Posterior Reversible Encephalopathy Syndrome (PRES): A reversible neurological disorder an analysis through case series

Sajith Sebastian[1,*], Jenny Susan Roy[2], Catherine[3]

[1]Consultant Physician, Star Hospital's, Kollam, Kerala, [2,3]Resident, Dept. of Periodontics, AJ Dental College

***Corresponding Author:**
Email: drsaseneeruvelil@gmail.com

Abstract

Background: Aim of the study was to analyze 3 cases of PRES, a relatively uncommon disorder which is completely reversible and presenting as encephalopathy during a period of two year in a tertiary neurological centre in South India.

Material and Method: Total of 3 cases of PRESyndrome were analysed retrospectively in a period of 2 yrs during January 1, 2014 to January 1, 2016 to assess the various presentations of this rare syndrome and their outcomes. Cases were studied in detail including complete history, clinical examination and other findings were recorded.

Result: out of f 3 patients diagnosed with PRES were females between age of 19-29 years and were 5-8 days post partum. 66.6% presented as GTCS. 33.3% had history of systemic hypertension while 33.3% had a history of PIH and GDM. Only 33.3% presented with very high blood pressures on admission. MRI was diagnostic and suggested sub cortical vasogenic edema in all. All of the 3 patients recovered completely without residual neurological deficit or else required long term treatments.

Conclusion: PRES should be considered in patients who present with symptoms of seizures / convulsions, alter in consciousness, visual disturbances and or headache, particularly if associated with acute hypertension. PRES has commonly seen in patients with chronic and acute kidney disease, solid organ transplantation, and use of immuno-suppressive drugs. Typical MRI findings include reversible, symmetrical, posterior subcortical vasogenic edema. If recognized and treated early, the rapid onset symptoms and radiologic features usually completely resolve within days to weeks.

Key words: Encephalopathy, Systemic Hypertension, Headache.

Introduction

Posterior reversible encephalopathy syndrome (PRES) is an increasingly recognized disorder, with a wide clinical spectrum of both symptoms and triggers, and yet it remains poorly understood. This syndrome (PRES) is also known as reversible posterior leukoencephalopathy.(1) If promptly recognized and treated, the clinical syndrome usually resolves within a week, and the changes seen in magnetic resonance imaging (MRI) resolve over days to weeks.(2,3) The development of seizures in a patient following caesarean section will prompt towards a rather common diagnosis of eclampsia, CVT or IC bleed. Though cases of posterior reversible encephalopathy syndrome (PRES) at term are reported we tend to overlook this increasingly reported neurologic syndrome[4]. However, hypertension is absent in about one-fourth of patients and even if present, does not usually reach the level of failed auto regulation.(5) Early diagnosis and treatment of PRES will prevent progression to irreversible neurological damage.(6)

It is more common in females and no age group is spared.(7) Predisposing conditions include eclampsia, organ transplantation, during infections, sepsis, shock, autoimmune disease, and it can also occur following cancer chemotherapy.(5) The pathogenesis of PRES remains unclear, but deranged cerebral autoregulation and endothelial dysfunction are believed to play a major role.(8) Neuroimaging is essential for the diagnoses of PRES. Abnormalities are often apparent on computed tomography scans but are best identified by MRI.(9) The common findings observed are symmetrical white matter oedema in the posterior cerebral hemispheres, particularly the parieto-occipital regions, but variations may occur.(10) Atypical imaging features, such as involvement of anterior cerebral regions, deep white matter and brainstem are also frequently seen. Vasoconstriction is common in vascular imaging. If this syndrome is recognised and treated early its completely reversible.

Material and Method

Patients with clinically and radiologically diagnosed as PRES were identified retrospectively from the files from tertiary neurological centre in South India during a period of 2 years from January 1, 2014, through January 1, 2016. The presence of all 3 of the following criteria was mandatory for inclusion:

1. Clinical history of acute neurologic change including headache, encephalopathy, seizure, visual disturbance, or focal deficit
2. Brain imaging findings of focal vasogenic edema and
3. Clinical proof of reversibility.

Hypertension was defined as a systolic blood pressure of 140 mm Hg or greater or a diastolic blood pressure of 90 mm Hg or greater. Patient records were assessed for demographic data, clinical presentation, peak systolic and diastolic blood pressure measurements, co-morbid and predisposing conditions, and outcome. The primary etiology of PRES was

determined for each case on the basis of the diagnosis of the attending neurologist.

Results

Table: PRES Clinical Profile

Characteristics	Patient 1	Patient 2	Patient 3
Sex	Female	Female	Female
Post LSCS (Days)	08	08	05
Presenting complaints	GTCS, Headache X 3 day & Blurring of vision	GTCS	Severe headache X 1 day, Vomiting X 1 day
Pregnancy history	P2L3A1	PlL1A0	P2L2A0
Pregnancy co-morbidities	PIH, GDM, Polyhydrominos	Nil	Nil
Past history of co-morbidities	Hypothyroidism	Hypertension	Nil

LSCS-Lower Section Cesarean Section, GTCS -Generalized tonic-clonic seizure, GDM-Gestational Diabetes Mellitus, PIH-Pelvic Inflammatory disease, P2L3A1-Pregnant twice gravida, Live kids 3, One abortion, PlL1A0-Pregnant once, Live kids 1, No abortion, PlL1A0- Pregnant twice, Live kids 2, No abortion

Only 33% of patients diagnosed with PRES had maximum BP> 200/110 mm of Hg while 66% had maximum BP 140/90 mm of Hg recorded during the hospital stay. All patients had normal Thyroid, Creatinine and other routine laboratory values. The CT images of all patients were reported as normal in all. None of them had any residual neurological deficit on arrival and their Electroencephalogram (EEG) and Magnetic resonance venography (MRV) was reported as normal. All of them showed subcortical edema s/o vasogenic edema in MRI commonly in parieto-occipital region. All of the patients received symptomatic treatment with anti-epileptics and control of blood pressures along with supportive care. All were admitted and less than a week and neither had any residual weakness during stay or after 3 months of follow up.

