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CASE REPORTS

Reversible cerebral vasoconstriction syndrome presenting as convexity subarachnoid hemorrhage and posterior reversible encephalopathy syndrome during postpartum: A case report and literature review

*Zejin Jia *MM*, *Wei Qin *MD*, Wenli Hu *MD*, Junliang Yuan *MD*

**Zejin Jia and Wei Qin contributed equally to this work and are joint first author.*

Department of Neurology, Beijing Chaoyang Hospital, Capital Medical University, Beijing, China

Abstract

Reversible cerebral vasoconstriction syndrome (RCVS) is characterized by thunderclap headache and reversible cerebral vasoconstriction, with other neurologic signs and symptoms. To the best of our knowledge, there were only a few cases of RCVS presenting both as both convexity subarachnoid hemorrhage (cSAH) and posterior reversible encephalopathy syndrome (PRES). Herein, we report a case of a 32-year-old woman with RCVS who presented with recurrent thunderclap headaches that occurred 50 days after delivery, with cSAH and PRES on magnetic resonance imaging (MRI). She had significant clinical and radiological recovery on 3 months' follow-up. The clinical coexistence of cSAH and PRES in our case with RCVS is quite rare. This case illustrates the importance of awareness of the diagnosis of RCVS among clinicians even when initial brain and vascular imaging are normal. Early diagnosis and treatment are crucial for better prognosis.

Keywords: Reversible cerebral vasoconstriction syndrome, convexity subarachnoid hemorrhage, posterior reversible encephalopathy syndrome, stroke

INTRODUCTION

Reversible cerebral vasoconstriction syndrome (RCVS) is a rare but increasingly recognized disorder. It is characterized by recurrent severe thunderclap headaches with or without other neurological symptoms, where diffuse segmental constriction of the cerebral arteries is present, usually resolving within three months.[1] This condition is increasingly supposedly to be due to a transient disturbance in the control of cerebral vascular tone with sympathetic overactivity. More than half of the cases occur after exposure to vasoactive substances or in the postpartum period.[2]

Acute non-traumatic convexity subarachnoid hemorrhage (cSAH) is increasingly recognized as a risk factor for future symptomatic intracerebral hemorrhage (ICH).[3] It may be related to a wide spectrum of etiologies.[4] The most common causes of cSAH are RCVS and cerebral amyloid angiopathy (CAA) without lobar hemorrhage.[5,6] Posterior reversible encephalopathy syndrome (PRES) is another clinical-radiologic syndrome characterized by reversible, posterior-predominant brain edema, usually associated with headache, altered mental status, seizures, and visual symptoms.[7] To date, there are only a few cases of RCVS presenting both with cSAH and PRES. We present here a rare case of a postpartum female with RCVS presenting with cSAH and PRES, with subsequent complete recovery of neurological and imaging findings.

CASE REPORT

A 32-year-old woman developed a severe thunderclap headache while defecating. Her blood pressure was elevated (180/100mmHg). She had undergone spontaneous vaginal delivery 50 days prior. Two days later, she had recurrent severe thunderclap headaches induced by coughing. Neurological examination revealed

Address correspondence to: Wenli Hu and Junliang Yuan, Department of Neurology, Beijing Chaoyang Hospital, Capital Medical University, 8th Gongti South, Chaoyang, Beijing 100020, China Tel/Fax: +86 10 85231376, email: Wenli Hu, wenlihu3366@126.com, Junliang Yuan, junliangyuan@ccmu.edu.cn

no abnormalities. Laboratory examinations were within normal limits.

Three days after the first episode of headache, initial computed tomography (CT) of the brain suggested bilateral cSAH over the parietal and temporal cortices, especially on the right side (Figure 1). At the same time, magnetic resonance imaging (MRI) of the brain also revealed bilateral cSAH and high-intensity FLAIR lesions located in the parietal and temporal cortices suggestive of PRES (Figure 2). However, magnetic resonance angiography (MRA) revealed no abnormalities. Digital subtraction angiography was also unremarkable six days after the first episode of headache. Transcranial Doppler showed elevated velocities in the bilateral middle cerebral artery, the right anterior cerebral artery, the left cerebral posterior cerebral artery and the left internal carotid artery 14 days after the first episode of headache.

After admission, she was treated with Nimodipine 60 mg 4-hourly, which effectively controlled the blood pressure. Nimodipine was first given by IV during in hospital. She was discharged well after two weeks. Nimodipine was continued by oral after discharge from the hospital.

