Original Article

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Clinico-radiological Spectrum of Posterior Reversible Encephalopathy Syndrome: A Study from Teaching Hospital in North Karnataka

Umesh G Rajoor[1], B N Seema[2]

[1]Associate Professor, Department of Medicine, Koppal Institute of Medical Sciences, Koppal, Karnataka, India, [2]Assistant Professor, Department of OBG, Shamnur Shivashankarappa Institute of Medical Sciences & Research Centre, Davangere, Karnataka, India

Abstract

Background: Posterior reversible encephalopathy syndrome (PRES), is characterized by neuroimaging findings of reversible vasogenic subcortical edema without an infraction. A clinical diagnosis of PRES includes the presence of encephalopathy, seizures, headache, and visual symptoms, as well as radiologic findings of focal reversible vasogenic edema, best seen on magnetic resonance imaging of the brain.

Objective: To retrospectively identify patients with PRES with a characteristic clinical presentation and neuroimaging abnormalities.

Materials and Methods: A 54 patients were included in the study. Medical records of these patients were reviewed for demographic data, clinical history, blood pressure measurements, laboratory investigations, predisposing condition and neuroimaging. The primary etiology of PRES was determined for each case on the basis of the diagnosis of the attending clinician's.

Results: Out of the 54 retrospectively identified cases, 48 were females and 6 were males. Mean age of the patients at presentation was 30.94 years. The most common clinical presentation was seizures, seen in 44 patients (81.48%). Primary etiologies of PRES included hypertension (*n* = 21 [38.88%]), normotension (*n* = 07 [12.97%]), pre-eclampsia, or eclampsia (*n* = 14 [25.92%]).

Conclusion: PRES is an under-diagnosed condition, needs a high degree of suspicion for diagnosis. In this study, females are commonly affected, and most of them were in a postpartum period and had a good prognosis.

Key words: Encephalopathy, Pre-eclampsia, Vasogenic

INTRODUCTION

Reversible posterior leukoencephalopathy syndrome also referred as posterior reversible encephalopathy syndrome (PRES), is characterized by neuroimaging findings of reversible vasogenic subcortical edema without the infraction.[1] The clinical diagnosis of PRES includes the presence of encephalopathy, seizures, headache, and visual symptoms, as well as radiologic findings of focal reversible vasogenic edema, best seen on magnetic resonance imaging (MRI) of the brain.[2] This syndrome can be triggered by pre-eclampsia or eclampsia, hypertensive emergencies, renal disease, sepsis, exposure to immunosuppressive agents, and rarely autoimmune disorders.[2-6] Despite the syndrome's name, radiographic lesions in PRES are rarely isolated to the "posterior" parieto-occipital white matter and instead of involve the cortex, frontal lobes, basal ganglia, and the brain stem.[1,7] The pathophysiology of PRES remains elusive. Several theories have been proposed.[2] The hypertension related PRES is due failure of cerebrovascular autoregulation, which in turn results in vasogenic edema.[8] Non-hypertensive PRES may be due to the immune response to various stimuli. Early recognition of PRES is important for the timely institution of therapy. Over the past 5-10 years, it has been diagnosed the more frequent due to greater awareness and the availability of

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Corresponding Author: Dr. Umesh G Rajoor, Department of Medicine, Koppal Institute of Medical Sciences, Koppal - 583 231, Karnataka, India. E-mail: drumeshrajoor@gmail.com

better non-invasive diagnostic techniques. Though several studies were done in India and elsewhere on PRES, it has not been extensively studied of late. The purpose of this study was to describe the demographics and clinic-radiologic profile of PRES.

MATERIALS AND METHODS

We retrospectively identified patients with PRES between September 2011 and August 2014. 54 patients were included in the study. Medical records of these patients were reviewed for demographic data, clinical history, blood pressure measurements, laboratory investigations, predisposing condition, and neuroimaging. The primary etiology of PRES was determined for each case on the basis of the diagnosis of the attending clinician.

Inclusion Criteria

The presence of all three of the following criteria was mandatory for inclusion.

1. Clinical history of acute neurologic change including a headache, encephalopathy, convulsions, visual symptoms, or focal deficit
2. MRI brain findings of focal vasogenic edema
3. Clinical or radiologic proof of reversibility.

