

Case Report

Posterior Reversible Encephalopathy Syndrome

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Abstract

Posterior reversible encephalopathy syndrome is one of the causes of convulsions in the postpartum period. It is a neurological condition which affects men and women and occurs in pregnancy both antepartum and postpartum. Herein we analyze two cases that presented in postpartum period with PRES and their management. We analysed patients who presented at a tertiary level medical college hospital with convulsions who were in pregnancy or Puerperium from 2007 to 2013. In this period we had 4315 deliveries in our hospital. We found that we had 18 cases of convulsions complicating pregnancy & puerperium giving us an incidence of 0.417%. Of these 12 were cases of eclampsia and 6 were cases of non-eclamptic convulsions. Of these six, 2 were cases of Posterior Reversible Encephalopathy Syndrome.

Key words : Posterior reversible encephalopathy syndrome (PRES)

Introduction

Posterior reversible encephalopathy syndrome is a neurological condition which is characterized by headache, nausea, vomiting, seizures, visual disturbances and altered sensorium. PRES can occur in pregnancy, both in antepartum and postpartum period. It is also seen in paediatric age group and adults, both men & women. PRES usually affects the cerebral white matter, but grey matter also can be affected. We analysed patients who presented at a tertiary level medical college hospital with convulsions who were in pregnancy or Puerperium from 2007 to 2013. In this period we had 4315 deliveries in our hospital. We found that we had 18 cases of convulsions complicating pregnancy & puerperium giving us an incidence of 0.417%. Of these 12 were cases of eclampsia and 6 were cases of non-eclamptic convulsions. Of these six, 2 were cases of Posterior Reversible Encephalopathy Syndrome.

Case1: 32 year old Para2Live2Abortion1 both Lower segment caesarean sections was brought on Post operative day-9 with history of one episode of convulsion (tonic clonic) at home which was preceded by complaints of severe headache for one week and blurring of vision for one day. Patient had one episode of vomiting and there was no history of fever. Previous menstrual history -3-4/30 days, regular. Married for 6 years, Non consanguinous marriage.

Obstetric history

1st pregnancy - Full term lower segment caesarean section, indication - Cephalo pelvic disproportion, wt-3.450kg, no Antenatal or postnatal complications.

2nd pregnancy -Dilatation and curettage done at

50days of amenorrhea.

Present pregnancy - Full term lower segment caesarean section, indication-previous LSCS with cephalopelvic disproportion.

On examination: Patient was conscious and oriented to time, place and person, afebrile, pedal edema present, no neck rigidity, Pupils equally reacting to light, Cardiovascular system and respiratory system were found to be essentially normal, BP on admission -164/90mmHg. Per abdomen-uterus well contracted and retracted, Deep tendon reflexes-brisk, clonus-negative, plantar- flexor, no motor or sensory deficit. No further convulsions noted after patient was brought to the casualty

Patient was further investigated and the reports are as follows:

Urine: albumin-nil, sugar-nil, microscopy-pus cells-1-3cells/ high power field, epithelial cells-1-2 cells per high power field. Blood: Hemoglobin-10.8 g/dl, Packed cell volume-33.6%, Total leukocyte count -6600cells /cmm, Differential count- Neutrophils -61%, Eosinophils -1.3%, Basophils-0.7%, Lymphocytes -31.4%, Monocytes-5.6%, Mean corpuscular volume-84. 8fL, Mean corpuscular hemoglobin- 27.2PG, Mean corpuscular hemoglobin concentration-32.1g/dl, RBC distribution width-11.7%, Platelet-2.51 lac/cmm, Blood urea nitrogen-6mg/dl, creatinine-0.65mg/dl, uric acid-4.3mg/dl, total protein-6.1g/dl, albumin-3.5g/dl, globulin-2.6g/dl, Albumin/Globulin ratio-1.3:1, bilirubin total-0.2mg /dl, bilirubin direct-0.03mg/dl, AST-26U/L, ALT- 41U/L, alkaline phosphatase -122U/L, gamma glutamyltransferase -22U/L, Bleeding Time -2mtsoosecs, Clotting Time-5mts30secs, Anti nuclear antibody (ELISA)-0.26(negative), Anti phospholipid antibodies- IgG-o .17, IgM-o .15, Activated partial thromboplastin time- control-29, test-