Fourteen days after the first episode of headache, repeat brain MRI showed complete resolution of the cSAH and high-intensity lesions in the bilateral occipital and parietal lobes (Figure 3). However, on this occasion, brain MRA showed multiple segmental constrictions of the cerebral arteries. A final diagnosis of the concurrence of RCVS associated with cSAH and PRES was made. Repeated brain MRA three months later showed absolute resolution of the vasoconstriction.

DISCUSSION

RCVS is an intracranial vascular manifestation of a wide variety of etiologies. RCVS is considered the second most common cause of thunderclap headache and the most common cause of recurrent severe secondary headaches. It is characterized by reversible segmental constriction and dilatation of cerebral arteries, typically associated with recurrent thunderclap headaches and often complicated by ischemic or hemorrhagic strokes.[7] Although RCVS may be spontaneous, it is often associated with the postpartum state or exposure to provocative drugs.[8] The prognosis of RCVS is favorable, and most patients may have a good outcome. However, the clinical outcome may be guarded when complicated with stroke.

RCVS has been reported to be the commonest cause of isolated cSAH in patients <60 years of age.[9] More than 30% of patients with RCVS suffer from cSAH.[10] RCVS sometimes occurs in concert with PRES. PRES is characterized by the acute onset of neurologic symptoms including headache, altered mental status, visual changes and seizures, with accompanying vasogenic edema

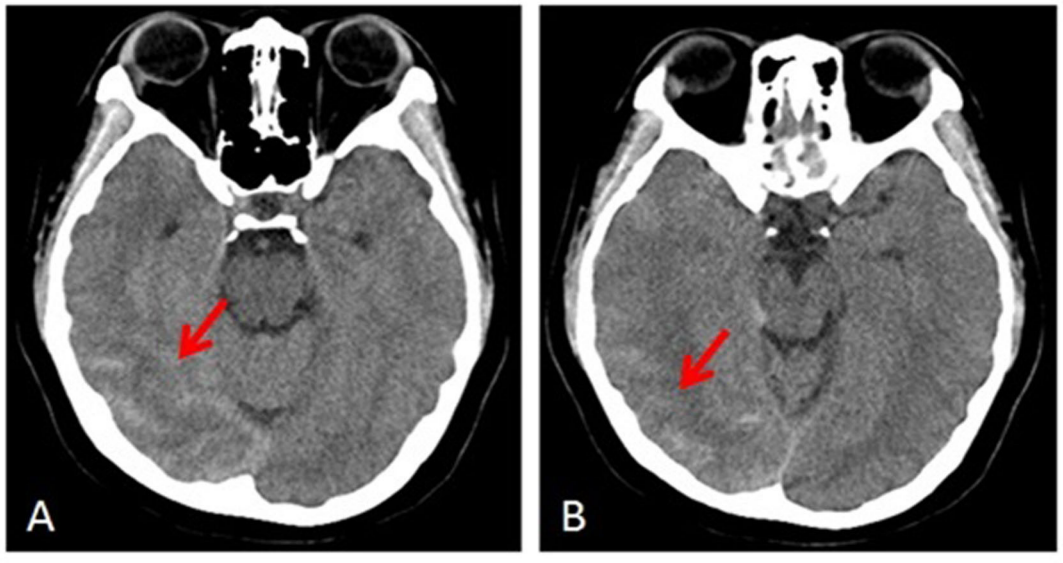

Figure 1. Brain CT suggested bilateral parietal and temporal cortices of cSAH, especially lateralized to the right hemisphere.

