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Case Report

Atypical presentation of posterior reversible encephalopathy syndrome: a case report

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ABSTRACT

Posterior reversible encephalopathy syndrome (PRES) is a reversible syndrome characterized by seizures, headache, altered mentation, and loss of vision associated with white matter changes on imaging. A 25-year-old primigravida presented at 31 weeks gestation with atypical PRES characterized by generalized seizures and altered mental status. Magnetic resonance brain imaging showed high-intensity lesions in bilateral corona radiata, capsulo-ganglionic region, genu and splenium of corpus callosum, cortical and subcortical white matter of bilateral high fronto-parietal, bilateral cerebral peduncle, pons and medulla. Cases of antepartum atypical PRES are rare. Patients with atypical PRES do not always show typical manifestations. Our case reports highlight the crucial role of a prompt diagnosis, the importance of rapid blood pressure reduction, with a multidisciplinary approach.

Keywords: Atypical PRES, Eclampsia, PRES

INTRODUCTION

Posterior reversible encephalopathy syndrome (PRES) was first described by Hinchey et al in 1996.[1] It characterised by a milieu of seizure activity, impaired consciousness, headaches, visual symptoms, nausea/vomiting and focal neurological signs. PRES can be associated with a number of conditions, all of which result in cerebral vasogenic oedema which seems to be the crucial pathogenic mechanism. As the name suggests, it is typically reversible once the underlying cause is removed.[2]

The global incidence of PRES is unknown. It has been reported in patients ranging from 4 to 90 years of age, with most cases occurring in young-aged to middle-aged adults. PRES occurs in association with a number of causes, most commonly hypertension (pre-eclampsia or eclampsia) and use of immunosuppressive agents. The characteristic findings of PRES on an magnetic resonance imaging (MRI) of brain are hyperintensities in parieto-occipital regions which represent vasogenic oedema.[3]

CASE REPORT

A 25-years-old female, primigravida, presented to labour room at gestation of 31-weeks and 1 day with complaints of one episode of generalized tonic clonic seizure (GTCS) with premonitory symptoms of severe throbbing headache and blurring of vision since early morning. She was a booked case and her antenatal care was uneventful. She has a monochorionic diamniotic twin pregnancy. She has no family or past history of hypertension or epilepsy.

Following an episode of GTCS, her blood pressure was 170/120 mm hg. She was stabilised with IV labetalol 20 mg, mannitol and loading dose of Magnesium sulphate and referred to our centre. Following admission, she remained irritable with a Glasgow coma scale (GCS) of 7/15, and delivered late preterm male babies of 1.5 kg and 1.8 kg through emergency lower segment caesarean section (LSCS). Blood and other lab investigations are with in normal limits.

Following LSCS, her blood pressure remained high up to 190/120 mm hg for which she was started on labetalol infusion. She could not be extubated, her general condition remained irritable with GCS of 5T/15 and was under continuous monitoring. Her blood pressure continued to remain high. An emergency computerized tomography (CT) brain was done that showed hypodense area in lower limb of right internal capsule, bilateral parieto-occipital region and left temporal region-possibility, suggestive of vasogenic oedema with differential diagnosis of PRES. Further MRI of brain as shown (Figure 1 and 2) revealed T2/FLAIR high signal intensity areas involving few areas in bilateral corona radiata and left cerebellar hemisphere showing diffusion restriction-suggestive of Atypical PRES. Magnetic resonance angiography (MRA) showed hypoplastic A1 segment of left anterior cerebral artery and hypoplastic left transverse sinus.

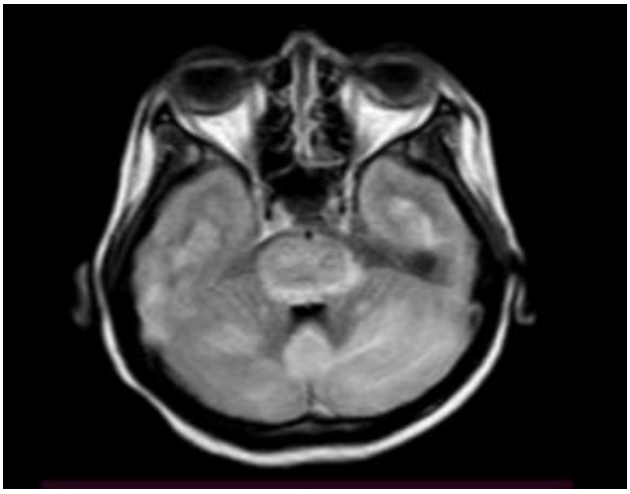

Figure 1: MRI brain shows high signal intensities on FLAIR sequence involving patchy areas of bilateral temporal lobes, pons, vermis and bilateral cerebellar hemispheres.

