Posterior reversible encephalopathy syndrome: Atypical and unusual imaging manifestations.

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Authors: J. Saad¹, F. Marrakchi², F. Harbi¹, S. Alghamdi¹; ¹Nejran/SA, ²Monastir/TN
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Learning objectives

1) illustrates the variable presentations of PRES, including cases with atypical imaging findings. We illustrate cases of PRES with varying distributions of vasogenic oedema as well as cases with atypical imaging findings, such as variations of haemorrhage and restricted diffusion.

2) review the best imaging approach in such cases.

Background

Posterior reversible encephalopathy syndrome (PRES) a neuro- radiological syndrome characterized by seizures, altered level of consciousness and visual disturbance. Regardless of the underlying cause, the main abnormality is cerebral vasogenic edema, the pathogenesis of which is still under debate [1, 2]. PRES is typically reversible once the cause is removed. However, patients with severe manifestations of PRES, such as coma and/or status epilepticus, may require admission to the intensive care unit (ICU) [9, 10]. Moreover, permanent neurological impairment or death occurs in a minority of patients [5, 7, 8].

PRES is well recognized because of its typical imaging appearance that is involvement of the parieto-occipital regions. Other brain regions may also be affected and unusual imaging manifestations are observed frequently. Atypical imaging findings should not dissuade the diagnosis of PRES in the appropriate clinical situation, and knowledge of the varied appearance and atypical findings of PRES allows the radiologist to make this diagnosis.

Findings and procedure details

Posterior reversible encephalopathy syndrome (PRES) [1, 2] is a clinicoradiological entity that was well described by Hinchey et al.

This condition has been designated by a variety of names (reversible posterior leukoencephalopathy syndrome, reversible posterior cerebral edema syndrome, and reversible occipital syndrome, reversible posterior cerebral edema syndrome, and reversible occipital parietal encephalopathy).
PRES is now the accepted term [1, 2, 3] but has been challenged recently based on the risk of neurological impairment and up to 15 % mortality rate [7, 8].

**Diagnosis**

PRES is a clinicoradiological entity. The intensity and severity of clinical manifestations vary and may require ICU admission. Imaging findings also vary in severity. Thorough familiarity with the imaging criteria is crucial to the diagnosis. The combination of suggestive clinical manifestations and radiological criteria establishes the diagnosis of PRES.

In doubtful cases, the clinical and radiological improvement that occurs once appropriate treatment is given confirms the diagnosis. Nevertheless, there are no consensual guidelines to validate diagnosis of PRES.

**Clinical Manifestations of PRES**

PRES is characterized by variable associations of seizure activity, consciousness impairment, headaches, visual abnormalities, nausea, vomiting, and focal neurological signs.

Acute hypertension is not usually described among the main signs of PRES. However, hypertension has been reported in most studies [3-5, 7, 8, 11, 12], in 67 % [11] to 80 % [3] of patients.

**Radiological Characteristics of PRES**

CT findings are often normal or nonspecific [11]. Hypodensities in a suggestive topographic distribution suggest PRES (predominantly in parieto occipital white matter (Fig 1).

MRI: Cerebral MRI is the key investigation for the diagnosis of PRES.

Proton-density and T2-weighted images show regions of high signal indicating edema. Fluid-attenuated inversion recovery (FLAIR) sequences also visualize the lesions. The use of FLAIR has been shown to improve the diagnosis of PRES and the detection of subcortical and cortical lesions in PRES [6]. T1-weighted images show low-intensity foci. Diffusion-weighted imaging (DWI) is normal but the apparent diffusion coefficient is increased [13]. Finally, enhancement is seen in about half the cases [11].
The cerebral imaging abnormalities are often symmetric and predominate in the posterior white matter.

Imaging typically shows areas of bilateral hemispheric edema affecting parietal and occipital lobes, followed by frontal lobes, inferior temporal occipital junctions and cerebellum (Fig. 2, 3).

**Unusual Locations**

- Involvement basal ganglia, brainstem, and deep white matter (external / internal capsule, corona radiata, splenium of corpus callosum) are a less common but recognized part of PRES particularly when associated with abnormalities in the typical location (Fig. 4, 5, 6).

