

Research Article

Posterior reversible encephalopathy syndrome: associated clinical and radiologic findings: a study from tertiary care hospital

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ABSTRACT

Background: Posterior reversible encephalopathy syndrome (PRES) is a syndrome with neuroimaging findings of reversible vasogenic subcortical edema without infraction. It is potentially reversible clinico-neuro-radiological syndrome featured by clinical symptoms of headache, altered mental status and seizures, visual perception defects in collaboration with radiological findings of posterior cerebral edema appearing as hypodense area on MRI and can also involve the brain stem, cerebellum and other cerebral areas. To identify the clinical associations and radiologic findings of posterior reversible encephalopathy syndrome (PRES).

Methods: One hundred twenty patients were included in the study. Demographic data, clinical history, blood pressure measurements, laboratory investigations, predisposing condition and neuroimaging were assessed. 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 one hundred twenty retrospectively identified cases, 98 were females and 22 were males. Mean age of the patients at presentation was 28.94 years. The most common clinical presentation was seizures, seen in 94 patients (78.3%). The most common location was the parieto-occipital 69 (57.5%), followed by temporal lobe 19 (15.8%), frontal lobe 14 (11.6%) and basal ganglia 08 (6.6%).

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

Keywords: Posterior reversible encephalopathy, Vasogenic edema, Preeclampsia

INTRODUCTION

Posterior reversible encephalopathy syndrome (PRES) a clinic-radiological term first described in 1996 by Hinchey and co-workers, as reversible posteriorleukoencephalopathy syndrome.¹ Classically the signs and symptoms are headache, visual disturbances, seizures, altered mental status and radiological findings of edema in the white matter of the brain areas perfused by the posterior brain circulation.² Most commonly seen in cases due to systemic hypertension (HTN) and other conditions identified as etiologic or risk factors in the absence of HTN are immunosuppressant drugs use,

nephrotic state, sepsis, and systemic lupus erythematosus (SLE).²⁻⁴

Hypertensive encephalopathy which complicates poorly controlled HTN, is the most common cause of PRES.^{5,6} In these patients the rate of increase of blood pressure is a more important factor in the development of PRES than the absolute BP levels.

The exact pathogenetic mechanism is not known. Nevertheless, regardless of the triggering factor, PRES involves the development of edema in the affected areas of the brain. In PRES, the autoregulatory response is

abnormal resulting in breakdown of the normal blood brain barrier (BBB) and in vasogenic brain edema.^{7,8} There is an apparent predisposition for edema to occur in the posterior CNS areas, particularly in the occipitoparietal areas.¹ This is thought to be secondary to the partial lack of sympathetic innervation of the vasculature that emerges from the basilar artery; however, other sites of the brain and cerebellum may be affected even when occipitoparietal areas are not involved.⁸

In non-hypertensive patients PRES may be due to immune response to various stimuli. Early recognition is important for timely initiation of therapy. Over the past 5 to 10 years it has been diagnosed more frequently due to greater awareness and the availability of better non-invasive diagnostic techniques. The purpose of this study was to describe the demographics and clinic-radiologic profile of PRES.

METHODS

We retrospectively identified patients with PRES between August 2014 to July 2015. One hundred twenty 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 etiology of PRES was determined for each case on the basis of the diagnosis of the attending clinician.

Inclusion criteria meeting all three

Clinical history of acute neurologic change including headache, encephalopathy, convulsions, visual symptoms, or focal deficit. MRI brain findings of focal vasogenic edema and clinical or radiologic proof of reversibility. Exclusion criteria were cases with alternative diagnosis, not favouring inclusion criteria were excluded from the study. Ethical clearance from the institutional ethics committee was gotten.

Statistical methods

The results were analysed by calculating percentages, and the mean values. Statistical software: The statistical software namely SPSS 15.0 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.

Out of the one hundred twenty patients retrospectively identified cases, 98 were females and 22 were males with ratio of 1:22. Mean age of the patients at presentation was 28.64 years (maximum - minimum, 17-77 years). The most common clinical presentation was seizures, seen in

94 patients (78.3%), including 76 with generalised tonic clonic seizures, 13 with focal seizures and 5 with status epilepticus. Other clinical presentations included altered sensorium in 66 cases (55%), headache in 35 patients (29.2%), vomiting in 29 patients (24.1%), and visual disturbances in 28 cases (28.3%).

Table 1: Demographics and baseline characteristics of the patients.

Variable	No. of patients (%)
Demographics	
Males	22 (28.4)
Females	98 (81.6)
Clinical presentation	
Seizures	94 (78.3)
Altered sensorium	66 (55)
Headache	35 (29.2)
Vomiting	29 (24.1)
Visual disturbances	28 (23.3)
Etiology	
Hypertension	59 (49.2)
Normotensive	08 (6.6)
preeclampsia/eclampsia	43 (35.8)
Sepsis	06 (5)
Renal failure	04 (3)
Neuroimaging findings	
Parieto-occipital lobes	69 (57.5)
Temporal lobe	19 (15.8)
Frontal lobe	14 (11.6)
Basal ganglia	08 (6.6)
Brainstem	07 (5.8)
others	03 (2.5)

Primary etiologies of PRES included hypertension 59 (49.2%), normotension 08 (6.6%), preeclampsia/eclampsia 43 (35.8%). Other causes were sepsis 6 (5%) and renal failure 4 (3%).

