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Case Report

Obstetrics and Gynaecology Section

Reversible Cerebral Vasoconstriction Syndrome along with Posterior Reversible Encephalopathy Syndrome in a Postnatal Eclamptic Patient: A Case Report

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ABSTRACT

Reversible Cerebral Vasoconstriction Syndrome (RCVS) and Posterior Reversible Encephalopathy Syndrome (PRES) are rare neurological disorders. Although rare, the diagnosis of these conditions is now possible due to increased clinical awareness and the availability of newer generation diagnostic methods such as Magnetic Resonance Imaging (MRI). The rarity of their occurrence can sometimes lead to misleading diagnoses, as both illnesses rarely coexist and share certain clinical and radiological characteristics. RCVS progresses rapidly and can result in chronic impairment or even in-hospital mortality in 5-10% of cases. On the other hand, PRES is a monophasic and reversible illness, although there have been instances of recurrence. In this case report, the authors present the case of a 22-year-old female who presented with post-natal seizures and severe headaches, suspected to have RCVS along with PRES. Since RCVS and PRES share some overlapping pathophysiological conditions, the findings of MRI, including Fluid-Attenuated Inversion Recovery (FLAIR), CR-Magnetic Resonance Angiography (MRA), Apparent Diffusion Coefficient (ADC), and Diffusion-Weighted Imaging (DWI), along with their correlation with clinical features, can help in differentiation. Vasoconstriction has been reported as the main cause; hence vasodilators are primarily suggested for controlling seizures. The patient survived and showed improvement. Initially, the patient was monitored every 15 days for one month, followed by monthly follow-ups up to 8 months after discharge. The purpose of presenting this case is to enhance understanding of RCVS with PRES, including associated risk factors, which can lead to better outcomes through timely diagnosis. Improved outcomes can be achieved through early recognition and improved clinical management.

Keywords: Hypoxaemic respiratory failure, Magnesium sulfate, Neurological disorders

CASE REPORT

A 22-year-old primigravida female patient presented to our emergency department on post-natal day 5 with complaints of post-natal seizures and a severe headache. She had no significant history of hypertension, hypothyroidism, or other notable conditions. The patient had delivered at the 28th week of gestation in a peripheral hospital due to severe hypertension and eclampsia. Upon admission to the peripheral hospital, her initial vitals were as follows: blood pressure of 200/110 mmHg, heart rate of 110/min, respiratory rate of 30/min, and SpO_2 of 94%.

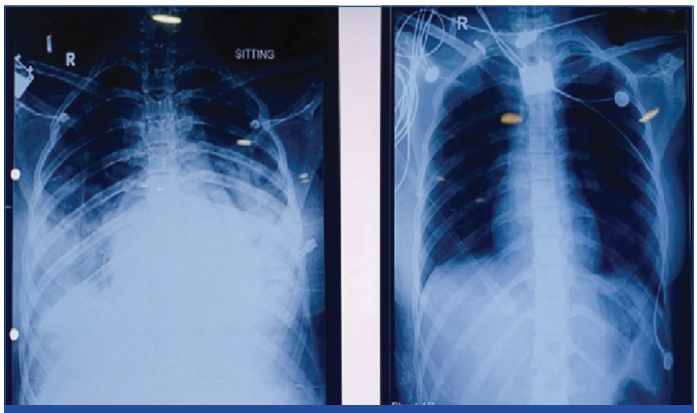
The case was managed in the critical care unit, where she received Magnesium sulfate and Labetol injection as anti-hypertensive treatment. However, the patient experienced her first seizure within 10 hours of admission. Consequently, a caesarean section was planned, during which the patient developed refractory seizures and experienced massive uterine bleeding post-delivery. The patient received anti-epileptics and uterine tocolytics, such as oxytocin and methylergometrine, and was subsequently transferred to the ICU. She was electively intubated and received mechanical ventilation, along with symptomatic treatments including anti-epileptics and antibiotics as needed. Extubation took place after 72 hours. The patient remained asymptomatic for 48 hours; however, she then reported an excruciating headache and hypoxaemic respiratory failure, followed by another seizure.

