Posterior reversible encephalopathy syndrome in children: MRI findings.

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Learning objectives

The purpose of our educational exhibit is to present the most important brain magnetic resonance imaging (MRI) findings of posterior reversible encephalopathy syndrome in children (PRES).

Background

Posterior reversible encephalopathy syndrome (PRES) is a clinical neuroradiological entity, characterized by rapidly progressive signs and symptoms, such as headache, seizures, consciousness disturbance, and/or visual disturbances.

PRES was first described by Hinchey et al in 1996 and is commonly associated with a sudden increase in blood pressure.

Risk factors include hypertension, infection/sepsis, autoimmune disease, chemotherapy - immunosuppressive drugs, renal dysfunction, systemic lupus erythematosus, transplantation and pre/eclampsia.

An early etiologic diagnosis permits immediate correction of the cause of PRES. Patients may need blood pressure control, withdrawal of cancer chemotherapy or immunosuppressive agents, dialysis, or other interventions.

The global incidence of PRES is unknown.

Disordered cerebral autoregulation fails to undergo vasoconstriction in response to rise in blood pressure, with resulting fluid exudation and vasogenic edema. The posterior circulation is more prone to PRES due to lesser sympathetic innervations. Endothelial dysfunction (breakdown of the blood-brain barrier) is related to PRES correlated to chemotherapy, immunosuppressant agents and sepsis, even in the absence of elevated blood pressure.

Basic PRES pattern resembles the brain watershed zones, with the cortex and subcortical and deep white matter involved to varying degrees.

Although PRES commonly involves the parietal-occipital region, four different imaging patterns of PRES have been described.

a. Holohemispheric watershed pattern. A swath of confluent vasogenic edema extends through the frontal, parietal, and occipital lobes. Involvement of the temporal lobes is less noticeable. This topography matches the watershed zone
between the anterior and posterior cerebral arteries, on the one hand, and the middle cerebral artery, on the other.

b. Superior frontal sulcus pattern. Patchy edema predominates in the frontal lobes along the superior frontal sulci. The parietal and occipital lobes are involved variably.

c. Dominant parietal-occipital pattern. In this pattern that was previously thought to be typical of PRES, the posterior part of the parietal and occipital lobes is predominantly involved. The edema alters in severity from mild to extensive.

d. Partial or asymmetric expression of the primary patterns. The partial form is described as bilateral absence of edema in either the parietal or the occipital lobes. The frontal lobes are often involved. The asymmetric form is characterized by unilateral absence of edema in either a parietal or an occipital lobe. Finally, in the partial and asymmetric form, there is both absence of involvement of either the parietal or the occipital lobes and asymmetric abnormalities in the affected parietal or occipital lobes.

Recognition of PRES has evolved with increasing availability of MRI. MRI findings consist of vasogenic edema (T2-weighted and FLAIR images show hyperintense areas, T1-weighted images show hypointense areas and ADC maps demonstrate increased ADC values) in the white matter of the posterior regions of the cerebral hemispheres, particularly in the parietooccipital regions. Accordingly, frontal cortical and subcortical white matter involvement is present in most of cases. Less commonly, the brainstem, basal ganglia, and cerebellum are involved.

The cortex, structurally more tightly packed than the white matter, resists accumulation of edema, hence predilection of abnormalities to be seen in the white matter.

Asymmetrical involvement is not unusual and at times the grey matter is also extensively affected.

Atypical MRI features concerning distribution, completely unilateral involvement, hemorrhage and contrast enhancement can be encountered in PRES.

The extent of combined T2 and DWI signal abnormalities correlate with patient outcome. In PRES, the regions of vasogenic edema are visualized as a hypointense or isointense signals on DWI and as markedly increased signals on ADC maps compared with normal brain tissue.

High DWI signal intensity and pseudonormalized ADC values are associated with cerebral infarction and may represent the earliest sign of nonreversibility as severe vasogenic edema progresses to cytotoxic edema. Hemorrhage (focal hematoma, isolated sulcal/subarachnoid blood, or protein) is also seen related to reperfusion injury.
and is another potential complication of blood-brain-barrier dysfunction and cerebral edema.\(^7,8\)

Many conditions may resemble PRES, including mainly post-ictal state (with or without status epilepticus), progressive multifocal leukoencephalopathy (PML), mitochondrial myopathy encephalopathy, neoplasms, encephalitis, inflammatory and infectious processes, demyelinating pathology and cerebrovascular events.

PRES is not so often in children. Manifestations of PRES in children include seizures, headache and altered mental status. Other symptoms include nausea, vomiting and blurring of vision.\(^9\)

**Findings and procedure details**

We retrospectively reviewed brain MRI studies of nine children (five girls and four boys) diagnosed with PRES. The age range was 4 months to 14 years.

The risk factors for PRES documented were: administration of cancer chemotherapy agents (n=5)\(^3\) three patients suffered from acute leukemia and two patients suffered from lymphoma, sepsis (n=2), H1N1 infection (n=1) and systemic lupus erythematosus (n=1) not having undergone appropriate treatment. (figure-grafima)

Three of the children undergoing chemotherapy had presented an episode of elevated blood pressure. MRI images were evaluated for lesion distribution, contrast enhancement, diffusion weighted/ADC features. Involvement of the cerebellum, brainstem and basal ganglia was also documented. Complications diagnosed radiologically at presentation of PRES were also recorded. Followup MRI images were correlated to initial MRI findings.