Prothrombin time-control- 12.6sec, test-13.3sec, INR-1.05, HIV/HCV/RPR - Non reactive, HbsAg - negative. MRI brain-axial T2/axial T2 FLAIR images (fig-1): white matter hyper intensities noted in bilateral parieto occipital and right frontal lobes. Mild tortuosity of both optic nerves noted more on the right side. Impression- features suggestive of posterior reversible encephalopathy syndrome.

Pt treated with Pritchard's regime (Magnesium sulphate regime) and Tablet Nifedipine. The patient recovered and was discharged normotensive and is on follow up with no sequelae.

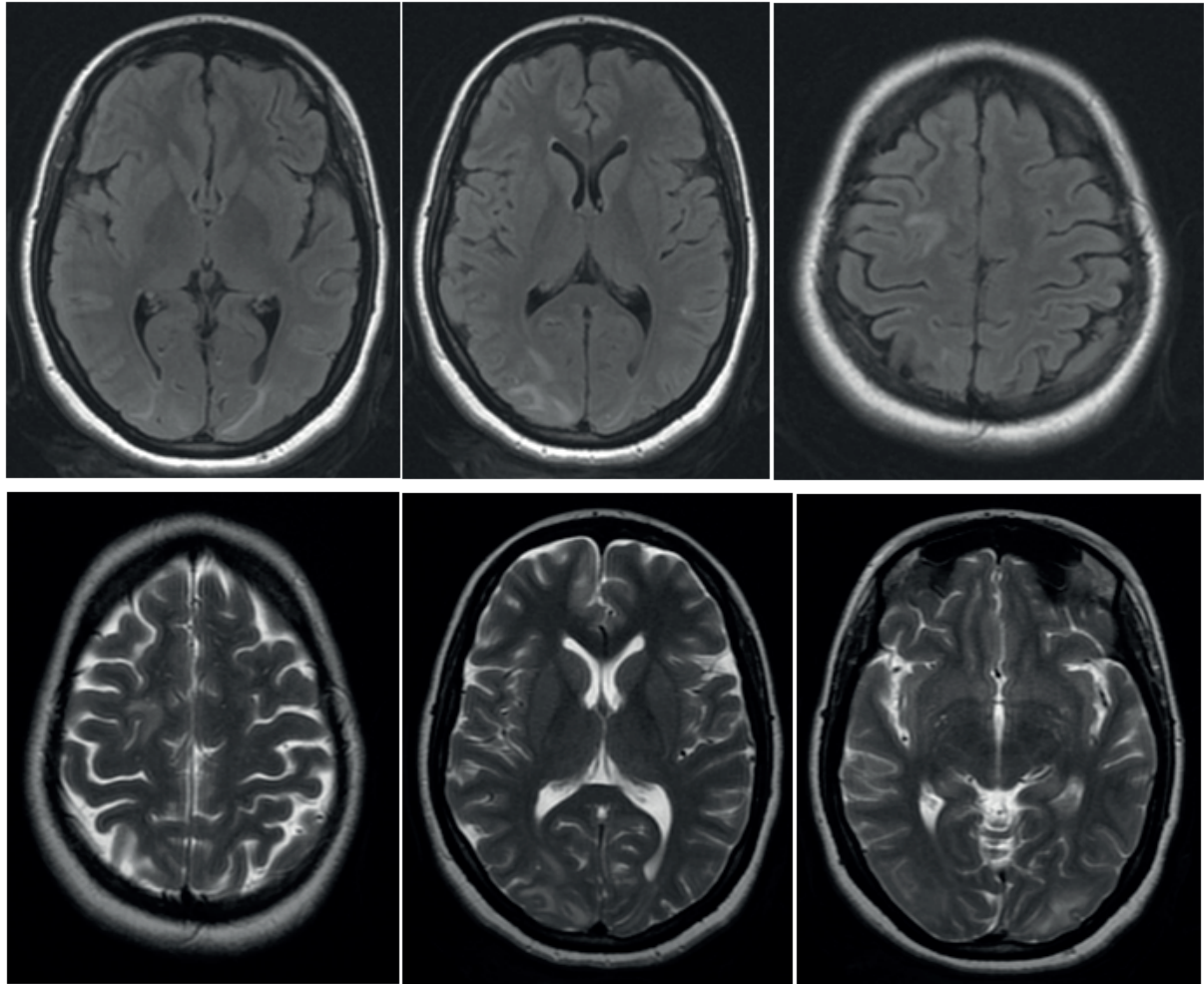


Fig-1: AXIAL T2 & AXIAL T2 FLAIR images:

Showing hyper intensities of white matter in the parietal, occipital and frontal lobes

Case2: 24 year old Para2Live2 both full term normal vaginal deliveries was brought on 4th post natal day with history of one episode of convulsion(tonic clonic) at home which was preceded by intense headache. Patient had no history of fever/vomiting/visual disturbances. Patient had delivered a girl baby weighing 2.98kg, Apgar score-8/10.

Previous menstrual cycle: 3-4/25 days, regular.

Married for 4 years, consanguineous marriage.

Obstetric history

1st pregnancy - Full term normal vaginal delivery, boy 3 yrs, alive and healthy

Present pregnancy- Full term normal vaginal delivery, girl baby four days old, alive and healthy.

On examination: Patient was conscious, oriented to time place & person, afebrile, no neck rigidity, Pupils equally reacting to light, pedal edema present, BP on

