

DOI: <http://dx.doi.org/10.18203/2320-1770.ijrcog20194388>

Case Report

Posterior reversible encephalopathy syndrome presenting with atypical findings: case study

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Received: 21 July 2019

Accepted: 30 August 2019

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ABSTRACT

PRES (posterior reversible encephalopathy syndrome) is a syndrome characterized by headache, confusion, seizures and altered mental status. A 26 years old woman came to emergency department with a history of hypertension, blurred vision and seizures. She was diagnosed as a G3P2L1 at 29 weeks + 1 day with IUGR with preeclampsia associated with deranged kidney functions later complicated by development of PRES. Patient underwent emergency LSCS and was kept on mechanical ventilator as condition worsened during immediate post-operative period. Dialysis was also done to regulate urea and creatinine levels. Patient was treated with anti-hypertensive, anti-epileptics, antibiotics, intravenous fluids and continuous monitoring of blood pressure. Patient's condition improved gradually, and her discharge was planned. PRES is a condition, if managed in initial phase can lead to early recovery and reduce mortality.

Keywords: Blurred vision, Deranged kidney function, Eclampsia, Focal seizures, Hypertensive retinopathy, Posterior reversible encephalopathy syndrome

INTRODUCTION

PRES (Posterior reversible encephalopathy syndrome) is a syndrome characterized by headache, confusion, seizures and altered mental status. Most of the time, it is accompanied by acute hypertension.¹ There may be visual disturbances and status epilepticus or tonic-clonic seizures. The common location is parieto-occipito lobe or posterior lobe of brain. Areas of edema (swelling) are commonly seen on MRI. Symptoms may get resolved after sometimes although visual changes last for longer duration or forever. Although, PRES is considered as reversible condition, but it may cause permanent neurological damage in rare cases.

CASE REPORT

A 26 years old multi-gravida woman who is a referred case brought to emergency department with a history of hypertension (HTN), deranged kidney functions since past 3 years presented with gradually increasing headache associated with blurred vision from last 3-4 days, along with two episodes of generalized abnormal body movements which was accompanied by uprolling of eyeballs and jaw clenching and one episode of vomiting. Patient's family revealed the history of on and off headache, bilateral pedal edema and was on medication i.e. Atorvas and Arkamin since last 3 years after she had

diagnosed with HTN during her last pregnancy and dead fetus was born with LSCS in 2016.

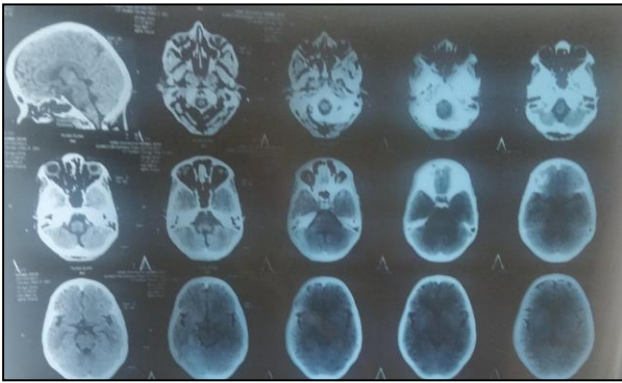


Figure 1: NCCT Brain depicting calcified granulomas are present in bilateral frontal lobes.

On her arrival to emergency department, her initial vital signs were: B.P = 170/120 mm/Hg, HR=88/min. and Temperature= 98.6°F. During initial examination, spontaneous conception was diagnosed by urine pregnancy test (UPT) which was later confirmed by ultrasonography (USG) for 8 months of gestation. After Obstetrics consultation, she was diagnosed as G3P2L1 at 29 weeks + 1 day with IUGR with HTN superimposed with eclampsia.

In emergency department patient was treated with Labetalol, Dexamethasone and MgSO₄. It was determined that an emergency LSCS may be needed to prevent fatal consequences and an emergency LSCS under GA was performed on 26th April 2019 (on the day of admission) and she delivered a male baby with birth weight 869gm and APGAR which was '0' initially and later came out to be '4'. Baby was admitted to NICU for close observation and further management. Unfortunately, patient was not able to come out of general anaesthesia and could not ventilate spontaneously. As an immediate treatment measures, she was shifted to ICU and kept on Mechanical ventilator. She was on SIMV mode (VC+PS) with GCS - 9 and infusion of Midazolam @ 2ml/hour and NTG @6ml/hour. She was also administered MgSO₄ @ 0.5ml/hour and DNS @ 800ml/24hours.

Investigations

Haematological examination revealed that patient's total leukocyte counts (26.33 thou/mm³), lymphocytes (15.71%), red blood cells count (2.758 mill/mm³), Haemoglobin (7.761gm/dl), hematocrit (24.72%), mean corpuscular haemoglobin concentration (31.4 g/dl), red cell distribution width (15.31%) and mean platelet volume (6.118 fL). Liver panel: LFT analysed alkaline phosphatase (467.4 U/L, serum protein (4.45 gm/dl). However, Kidney panel: KFT revealed that urea (112.56 mg/dl) and creatinine (5.17mg/dl) levels were raised.

NCCT brain was done which reported that calcified granulomas are present in bilateral frontal lobes. There is presence of ill-defined white matter hypo-density in right parietal lobe with preserved grey matter along with white matter differentiation. Findings were suggestive for PRES syndrome (Figure 1). Ophthalmology consultation was done and found cotton wool spots in right eye and flame shaped haemorrhage in left eye which clearly depicts that patient had hypertensive retinopathy (Grade III).