- Isolated unusual location in PRES, e.g central PRES with only basal ganglia and brainstem involvement, is rare but well known to occur.

- Medulla oblongata and spinal cord at least 4 cases described, all are associated with hypertension(Fig. 7).

- PRES may occasionally present with minimal or no detectable parietooccipital edema. In such cases, it is necessary to exclude other causes, such as myelinolysis or encephalomyelitis using clinical history and follow-up imaging, when necessary.

**Atypical manifestations of PRES**

- Enhancement (up to 37%) : cortical, leptomeningeal, parenchymal or pachymeningeal (Fig. 8, 9).

- Restricted diffusion (11-26%) (Fig. 10).

- Hemorrhage (10.5-17.1%) : parenchymal or subarachnoid(Fig. 11, 12).

- Unilateral hemispheric involvement (2.6%) (Fig. 13).

- Altered brain perfusion : regional decreased or increased, depends on disease time course (Fig. 14, 15).
Complications diagnosed radiologically at presentation of PRES

- Cerebral ischemia: Cerebral infarction is seen as high DWI signal intensity with a decrease in the apparent diffusion coefficient below 20%. Cerebral infarction is among the early signs of non-reversible damage associated with adverse outcomes [13]. This complication was present at the acute phase of PRES in 9 (10%) of the 82 patients with available DWI in one study [11] and in 5 (23%) of 22 patients in another [13]. In this setting, every effort must be taken to exclude a reversible cerebral vasoconstriction syndrome defined as at least two narrowings per artery on two different cerebral arteries at brain magnetic resonance angiography (MRA) or at conventional angiography [15]. Ducros et al. reported a 9% incidence of PRES in reversible cerebral vasoconstriction syndrome [15].

- Cerebral hemorrhage: Cerebral hemorrhage is uncommon in PRES (5% [7] to 17% [14] of patients). Reported cases were about evenly distributed in three categories, parenchymal hematoma, subarachnoid hemorrhage, and focal intraparenchymal hemorrhage [14, 16]. Cerebral hemorrhage may be more common among patients with allogeneic bone marrow transplantation or anticoagulant treatment, whereas blood pressure levels may have no influence on the bleeding risk [16]. A statistically significant association has been reported between edema severity on FLAIR sequences and bleeding risk [14]. In the presence of cerebral or subarachnoid hemorrhage, vascular imaging must rule out cerebral aneurysm and reversible cerebral vasoconstriction syndrome.

- Cerebral herniation: Posterior edema, particularly when located in the cerebellum and brainstem, may cause transtentorial cerebral herniation [17].

Retrospective Diagnosis of PRES after Regression of the Initial Clinical and Radiological Abnormalities
In some cases, the diagnosis of PRES remains in doubt. In this situation, regression of the clinical and radiological abnormalities with appropriate treatment supports the diagnosis. Thus, repeated brain imaging is helpful.

**Differential Diagnosis**

The non-specific clinical manifestations and multiplicity of radiological patterns raise diagnostic challenges. Many conditions may resemble PRES, including ictal or post-ictal state (with or without status epilepticus), progressive multifocal leukoencephalopathy (PML), severe leukoaraiosis, cerebral autosomal dominant arteriopathy with subcortical infarcts and leukoencephalopathy (CADASIL), infectious encephalitis, acute disseminated encephalomyelitis, mitochondrial myopathy encephalopathy lactacidosis and stroke-like episodes syndrome (MELAS), vasculitis, Creutzfeld-Jakob disease, cerebral venous sinus thrombosis, and ischemic stroke (watershed or posterior cerebral artery territory) [18, 19].

**Diagnostic Strategy**

The diagnostic strategy for PRES is fairly well standardized. After a careful history and thorough physical examination, investigations should be performed as appropriate, starting with the simplest and moving to the more sophisticated.

CT may be easier to obtain first. However, MRI must be performed, either as the first or as the second imaging study. MRI is considerably better than CT for the diagnosis of PRES and can provide information regarding many of the causes of PRES [1-3, 6, 7, 11, 13]. MRA must be added to MRI to identify an associated cerebral reversible vasoconstriction syndrome.

