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The imaging spectrum of posterior reversible encephalopathy syndrome: A pictorial review

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ABSTRACT

Posterior reversible encephalopathy syndrome (PRES) is characterized by the acute onset of neurologic symptoms (headache, altered mental status, visual changes, seizures) with accompanying vasogenic edema on brain imaging. Risk factors for PRES include infection, uremia, malignancy, autoimmune disorders, the peripartum state and hypertension. PRES is classically described as being posterior (i.e. parieto-occipital) but radiologic variants are increasingly recognized. This pictorial review demonstrates the heterogeneity of the different radiologic presentations of PRES in reference to lesion distribution, hemorrhage, diffusion restriction, contrast enhancement, and other associated findings.

1. Introduction

Posterior reversible encephalopathy syndrome (PRES), also commonly referred to as reversible posterior leukoencephalopathy syndrome (RPLS), is a clinical and radiologic entity that involves the acute onset of neurologic symptoms accompanied by vasogenic edema, classically in a bilateral parieto-occipital distribution. PRES is associated with a wide variety of predisposing conditions, including acute hypertension, exposure to cytotoxic or immunosuppressant drugs, systemic infection, uremia, pregnancy, and autoimmune disorders. Cyclosporine is most commonly associated with PRES, but other medications including tacrolimus, cisplatin, rituximab, and bevacizumab have also been implicated [1–3]. The overall incidence of PRES is unknown, though it is observed to have a female predominance, likely secondary to its association with peripartum conditions and autoimmune diseases [1,4]. PRES affects both adult and pediatric populations, with reports in children as young as two years old and adults as old as 90 years old. Most cases are reported in individuals from 20 to 65 [5–7]. Neurologic symptoms typically include altered mental status, visual changes, headache, focal neurologic deficits, and seizures [1].

While the pathophysiology of PRES is debated, it is believed to involve dysfunction of cerebrovascular autoregulation and/or endothelial damage [6,8]. The leading theory of autoregulation and dysregulation posits that acute hypertension overwhelms the autoregulatory response and results in hyperperfusion, blood brain barrier dysfunction, and extravasation of plasma and macromolecules into the brain parenchyma, resulting in vasogenic edema. This hypothesis is supported by the fact that the distribution is often posterior, as it has been reported that the posterior circulation has less sympathetic innervation and ability to autoregulate [9]. In contrast, frequent distribution of vasogenic edema in watershed patterns has raised the possibility that hypoperfusion may play a role, perhaps secondary to exaggerated vasoconstriction and ischemia-related endothelial damage [10]. Additionally, there have been reports of PRES in normotensive patients, most often in the setting of exposure to cytotoxic or immunosuppressant drugs or sepsis, suggesting that inflammation and direct endothelial damage may be involved [3,11].

The acronym PRES is typically favored, but it is somewhat of a misnomer. While the classic presentation of PRES is posterior (i.e. parieto-occipital), atypical presentations have been increasingly appreciated [3,4,12]. Additionally, while PRES is largely reversible if diagnosed and treated promptly, the possibilities of irreversible parenchymal damage and poor clinical outcomes are seen in some patients [6,13,14]. The purpose of this paper is to demonstrate the heterogeneity of radiologic presentations and to emphasize that atypical imaging findings should not discourage a diagnosis of PRES in the appropriate clinical context. The clinical significance of certain imaging findings is also discussed.

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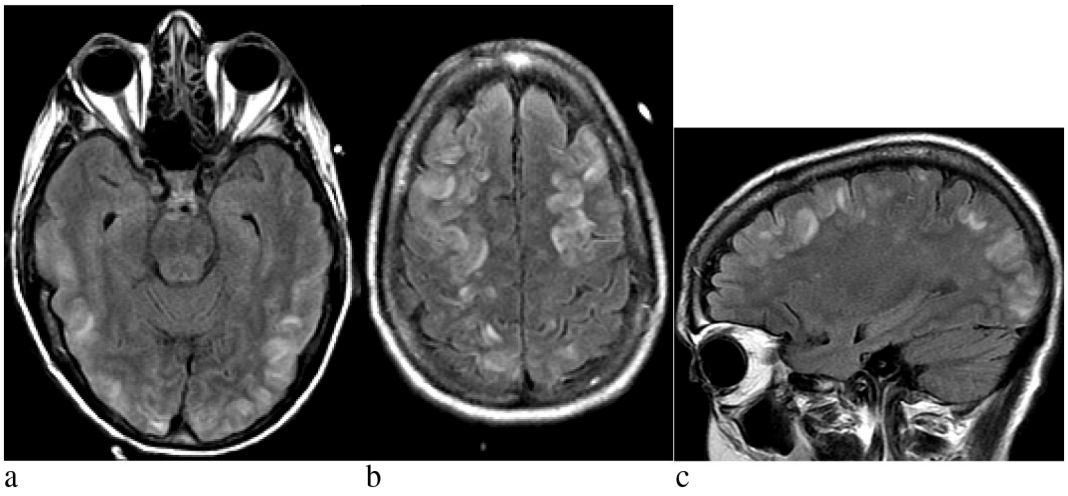

Fig. 1. MR images of a 66 year-old woman with metastatic rectal cancer on chemotherapy who presented to the ED with a tonic-clonic generalized seizure following 3 days of lethargy at home. Blood pressure was 181/87. Axial T2 FLAIR images demonstrate extensive vasogenic edema in a classic parietal-occipital distribution (a) as well as frontal lobes and along the superior frontal sulci (b, c).

