Reversible Cerebral Vasoconstriction Syndrome in Postpartum Patient With Spontaneous Intracranial Hypotension: A Case Report

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Case report

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Abstract

Background

Reversible cerebral vasoconstriction syndrome (RCVS) is a rare secondary headache disorder. It may have possibly fatal consequences but its pathophysiological basis is still debated. Herein, we presented a rare case of postpartum patient with spontaneous intracranial hypotension (SIH) suffered RCVS.

Case presentation

A 29-year-old nulliparous female underwent SIH as a result of painless labor and the clinical symptom of postural headache was relieved with a conservative treatment. Two days later, she suffered new-onset thunderclap headache with cortical blindness and generalized seizure; meanwhile, arterial pressure was higher than the normal value. MRI scan found FLAIR hyperintensities in the right cerebellum, left caudate nucleus and bilateral frontotemporal parietal occipital lobes with predominant vasogenic edema. Simultaneously, MR angiography revealed segmental vasoconstriction involving arteries of anterior and posterior cerebral circulations. Taken together, these findings were in accordance with the diagnostic criteria of RCVS and the abnormal MRI hypersignals were mostly consistent with posterior reversible encephalopathy syndrome (PRES). On day 10, she recovered completely and previous abnormal signals disappeared markedly with the treatment of bed rest, high-dose fluids, non-steroidal drug, calcium channel blocker and antiepileptic drug.

Conclusions

RCVS can occur in postpartum patients with SIH, and PRES may also encounter in these cases. A heightened awareness of the disease accompanied by typical clinical and imaging features are associated with an early diagnosis and a better prognosis.

Key Findings Bullet Points

Identification of occurrence of RCVS in postpartum patient with SIH.

Emphasis the uncommon complication of PRES in patient with RCVS.

Highlights the differentials diagnosis and early therapy for RCVS.

Background

Reversible cerebral vasoconstriction syndrome (RCVS) is a rare but increasingly recognized disorder. The syndrome is characterized by thunderclap headache with or without additional focal neurological deficits or seizures, and diffuse segmental narrowing of the cerebral arteries that resolves spontaneously within 3 months[1,2]. It usually follows a benign, monophasic, and self-limited course, but catastrophic forms such as ischemic stroke, intracranial hemorrhage, cortical subarachnoid haemorrhage, posterior reversible
encephalopathy syndrome (PRES) and death may also occur in a small minority of patients[3,4,5]. The vasospastic disorder may firstly involve small distal arteries responsible for thunderclap headache, haemorrhagic strokes and PRES, and then progresses towards medium sized and large arteries responsible for ischaemic events[6]. The pathological process of RCVS remains mostly unknown but a transitory cerebral vascular autoregulation dysfunction and blood–brain barrier breakdown might be underlying mechanisms [7,8]. Spontaneous intracranial hypotension (SIH) is a treatable cause of headache which is predominantly presenting with low cerebrospinal fluid pressure and postural headache[9]. An early diagnosis and therapy may guarantee a good outcome, while delays may result in irreversible poor outcome in some cases. The certain precipitants of RCVS are still not well established, although a report has proposed RCVS may trigger by intracranial hypotension as in postpartum patient[10]. Here, we present a rare case of postpartum patient with SIH who subsequently suffers RCVS.

