PRES Syndrome

Reversible Posterior Leukoencephalopathy Syndrome

Introduction

Reversible posterior leukoencephalopathy syndrome (also known as reversible posterior leukoencephalopathy syndrome) is a **clinical radiographic syndrome** of heterogeneous etiologies that are grouped together because of similar findings on neuroimaging studies. It is characterized clinically by headache, abnormalities of mental status and visual perception, and seizures. Altered consciousness ranges from mild somnolence to confusion and agitation, progressing to stupor or coma in extreme cases. The headache is typically constant, nonlocalized, moderate to severe, and unresponsive to analgesia.Visual perception abnormalities are often detectable. Hemianopia, visual neglect, auras, visualhallucinations, and cortical blindness may occur. Seizures are usually generalized tonic-clonic. they may begin focally and often recur.

Despite its diverse causes, common precipitating factors are defined as abrupt elevations of blood pressure, renal decompensation, fluid retention, and immunosuppressive therap. It is often—but by no means always—associated with acute hypertension . Press is not always reversibe but If promptly recognized and treated, the clinical syndrome usually resolves within a week, and the changes seen in magnetic resonance imaging (MRI) resolve over days to weeks.

The cause of PRES is not clearly understood, but two main theories have been suggested regarding the mechanism of the disease process. One theory posits a **hypertension-induced autoregulatory failure**. The failure causes vasodilation and subsequently increases capillary hydrostatic pressure, leading to vasogenic edema. A second theory posits that excessive arteriolar **vasoconstriction** results in decreased blood flow, ischemia, and cytotoxic edema. The preferential involvement

of the parietal and occipital lobes is thought to be related to the relatively poor sympathetic innervation of the posterior circulation.

Etiology include severe hypertension (postpartum eclampsia/pre-eclampsia,acute glomerulonephritis) , hemolytic-uremic syndrome (HUS), thrombocytopenic thrombotic purpura (TTP), systemic lupus erythematosus (SLE) **and** drug toxicity

Complications

- Ischemia
- Intracranial hemorrhage (Figure 1)

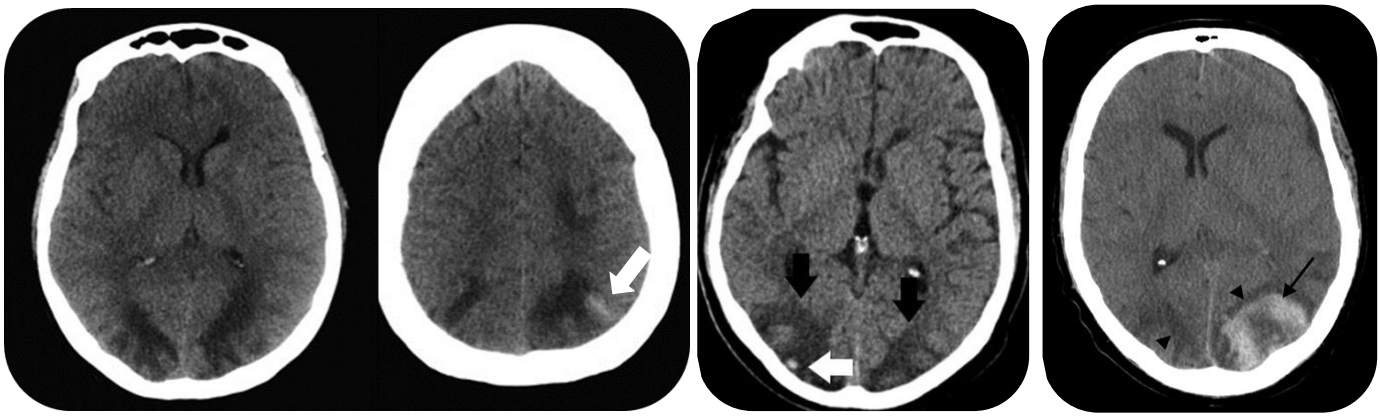

Figure 1

Diagnosis

There are no specific diagnostic criteria for PRES and in the appropriate clinical setting (in particular hypertension, immunosuppressive or cytotoxic therapy, kidney disease), clinicians should recognize the neurologic syndrome of headache, visual symptoms, confusion, and seizures, and order MRI, which typically supports the diagnosis. DWI, if available,adds considerable diagnostic and prognostic information.