admission was 190/106 mm Hg. Cardiovascular system and Respiratory system were found to be normal. Per abdomen: soft and uterus involuting. Central nervous system: deep tendon reflexes-brisk, clonus negative, plantar-flexor, no motor or sensory deficit. Patient had the second episode of convulsion in the casualty. Pritchard's regime was started. Papilledema was ruled out after an Ophthalmologist' opinion. Neurologist opinion was obtained and was advised to continue the same treatment and to investigate for the cause of hypertension.

Patient was further investigated and the reports are as follows:

Urine: albumin-nil, sugar-nil, Pus cells 1-2 cells per high power field, Epithelial cells-2-4 cells per high power field, Blood: Hemoglobin-12.5g/dl, Packed cell volume- 37.7%, Total leukocyte count- 13100 cells/cubic mm.

Differential count: Neutrophil 76.9%, Lymphocyte-16.1%, Monocytes-3.6%, Basophils-1%, Eosinophils-2.4%, Mean corpuscular volume-90fL, Mean corpuscular hemoglobin-29.8PG, Mean corpuscular hemoglobin concentration-33.1g/dl, RBC distribution width-10.2%, Random blood sugar-116mg/dl, Blood urea nitrogen-13mg/dl, uric acid- 5.5mg/dl, total protein-6.6g/dl, Albumin-4.1g/dl, Globulin-2.5g/dl, SGOT-21U/L, SGPT-41U/L, Serum alkaline phosphatase-160U/L, Gamma glutamyltransferase-21U/L, Bilirubin total- 0.2mg/dl, bilirubin direct-0.05mg/dl, sodium 143mEq/L, potassium 4.5mEq/L, HIV/HCV/RPR-Non reactive,

HbsAg- negative, blood group- O positive, MRI- axial T2 and axial T2 flair images(fig-2): hyper intensities noted in bilateral occipital and parietal lobes, no signal intensity changes noted in gradient or diffusion weighted images, signal intensity changes noted in left optic nerve. Impression: features suggestive of Posterior reversible encephalopathy syndrome [PRES].

Patient treated with Pritchard's regime and Tablet Nifedipine. Patient became normotensive & recovered completely.