Case Presentation

A 29-year-old nulliparous female with an uneventful pregnancy underwent painless labor by continuous peridural analgesia-anesthesia induced at gestational age 39 weeks + 6/7. She initially developed postural headache in 8 hours after delivery without other neurological symptoms and signs. SIH was considered according to the patient’s medical history and her typical clinical manifestations. Conservative approaches include strict bed rest, increasing fluid intake, intravenous hydration and lornoxicam were administered to the patient. After that, orthostatic headache was relieved for the next two days. However, the patient suffered severe bilateral parietal occipital excruciating headache that peaked in less than 1 min. This headache was qualitatively and quantitatively different from her previous headache attributed to SIH. Meanwhile, she also complained a blurred vision and developed a generalized tonic-clonic seizure lasting for approximately one minute. Simultaneously, abnormal arterial pressure was found which was significantly higher than the normal value (with a systolic blood pressure>170mmHg and a diastolic blood pressure>100mmHg). Neurological examination revealed cortically blind but denied any other positive neurologic examination result. A full blood count, markers of inflammation, and basic metabolic panel were normal. The postpartum patient with SIH suffered acute severe headache, binocular visual impairment, and generalized seizure, which suggested the lesions emerging were most probable in the bilateral cerebral cortex. Subarachnoid hemorrhage (SAH) owing to ruptured aneurysm was firstly suspected due to the sudden severe headache. However, brain CT scan was normal and SAH was less probable. Further, enhanced MRI suggested the diagnosis of SIH which showed diffuse pachymeningeal gadolinium enhancement, downward displacement of the brain, and engorgement of venous structures (Fig. 1). Patients with SIH may present with orthostatic headache, reduced visual acuity, and generalized seizure; however, severe new-onset headache and elevated blood pressure in our patient must be assessed urgently which were rare clinical symptoms of SIH. FLAIR MR sequence showed hyperintensities in the right cerebellum, left caudate nucleus and bilateral frontotemporal parietal occipital lobe, with no obvious diffusion restriction on diffusion weighted imaging (Fig. 2). However, the differential diagnosis of acute intracranial multifocal lesions was wide. The presence of acute headache, visual impairment, generalized seizure, and intracranial multifocal lesions of the peripartum patient
should raise the possibility of cerebral venous sinus thrombosis, but the result of MR venography was normal. Thus, venous cerebral infarction was excluded. Transient peri-ictal MRI abnormalities (TPMA) might occur during the acute ictal and postictal phases of seizures. But it was strongly associated with prolonged status epilepticus and lateralized periodic discharges. Meanwhile, electroencephalography monitoring was normal which didn’t suggest a diagnosis of TPMA. RCVS may be another possible diagnosis as it could present with similar symptoms and elevated arterial blood pressure. MR angiography (MRA) was performed which clearly revealed diffuse, multifocal, segmental narrowings involving large and medium-sized arteries in the anterior and posterior circulations, with occasional dilated segments, like ‘strings and beads’ (Fig. 2). Furthermore, distinguishing primary angitis of the central nervous system (PACNS) and RCVS could also be a challenge in clinical practice. However, the clinical features of female, postpartum and thunderclap headache were most frequent in RCVS. Based on the above, the clinical features of the patient were in accordance with the diagnostic criteria of RCVS and the abnormal MRI FLAIR hypersignals with predominant vasogenic edema were mostly consistent with PRES. Except for a treatment with bed rest, appropriate liquid, and non-steroidal drug, calcium channel antagonist (nifedipine), intravenous administration of diazepam and antiepileptic drug (levetiracetam) were prescribed to our patient. She was seizure freedom quickly and the symptoms of postural headache and cortically blind resolved completely within a week. A follow-up MRI was conducted 10 days after the initial MR imaging which showed complete remission of the previous abnormal signals; meanwhile, head MRA revealed diffuse segmental constriction of cerebral arteries markedly resolved (Fig. 2).

**Discussion And Conclusions**

Postpartum patient with SIH suffering RCVS is rarely reported. Here, we described a young postpartum patient initially developed SIH after painless labor. Subsequently, she suffered thunderclap headache with cortical blindness and generalized seizure. MRI FLAIR sequence found predominantly bilateral regions of subcortical lesions and MRA showed diffuse segmental constriction of cerebral arteries which were consistent with RCVS. With an aggressive treatment, the patient recovered completely and abnormal MRI signals disappeared markedly in ten days.

RCVS is a rare but still poorly understood syndrome which may occur spontaneously or be provoked by postpartum and exposure to adrenergic or serotonergic drugs[1]. Late pregnancy and early puerperium with a high probability of vasoconstriction are common conditions linked to RCVS[11]. Patients who expose to licit or illicit drugs, ergot alkaloid derivatives, immunosuppressant or immunomodulator drugs and blood products are associated with RCVS; other clinical conditions such as acute head and neck conditions, cerebral vascular associations, catecholamine secreting tumors, and headache disorders may also encountered[6,7,8]. However, the certain precipitants of RCVS are still not well established and the pathological process is unknown. SIH occurred in postpartum setting is a relatively rare and easily overlooked condition associated with RCVS, but RCVS may lead to functional disability and death due to postpartum angiopathy[11,12]. In patients with SIH, cerebrospinal fluid (CSF) leakage may lead to traction on intracranial meningeal vessels and a loss of venous blood pooling[12].Cerebral vasospasm is probably triggered by the anatomical displacement of the brain caused by the decrease in CSF
volume[13]. Emotional or pain stimuli due to SIH may trigger an activation of the adrenergic system and vasospasm of the cerebral vessels can occur. Meanwhile, the loss of intracranial volume due to CSF leakage with compensatory dilatation of intracranial venous system could result in adrenergic overstimulation which is also associated with cerebral vasospasm and the occurrence of RCVS.