Exclusion Criteria

Cases with alternative diagnosis, not favoring inclusion criteria were excluded from the study.

Statistical Methods

The results were analyzed by calculating percentages, and the mean values.

Statistical Software

The statistical software namely SPSS 15.0, STATA 8.0, MEDCALC 9.0.1, and SYSTAT 11.0 were used for the analysis of the data and Microsoft word and excel have been used to generate the tables.

RESULTS

Demographics and baseline characteristics of the patients are depicted in Table 1.

Table 1: Demographics and baseline characteristics of the patients

Variable	Number of patients (%)
Demographics	
Males	13 (24.07)
Females	41 (75.93)
Male:female	1:8
Mean age	30.94
Clinical presentation	
Seizures	44 (81.48)
Headache	36 (66.67)
Vomiting	24 (44.44)
Altered sensorium	22 (40.74)
Visual disturbances	09 (16.67)
Etiology	
Acute hypertension	21 (38.88)
Normotensive	07 (12.97)
Pre-eclampsia or eclampsia	14 (25.92)
Sepsis	06 (11.11)
Renal failure	04 (7.40)
Immunosuppression	02 (3.7)
Neuroimaging findings	
Parieto-occipital lobes	27 (50)
Frontal lobe	14 (25.92)
Temporal lobe	09 (16.67)
Cerebellum	08 (14.81)
Basal ganglia	05 (9.25)
Brainstem	03 (5.55)
Subcortical	20 (37.03)
Cortical	03 (5.55)
Deep white matter	07 (12.96)

Demographic Profile

Out of the 54 retrospectively identified cases, 48 were females, and 6 were males. The mean age of the patients at presentation was 30.94 years (maximum-minimum, 17-80 years).

Clinical Profile

The most common clinical presentation was seizures, seen in 44 patients (81.48%), including 39 with generalized tonic-clonic seizures, 3 with focal seizures, and 2 with status epilepticus. One had a history of epilepsy. Other clinical presentations included headache in 36 patients (66.67%), vomiting in 24 patients (44.44%) and altered sensorium in 22 cases (40.74%), visual disturbances in 09 cases (16.67%).

Etiological Profile

Primary etiologies of PRES included hypertension (n = 21 [38.88%]), normotension (n = 07 [12.97%]), and pre-eclampsia or eclampsia (n = 14 [25.92%]). Out of 41 females, 26 were in postpartum period. Other causes were sepsis (n = 6 [11.11%], 3 had pneumonia and 1 had urosepsis), renal failure (n = 4 [7.40]), and 2 were on immunosuppressive medications.

Radiologic Findings (Figures 1-3)

The most common location was the parieto-occipital and cerebellar region (n = 35 [64.81%]), followed by frontal lobe (n = 14 [25.92%]), temporal lobe (n = 09 [16.67%]), and basal ganglia (n = 05 [9.25%]). 20 (37.03%) had cortical involvement, and 3 (5.55%) had subcortical involvement. The lesions were asymmetrical in 40 cases and symmetrical in 14 cases.

Outcome

53 patients recovered with the appropriate treatment, and one expired. Expired patients had severe sepsis with multiorgan dysfunction.

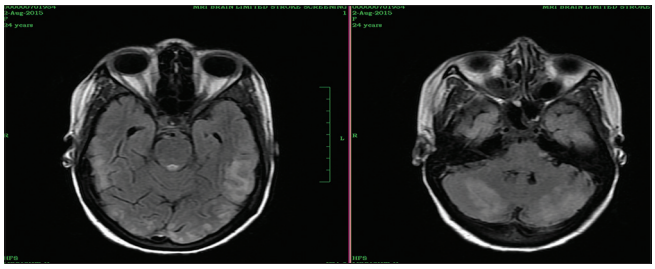

Figure 1: Patient 1-areas of altered signal intensity appearing hyperintense on fluid attenuation inversion recovery images seen in bilateral parieto-occipital region