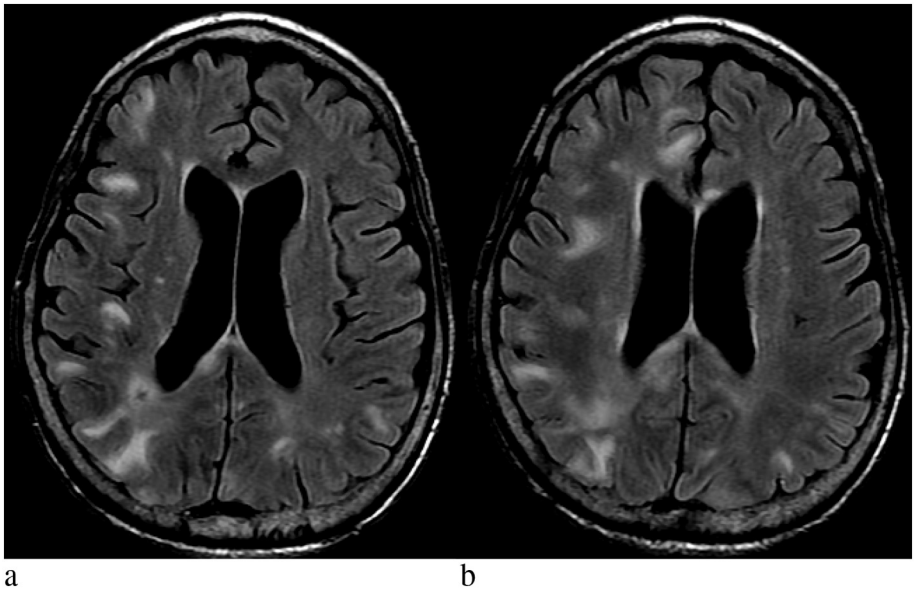

Fig. 2. MR images of a 65 year-old woman with follicular lymphoma with prior bone marrow transplant who was taking tacrolimus. She presented with altered mental status and fever and was found to have micrococcus bacteremia and a *Klebsiella* urinary tract infection. Blood pressure was 180/107. Axial T2 FLAIR images show extensive vasogenic edema involving the frontal, parietal, and occipital lobes as well as the corpus callosum (a–b).

2. Heterogeneity of imaging findings

2.1. Distribution of edema

Subcortical and cortical vasogenic edema is the hallmark of PRES and can be detected with high sensitivity using T2 Fluid-Attenuated-Inversion-Recovery (FLAIR) sequences. Parieto-occipital involvement is classic and seen in the majority of cases [4,12]. However, other locations including frontal and temporal lobes, cerebellum, brainstem and deep brain structures such as basal ganglia, thalamus, and corpus callosum are also involved to varying degrees. In particular, frontal lobe involvement has been reported in 50–79% of cases of PRES [4,12,15,16]. Frontal lobe abnormalities are typically located along the mid to posterior aspect of the superior frontal sulcus (Figs. 1, 2). According to Bartynski and Boardman, the superior frontal sulcus pattern is on a continuum of a more extensive holohemispheric watershed pattern, which was observed in 23% of their sample of 136 patients with PRES. In this distribution, a linear pattern spanned the frontal, parietal and occipital lobes, and to some extent the temporal lobes, likely at a watershed zone between the anterior cerebral artery and the posterior cerebral artery (ACA/PCA) and middle cerebral artery (MCA) territories [4]. While less common, atypical posterior fossa variants have been reported. Cerebellar involvement is seen in up to 34–53% of cases [12,17,18]. Similarly, brainstem involvement has been reported in 13% to 21% of cases [13,14]. Radiologists should exclude other causes of brainstem edema such as myelinolysis or encephalomyelitis, but PRES-related edema in the brainstem has been increasingly recognized as an atypical variant [12,19] (Figs. 3, 4). Thalamic and basal ganglia involvement has also been reported in 20–30% and 12–34% of cases, respectively [4,12,15,17,18] (Fig. 5).

The radiologic characteristics of pediatric PRES do not appear to be a distinct entity as compared to adults. While some studies have suggested a higher incidence of frontal lobe involvement, other atypical findings including involvement of the brainstem, cerebellum, and deep gray structures appear to be comparable to adults [5,20,21]. Similarly, differences in the radiologic appearance of PRES between men and women have not been reported.

2.2. Extent of vasogenic edema and severity classification

Extent of vasogenic edema has been classified with several different systems. Several studies employ a system of grading vasogenic edema