Original Article

Posterior reversible encephalopathy syndrome

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Abstract

Objective: To evaluate the Magnetic Resonauce Imaging (MRI) features in patients having Posterior Reversible Encephalopathy Syndrome.

Methods: This is a retrospective study from 8th June 2005 to 26th July 2009. Twelve patients were included who were confirmed to have Posterior Reversible Encephalopathy Syndrome, per imaging and clinical follow-up. Two neuro-radiologists blinded to the clinical condition retrospectively reviewed each image. Standard sequences were unenhanced Fluid Attenuated Inversion Recovery (FLAIR), T1-weighted, T2- weighted images followed by diffusion-weighted imaging and contrast-enhanced T1-weighted imaging. The regions involved were recorded on the basis of these sequences.

Results: Abnormal T2-weighted hyperintense signals (indicating vasogenic oedema) were consistently present in the parietal or occipital regions in 5 (41.6%), but other locations were also involved, including the deep white matter in 3 (25%), frontal lobes in 1, inferior temporal lobes in 1, cerebellar hemispheres in 1, and basal ganglia in 1 (8.3% each). On follow-up examination after 5-7 weeks, the patients showed marked improvement clinically and on neuro-imaging, and were discharged in a stable condition. After administration of gadolinium contrast, there was no area of abnormal enhancement in 11 cases and minimal enhancement was seen in 1 case. In our series, 3 patients had follow-up MRI examination which revealed the resolution of previously seen changes as well as the resolution of clinical symptoms. However, the diagnosis of Posterior Reversible Encephalopathy Syndrome was established in 9 other patients by resolution of clinical symptoms alone in 2-3 weeks.

Conclusion: Awareness of diverse clinical and radiographic presentation of acute Posterior Reversible Encephalopathy Syndrome is essential to avoid misdiagnosis and treatment delay. Moreover, the syndrome is reversible with prompt treatment and has good outcome. This case series confirmed clinical improvement and recovery in most patients within weeks.

Keywords: Posterior Reversible Encephalopathy Syndrome, Imaging patterns, Magnetic resonance imaging. (JPMA 62: 657; 2012)

Introduction

Posterior Reversible Encephalopathy Syndrome (PRES) is most commonly identified in patients with eclampsia and in those who have undergone organ transplantation.[1] It may also occur in patients with other systemic conditions such as Wegener granulomatosis, systemic lupus erythematosus (SLE), non-specific renal inflammatory conditions (glomerulonephritis, hepatorenal syndrome), hypertension and post-chemotherapy.[2] Clinically patients with PRES have seizures, visual disturbances, altered mental status and headache.[3]

W.S. Bartynski et al in 2007, described vasogenic oedema in parietal or occipital regions 98%, frontal lobes 68%, inferior temporal lobes 40%, cerebellar hemispheres 30%, basal ganglia 14%, brainstem 13%, deep white matter 18%, splenium 10%.[4] Three major patterns of PRES were noted: the holohemispheric watershed 23%, superior frontal sulcal 27%, parietal occipital 22%, with additional common partial or asymmetric expression of these primary PRES patterns 28%. These demarcate lateral hemispheric blood supply (middle cerebral artery, MCA) and medial hemispheric supply (anterior cerebral artery ACA), posterior cerebral artery, PCA) and further reflect the junctional/watershed nature of PRES.

The findings on neuro-imaging in PRES include non-enhancing abnormal signals that appear as areas of low attenuation on CT scan and appear hypointense on T1-weighted imaging and hyperintense on T2-weighted imaging MRI. These abnormalities partially or completely resolve on follow-up scanning, thereby suggesting subcortical oedema without infarction.[5]

Lamy C et al in 2004 described highly characteristic neuro-imaging features include abnormal increased T2W/FLAIR signal, thought to be vasogenic oedema,

primarily within the white matter of the posterior circulation territories with typical sparing of the calcarine and paramedian regions of the occipital lobes.[6] The frontal/temporal white matter may be involved as well as gray matter structures such as the cortex and basal ganglia. Contrast enhancement has been described in the literature, but most of the reported contrast enhancement is secondary to complications such as subacute infarction.[6]

The hypothesis is that the typical changes noted in cases of PRES are reversible and is also associated with clinical improvement.

This retrospective study was conducted to see if the findings of earlier studies done globally in this regard were applicable on the local population in Karachi, Pakistan.