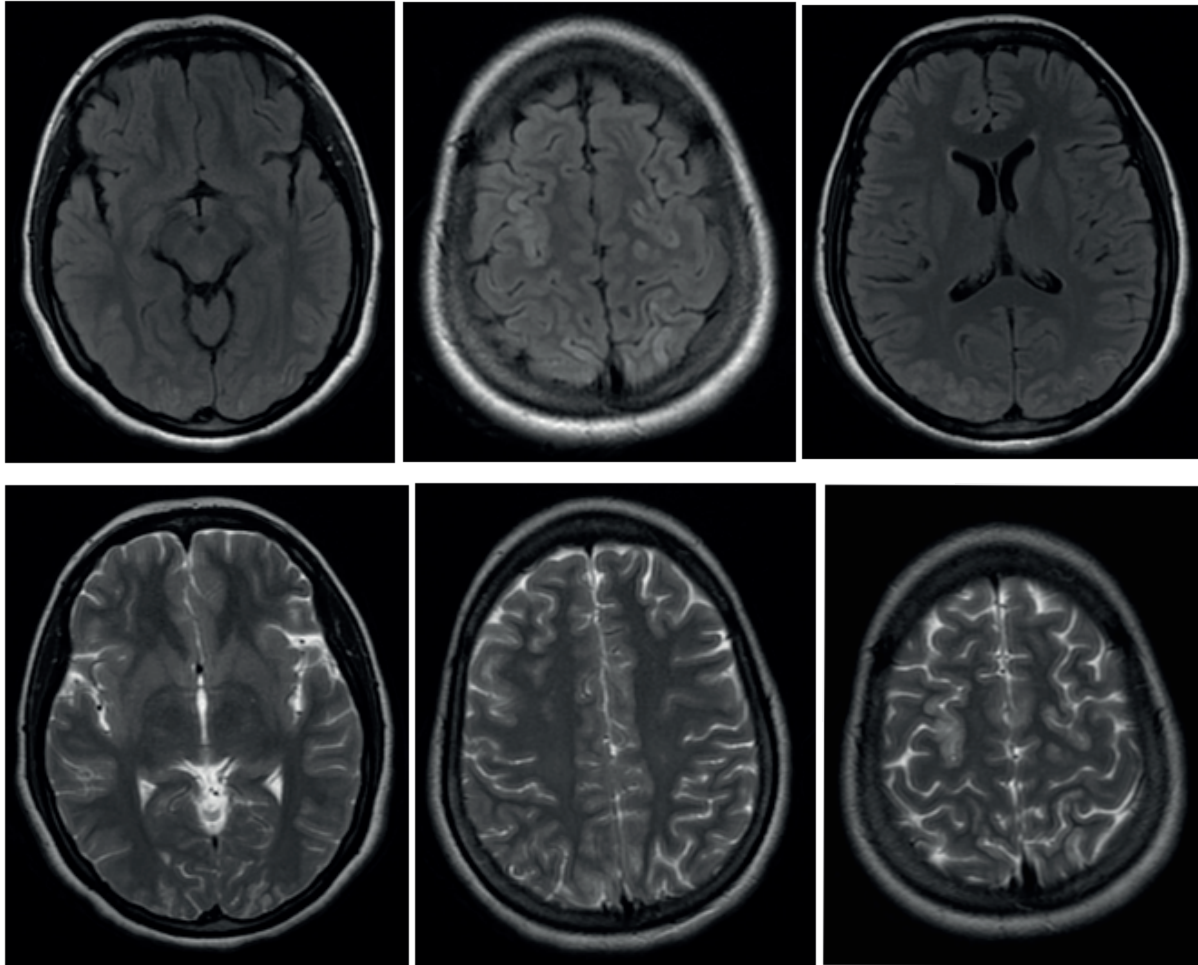


Fig-2: AXIAL T2 & AXIAL T2 FLAIR images: showing hyper intensities in the bilateral parietal and occipital lobes.

Discussion

Posterior reversible encephalopathy syndrome is a neurological condition which is characterized by headache, nausea, vomiting, seizures, visual disturbances and altered sensorium. PRES can occur in pregnancy, both in antepartum and postpartum period. Cases of PRES have been reported in children and adults, men & women. PRES usually affects the cerebral white matter, but grey matter can also be affected. Parietal and occipital regions are most commonly involved; sometimes the lesions can extend into basal ganglia, brain stem and cerebellum.