In our case, the new-onset thunderclap headache (TCH) must be assessed urgently and underlying fatal causes must be considered. SAH, ischaemic stroke, cerebral venous sinus thrombosis, cervical artery dissection, intracranial infection, PRES and RCVS may be potential causes of TCH[14]. It often reaches maximal intensity in under one minute, with or without seizures and focal neurological deficits in patients with RCVS[15]. TCH initially requires a non-contrast brain CT to exclude aneurysmal SAH; if negative, a lumbar puncture should be performed to exclude intracranial infection and untypical SAH[7,8,14]. Non-invasive imaging of MRI, MRA, and CTA of the brain may help exclude other important differentials and reveal the features of RCVS which may find segmental vasoconstriction involving arteries of the circle of Willis that resolves spontaneously within 3 months; while digital subtraction angiography is gold standard diagnostic investigation which can clearly display small distal arteries[16,17]. Except for TCH and seizures, other neurological symptoms, including altered cognition, visual disturbances, motor and sensory deficits, or ataxia, can occur secondary to ischemic or hemorrhagic stroke in brain regions that are perfused by sustained cerebral vasoconstriction[7,15,18]. Transient hypertension can also occur just like in our case, and more than one third of patients have blood pressure surges during headache attacks[16,19]. Brain imaging with either CT or MRI of RCVS may be normal, but cortical surface subarachnoid hemorrhage (cSAH), PRES, intracranial hemorrhage, and ischemic stroke might encounter in some patients. As in our case, elevated arterial blood pressure and vasoconstrictions in major cerebral arterial segments of M1 and P2 were important determinants for PRES in patients with RCVS[16,20].

RCVS is a rare neurological disorder which has become increasingly recognized owing to improved imaging techniques and clinical awareness. However, accurate diagnosis can be a challenge in clinical practice. SAH from a ruptured cerebral aneurysm is a well-recognized and life-threatening differential diagnosis which may be confused with cortical SAH. Cortical SAH has received limited attention, and in younger patients the commonest cause is RCVS[21]. Younger age, recurrent TCH, chronic headache disorder, prior depression, and the presence of bilateral arterial narrowing are predictors for RCVS-SAH[7,22]. PACNS and RCVS are invariably considered in the differential diagnosis of new cerebral. PACNS requires long-term immunosuppression therapy while steroids may be deleterious in RCVS, further underscoring the need for proper early distinction between RCVS and PACNS. Patients with RCVS are often female, with migraines, and postpartum. TCH, cortical SAH, border zone infarcts and vasogenic edema have the highest positive predictive value for diagnosing RCVS; but multiple small deep infarcts, extensive deep white matter lesions, tumor-like lesions, or multiple gadolinium-enhanced lesions are observed only in PACNS[23,24]. Meanwhile, cerebral arteriopathies such as intracranial atherosclerosis, infectious arteritis and fibromuscular dysplasia should be considered and excluded. Just like in our case, PRES-like reversible cerebral edema is not uncommon in RCVS which may often occur in the first week and a rapid increase in blood pressure is supposed to be key factor for the development of PRES[20].
However, PRES and RCVS may share some common clinical and radiologic features and occasionally occur in the same patient which is difficult to make differential diagnosis.

In conclusion, RCVS can occur in postpartum patient when one suffered thunderclap headache with or without other acute neurological symptoms. A heightened awareness accompanied by typical clinical features and reversible diffuse segmental constriction of cerebral arteries are associated with an early diagnosis and a better prognosis.

Declarations

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Authors’ contributions

YY contributed to the concept, drafting, and reporting of the case. ZJC, LSH, and ZSS acquired clinical data. DJF, ZY, TYF contributed to revision of the manuscript. All authors have read and approved the final manuscript.

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Availability of data and materials

All data related to this case report are documented within this manuscript.

Ethics approval and consent to participate

Informed consent was obtained from the patient to publish her case, and approval for this study was provided by the Research Ethics Committee of Mianyang Central Hospital.

Consent for publication

Written informed consent for publication of this case report was obtained from the patient.

Competing interests

The authors declare that they have no competing interests.

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**Figures**

![Figure 1](image-url)
Enhancement MRI scan revealing diffuse pachymeningeal gadolinium enhancement (A, B), downward displacement of the brain (C), and engorgement of venous structures (D).

Figure 2

FLAIR sequence showed hyperintensities in the right cerebellum, left caudate nucleus and bilateral frontotemporal parietal occipital lobe, without obvious diffusion restriction (E), and MRA showed a ‘string and beads’ appearance of intracranial vessels (F). A follow-up MRI and MRA (10 days later) found complete remission of the previous abnormal MRI signals (G); and MRA revealed diffuse segmental constriction of cerebral arteries markedly resolved (H).