- **MRI imaging patterns of PRES of our study group.**

  Holohemispheric watershed pattern was found in six children (66.6\%) (Figure 1). Dominant parietal-occipital pattern was present in two children (22.2\%) (Figure 2) while partial expression of the three primary patterns was found in only one child (11.1\%) (Figure 3). Superior frontal sulcus pattern was not encountered in any patient.

- **Contrast enhancement.**

  Contrast enhancement was noted in two of nine children (22.2\%) (Figures 4, 5), presumably reflecting disruption of the blood-brain barrier.

- **Diffusion Weighted Imaging (DWI) and Apparent Diffusion Coefficient (ADC).**
Normal DWI and increased ADC in PRES lesions, indicative of classic PRES appearance was demonstrated in initial MRI in six children (66.6%) (Figure 6). The remaining three children (33.3%) presented restriction of diffusion (high-signal intensities in DWI) (Figure 7).

Patients presenting diffusion restriction may have non-reversible damage.

· **Involvement of cerebellum, basal ganglia and brain stem.**

Cerebellum was affected by PRES in two children (22.2%) (Figures 6, 8). One of the previous children also demonstrated PRES in the basal ganglia (Figure 9). Two children in total presented lesions indicative of PRES in the basal ganglia (22.2%). An other child (11.1%) presented PRES at the brain stem (Figure 10).

· **Complications of PRES**

One child (11.1%) suffering from leukemia in the follow up MRI presented restriction of DWI (Figure 11) and new PRES lesions (Figure 12), indicative of complicated PRES.

Two of the children (22.2%) that presented diffusion restriction on the initial MRI scan also presented foci of intraparenchymal hemorrhage (Figure 13).

· **Follow-up**

Four of nine children (44.4%) underwent follow up MRI exams. PRES imaging findings resolved in three of four patients (Figure 14). One child presented major complications.

**Images for this section:**

![Images](image.png)

**Fig. 1:** A thirteen year old girl suffering from pneumonia presented an episode of elevated blood pressure. Axial T2 and FLAIR images show areas of bilateral hyperintense signal intensity characteristic of vasogenic edema of the frontal and parietal lobes, indicative of holohemispheric watershed pattern of PRES.
Fig. 2: Axial T2 and T1 images of a 4 1/2 year old girl demonstrates bilateral vasogenic edema in the white matter of the occipital and parietal lobes. This pattern is characterized as "classic".

Fig. 3: Eight year old boy suffering from acute lymphocytic leukemia undergoing chemotherapy. This patient presented neutropenic colitis due to cyclosporine and methotrexate. Axial FLAIR depicts asymmetrical and unilateral hyperintense signal intensities (vasogenic edema) of white matter in the frontal and parietal lobes of the right hemisphere.
Fig. 4: Axial T1 and T1 post contrast administration of a thirteen year old girl depicts linear enhancement within area of vasogenic edema at the left parietal lobe.

Fig. 5: Sagittal and coronal T1 images post contrast administration of a thirteen year old girl reveals nodular foci of enhancement of PRES lesions of the frontal, temporal and parietal lobes bilaterally.

Fig. 6: This thirteen year old boy with history of systemic lupus erythematosus not undergoing appropriate treatment presented multiple seizures. DWI does not depict diffusion restriction while ADC map shows elevated signal intensities at the left parietal lobe, the cerebellum and the basal ganglia on the left.

Fig. 7: DWI and ADC map of a 4 ½ year old girl with parietal-occipital pattern of PRES. DWI demonstrates, in addition to lesions of vasogenic edema, areas of diffusion
restriction while ADC depicts decreased signal intensity, indicating presence of cytogenic edema.
Fig. 8: This thirteen year old boy with history of systemic lupus erythematosus not undergoing appropriate treatment presented multiple seizures. Axial FLAIR image demonstrates hyperintense areas in the white matter of both cerebellar hemispheres.
**Fig. 9:** Four month old girl with septicemia and multiple organ dysfunction syndrome. Areas of high signal intensity on axial FLAIR image are depicted in basal ganglia bilaterally.

![Image of brain MRI showing septicemia and multiple organ dysfunction syndrome](image)

**Fig. 10:** Twelve year old girl with malignancy undergoing therapy presents holohemispheric watershed pattern of PRES. Note involvement of brainstem.

![Image of brain MRI showing PRES in a malignancy patient](image)

**Fig. 11:** Eight year old boy suffering from acute lymphocytic leukemia undergoing chemotherapy with diagnosis of PRES. Follow-up MRI performed due to deterioration of neurologic findings demonstrates restricted diffusion in the right hemisphere (parietal and frontal lobe).

![Image of brain MRI showing PRES in a leukemia patient](image)
Fig. 12: Eight year old boy suffering from acute lymphocytic leukemia undergoing chemotherapy. Follow-up MRI depicts additional PRES lesions.

Fig. 13: Four month old girl with septicemia and multiple organ dysfunction syndrome. Areas of restricted diffusion in basal ganglia, indicative of cytotoxic edema (complicated PRES) present foci of intraparenchymal hemorrhage (high signal intensity also on axial T1 image).
**Fig. 14:** Seven year old boy with H1N1 infection. Initial (A) and serial follow up (B and C) axial FLAIR images show gradual resolution of PRES lesions, with almost MRI normalization of brain parenchyma findings.
Conclusion

Prompt diagnosis of PRES in children, although not so often, is essential to avoid irreversible brain damage, such as cerebral infarction, bleeding, and death. MRI is crucial for diagnosing PRES, monitoring the course, and assessing treatment effectiveness.

Personal information

References