Patients and Methods

The retrospective study included cases between 8th June 2005 and 26th July 2009. Twelve patients were included as having confirmed PRES, per imaging and clinical follow-up. Standard sequences were unenhanced FLAIR, T1-weighted, T2-weighted images in all patients followed by diffusion weighted imaging and contrast-enhanced T1-weighted imaging. The regions involved were recorded on the basis of these sequences. All MRI scans had been performed on a SIEMENS AVANTO (1.5 Tesla) MRI scanner. The diagnosis of PRES was established if the follow-up imaging showed resolution of changes or if the patient's clinical condition resolved.

Inclusion criteria were records of all patients referred to the radiology department of Aga Khan University Hospital Karachi, with relevant clinical symptoms and/or signs and symptoms of PRES. Records of patients who were claustrophobic or had a pacemaker or an intracerebral aneurysm clip were excluded from the study.

All MRI records were reviewed by 2 neuro-radiologists - one with clinical experience of radiology of about 20 years and the other with experience of three years. If there was an initial disagreement, the final decision was reached by consensus.

Results

We identified 12 patients of PRES. All of them were females in the age range of 10-39 years. PRES occurred in association with eclampsia in 10, SLE in 1, and in post-renal transplant in 1 patient. Presenting symptoms included uncontrolled hypertension, clinical seizures, encephalopathy and headache. Abnormal T2-weighted hyperintense signal (indicating vasogenic oedema) was consistently present in the parietal or occipital regions in 5 (41.6%), but other locations were also involved, including the deep white matter in 3 (25%), frontal lobes in 1 (8.3%), inferior temporal lobes in 1 (8.3%), cerebellar hemispheres in 1 (8.3%) and basal ganglia in 1 (8.3%) (Table-1). These changes showed isointense to hypointense signal on T1-weighted, hyperintense on T2-weighted and FLAIR images (Figure). On diffusion-weighted images, there was no evidence of diffusion restriction. After administration of gadolinium contrast, there was no area of abnormal enhancement in 11 cases and minimal enhancement was seen in 1 case. The diagnosis of PRES was established if the follow-up imaging showed resolution of changes or if the patient's clinical condition resolved. In our series, 3 patients had follow-up MRI examination which revealed the resolution of previously seen changes as well as the resolution of clinical symptoms. However, the diagnosis of

Table-1: Distribution of abnormal signals in PRES patients (n=12).

Location	No. of patients	% Patient
Parietal Occipital	5	41.6 %
Deep White Matter	3	25 %
Frontal Lobes	1	8.3 %
Temporal Lobes	1	8.3 %
Cerebellum	1	8.3 %
Basal Ganglia	1	8.3 %

Table-2: Comparison of mr sequences in PRES patients with radiologic and clinical findings.

Case No	Age (year)	Sex	Cause	Radiological Distribution of Disease	Post Contrast	F/U Clinical	F/U Neuroimaging
1.	32	F	Eclampsia	Deep White Matter	No enhancement		Resolved
2.	20	F	Eclampsia	Deep White Matter	No enhancement	Improved	
3.	34	F	Eclampsia	Frontal	No enhancement	Improved	
4.	24	F	Eclampsia	Parieto-Occipital	Minimal en hancement		Resolved
5.	19	F	Eclampsia	Temporal	No enhancement		Resolved
6.	39	F	Post Renal Transplant	Deep White Matter	No enhancement	Improved	
7.	10	F	SLE	Parieto-Occipital	No enhancement	Improved	
8.	23	F	Eclampsia	Parieto-Occipital	No enhancement	Improved	
9.	20	F	Eclampsia	Cerebellum	No enhancement	Improved	
10.	25	F	Eclampsia	Basal Ganglia	No enhancement	Improved	
11.	19	F	Eclampsia	Parieto-Occipital	No enhancement	Improved	
12.	30	F	Eclampsia	Parieto-Occipital	No enhancement	Improved	