Neuroimaging

Neuroimaging is essential to the diagnosis of RPLS. While a noncontrast MRI should be performed in all cases, a noncontrast CT is often the first study performed in the emergency department. Neuroradiographic abnormalities of RPLS are often apparent on CT scans but are best depicted by MRI

Radiographic features

Most commonly there is **vasogenic edema** within the **occipital** and **parietal** regions. The edema is usually **symmetrical**. Despite being termed posterior, posterior reversible encephalopathy syndrome can be found in a **non-posterior distribution**, mainly in watershed areas, including within the frontal, inferior temporal, cerebellar, and brainstem regions. (Figure 2) Both cortical and subcortical locations are affected. The distribution of abnormalities is usually not confined to a single vascular territory.

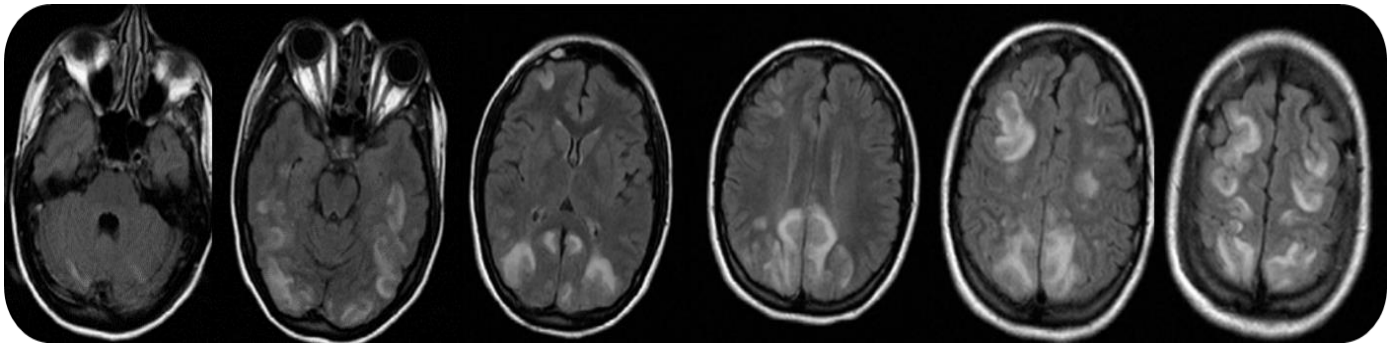

Figure2: FLAIR sequence showing bilateral high-signal foci in the cerebellum, basal ganglia, and occipital, parietal, frontal, and temporal lobes

There are three main imaging patterns:

- Holohemispheric at watershed zones
- Superior frontal sulcus
- Parieto-occipital dominance
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In **CT scan** the affected regions are hypoattenuating (Figure 3)

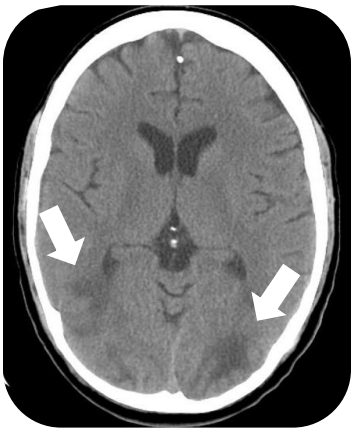


Figure 3

In **MRI** signal characteristics of affected areas usually reflect vasogenic edema. The most commonly observed abnormalities on MRI are focal or confluent areas of increased signal on T2-weighted images (Figure 4)

T1: hypointense in affected regions, **T2**: hyperintense in affected regions**, FlAIR** sequences improve sensitivity and detect subtle peripheral lesions**, DWI**: usually normal, sometimes hyperintense due to edema (T2 shine-through) or true restricted diffusion, **ADC**: usually increased signal due to increased diffusion, but restricted diffusion is present in a quarter of cases