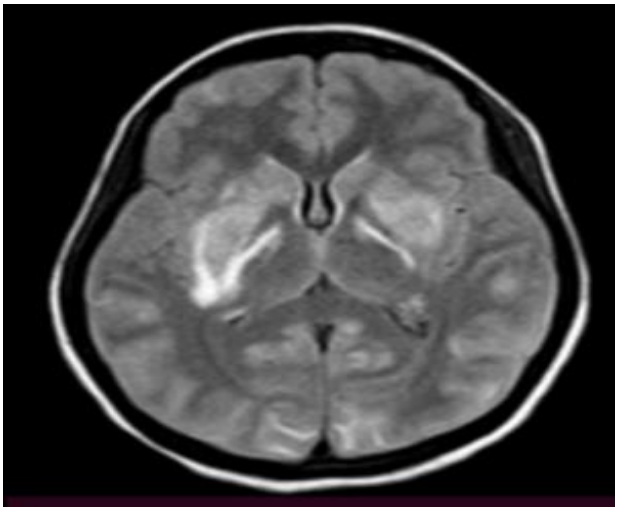

Figure 2: MRI brain shows high signal intensities on FLAIR sequence involving bilateral capsulo-ganglionic regions and occipital regions.

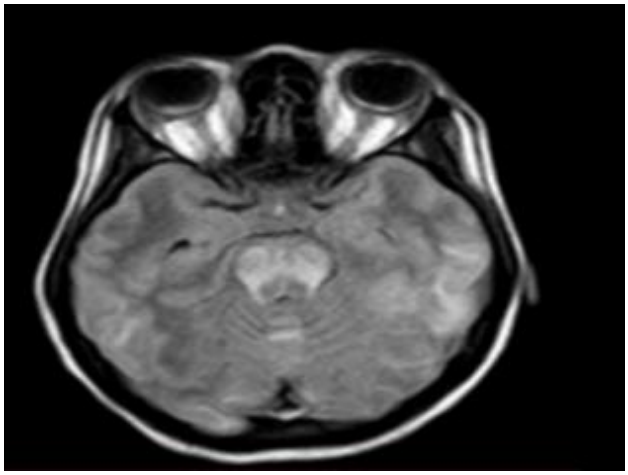

Figure 3: MRI brain shows high signal intensities on FLAIR sequence predominantly involving left temporal region and pons.

Differential diagnosis

Neurophysician opinion was sought and midazolam infusion was started. On post-delivery day (POD) 4, her general condition improved and was extubated and midazolam was stopped. Post extubation, mannitol was tapered and stopped. Oral clinidipine 20 mg/day along with oral labetolol 300 mg/day in divided doses were given for control of hypertension. Oral levetiracetam 1000 mg/day in divided doses were continued as an antiepileptic.

Outcome and follow-up

On POD 12, She was discharged in a stable condition and was continued on oral labetalol 200 mg/day and levetiracetam 1000 mg/day in divided doses. On POD 20, she remained stable when her blood pressure within the normal range. Hence, labetalol was tapered and stopped by POD 30 and advised to continue only on levetiracetam for 3 months.

DISCUSSION

PRES generally involves parietal and occipital lobes with vasogenic oedema.[4] In our patient, MRI findings (Figure 1-3) were bilateral corona radiata, capsulo-ganglionic region, genu and splenium of corpus callosum, and subcortical white matter of bilateral high fronto-parietal, bilateral cerebral peduncle, pons and medulla with few areas in bilateral corona radiata and left cerebellar hemisphere with MRA showed hypoplastic A1 segment of left anterior cerebral artery and hypoplastic left transverse sinus. A large area of brain was involved which is atypical in PRES.[5,6]

Atypical PRES is an interesting yet confusing finding associated with eclampsia. PRES can be considered as a differential diagnosis in pregnant women presenting with seizures, headache, and altered mentation.[7] Cases of

antepartum atypical PRES are rare and do not always show typical manifestations.[8] A multi-disciplinary approach is quintessential and life-saving in these critical cases.[9] Our patient had additional risk factors of being thin built (BMI of 18 kg/m^2), third degree consanguineous marriage and twin gestation. She couldn't be extubated immediately after caesarean and continued to be medically unstable. Antihypertensive infusion alone couldn't help to control her high blood pressure and required mannitol to reduce vasogenic oedema, which in turn controlled her high blood pressures and improved her general condition. Sedation with midazolam infusion was required to control her irritability. Magnesium sulphate as an antiepileptic which is choice of treatment in eclampsia was stopped and levetiracetam was given to control her seizures.

CONCLUSIONS

Cases of antepartum atypical PRES are rare and do not always show typical manifestations. Our case reports highlight the crucial role of a prompt diagnosis, the importance of rapid blood pressure reduction, with a multidisciplinary approach.

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