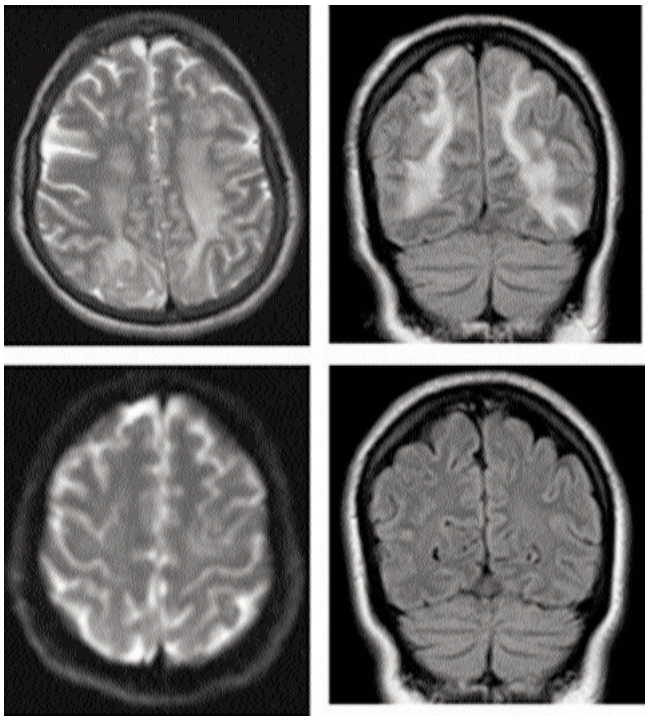

Figure: (a)- T2-weighted axial image shows diffuse hyperintense signal in deep white matter and subcortical white matter in bilateral frontal and occipital regions. (b)- Coronal FLAIR image shows diffuse hyperintense signal in deep white matter and subcortical white matter along the frontal and occipital regions. (c)- Diffusion-weighted image shows no evidence of diffusion restriction. (d)- Follow-up coronal FLAIR image shows complete resolution of previously noted abnormal signals.

PRES was established in the 9 other patients by resolution of clinical symptoms alone in 2-3 weeks and no follow-up imaging was done (Table-2).

Discussion

PRES is usually a reversible neurologic condition.[5] It has been suggested that in the pathogenesis of PRES there is a temporary failure of autoregulatory capabilities of the cerebral vessels, leading to hyperperfusion, a breakdown of blood brain barrier and consequent vasogenic oedema.[6] The typical imaging findings of PRES are most apparent in the parieto-occipital and posterior frontal cortical and subcortical white matter.[7] The changes of PRES can progress to ischaemia, cerebral infarction and even death.[8] On CT SCAN, abnormalities of PRES are seen as symmetric, bilateral areas of low attenuation of white matter.[9] On MRI, T1-weighted images show hypointense and hyperintense areas on T2-weighted and FLAIR images.[9] Atypical imaging appearances include contrast enhancement, haemorrhage and restricted diffusion on MRI.[10] If other than the parieto-occipital lobes are involved, the syndrome can be called atypical. In such cases, a diffusion weighted MRI with ADC mapping shows increased ADC values representing vasogenic oedema, thus differentiating atypical PRES from other brain disorders.[11] On MRA, reversible "vasculopathy" (diffuse/focal vasoconstriction) or vessel pruning is noted.[12] MRV tends to be normal. On Proton MR Spectroscopy reduced Sodium Acetylaspartate: Choline and Sodium Acetylaspartate: Creatine ratios have been described in regions of PRES vasogenic oedema.[13] Decreased perfusion has been noted on both brain single -photon emission CT technetium Tc 99m hexamethylpropyleneamine oxime imaging and by MR perfusion.[14]

Histologic evaluation of PRES is uncommon and often obtained late in the course of complex systemic disease.[15]

Rapid diagnosis of PRES is essential to prevent complications such as infarction and haemorrhage.[16]

A study of 76 patients by Alexander M. McKinney et al[17] in 2007, showed that the incidence of regions of involvement was parieto-occipital 98.7%, temporal 68.4%, thalamus 30.3%, cerebellum 34.2%, brainstem 18.4%, and basal ganglia 11.8%. The incidence of less common manifestations was enhancement 37.7%, restricted diffusion 17.3%, haemorrhage 17.1% and a newly described unilateral variant 2.6%.[17]

In our study the diagnosis of PRES could only be confirmed by resolution of the changes on follow-up imaging in 3 patients and in 9 patients clinical symptoms resolved after 2-3 weeks. Abnormal T2-Weighted hyperintense signal (indicating vasogenic oedema) was consistently present in the parietal or occipital regions in 5 (41.6%), but other locations were also involved including the deep white matter in 3 (25%), frontal lobes in 1 (8.3%), inferior temporal lobes in 1 (8.3%), cerebellar hemispheres in 1 (8.3%) and basal ganglia in 1 (8.3%). After administration of gadolinium contrast, there was no area of abnormal enhancement in 11 cases and minimal enhancement was seen in 1 case. We found no correlation between clinical characteristics and the extent of vasogenic oedema seen on brain imaging in PRES. We also found no overall difference in lesion location on the basis of suspected PRES etiology.

Conclusion

Awareness of the diverse clinical and radiographic presentation of acute PRES is essential to avoid misdiagnosis and treatment delay. It is imperative that the syndrome of PRES is correctly recognized on neuro-imaging, as the condition is reversible and potential complications can be avoided with appropriate therapy. This series of PRES cases have confirmed neuro-imaging and clinical improvement. Clinical recovery occurred in most patients within weeks.

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