Table 2: Parameters of the subjects

Parameters	Patient 1	Patient 2	Patient 3
BP (mmHg)	210/110	140/90	140/90
PR (/mt)	106	80	76
Neurological deficit	Nil	Nil	Nil
S. Cr (mg/dl)	0.9	0.6	0.6
TSH	2.08	1.73	1.5
RBS (mg/dl)	117	98	112
CT Brain	Nil Special	Nil Special	Nil
MRI	T2 and Flair hyperindensities on cortex white matter of B/1 parietal and occipital	T2 and Flair intensities in b/1parieto occipital region without diffusion restriction	T2 and Flair intensities showing vasogenic edema on left parietal
MRV	Normal	Normal	Normal
EEG	Normal	Normal	Normal
Duration of Hospitalization	6 days	7 days	6 days
Residual squeal	Nil	Nil	Nil

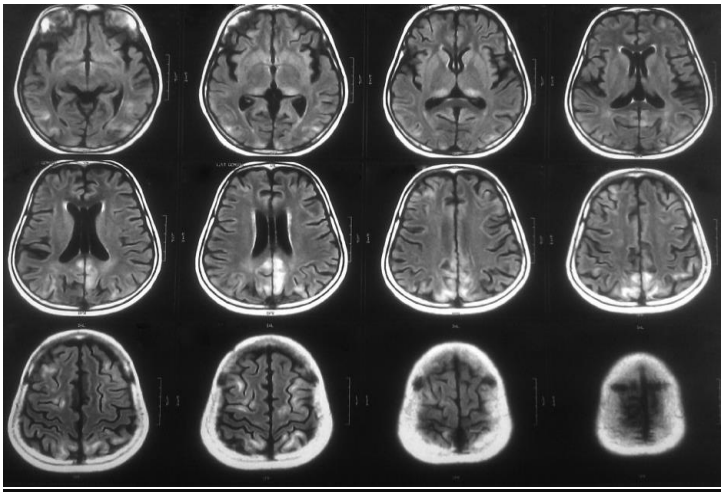

Fig. 1: MRI brain showed bilateral asymmetrical T2, FLAIR hyperintense lesions in cortical and subcortical location of parieto-occipital, temporal lobes, bilateral thalami, and cerebellum

Discussion

The cause of PRES still remains controversial, but the most popular theory is that severe hypertension causes interruption to brain auto regulation.[8,9] Breakdown in cerebral autoregulation occurs above a mean arterial blood pressure of 150 - 160 mmHg; in chronic hypertension, it occurs at higher pressures.[8] Uncontrolled hypertension leads to hyperperfusion and cerebral vessel endothelial damage, resulting in interstitial extravasation of proteins and fluids, causing vasogenic edema. Irreversible damage is commonly seen at mean arterial pressures above 200 mmHg[8]. Conditions commonly co-existing in PRES, such as chronic hypertension and atherosclerosis, are known to reduce the effectiveness of cerebral auto regulation. However, the auto regulation theory does not explain why blood pressure in PRES sometimes does not usually reach the upper limit of auto regulation[1] why PRES occurs in the absence of hypertension, and why the extent of the edema is not directly related to the severity of the blood pressure.[8,10] Furthermore, although this theory suggests brain hyperperfusion, however the positron-emission tomography studies shows demonstrates cerebral hypo perfusion in some cases which cannot be explained by this theory alone. An alternative theory is that PRES is a result of a systemic inflammatory state leading to endothelial dysfunction. That postulate is supported by the observation that PRES is usually associated with a systemic inflammatory process such as sepsis, eclampsia, transplantation, and autoimmune disease. Angiography in PRES demonstrates reversible focal and or diffuse abnormalities,[8] possibly reflect endothelial dysfunction. When blood pressure is high, the vasoconstriction that occurs during auto regulation could exacerbate such a pre-existing inflammatory endothelial dysfunction, causing hypoxia and subsequent vasogenic edema.[11] This mechanism would explain that the control of hypertension favours for recovery, but this fact does not explain why some cases of PRES seem to occur in the absence of any inflammatory state.[10] However, further researches are required to understand this potentially devastating but truly reversible and treatable condition.

Conclusion

PRES should always be considered in patients who present with seizures, altered consciousness, visual disturbance, or headache, particularly in the context of acute hypertension. PRES has been commonly associated with chronic and acute kidney disease, PIH, solid organ transplantation, and use of Immuno-suppressive drugs. Typical MRI findings include reversible, symmetrical, posterior subcortical cacogenic edema .If recognized and treated promptly, the rapid-onset symptoms and radiologic features usually fully resolve within days to weeks.

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