**Images for this section:**
Fig. 1: 28 years old female patient presented isolated headache followed by a generalized tonico-clonic seizure. Brain CT with injection of contrast was performed showing bilateral and symmetrical parieto-occipital hypodensities.
Fig. 2: The brain MRI performed after brain CT scan on the same day demonstrated bilateral parieto-occipital cortical and subcortical hyperintense lesions seen on T2 ad FLAIR weighted images (a, b). The apparent diffusion coefficient (ADC) was increased (c).

Fig. 3: A 12 yr-old-girl developed PRES after post streptococcal glomerulonephritis and hypertension. FLAIR images show high signal symmetrically involving bilateral parieto-occipital, posterior frontal, and temporo-occipital regions.
Fig. 4: A 6-year old boy with sickle cell anemia presented with alteration of consciousness, headaches and seizures, and found to have hypertension (200/100). FLAIR images show high signal symmetrically involving bilateral parieto-occipital lobes, frontal lobes, bilateral cerebellar hemispheres and splenium of corpus callosum.

Fig. 5: 39 year-old-female with severe hypertension, developed severe headache and visual disturbances. FLAIR images show high signal involving bilateral parieto-occipital lobes, frontal lobes, temporal lobes, bilateral basal ganglia, left thalamus, pons and cerebellum.
Fig. 6: A 10-year old boy with pyoderma gangrenosum, presents with headache and seizures. FLAIR images show high signal in parasagittal frontal lobes, right temporo-occipital lobe, midbrain, right basal ganglia and thalamus, genu of corpus callosum and cerebellum.
Fig. 7: 23 yr-old-male cocaine-induced malignant hypertension, presenting with headaches, confusion and spinal cord syndrome. T2W images show high signal in bilateral parietal lobes (a), medulla oblongata and cervical cord (b). Four weeks later follow up shows resolution of cord lesions (c).
Fig. 8: A 66 yr-old-female developed PRES after severe hypertension. FLAIR images (a,b) show high signal in bilateral parieto-occipital lobes, cerebellum and splenium of corpus callosum. Post gadolinium T1WIs (c,d) show cortical enhancement of lesions seen in FLAIR.
**Fig. 9:** A 9 year-old-boy developed PRES due to severe hypertension. FLAIR images (upper row) show high signal in bilateral parieto-occipital and frontal lobes. Post gadolinium images (bottom row) show leptomeningeal enhancement.

**Fig. 10:** A 66 yr-old-female developed PRES after severe hypertension. FLAIR image (a) shows high signal involving bilateral parieto-occipital lobes and splenium of the corpus callosum with restricted diffusion on DWI (b) and ADC map (c).
Fig. 11: 50 yr-old-female developed PRES after severe hypertension. FLAIR image (A, B) show high signal in bilateral occipital lobes, abnormal sulcal signal in left frontal region. There is low signal on GRE T2*W (C), corresponding with sulcal hyper attenuation on CT (D) due to subarachnoid hemorrhage.
**Fig. 12:** 52 yr-old-female post cardiac transplant developed PRES possibly caused by tacrolimus toxicity. FLAIR images (a,b) show high signal in bilateral parieto-occipital lobes. CT (c) done next day shows parenchymal hemorrhage in previous affected areas.

**Fig. 13:** 12 yr-old-male presenting with headache and seizures, found to have severe hypertension. T2W images show high signal predominantly in the left parieto-occipital and left temporal regions.
**Fig. 14:** A 10-year old boy with pyoderma gangrenosum, presents with headache and seizures. FLAIR images (a) show unusual locations of PRES with minimal involvement of fronto parietal lobes. ASL map (b) shows decreased perfusion (blood flow) in bilateral cerebral hemispheres, more on the left side.

**Fig. 15:** A case of 33 yr-old-female with PRES. DWI (a) and ADC (b) map show restricted fluid diffusion in bilateral occipital lobes. ASL map (c) shows increased perfusion in affected regions.
Conclusion

Neuroradiologists should be aware that atypical imaging manifestations of PRES are more common than commonly perceived. Recognition of atypical variants of PRES can be helpful to manage these patients and avoid complications in a timely manner.

Personal information

References


