ABSTRACT

Posterior reversible encephalopathy syndrome (PRES) is a clinical-neuroradiological entity characterized by headache, vomiting, confusion, seizures and blurred vision along with images suggesting white gray matter edema in posterior regions of the brain as shown by magnetic resonance imaging (MRI). The term PRES describes a potentially reversible imaging appearance and may occur in diverse situations, including hypertension, eclampsia, pre-eclampsia, immunosuppressive medications, such as cyclosporine, various antineoplastic agents, severe hypercalcemia, thrombocytopenic syndromes, Henoch-Schönlein purpura, systemic lupus erythematosus (SLE), renal failure, post-transplantation, infection and sepsis. We report two cases of acute PRES who had eclampsia and presented with recurrent episodes of seizures and hypertension. The authors emphasize that even though PRES is usually reversible, the early recognition and management of this syndrome is important to prevent permanent neurological sequelae. Treatment of PRES needs to be early and aggressive with rapid control of convulsions and arterial hypertension. Although prognosis is favorable, delay in treatment can sometimes lead to cerebral ischemia and infarct.

Keywords: Cortical blindness, Eclampsia, Hypertensive emergency, Reversible encephalopathy.


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INTRODUCTION

Posterior reversible encephalopathy syndrome (PRES), also known as reversible posterior leukoencephalopathy syndrome (RPLS), is a syndrome characterized by headache, confusion, seizures and visual loss. It may occur due to a number of causes, predominantly malignant hypertension, eclampsia and some medical treatments. It occurs due to elevated blood pressure which exceeds autoregulatory capacity of brain vasculature. Posterior reversible encephalopathy syndrome is characterized by headache, confusion, seizures, and altered mental function.

CASE REPORTS

Case 1

Twenty-six years old primigravida at 33 weeks and 3 days, admitted with one episode of increased blood pressure for evaluation. There was no past history of hypertension, cardiac disease, seizures or vision abnormalities. On examination, she was conscious, riented, afebrile with bilateral pitting pedal edema. Pulse 86 per minute, blood pressure 160/90 mm Hg. Uterus corresponding to 34 weeks, relaxed, cephalic, fetal heart 140 per minute. She complained of severe headache, blurring of vision after admission immediately following which she developed generalized tonic clonic convulsions. Magnesium sulfate 4 gm intravenous loading dose was given immediately followed by infusion 1 gm per hour to prevent further seizures. Emergency cesarean section was done under general anesthesia in view of unfavorable bishop score and recurrent seizures despite magnesium sulfate. Post-operatively, she was shifted to intensive care unit (ICU) in view of persistently elevated blood pressure. She had loss of vision and headache 5 hours after cesarean section. Patient had recurrent seizures every 3 to 4 hours inspite of magnesium sulfate infusion. Her treatment included magnesium sulfate infusion, labetalol infusion 10 mg per hour, intravenous midazolam 2 mg per hour, intravenous fosphenytoin. Injection Levetiracetam 500 mg intravenous twice a day was started after neuromedicine opinion. Magnetic resonance imaging (MRI) brain showed diffuse hyperintense abnormal signal intensities on T2-weighted and fluid-attenuated inversion recovery (FLAIR) sequences involving bilateral parietal, occipital, temporal and frontal lobes (Figs 1 and 2). No evidence of infarct or hemorrhage. Venogram showed no evidence of venous sinus thrombosis. She was diagnosed to have posterior reversible encephalopathy syndrome (PRES).
and was monitored in ICU. She continued to have blurred vision and regained normal vision on 3rd postoperative day. She was extubated on 4th postoperative day and changed to oral medications labetalol, nifedipine and phenytoin. She had an uneventful recovery and follow-up.

Case 2

A 24-year-old primigravida, 32 weeks with dichorionic diamniotic (DCDA) twins, conceived with in vitro fertilization (IVF), referred as a case of severe pre-eclampsia. Patient was on labetalol 100 mg twice a day for 3 weeks before referral. She was given labetalol and nifedipine to control blood pressure. Emergency cesarean section was done on 3rd day of admission in view of uncontrolled hypertension 160/110 mm Hg and imminent eclampsia. On second postoperative day, she had an episode of generalized tonic clonic convulsion and visual disturbances. She was started on magnesium sulfate. She had two episodes of recurrent seizures in spite of 4 hours of magnesium sulfate. She was shifted to ICU and given injection fosphenytoin and injection midazolam on advice of neurologist. She was on tablet labetalol 200 mg twice a day and tablet nifedipine 20 mg thrice a day. Due to recurrent seizures, she was given injection levetiracetam 500 mg iv thrice a day. Magnetic resonance imaging brain showed abnormal signal intensities in parieto occipital region associated with edema and effacement of ipsilateral cortical sulci (Figs 3 and 4). All these changes showed hypointense signal on T1-weighted and hyperintense signal on T2-weighted and FLAIR images.

MR angiogram and MR venogram were normal. On follow-up she showed marked improvement clinically with control of hypertension and was discharged in stable condition. Her visual disturbances resolved with no sequelae on follow-up after 6 weeks. Fundus examination was normal. She was given tablet levetiracetam 500 mg thrice a day for 6 weeks.
DISCUSSION

Posterior reversible encephalopathy syndrome is a recently described syndrome in literature, which constitutes a recognizable syndrome characterized by headache, altered mental status, seizures and visual loss. Altered mental status could range from lethargy, somnolence, restlessness, agitation, confusion to stupor and coma. Multiple seizures are more common than a single event.

Rapid diagnosis of posterior reversible encephalopathy syndrome is essential to prevent complications, such as infarction and hemorrhage. Proper diagnosis requires careful attention to clinical and radiographic presentation.

Multiple theories have been proposed on the pathophysiology of PRES, the most accepted being vasogenic edema. Cerebral autoregulation maintains a constant blood flow to the brain despite alterations in the systemic pressures. Once this mechanism gets disrupted, increased perfusion pressure is sufficient to overcome the blood brain barrier allowing extravasation of fluid. So, PRES represents vasogenic rather than cytotoxic edema in the majority of cases.

We have presented this case series as awareness of the diverse clinical and radiographic presentation of acute PRES is essential to avoid misdiagnosis and treatment delay. Posterior reversible encephalopathy syndrome is a reversible condition and should be considered in patients with seizures and visual disturbances. Multiple seizures are more common than single event. Pre-eclampsia and eclampsia may be the most common causes of PRES and most causes are managed without neuroimaging, hence, the incidence remains unknown.

Clinical improvement follows the treatment of elevated blood pressure. Magnesium therapy should be initiated as soon as eclampsia or PRES in pregnancy is suspected. Even mild fluctuations in blood pressure during or after anesthesia or changes in serum electrolytes, notably magnesium, may be sufficient to precipitate PRES in susceptible patients. Early treatment results in complete reversal of the deficits over few days to several weeks. Follow-up neuroimaging was not considered in these cases in view of rapid clinical recovery of the patient. Thus, this case report emphasizes the need for early diagnosis and prompt treatment of PRES to avert short- and long-term neurological sequela.

REFERENCES