Management

During initial 2nd post-operative day, patient had altered sensorium and eventually developed focal seizures, had impaired urine output (20-25ml/hours). Urgent dialysis was done, and she was on drugs i.e. NTG, MgSO₄, Labetalol, Midazolam, Mannitol and Fetanyl. She was extubated on 30th April 2019 as she could manage to ventilate spontaneously with oxygen therapy. Patient also developed post-ictal confusion and delirium due to renal causes and psychiatric consultation was done for the same. Tab. Risperidone (0.5mg HS) was prescribed. On 1st May 2019 patient was capable enough to ventilate on room air and there was a slight change in her treatment. Ceftriaxone, Metrogyl, Pantop, Tranexa, Amlodipin, Lasix and Risperidone are the drugs given for further management. Patient was responding well to treatment and her condition improved gradually.

DISCUSSION

Posterior reversible encephalopathy syndrome PRES is a reversible neurotoxic state that happens secondary to the shortcoming of the posterior circulation to autoregulate in response to acute changes in vital sign (blood pressure). Hyper perfusion associated with interference of the blood brain barrier results in vasogenic swelling within the parieto-occipital regions, most of the time without infarction.¹⁻³ One of the hallmark of this disease is its reversibility of symptoms.⁴ In few typical cases like coma, status epilepticus, patients may require intensive treatment and it may also lead to permanent neurological impairment or death in very rare cases.^{5,6}

The cause of PRES is still a debatable matter, but most common hypothesis state that it occurs due to disruption of brain auto-regulation which is primarily caused by hypertension. Hyper-perfusion and cerebral vessel damage occurs as result of uncontrolled hypertension which further leads to vasogenic edema because of interstitial extravasation of proteins and fluids.^{3,4} Mean arterial pressures is an important parameter as increased MAP>200mmHg may cause irreversible damage. Another hypothesis state that it occurs in case of systemic inflammatory state including sepsis, eclampsia, transplantation and auto-immune disease which causes endothelial damage.¹⁻³ It can be stated here that further research is still needed to find out the exact cause and prevent this treatable yet threatening condition. In present

case, patient had history of hypertension since last 3 years and also had previous history of eclampsia during the time of last childbirth.

It is considered as a syndrome as it includes group of symptoms like headache, visual disturbances, seizures, and altered consciousness, which are often associated with distinctive neuro-imaging findings of edema in affected areas of brain.^{2,3} The symptoms can vary from mild to severe in intensity like visual loss can progress to cortical blindness, retinal detachment and confusion or agitated client can go into coma.^{1,3,7} Sometimes, patient experience non-convulsive seizures like staring, eye blinking, or head turning.^{8,3} Acute hypertension is identified as a trigger for PRES, although patients may have other co morbidities as renal failure, autoimmune disturbances which increases the risk of PRES during pregnancy.^{1,3} It is common among women because the major causes of PRES is pre-eclampsia/eclampsia developed during pregnancy.⁶ Our patient presented with headache, blurred vision, two episodes of generalized tonic-clonic seizures and bilateral pedal edema. Clinical picture of our patient and computed tomography findings guided to a diagnosis of PRES. She developed focal seizures, retinopathy and post-ictal confusion in post-operative phase which was then managed actively to prevent further damage.

Stroke, demyelinating lesions of brain, stroke, cerebral venous thrombosis is some of the differential diagnosis of PRES. Computed tomography (CT) is not 100% sensitive for diagnosis of PRES, but it is commonly used for rapid assessment in case of emergency. In PRES and many other cases, CT imaging can be normal and therefore it is not reliable.⁷ MRI is helpful in differentiating vasogenic edema from cytotoxic edema that is why mostly preferred to make final diagnosis.⁹ For this patient, NCCT was done and findings reported presence of ill-defined white matter hypo-density in right parietal lobe with preserved grey matter along with white matter differentiation.

Prevention is always better than cure, early identification and diagnosis is the mandatory to prevent irreversible damage and long term complications. Screening for patients should be done for all patients with pre-eclampsia and HELLP syndrome. Management of patient in early stage lead to faster recovery and patients' condition improve dramatically within few days or week. In case of PRES, it is important that how was patient dealt in acute phase for management.

Management of blood pressure is first priority in management and it is important to rule out the underlying pathology.^{1,7} Major goal of blood pressure management with intravenous anti-hypertensive therapy is not only to control blood pressure but also to decrease 20-25% mean arterial pressure (MAP) during initial 2 hours of treatment.^{6,7,10} Patients with PRES may also require anaesthesia and ventilation in case of generalized status epileptics and airway management, anti-epileptic for

management of seizures. Blood transfusion, administration of thiamine and management for impaired kidney functions including dialysis is also a part of standard treatment.

It is required to be sensitive while administering anti-hypertensives because quick reduction of blood pressure can cause impairment in cerebral perfusion pressure and promote ischemia. It is suggested that appropriate choice of anti-hypertensive drugs include labetalol, cardene, or fenoldopem.^{6,7} In present case, patient was treated with NTG initially along with MgSO₄ and labetalol therapy. Patient was also given midazolam, mannitol and fetanyl for management of convulsions. It has been reported in many case studies that patient's condition improve quickly in response to anti-hypertensive, anti-epileptics and other supportive care and management.^{3,6,7}

CONCLUSION

The patient described in this case was a woman with uncontrolled hypertension history and diagnosed with PRES. She was admitted to ICU as she was on continuous infusion of anti-hypertensives, anti-epileptics and management of airway. Later, she underwent dialysis and her condition improved rapidly. Patient became normal after two weeks of treatment and plan for discharge was initiated with follow-up in OPD.

Funding: No funding sources

Conflict of interest: None declared

Ethical approval: The study was approved by the Institutional Ethics Committee

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Cite this article as: Gaur R, Thakur K, Mudgal SK. Posterior reversible encephalopathy syndrome presenting with atypical findings: case study. *Int J Reprod Contracept Obstet Gynecol* 2019;8:4090-3.