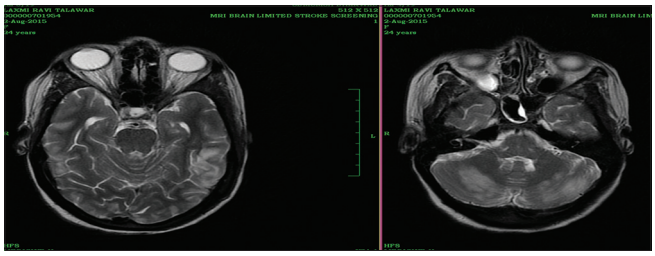

Figure 2: Patient 17-areas of altered signal intensity appearing hyperintense on fluid attenuation inversion recovery images seen in bilateral parieto-occipital region

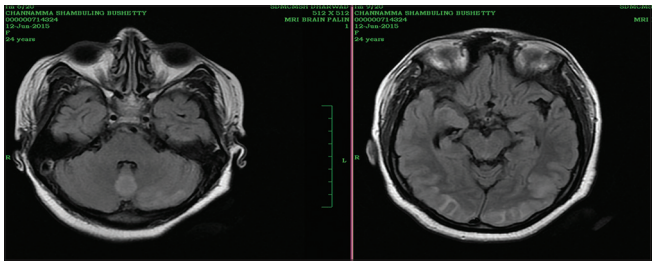

Figure 3: Patient 33-bilateral asymmetrical areas of hyperintense signal are seen in bilateral parieto-occipital areas

DISCUSSION

As the name implies, PRES is classically associated with the features of subcortical vasogenic edema, patchy symmetrical bilateral involvement with the preferential involvement of the posterior head regions, and complete clinical and radiological resolution.[9] The clinical signs and findings on the neuroimaging in patients with the PRES are consistent enough that this entity should be readily recognizable. Its causes are diverse, but common precipitants are acute elevations of blood pressure, renal decompensation, fluid retention, and treatment with immunosuppressive agents.[9]

The pathophysiology of PRES remains elusive. Several theories have been proposed, the most wide accepted of which states that rapidly developing hypertension leads to a breakdown in cerebral autoregulation, particularly in the posterior head region where there is a relative lack of sympathetic innveration. Hyperperfusion ensues with protein and fluid extravasation, producing focal vasogenic edema.[2,10] An alternative theory, which has been best characterized in pre-eclampsia, eclampsia, and sepsis implicates endothelial dysfunction. A third theory proposes that vasospasm with subsequent ischemia may be responsible.

Early recognition of PRES is important for the timely institution of therapy, which typically consists of gradual blood pressure control and withdrawal of potentially offending agents. Although reversible by definition, secondary complications, such as status epilepticus, intracranial hemorrhage, and massive ischemic infraction, can cause substantial morbidity and mortality.[11]

The observations are compared with the studies done by others on the PRES. In our study of 54 patients, maximum numbers of cases (76.93%) were seen in females in the age group of 21-40 years. This correlates well with a similar study by Fugate *et al.*[2] (65%). The mean age of onset in the present study was 30.94 years which is comparable with Patil *et al.*[12] study (29.90 years).

The most common clinical presentation was seizures, seen in 44 patients (81.48%), including 39 patients with generalized tonic-clonic seizures. These findings are comparable with Fugate *et al.*[2] study (74%).

Cho and Lee[13] in their study reported that PRES associated with the pregnancy, in the peripartum period presented with the seizures - Generalized tonic-clonic type, headache and visual disturbances. Similarly in our study, PRES is predominantly affected in postpartum female population.

Pedraza *et al.*[14] reported that PRES is most common associated with hypertension, pre-eclampsia-eclampsia, and HELLP syndrome. Similarly, findings are noted in our study also.

The most common location in the neuroimaging in this study was the parieto-occipital and cerebellar region (64.81%), followed by frontal lobe (25.92%), temporal lobe (16.67%), and basal ganglia (9.25%). These findings are comparable with Fugate *et al.*[2] study and McKinney *et al.*[15] study.

CONCLUSION

PRES is a challenging condition because of its variability of clinical symptoms and signs. A high index of clinical suspicion is needed to diagnose PRES. Data from observational studies on outcome and mortality in PRES scarce. We found many of the classic etiologies and triggering factors known to be

associated with PRES, including acute hypertension, pre-eclampsia-eclampsia, and sepsis. The overall prognosis for survival and functional independence is better in PRES when compare with other neurological conditions.

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