Seizures in pregnancy are usually considered to be

manifestations of eclampsia. Eclampsia is defined as occurrence of convulsions or coma with hypertension, proteinuria and or pedal edema during pregnancy between 20 wks of gestation and 48hrs postpartum without any pre existing neurological disorders. Seizures occurring beyond 48hrs but less than 4 wks after delivery is accepted as late postpartum eclampsia. The causes for postpartum seizures or altered sensorium other than eclampsia include cerebral infarction, hemorrhage, hypertensive encephalopathy, cerebral venous thrombosis, cerebral malaria, meningitis, intra cranial tumors and many others¹.

Etiology : The common causes of posterior reversible encephalopathy syndrome are hypertension,

eclampsia, renal failure, following organ transplantation, use of immunosuppressive and cytotoxic drugs like Cyclosporin A, Interferon alpha, intravenous immunoglobulins, Cisplatin, Tacrolimus etc,. Immunological disorders like Systemic lupus erythematosus, Porphyria, Behcet's syndrome, Polyarteritis nodosa are some rare causes of posterior reversible encephalopathy syndrome.

Pathogenesis : The pathogenesis of PRES is thought to be due to failure of cerebral auto regulation and endothelial dysfunction. Increase in blood pressure causes disruption of brain's normal auto regulation of cerebral blood flow. This disturbance of auto regulation produces dilatation of cerebral arterioles with opening up of endothelial tight junctions and leakage of plasma cells into the extra cellular space causing cerebral edema. Vasogenic edema occurs if cerebral white matter is affected. The cerebral white matter is composed of myelinated fibres in a cellular matrix of glial cells, arterioles and capillaries due to which there can be fluid accumulation leading to vasogenic edema. There is deficiency of adrenergic innervation of cerebral blood vessels, mainly in the posterior cerebral area, so it is affected the most.²

Clinically, PRES is characterized by seizures, headache, altered sensorium, and visual defects like blurring of vision, hemianopia and even cortical blindness. Seizures are of generalized tonic clonic type and can be single or multiple. Hypertensive encephalopathy presents with the same clinical features as PRES.

So, the complete diagnosis can only be arrived in conjunction with radiological evidence.³ In PRES, T2 weighted MR images and T2FLAIR images show hyper intense lesions in the cerebral white matter mainly in the parieto occipital region, sometimes grey matter can also be affected. Hypertensive encephalopathy shows all the features of PRES along with empty sella and optic nerve hydrops.

PRES is reversible if the blood pressure is controlled and treated early. Anti epileptic therapy is not needed. Controlling the blood pressure can reverse all the clinical effects and MRI abnormalities of PRES. If not treated in time, patients may develop irreversible neurological deficits, vision loss and the condition may even be fatal.

References

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Cure for Effects of Dangerous Decibels?

Noise pollution induced damage is an almost unavoidable consequence of modern living. Paradoxically, most of the noise is produced by those seeking pleasure (loud music), or politics (loud speakers) or comfort (machinery). Until now, how noise induced the hearing loss had not been fully explained. Now in a study conducted on mice (*FASEB J.* 2013 Sep;27(9):3730-40. doi: 10.1096/fj.13-232892. Epub 2013 May 31), researchers from Oregon Hearing Research Centre have demonstrated that perivascular resident macrophages (PVM/Ms; a hybrid cell type with characteristics of both macrophages and melanocytes) are damaged by noise leading to decreased production of pigment epithelium derived factor (PEDF) and increased leakiness of endothelial cells. The key event appears to be the down-regulation of PEDF. The latter is necessary for the integrity of interstitial-blood barrier. Breach of the latter leads to hearing loss. The researchers have shown that the delivery of PEDF to damaged ear can reverse the situation. Now that we have a solution, the noise pollution may become worse.

- Dr. K. Ramesh Rao