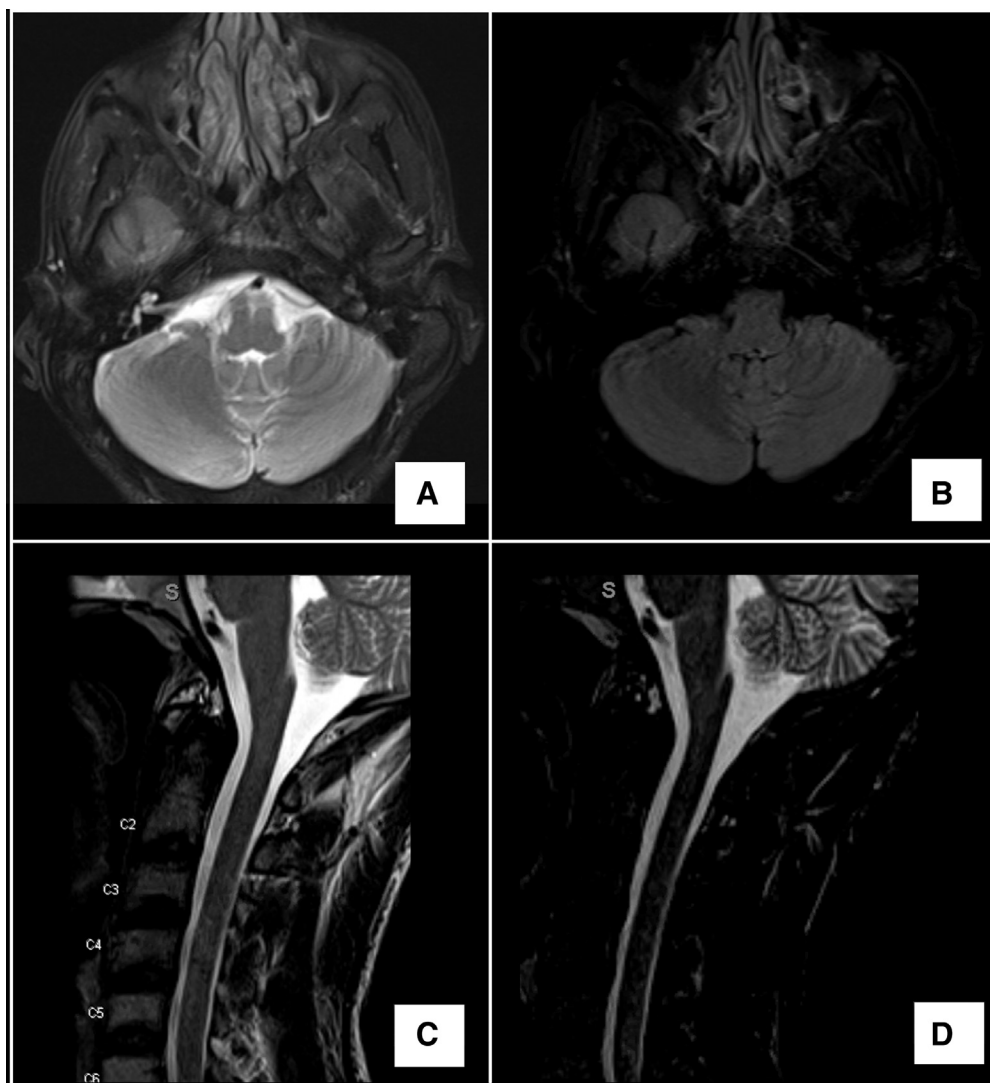


Fig. 3 – A follow up MRI (after 60 days) images show resolution of the brainstem, cerebellum, and spinal cord lesions.

a pre-existing chronic small vessel ischemic disease due to chronic uncontrolled hypertension or chronic renal disease.

In conclusion, we reported a rare case of variant-type PRES with isolated involvement of infratentorial brain structures highlighting the importance of recognizing this benign, treatable condition. Physicians should suspect PRES with spinal cord involvement in extreme elevation of blood pressure with MRI findings or PRES involving the infratentorial structures and cervico-medullary junction. Appropriate clinical and radiological correlation is crucial to avoid misdiagnosis of myelitis in these cases.

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