Case Report

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Reversible Cerebral Vasoconstriction Syndrome with Subarachnoid Hemorrhage in the Postpartum Period: A Rare Case Scenario

Authors:- Sachin Patil[1], Kotli Nagaraj[2], Susheel[3], Rahul[4], Hemanth Reddy Nukala[5]

[1,3,4,5]Junior Resident, [2]Professor, Dept Of General Medicine, M R Medical College Kalaburagi, India.

Abstract

Reversible cerebral vasoconstriction syndrome (RCVS) is a rare, reversible condition characterized by segmental narrowing of cerebral arteries, often presenting with thunderclap headaches and various neurological deficits. This case report describes a 29-year-old multigravida who presented in the postpartum period with sudden onset of severe headache, seizures, and right-sided hemiplegia. Imaging revealed subarachnoid hemorrhage (SAH), infarctions in watershed areas, and segmental vasoconstriction of cerebral arteries on MRA. The patient was managed with antiepileptics, antihypertensives, and supportive care, resulting in full recovery within one month. This case underscores the importance of considering RCVS in postpartum patients with neurological symptoms to ensure prompt diagnosis and management. Similar cases in the literature emphasize the variable presentation and the need for high clinical suspicion to prevent misdiagnosis and inappropriate treatment.

Keywords:- Reversible cerebral vasoconstriction syndrome, Postpartum hemorrhage, Subarachnoid hemorrhage, Seizures.

INTRODUCTION

Reversible cerebral vasoconstriction syndrome (RCVS) is an uncommon, but increasingly recognized, clinical and radiological syndrome characterized by reversible segmental narrowing of cerebral arteries, often presenting with thunderclap headaches. This syndrome can lead to various neurological complications such as transient ischemic attacks (TIA), ischemic strokes (IS), intracerebral hemorrhage (ICH), and subarachnoid hemorrhage (SAH).[1]

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Corresponding Author:
Dr Sachin Patil
Junior Resident, Department Of General Medicine, M R Medical College Kalaburagi, Karnataka, India.

The pathophysiology of RCVS is not entirely understood, but it is believed to involve a transient disturbance in the regulation of cerebral arterial tone potentially triggered by various factors including vasoactive substances, pregnancy and the postpartum state.[2]

The postpartum period is a recognized risk factor for RCVS, accounting for 7-9% of all cases.[3] This is thought to be related to the significant physiological changes that occur during pregnancy and around puerperium including fluctuations in blood pressure, blood volume, and hormonal levels. All these factors are thought to be contributing to alterations in cerebral arterial tone. In particular, the sudden changes in female reproductive hormones after delivery are believed to play a key role in the development of RCVS in postpartum period. The use of vasoactive drugs during this period to manage postpartum hemorrhage or suppress lactation may further exacerbate the condition.[4]

Clinically, RCVS is most commonly characterized by a sudden, severe headache, often described as a "thunderclap." However, as seen in this case, RCVS can also present with other neurological symptoms, such as seizures and focal neurological deficits. Imaging studies, including magnetic resonance imaging (MRI) and magnetic resonance angiography (MRA) are crucial for diagnosis, revealing characteristic findings such as focal or segmental vasoconstriction as well as evidence of infarction or hemorrhage. The condition is self-limiting, with symptoms typically resolving within weeks to months and imaging abnormalities normalizing over time.[5]

In this report, we present a rare case of postpartum RCVS complicated by subarachnoid hemorrhage (SAH) and focal neurological deficits, highlighting the importance of considering RCVS in every patient presenting with postpartum neurological symptoms.

CASE REPORT

A 29-year-old multigravida (G4P3L3) was brought to the emergency department at 9 months of amenorrhea with a sudden onset of severe headache followed by a generalized tonic-clonic seizure and tongue bite. She had no significant history of hypertension or proteinuria during her pregnancy.

On presentation, her blood pressure was elevated at 170/100 mmHg, but she was conscious and oriented, with no motor deficits.

Two hours after admission, the patient delivered vaginally without complications. However, 24 hours post-delivery, she developed sudden right-sided hemiplegia, which was non-progressive. Neurological examination revealed right hemiplegia with hypotonia and absent plantar reflex on the right side, without any cranial nerve involvement or neck stiffness. Laboratory investigations showed anemia (hemoglobin 6.7 g/dL) and leucocytosis (WBC count 12,000/µL). Other laboratory parameters, including renal function tests, liver function tests, and electrolytes, were within normal limits and on urine examination there was no proteinuria.

Imaging studies were performed, including MRI T2/FLAIR, which revealed high signal intensity in the cortical and subcortical white matter of the bilateral fronto-parieto-occipital regions, posterior limb of the internal capsule, external capsule, and cerebellar hemispheres, suggestive of white matter edema. An acute infarct was also noted in the bilateral fronto-parieto-occipital regions and the left corona radiata. Additionally, subarachnoid hemorrhage was observed in the left high frontal region.