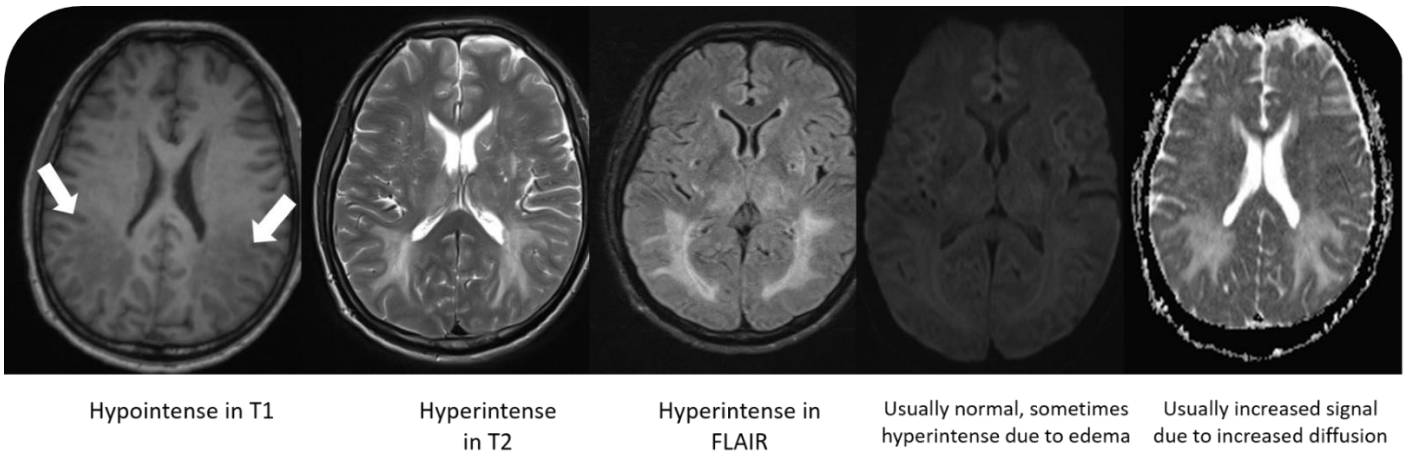


Figure 4

DWI aids in the distinction of **RPLS** from **top-of-the-basilar stroke**

The vasogenic edema that is characteristic of RPLS is usually visualized as a hypo- or isointense signal on DWI (sometimes slightly hyperintense due to T2 shine-through) and increased signal on apparent di usion coeficient

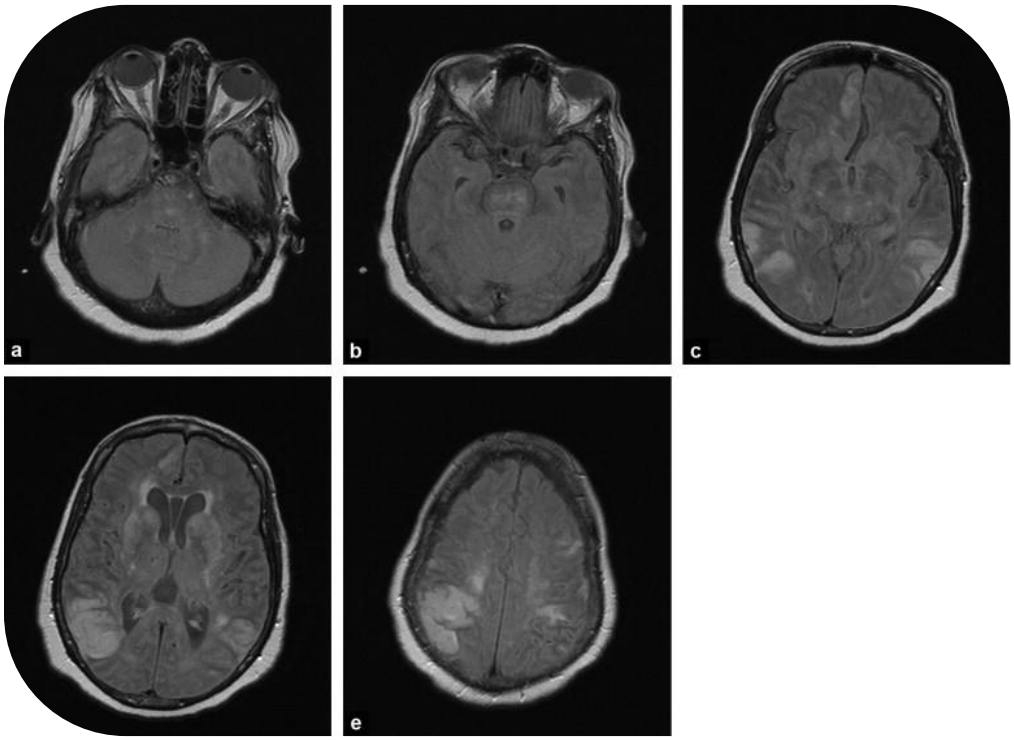
By contrast, acute cerebral infarction produces marked hyperintensity on DWI and hypointensity on ADC maps

Although the white matter edema is typically most prominent in both posterior cerebral hemispheres, the calcarine and paramedian parts of the occipital lobe are usually spared, helping to distinguish RPLS from bilateral posterior cerebral infarctions

Relative sparing of the cortical gray matter in RPLS also distinguishes this from posterior cerebral artery infarction

Atypical presentations

In rarer cases, the lesions can extend to the basal ganglia (14%), the brain stem (13%) and the deep white matter, in particular the splenium of the corpus callosum (10%). Even though involvement is unilateral, this must not mean that the diagnosis does not stand.



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