Case Series

Posterior Reversible Encephalopathy Syndrome with Atypical Presentations: Case Series

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ABSTRACT

Posterior reversible encephalopathy syndrome (PRES) is a novel clinic-radiological entity characterized by headache, visual disturbances, encephalopathy and seizures. Radiological features typically include bilateral symmetrical oedema of posterior cerebral region especially of parieto-occipital lobe. Various conditions have been associated with PRES but toxaemia of pregnancy, organ transplantation, immunosuppressive treatment, and hypertension are most commonly described. The pathophysiology of PRES is still debatable. Basically, it represents vasogenic oedema resulting from the breakdown of cerebral autoregulation which if not managed promptly may lead to cytotoxic oedema. Treatment of severe hypertension, seizure control, withdrawal of the causative agent and antioedema drugs are the hallmark of specific therapy in PRES. Delay in diagnosis and treatment may lead to permanent neurological impairment. Therefore, awareness of PRES is important for both obstetrician and radiologist. This article reports three cases of PRES which illustrate clinical features, diagnosis and management with the aim to draw attention of clinicians and radiologist to the existence of this clinic-radiologic entity associated with preeclampsia-eclampsia.

Key words: Pre-eclampsia, eclampsia, encephalopathy.

INTRODUCTION

In 1996, Posterior reversible encephalopathy syndrome [PRES] was introduced as a new entity in medical literature.[1] The accurate statistics for global incidence of PRES is largely unknown. It is characterized by impaired consciousness, seizures, headache, visual abnormalities, nausea/vomiting, and focal neurological signs.[2] The consciousness impairment may range from confusion, somnolence, and lethargy to encephalopathy or coma.[3] Majority of women [around 92%] present with seizures and multiple seizure episodes are more frequent than a single event.[4,5] Visual disturbances may be in the form of blurred vision, hallucinations, homonymous hemianopsia and cortical blindness.[3] Although MRI (Magnetic Resonance Imaging) is the investigation of choice but CT (Computerized Tomography) scan can also clinch the diagnosis as in the reported case series. Here three cases of PRES have been reported. Two of them presented as antepartum eclampsia with recurrent seizures and altered sensorium while one case presented as late postpartum seizures in the setting of septicemia. With these cases, we intend to draw attention towards the existence of this novel clinico-radiological entity not only in relation to preeclampsia and eclampsia but also with septicemia.

CASE REPORT 1

A 19 year old primigravida presented with four episodes of generalized tonic-clinic seizures(GTCS) at 34 week of gestation. She had headache, epigastric pain, and vomiting for last 1 day. There was no past history of hypertension and epilepsy. On examination, she was in altered sensorium, drowsy and not responding to verbal commands. Her pulse rate (PR) was 116 beats per minute (bpm) and blood pressure (BP) was 220/120 mm of Hg. Cardiovascular and respiratory

system examination was unremarkable. Obstetrical examination revealed uterus corresponding to 34 weeks size, with single fetus in cephalic presentation and absent fetal heart rate with mild uterine contractions. Speculum examination did not reveal any bleeding or leaking through vagina. Vaginal examination revealed soft anterior cervix with 1.5 cm of dilatation and 70% effacement with intact membranes, Vertex was at –2 station and pelvis was adequate. On investigations, urine albumin by dipstick was 4+. Her complete blood count revealed Hb of 8.4 gm/dL, TLC of 21000/mm^3 and platelet count of 66,000/mm^3. There was evidence of hemolysis on peripheral smear. Blood urea was 22 mg/dL and serum creatinine was 0.8 mg/dL. Liver function test showed serum bilirubin 0.5 mg/dL with raised liver enzyme (Aspartate aminotransferase (AST)-369 U/L and Alanine transaminase(ALT)-314 U/L). Serum electrolytes (Na:131 mEq/L), K: 3.9 mEq/L), blood sugar (fasting: 93 mg/dL, Postprandial: 108 mg/dL) were normal. Serum LDH was raised to 1005 U/L. Coagulation profile was normal with INR (international normalized ratio) of 1.14. Fundus examination revealed early papilledema. Hence, a provisional diagnosis of eclampsia with HELLP syndrome was made. Loading dose of magnesium sulphate was given (total 14 grams) followed by maintenance dose (5 gm) every 4 hours. Intravenous labetalol 20 mg was given for control of hypertension. Labour was augmented with oxytocin. She delivered vaginally a dead male baby of 2.25 kg. Postnatally, blood pressure was controlled on oral labetalol. In view of persistent stupor contrast enhanced CT (CECT) scan was done which revealed subcortical hypodensity in bilateral parieto-occipital region suggestive of PRES. Patient was started on intravenous mannitol (250 ml of a 20% solution over 20 minutes followed by 100 mg intravenous 8 hourly). She responded to treatment and regained full consciousness after 2 days. Oral glycerol was started and tapered off by 7th postnatal day. Blood pressure became normal after 5 days of delivery and the patient was discharged on 8th postnatal day after complete clinical recovery.

