Posterior Reversible Encephalopathy Syndrome in children: report of 10 cases

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Purpose

Posterior Reversible Encephalopathy Syndrome (PRES), named by Hinchey in 1996, is a clinical neuroradiological syndrome characterized by vasogenic subcortical edema without infarction that is often missed by clinicians. It may arise as a consequence of different conditions associated with hypertension or as an adverse effect of immunosuppressive treatment. The pathophysiology of PRES is unclear. According to the most widely accepted theory rapidly developing hypertension leads to a breakdown of cerebral autoregulation mechanisms, hyperperfusion and protein and fluid extravasation, resulting in focal vasogenic edema. According to other theories ischemia is the consequence of endothelial dysfunction or vasospasm.

The name Predominantly Reversible Encephalopathy Syndrome has recently been proposed as a more accurate description, because the radiographic lesions of PRES are rarely confined to the posterior parieto-occipital white matter, and often involve the cortex, frontal lobes, basal ganglia and brainstem (Fig 1 on page ).

The aim of this work was to define the typical radiological features of PRES and highlight the role of the radiologist and of brain MRI in the diagnostic work-up. The radiologist has an early opportunity to raise a diagnostic suspicion and an essential role in the timely recognition of PRES, allowing appropriate treatment in the reversible phase and reducing the risk of long-term consequences.
Methods and Materials

We present the cases of 10 pediatric patients who were diagnosed with PRES on neuroradiological findings and review their early and follow-up bio-clinical and MRI data.

- 6 patients (group 1) were apparently in good health until the onset of headache, fever, and vomiting, which were followed by partial seizures and visual disorders (diplopia, gaze deviation to the left, horizontal nystagmus);
- 4 patients (group 2) were receiving immunosuppressive treatment for acute leukemia and had sudden seizures; the most severely affected patient had severe oliguria, amaurosis and tonic-clonic seizures.

All 10 patients underwent brain MRI with turbo spin-echo DP-T2 and FLAIR sequences and spin-echo T1-weighted (T1W) sequences before and after gadolinium administration both in the work-up and in the follow-up phase (Figs 2a and 2b). Diffusion-weighted imaging (DWI) was also performed in 3 patients and brain CT in 4. Clinical, laboratory and EEG data were available for all patients.
Results

The CT scans demonstrated bilateral, predominantly subcortical low attenuation areas in the posterior temporo-parietal and occipital lobes (Figs. 3a and 4a) in 3/4 patients. In all 10 patients MRI disclosed non-enhancing bilateral, asymmetric areas of hyperintense signal, mainly located in subcortical areas and involving temporo-parietal and occipital areas on FLAIR and T2W sequences, and reduced signal on T1 sequences. DWI showed hyperintense signal due to a T2 shine-through effect, in the same regions, in 3/3 patients (Figs 2a,3b,4b,5a on page 6,7a).

It is important to emphasize that DWI with apparent diffusion coefficient (ADC) mapping can distinguish vasogenic edema related to PRES from the cytotoxic edema of acute ischemia. DWI could therefore be used for early diagnosis and follow-up of children with suspected PRES, to prevent progressive and irreversible neurological deterioration, ischemia and infarction. Whereas the cytotoxic edema related to acute ischemia involves reduced ADC and heightened signal on DWI, due to increased intracellular and decreased extracellular fluid, vasogenic edema is associated with elevated ADC values, and DWI documents iso-hypointense or, more frequently, hyperintense signal due to a T2 shine-through effect. ADC mapping is therefore mandatory to differentiate between the two types of edema.

The MRI findings of our patients appeared to be consistent with PRES and directed the subsequent work-up. In group 1 patients the MRI findings, added to the clinical data and the patient's history, led to investigation into the possible causes of PRES. All group 1 patients had hypertension, renal function abnormalities and bilateral slow wave activity on the EEG. Evaluation of C3, C4 and TAS titer led to a diagnosis of post-streptococcal glomerulonephritis in 5 patients. The more severe clinical condition of the sixth patient, who also had nephrocalcinosis and microscopic hematuria, was consistent with Dent syndrome and prompted a search for chloride channel gene mutations. In all 6 patients appropriate treatment led to complete lesion resolution on MRI (Figs 2a-b and 3b-c).

In the 4 leukemia patients, temporary suspension of the immunosuppressive treatment led to complete clearance of the encephalopathy lesions (Figs 7 and 4), in all but the most severely affected patient (Fig 5a-b).
Conclusion

Failure to recognize and treat PRES can have devastating consequences. In the presence of neurological symptoms, brain MRI rapidly confirms the diagnostic suspicion even in the absence of known disease, thus directing work-up and treatment. Brain MRI is highly sensitive to the diffuse changes related to vasogenic edema typical of PRES. In our pediatric patients it allowed early diagnosis and improved the neurological outcome, avoiding the long-term sequelae in most patients. We found no correlation between clinical characteristics and extent of the vasogenic edema seen on MRI. Also, we found no significant difference in lesion site in relation to suspected PRES etiology. However, despite the small patient sample, lesions were predominantly located in the right hemisphere in both groups (Fig 6), and in patients receiving immunosuppressive treatment they were predominantly found in the posterior and mesial portions of the brain (Fig 7a and 5a on page ), often also involving the cerebellum (Fig 8 on page ).
References
