INTRODUCTION

Posterior reversible encephalopathy syndrome (PRES) also known as reversible posterior leukoencephalopathy syndrome (RPLS) was first described by Hinchey in 1996.1 It is considered to be a potentially reversible clinico-neuro-radiological syndrome characterized by clinical symptoms of headache, visual perception defects, altered mental status and seizures, in conjunction with radiological findings of mainly posterior cerebral as oedema appears as hypodense area on MRI and can also involve the brain stem, cerebellum and other cerebral areas.2,3

The main aim of this report is to describe the association of PRES syndrome with hypertensive states in pregnancy as timely diagnosis and treatment would prevent permanent neurological sequelae.

CASE REPORT

A 30 years old women, third gravida, second para presented to the Emergency Department of Liaquat National Hospital, Karachi, on the 7th day postpartum after she experienced headache, blurring of vision, generalized body weakness, nausea, low grade fever and one episode of generalized tonic clonic fit.

She was booked at the end of second trimester and had regular antenatal visits thereafter. All antenatal laboratory values were within normal limits but her blood pressure was recorded on and off in mild high range (130-140/85-90 mmHg) and was diagnosed as a case of pregnancy induced hypertension, though she was never prescribed any antihypertensive medications. At 38 weeks of gestation, she electively underwent caesarean section due to previous two caesarean deliveries. An alive baby girl of 2.9 kg with good Apgar scores was delivered and discharged on request on the third postoperative day. On the fifth postoperative day, she had developed severe headache and high blood pressure, ranging between 160-180/100-110 mmHg and consulted local general physician. On the sixth postoperative day due to same complaints of headache and persistently high blood pressure she was admitted at peripheral hospital where she had one episode of generalized tonic clonic fit and received initial treatment. Due to non-availability of tertiary facilities she was referred to Liaquat National Hospital, Karachi.

On admission to the Emergency Department, the patient was fully conscious, ambulant and oriented to time, place and person. She had a pulse of 110 /minute, blood pressure of 150/100 mmHg, respiratory rate 28 breaths/minute and temperature of 100°F. General physical examination was normal. Pupillary reactions and fundoscopic examination were also normal and Plantars were flexor in response. All systemic examinations including abdominal, respiratory, cardiovascular were all unremarkable.

The complete blood count, renal function tests, liver function tests, ECG and clotting profile were within normal limits. Electroencephalogram (EEG) did not show any underlying seizure disorder. MRI of brain showed bilateral symmetrical and multifocal cortical and sub-cortical abnormal signal intensity involving parietal and occipital regions in T1, T2 weighted and FLAIR sequences (Figures 1 and 2), consistent with PRES. MRA (magnetic resonance angiography) and MRV (magnetic resonance venography) were normal. She was admitted in the medical HDU (high dependency unit). Her blood pressure was stabilized on anti-hypertensive medications, Apresoline, 250 mg TDS and Minipress, 1 mg BD and loaded with anti-epileptic, Phenytoin 1.2 gms intravenous and was continued as maintenance in a dose of 500 mg BD. She was discharged on same medications on 8th day post-
admission after clinical improvement in symptoms and followed-up in the outpatient department. MRI at one month of initiation of treatment showed resolution of pathological lesions.

**DISCUSSION**

PRES is a clinico-radiologic syndrome with symptoms of headache, visual disturbance, altered consciousness or fits. It may occur in diverse situations, including hypertension, eclampsia, pre-eclampsia, immunosuppressive medications such as cyclosporine, various anti-neoplastic agents, severe hypercalcaemia, thrombocytopenic syndromes, Henoch-Schonlein purpura, haemolytic uraemic syndrome, amyloid angiopathy, systemic lupus erythomatosus (SLE), renal failure, post-transplantation, infection, sepsis (gram positive organisms predominate) and shock.°

Parieto-occipital regions were the most commonly involved (94%) followed by the frontal lobe (77%), temporal lobe (64%), and cerebellum (53%). Cerebellar involvement was significantly more frequent in patients with a history of autoimmunity and patients with sepsis were more likely to have cortical involvement.°

The concept that eclampsia can cause PRES has arisen from numerous similarities in clinical presentation. These include comparable imaging findings on computed tomography (CT) and magnetic resonance imaging (MRI), the same neurologic symptoms (headache, vomiting, cortical blindness, and seizures) and prompt reversibility of symptoms after blood pressure being brought to normal.

The underlying pathophysiology of PRES remains elusive. Several theories have been proposed. The most widely 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 innervation). Hyperperfusion ensues with protein and fluid extravasation, producing focal vasogenic oedema. 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 ischaemia may be responsible.°

Diffusion-weighted MRI has provided insights into the pathogenesis of reversible posterior leukoencephalopathy syndrome by showing that signal abnormalities on T2-weighted MR images are associated with vasogenic rather than cytotoxic oedema. However, on diffusion-weighted images, lesions of reversible posterior leukoencephalopathy syndrome often appear isointense rather than hypointense as expected in vasogenic oedema. This finding is probably caused by the net effect of a combination of decreased signal intensity on diffusion-weighted images (from vasogenic oedema) and increased signal intensity caused by T2 prolongation effects (T2 "shine-through" effect).° Apart from a significant higher number of involved brain regions and a tendency for basal ganglia involvement in patients with PRES associated with pre-eclampsia-eclampsia, the MRI appearance of patients with PRES does not seem to be influenced by the predisposing risk factors.°

As PRES is potentially reversible so, appropriate therapy usually results in complete resolution of the deficits over several days to weeks, although partial resolution has been reported and the disease can be fatal. Treatment of arterial blood pressure with antihypertensives, control and prevention of seizures with magnesium sulphate or phenytoin or withdrawal of the offending agent is the treatment of choice.°

Acute hypertension, eclampsia, immunosuppressive medications, infections or autoimmune diseases can all result in the clinical syndrome of vasogenic oedema in the central nervous system like in PRES leading to
headache, seizure, confusion and frequent visual loss. The biologic basis for the syndrome is likely to be an insult to cerebral vascular autoregulation; so MRI of the brain is an essential tool in diagnosing a cerebral cause for these symptoms as the changes are reversible in PRES.

REFERENCES