Oxygen support was provided, and conservative management was continued. Additionally, the patient developed a high-grade fever and experienced decreased urine output. In an emergency situation, she was referred to our hospital for further clinical management. Upon arrival, the patient exhibited respiratory distress, altered sensorium, and non-responsiveness to verbal commands. Her vital signs were as follows: blood pressure of 188/100 mmHg, heart rate of 142/min, respiratory rate of 36/min, and SpO_2 of 80% with 15 L/min O_2 support. A summary of her laboratory parameters is provided in [Table/Fig-1]. The laboratory findings suggested

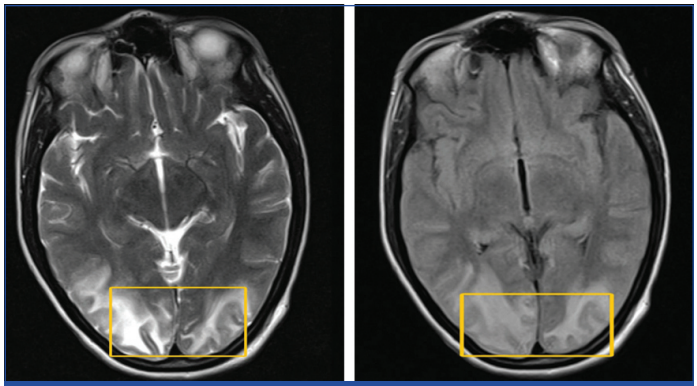
Parameter	Value
Hb	6.2 gm%
Platelets	16000/cumm
TLC	21000/cumm
Liver function test	
SGOT	278 IU/L
SGPT	198 IU/L
ALP	304 U/L
Total protein	3.6 gm/dL
Albumin	1.7 gm/dL
Globulin	1.9 gm/dL
Total bilirubin	3.4 mg/dL
Conjugated	1.0 mg/dL
Unconjugated	2.4 mg/dL
Kidney function test	
Creatinine	2.44 mg/dL
Blood urea	117.6 mg/dL
Sodium	134 mmol/L
Potassium	5.3 mmol/L
Urine	Proteinuria (3+)
LDH	786 U/ L

[Table/Fig-1]: Laboratory parameters of the patient.

haemolytic anaemia, as evidenced by a drop in Hb values from 12.6 gm% to 6.2 gm% over a period of five days without active bleeding, as well as abnormal Red Blood Cells (RBCs) observed on the peripheral smear and elevated LDH (Lactate Dehydrogenase) levels. Based on the presence of seizures, hypertension, and proteinuria (characteristic of eclampsia), as well as Haemolysis, Elevated Liver Enzymes, and Low Platelet Count (characteristic of HELLP Syndrome), a provisional initial diagnosis of Eclampsia with HELLP syndrome was made. The patient received management in the ICU with a multidisciplinary approach. She was reintubated and placed on mechanical ventilation support due to decreased SpO_2. An immediate chest X-ray was performed, revealing pleural effusion and bilateral lower zone haziness [Table/Fig-2]. Considering the patient's history of post-natal seizures and elevated blood pressure, an MRI of the brain was conducted. The MRI showed confluent signal intensity areas in the bilateral parieto-occipital lobes, bilateral cerebellar hemispheres, and the right side of the Pons, appearing hyperintense on T2 [Table/Fig-3] and FLAIR sequences, indicative of PRES due to the presence of symmetric lesions [Table/Fig-4]. The post-contrast study revealed multiple segmental areas of significant narrowing involving the circle of Willis, suggesting associated RCVS [Table/Fig-5]. The rest of the brain parenchyma appeared normal, and there was no evidence of a haemorrhagic stroke.

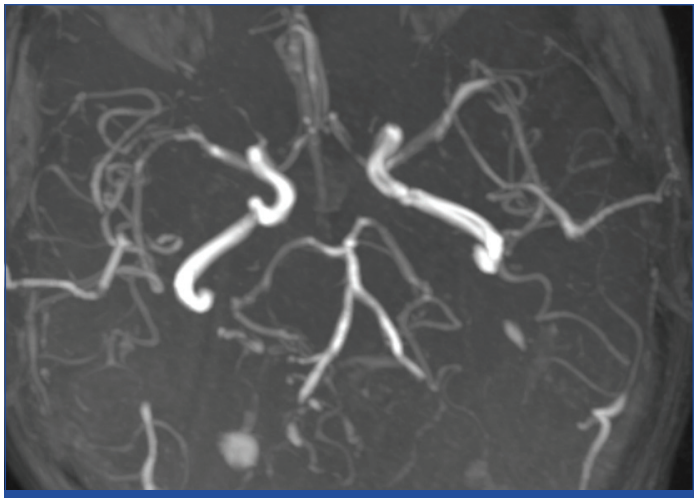


[Table/Fig-2]: a) The left-Bilateral mid and lower zone of homogeneous opacity and blunting of bilateral costophrenic angles suggestive of pulmonary oedema and bilateral pleural effusion; b) Right-Radiological improvement post-treatment. (Images from left to right)



[Table/Fig-3]: Confluent abnormal hyperintense lesion in bilateral parieto-occipital lobes on T2. **[Table/Fig-4]:** Confluent abnormal hyperintense lesion in bilateral parieto-occipital lobes on FLAIR. (Images from left to right)

Due to eclampsia, PRES was diagnosed, and the additional radiological and laboratory findings led to the conclusion of RCVS. Transfusion was performed due to decreased haemoglobin and platelet counts. In consultation with a neurologist, the patient was treated with the anti-epileptic drug Levetiracetam. The anti-hypertensive drug Injection Labetalol was initiated, and antibiotics were administered based on the urine culture report, which showed the growth of *E.coli* bacteria. Diuretics and other supportive treatments were provided. The patient showed clinical improvement, with improvements observed in the chest X-ray and urine output.



