Case Report

Indian Journal of Emergency Medicine
Volume 2 Number 1, January - June 2016
DOI: http://dx.doi.org/10.21088/ijem.2395.311X.2116.7

Recurrent Posterior Reversible Encephalopathy Syndrome

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Abstract

Posterior reversible encephalopathy syndrome (PRES) is a reversible neurotoxic state that occurs secondary to the inability of posterior circulation to auto-regulate in response to acute changes in blood pressure. It is characterized by seizure, headaches, visual symptoms, impaired consciousness and other focal neurological findings. It is coupled with a unique CT or MR imaging appearance.

Recurrent PRES has been reported in severe hypertension and after allo-BMT [1,2]. In a recent reported series, recurrent PRES was noted in 3.8% of patients [3].

We describe a case of 62 year old woman with chronic obstructive pulmonary disease and hypertension who presented with two days history of blurring of vision, altered sensorium, headache and developed 2 episodes of generalized tonic clonic seizures after coming to hospital. On MRI Brain, hyper intense lesions seen in the bilateral parieto-occipital lobes with diffusion restriction, suggesting possibility of Posterior Reversible Encephalopathy Syndrome. Patient had history of similar episode 17 months back. CECT brain done at that time had features persistent with Posterior Reversible Encephalopathy Syndrome.

Keywords: Posterior Reversible Encephalopathy Syndrome; Hypertension; Seizure; Recurrent PRES; Hypertensive Encephalopathy.

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Introduction

Posterior reversible encephalopathy syndrome (PRES) is a clinical and radiologic entity characterized by headache, variable mental status, epilepsy, visual disturbances, and typical transient changes in the posterior cerebral perfusion [4].

More than 70% of patients with PRES are hypertensive, though a significant proportion have normal or only mildly raised blood pressure. Peak systolic blood pressure is usually between 170 mmHg and 190 mmHg [5,6]. It evolves over a matter of hours, with the most common presenting symptoms being seizures, disturbed vision, headache, and altered mental state [5].

The lesions generally disappear with appropriate treatment, although reversibility of the lesions depends on the underlying disease and location of the lesions.

Case Report

A 62 year old female with chronic obstructive pulmonary disease and hypertension presented with two days history of blurring of vision, altered sensorium, headache and developed 2 episodes of generalized tonic clonic seizures after coming to hospital.

On examination, the blood pressure was 174/96 mmHg and other vital signs were normal. Neurological findings showed no abnormalities. Laboratory data was also within normal limits.

Patient was started on antiepileptics. She was on mechanical ventilator support for 2 days.

On MRI Brain with contrast, T2 and FLAIR hyper intense lesions noted in the bilateral parieto-occipital lobes with diffusion restriction, suggesting possibility of Posterior Reversible Encephalopathy Syndrome.

Patient was managed conservatively and improved symptomatically. Repeat MRI done after 72 hours was normal.

Patient had history of similar episode 17 months back for which CECT brain was done. On CECT brain, hypodensity noted in the white matter of bilateral parieto-occipital lobes without any contrast enhancement. These features are persistent with Posterior Reversible Encephalopathy Syndrome.

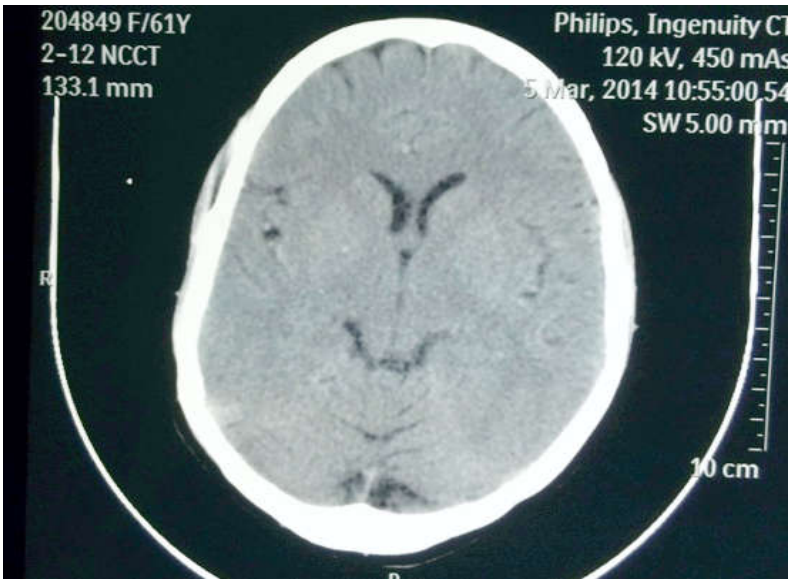

Fig. 1:

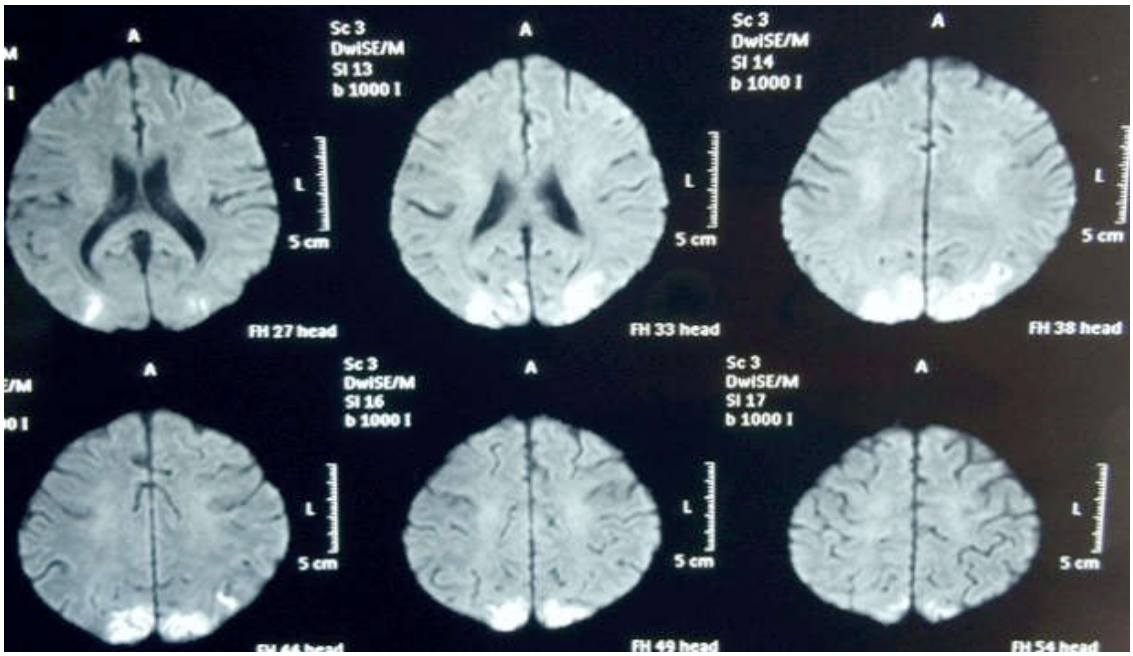

Fig. 2:

Discussion

PRES is defined by reversible cerebral edema due to dysfunction of the cerebrovascular blood-brain barrier unit. The pathophysiology of PRES is thought to result from abnormalities in the transmembrane flow of intravascular fluid and proteins caused by two phenomena: (1) cerebral autoregulatory failure and (2) loss of integrity of the blood-brain barrier [7]. A sudden rise of blood pressure, which exceeds the autoregulatory capability of the brain, causes regions of vasodilation and vasoconstriction to develop. The endothelial dysfunction causes a breakdown of the

blood-brain barrier allowing for focal transudation of fluid and edema formation [4,8,9]. This creates hypertensive encephalopathy with vasogenic edema with clinical and imaging manifestations that resolve with blood pressure control [4]. A range of precipitating etiologies has been described in literature including hypertension, preeclampsia/eclampsia, hypercalcemia, uremia, porphyria, and neurotoxicity secondary to immunosuppressants such as cyclosporine [4].

Neuroimaging is essential to the diagnosis of PRES. Neuroradiographic abnormalities of PRES are often apparent on CT scans but are best depicted by MRI. It can be diagnosed with reversible hyperintensities on T2-weighted cranial MR-images. Predominantly, it affects the territory of the posterior circulation and the clinical hallmarks are headache, confusion, seizures, cortical visual disturbances or blindness and, less common, other focal neurological signs. Headache is usually constant, nonlocalized and unresponsive to analgesia. The fundoscopic examination is usually normal but papilledema may be present with flame-shaped haemorrhages and exudates. The deep tendon reflexes are frequently brisk with babinski sign often present. There are no specific diagnostic criteria for PRES. It mostly is a benign, reversible condition, especially once the causative factor can be eliminated.

Recurrence of PRES due to various etiologies, though uncommon, does occur. It is associated with a rapid rise in blood pressure and affects patients that may be predisposed to this cerebral dysregulation. Recurrent PRES has been anecdotally reported in severe hypertension and after allo-BMT [1,2]. In a recent reported series, recurrent PRES was noted in 3 (3.8%) of 78 patients and was associated with sickle-cell disease with infection, allo-BMT with infection, or atypical autoimmune disease and possible viral infection [3].

References

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