CASE REPORT 2

26 year old gravida 2, para 1 with 1 live issue at 33 weeks of gestation presented with 2 episodes of GTCS at home. There was no history of preceding headache, blurring of vision or epigastric pain. Her previous blood pressure records were normal. The patient was conscious but disoriented. Her PR was 120 bpm and BP was 150/104 mmHg .There was mild pedal edema. Respiratory and cardiovascular system examination was normal. Abdominal examination revealed 32 weeks fundal height with single live fetus in cephalic presentation. Fetal heart rate was 140/minute. Vaginal examination revealed firm cervix in mid position, 1.5 cm dilated, early effaced with intact membranes and adequate pelvis. Urine albumin by dipstick was 2+. All the blood investigations including liver and kidney investigations were normal (Hb-9.4 gm%, TLC- 8600/ mm^3, platelet count-1.2 lakh/ mm^3, serum urea- 24 mg/dL, serum creatinine-0.6 mg/dL, serum bilirubin -0.5 mg/dL, AST-46 U/L, ALT-54 U/L, blood sugar fasting -86 mg/dL, postprandial-112 mg/dL and INR-1.14). Fundus examination was also normal. Anticonvulsant therapy in the form of magnesium sulphate was given. Tab labetalol 100 mg three times in a day started for control of hypertension. Termination of pregnancy with dinoprostone gel followed by oxytocin was done. A male baby of 1.8 kg was delivered. Post delivery blood pressure came to normal thus tablet labetalol was stopped. On 3rd postnatal day, patient complaint of weakness in right upper limb and lower limb. There was no bladder and bowel complaint or any headache, epigastric pain or vomiting. On examination, patient was conscious, well oriented and normotensive. Neurological examination revealed right sided hemiparesis. All the baseline investigations were repeated and found to be within normal limits except for hemoglobin which was 8.4 gm/dL**.** Fundus examination was also normal. CECT head revealed ill-defined hypodensities along the gray white matter junction in bilateral high frontal and parieto-occipital regions suggestive of PRES. There was no evidence of midline shift and bilateral ventricular system were normal. Osmotic diuresis with intravenous mannitol was done in a same manner as in previous case. Neurological symptoms gradually improved and completely resolved by 9th post natal day and the patient was discharged without any neurological sequalae.

CASE REPORT 3

19 year old para one with one live issue, postpartum women delivered at home by dai, presented on 9th postpartum day, with complaints of abdominal pain, abdominal distension and breathlessness for three days. She belonged to the low socioeconomic status and never

had any antenatal or postnatal visits at any hospital. The general condition of the patient was poor. She was dehydrated and had tachycardia. Her PR was 130 bpm, BP was 80/40 mmHg, respiratory rate was 40/minute. Respiratory and cardiovascular examination were normal. Abdominal examination revealed distension and diffuse tenderness with 16 weeks uterus. Speculum examination revealed foul smelling lochia. Vaginal examination revealed a soft tender sub-involuted uterus with fullness and tenderness in both the fornices. Complete blood count revealed Hb of 9.8 gm/dL, total leucocyte count 28,000/mm^3, platelet count of 1.88 lacs/mm^3 . Blood urea was 36 mg/dL, Serum creatinine was 0.8 mg/dL, Serum bilirubin-0.8 mg/dL, AST-58 U/L, ALT-49 U/L, Random blood sugar was 110 mg/dL and INR was 1.45. Ultrasonography revealed hypoechoic collection of 800-900 ml in the abdomen. Emergency exploratory laparatomy was done with drainage of 1 litre of pus. Patient was kept on mechanical ventilation because of poor general condition. Despite control of sepsis and hemodynamic stability with time, she could not be weaned off the mechanical ventilation because of stupor state. On 7th postoperative day (POD), patient had GTCS and hypertension along with passage of tapeworm in the stools which pointed towards diagnosis of neurocysticercoisis. CT head showed hypodensities in bilateral occipital lobes suggestive of PRES. Patient was started on tab amlodipine along with dexamethasone injection 8 mg. Patient responded to treatment and could be extubated on 11th POD and shifted to ward on 15th POD. Patient was finally discharged without any psychological and neurological sequalae on 17th POD.