[Table/Fig-5]: Multiple areas of significant narrowing involving the circle of Willis/MR angiography.

During the hospital stay, magnesium sulfate was administered according to the PRITCHARD regimen for 24 hours, including a loading dose and maintenance dose. Inj. Labetalol infusion was initiated, and blood pressure was maintained at 126-130/80 90 mmHg for 72 hours. The tapering infusion was stopped after 24 hours. The patient regained consciousness after four days and was subsequently extubated following an adequate spontaneous breathing trial and T-piece trial. Tablet Amlodipine (5 mg) OD was initiated, with continuous blood pressure monitoring every four hours for 72 hours. The patient's vitals normalised, blood pressure was under control, respiratory rate was within normal limits, and there were improvements in the radiological and laboratory profiles. After a 15-day hospitalisation, the patient was discharged with a scheduled follow-up after two weeks and regular follow-up visits every month for the next eight months.

DISCUSSION

There is a recognised association between PRES and RCVS. PRES is characterised by clinical symptoms such as headache, seizures, vision disturbances, and focal neurological deficits, typically linked to conditions like eclampsia, immunosuppressive drugs, and hypertensive and uremic encephalopathy. It is typically identified by vasogenic oedema [1,2]. On the other hand, RCVS is a reversible condition characterised by thunderclap headaches, primarily caused by constriction of cerebral blood vessels, which can result in various neurological symptoms including visual disturbances, focal neurological deficits, and rarely, seizures or encephalopathy [3,4]. The underlying mechanisms linking RCVS and PRES are not yet fully understood, but occasionally these conditions are observed together. It is believed that vasoconstriction in RCVS can contribute to the development of PRES by disrupting normal blood flow and causing increased pressure within cranial blood vessels [5].

A differential diagnosis for sinus venous thrombosis was considered but ruled out based on the evaluation of clinical parameters, neuroimaging (MRI brain), and blood tests. Unlike in other patients, there was no evidence of haemorrhage, but the presence of eclampsia led to the diagnosis of PRES. The patient did not experience postdural puncture headache or any other side effects, likely due to already being eclamptic during the antenatal period. The presence of eclampsia was determined to be the cause of development for both PRES and RCVS [6,7].

PRES is characterised by symmetric lesion distribution with a parieto-occipital pattern of oedema, unlike RCVS. RCVS, on the other hand, presents with asymmetric lesions, distal vascular pruning, vasoconstriction, and ischaemic lesions as common radiological findings. The postpartum presentation of RCVS with PRES is a

rare clinical condition. Severe headache, seizures, hypertension, and vasogenic oedema are the main clinical symptoms reported in postpartum women developing RCVS with PRES [8,9]. Neuroimaging, such as MRI, can help differentiate between RCVS and PRES. Pop A et al., have reported that MRI-FLAIR, MRI-ADC, and MRIDWI can be useful in distinguishing between PRES and RCVS or in making a co-diagnosis of both conditions [10].

The main purpose of this case presentation is to raise awareness about postpartum RCVS and PRES occurring together, similar to our patient. Treating the underlying causes (hypertension, pain management, seizure control, calcium channel blockers, antibiotics, kidney function improvement, and other supportive care) remains the mainstay of treatment in such cases. Research literature reports comprehensive clinical management in postpartum females with PRES and RCVS, requiring collaboration between specialists such as obstetricians, neurologists, radiologists, and others. This collaboration is necessary to guide the complete treatment plan [9-11]. It has been reported that patients with postpartum complications can achieve clinical normalisation, and follow-up radiological imaging has shown normal findings [8,11].

CONCLUSION(S)

In pregnant or postpartum patients presenting with classic neurological symptoms, the possibility of PRES and RCVS should not be overlooked. In emergency situations, an MRI of the brain should be promptly performed if a CT scan is inconclusive. Timely diagnosis of cases with both PRES and RCVS is crucial, as advancements in diagnostic methods and increased awareness of the pathophysiology of PRES and RCVS contribute to optimising disease management.

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