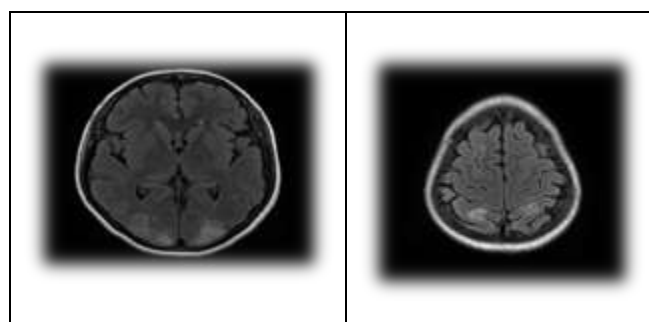


Figure 1: Areas of altered signal intensity appearing hyper intense on flair images seen in bilateral parieto-occipital region.

Radiologic findings

(Figure 1-3) the most common location was the parieto-occipital 69 (57.5 %), followed by temporal lobe 19

(15.8%), frontal lobe 14 (11.6%) and basal ganglia 08 (6.6%). The lesions were asymmetrical in 72 cases.

116 patients recovered with the appropriate treatment and 4 expired. Expired patients had multiorgan dysfunction and sepsis.

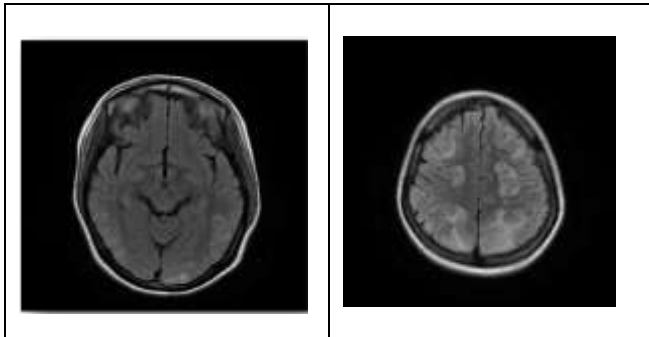


Figure 2: Demonstrates extensive holohemispheric pattern with vasogenic edema at the junction between the medial hemispheric (ACA, PCA) and lateral hemispheric (MCA) branches. The pattern outlines the entire supratentorial watershed or borderzone in the frontal, parietal, occipital and temporal lobes.

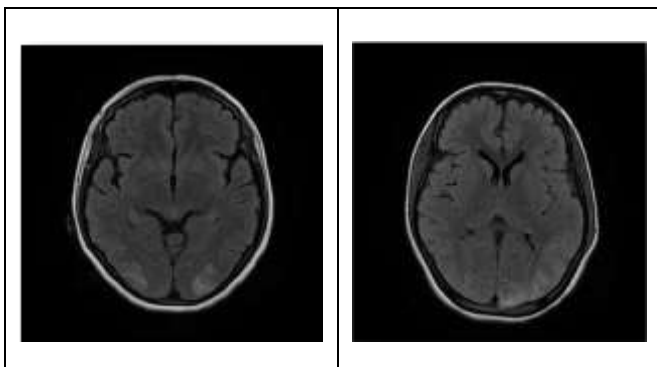


Figure 3: Patchy vasogenic edema in the bilateral occipital lobes and patchy edema in unilateral parietal lobe.

DISCUSSION

PRES is a clinico-radiologic syndrome characterized by headache, visual disturbance, altered consciousness or seizures. It occurs in hypertension, eclampsia, pre-eclampsia, immunosuppressive various anti-neoplastic agents, severe hypercalcaemia, thrombocytopenic syndromes, Henoch-Schonlein purpura, systemic lupus erythematous (SLE), renal failure, post-transplantation, infection, sepsis (gram positive organisms predominate) and shock.

PRES is usually associated with sub cortical vasogenic edema, patchy symmetrical bilateral involvement of the posterior head regions, and complete clinical and radiological resolution.⁹

The pathophysiology of PRES remains still a topic of discussion. Out of the several proposed theories, the most widely accepted theory states that rapidly developing hypertension leads to a breakdown in cerebral auto regulation, particularly in the posterior region where there is a relative lack of sympathetic innervation. Hyperperfusion ensues with protein and fluid extravasation, producing focal vasogenic edema.^{10,11} An alternative theory, which has been best characterized in preeclampsia, eclampsia, and sepsis implicates endothelial dysfunction. Early recognition leads to 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 infarction, can cause substantial morbidity and mortality.¹²

Our study of 120 patients were maximum numbers of cases 98 (81.66%) were seen in females in the age group of 21-40 years. This correlates well with a similar study by Fugate et al (65%).¹¹ Mean age of onset in the present study was 28.64 years which is comparable with Patil VC et al study (29.90 years).¹² The most common clinical presentation was seizures, seen in 94 patients (78.3%), including 76 with generalised tonic clonic seizures, 13 with focal seizures and 5 with status epilepticus. These findings are comparable with Fugate et al study (74%).² Cho et al in their study reported that PRES associated with the pregnancy, in the peripartum period presented with the seizures- generalised tonic clonic type, headache and visual disturbances.¹³ Similarly in our study PRES is predominantly affected in postpartum female population.

Pedraza R et al reported that PRES is most commonly associated with hypertension, preeclampsia-eclampsia and HELLP syndrome.¹⁴ Similarly findings are noted in our study also.

The most common location was the parieto-occipital 69 (57.5%), followed by temporal lobe 19 (15.8%), frontal lobe 14 (11.6%) and basal ganglia 08 (6.6%). The lesions were asymmetrical in 72 cases. These findings are comparable with Fugate et al study and Alexander et al study.¹⁵

CONCLUSION

PRES is more prevalent in women. It can occur over a wide age range. Brain edema at MR imaging in patients with PRES was not associated with hypertension level. Although the parietal and occipital lobes were the most frequently abnormal regions in our patients, involvement of the frontal and temporal lobes and basal ganglia are also common in PRES. Overall prognosis for survival and functional independence is better in PRES when compare with other neurological conditions.

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Conflict of interest: None declared

Ethical approval: The study was approved by the Institutional Ethics Committee

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