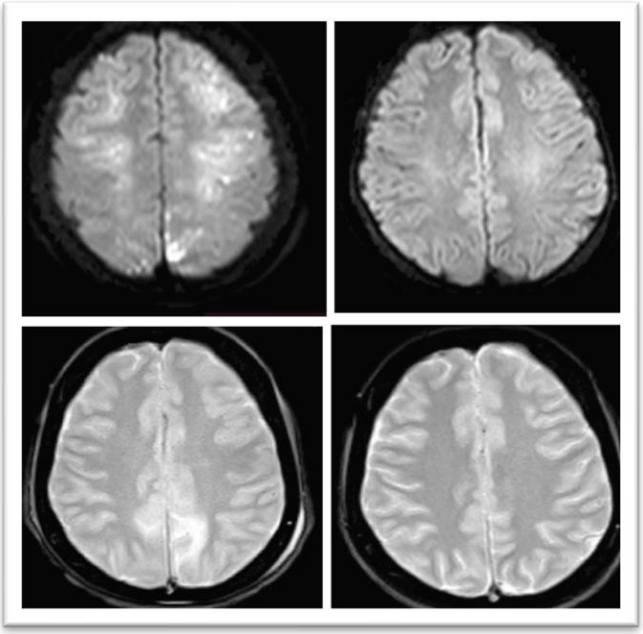

Figure 1: Magnetic Resonance Imaging showing high signal intensity in the cortical and subcortical white matter of the bilateral fronto-parieto-occipital regions.

MRA revealed focal vasoconstriction and steno-occlusion of the C6 segment of the right internal carotid artery, M2 segments of bilateral middle cerebral arteries (MCA), A1 and A2 segments of bilateral anterior cerebral arteries (ACA), and P1 and P2 segments of posterior cerebral arteries (PCA). MR venography (MRV) showed a hypoplastic left transverse sinus.

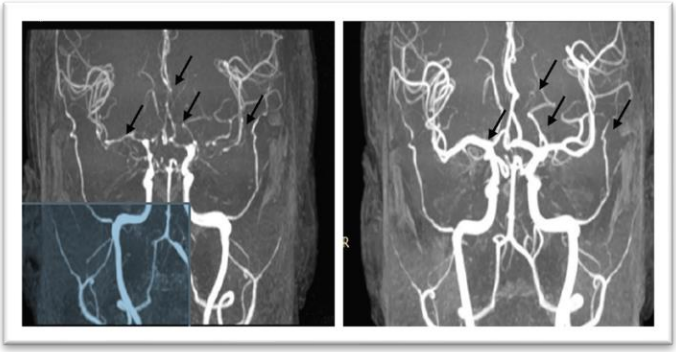

Figure 2 :- Magnetic Resonance (MR) angiography showing diffuse narrowing of cerebral vessels (Left). Note complete resolution of abnormalities at the time of 1 month follow up.

The patient was managed with antiepileptics, antihypertensives (labetalol and cilnidipine), and supportive measures. Remarkably, her motor strength began to improve two days after treatment, and she regained full motor function by day five. She was discharged one week later in stable condition. On follow-up after one month, the patient was asymptomatic, and repeat MRI showed complete resolution of the imaging abnormalities, with a normal angiogram.

DISCUSSION

Postpartum RCVS is a rare but significant neurological condition that can mimic other serious conditions such as eclampsia, cerebral venous thrombosis, or primary angiitis of the central nervous system (PACNS).[6] The hallmark of RCVS is the reversible narrowing of cerebral arteries, which can lead to a range of complications, including SAH, as seen in this case. The rapid resolution of both clinical symptoms and imaging findings is a distinguishing feature of RCVS setting it apart from other vasculopathies.[7]

Several similar cases have been reported in the literature. Sharma et al described a postpartum case of RCVS with radiological features of a haemorrhagic stroke, which resolved following appropriate management.[8] Similarly, Skeik et al analyzed multiple cases of postpartum RCVS and highlighted the importance of recognizing this condition early to prevent misdiagnosis and inappropriate treatment.[9] Ducros et al provided a comprehensive overview of the clinical and radiological spectrum of RCVS, underscoring the variable presentations and the necessity for high clinical suspicion.[10]

In this case, the diagnosis of RCVS was supported by the typical imaging findings of segmental vasoconstriction on MRA and the clinical course, which included a dramatic response to treatment with full recovery. The presence of SAH and infarctions in watershed areas further supports the diagnosis, as these are common complications of RCVS. The patient's complete recovery within one month, both clinically and radiologically, is consistent with the self-limiting nature of the syndrome.

This case highlights the need for awareness of RCVS among clinicians, particularly in the postpartum setting, where it may be easily confused with more common conditions like eclampsia. Prompt diagnosis and appropriate management are crucial in preventing long-term neurological sequelae.

CONCLUSION

Reversible cerebral vasoconstriction syndrome (RCVS) is a rare but important diagnosis to consider in postpartum patients presenting with neurological symptoms. Early recognition and treatment are essential to prevent complications and ensure complete recovery, as demonstrated in this case.

Conflict Of Interest : None

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