DISCUSSION

Pregnant female presenting with seizures and altered sensorium puts every obstetrician in the challenging dilemma of various possibilities. The differential diagnosis ranges from the most common eclampsia to various medical disorder i.e. epilepsy, cerebrovascular accidents complicating pregnancy like cerebral infarct, cerebral venous sinus thrombosis, subarachnoid hemorrhage or central nervous system infections or it could be the clinical syndrome of posterior reversible encephalopathy syndrome.[6,7] PRES is a novel but a diverse clinic-radiological syndrome with diverse clinical presentations but characteristic imaging pattern.

Although still not completely elucidated, vasogenic edema remains the most accepted pathophysiological mechanism of PRES.[1] Cerebral autoregulation maintains a constant blood flow to brain despite alterations in the systemic mean arterial pressures. If this mechanism gets disrupted, the increased perfusion pressure is sufficient to overcome the blood brain barrier, allowing extravasation of fluid and macromolecules including red blood cell leading to vasogenic edema. A persistent vasogenic edema or extensive damage of the brain may finally lead to cytotoxic edema and resultant irreversible damage to neurons.[8] The maternal mortality has been reported to be as high as 15%.[3,4]

The presenting symptoms may range from headache, nausea, altered mental status, visual disturbances to loss of consciousness, generalized seizures and coma.[4] Symptoms may develop acutely or gradually over a period of several days. In 70-80% of the cases of PRES, hypertension is present as seen in first case report.[1,3,4] However in 20-30% of the cases patient may be normotensive initially as in last 2 cases[1,9].

PRES is seen with a wide variety of associated conditions like chronic renal insufficiency, preeclampsia, eclampsia, thrombotic thrombocytopenic purpura, autoimmune disorders alone or together with immunomodulating agents, infections or sepsis.[1,2,5,6] However regardless of the underlying cause, the main abnormality is the cerebral vasogenic edema. The pathogenesis of vasogenic edema may be endothelial dysfunction.[10,11] The predominance of the posterior involvement is because of its comparatively lesser innervations making it more vulnerable for blood pressure fluctuations and autoregulation breakdown.[12]

Radiological investigations are essential for confirming the diagnosis of PRES. MRI is the key investigation for the diagnosis of PRES and it allows more precise characterization and recognition of PRES than CT although we could not do this test because of unavailability of this facility at our centre.[13,14] The characteristic radiologic feature is bilateral and symmetrical regions of edema typically located in the white matter and predominating in the posterior parietal and occipital lobes. MRI shows high signal intensity on T2 weighted images and FLAIR sequences in the characteristic topographic distribution.[15] However atypical distribution involving cerebellum, brainstem, basal ganglia as well as the asymmetric

location or involvement of grey matter can add to the diagnostic dilemma.

CT scan is usually negative in early stage of PRES. However, in advanced cases, it shows hypodensities in the characteristic topographic distribution [16]. CT scan is usually the initial imaging modality readily available even in low resource settings. In all the three cases, CT scan revealed the characteristic hypodensities in the posterior parieto-occipital region suggestive of PRES. Whatever may be the initial imaging modality, the hallmark of PRES is the resolution of the radiographic lesion after complete clinical recovery following adequate treatment.

Management of PRES involves early recognition and correction of the underlying cause, control of hypertension, anticonvulsant therapy and termination of pregnancy. In first 2 cases, component of hypertension was present pointing towards baseline endothelial dysfunction which might have lowered the threshold of autoregulation breakdown and even with a normal blood pressure, patient had vasogenic edema leading to neurological deficit. In both these cases osmotic diuretics were used to decrease the cerebral edema. The role of osmotic diuretic has not yet been elucidated in any of the previous studies related to PRES.

Third case was also atypical as the patient presented with septicemic shock and only modest elevation of blood pressure. Septicemia can be associated with PRES in 8%-24% of the cases.[17]

CONCLUSION

PRES is a novel entity and in obstetric practice, it should be suspected in patients of eclampsia or septicaemia with prolonged altered sensorium and in presence of transient focal neurological deficit. Although MRI is diagnostic, but CT scan in a low resource setting is equally effective in clinching the diagnosis. The hallmark of management of PRES is identification and removal of the underlying cause, antihypertensives, anticonvulsant, and termination of pregnancy. Osmotic diuretic may have a promising role in improving the outcome of patients with PRES.

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