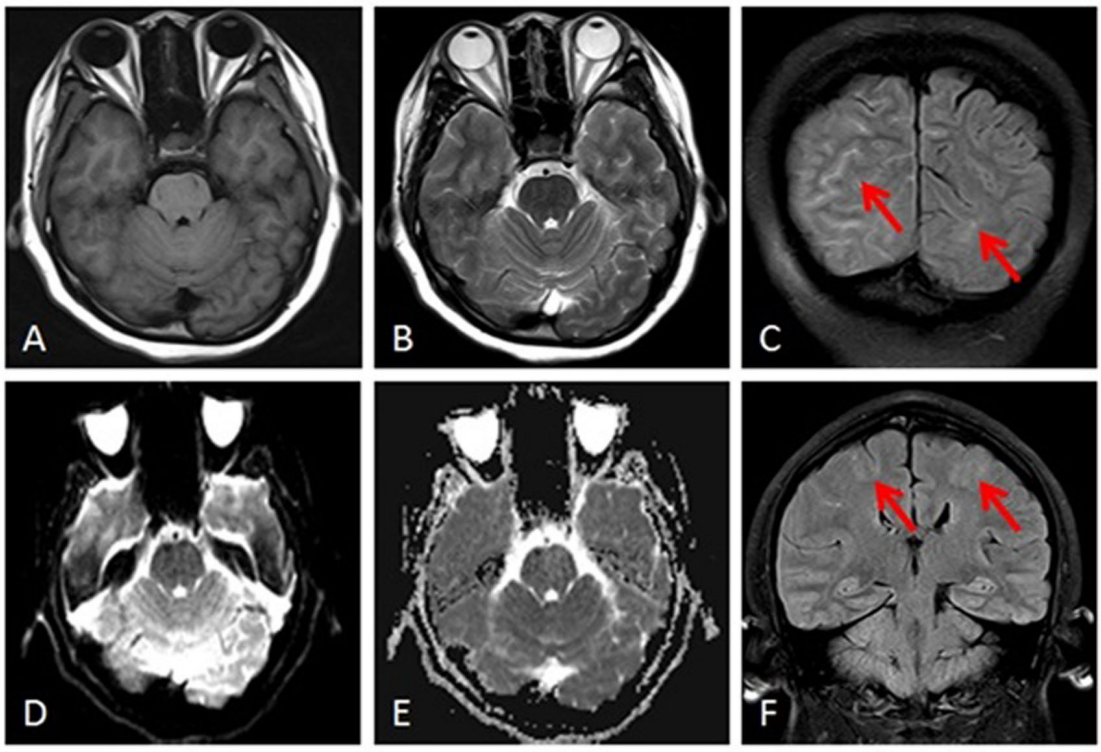

Figure 2. Brain MRI revealed abnormal high-intensity lesions in bilateral parietal and temporal cortices.

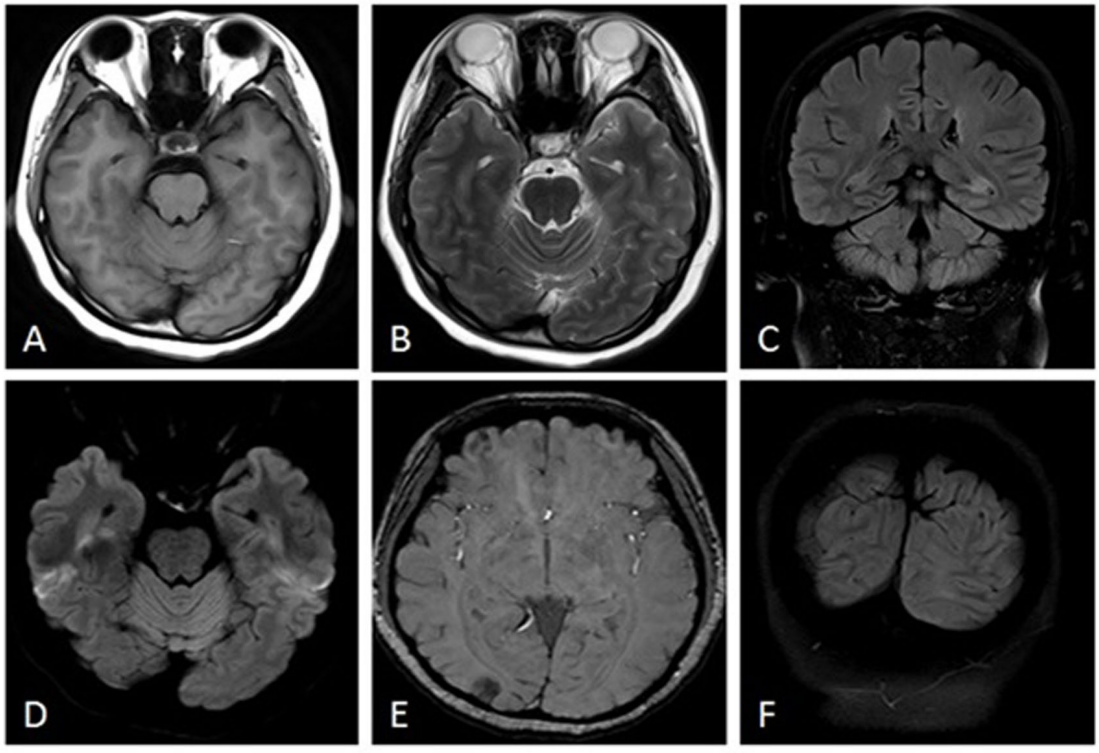

Figure 3. Brain MRI indicated complete resolution of the cSAH and high-intensity lesions in bilateral parietal and temporal cortices 14 days after the first episode of headache.

on neuroimaging. Risk factors of PRES include infection, uremia, malignancy, autoimmune disorders, the peripartum state and hypertension. PRES shares similar clinical and radiological features with RCVS and can coexist with it. They may also share a common pathophysiology.[7]

To the best of our knowledge, the co-occurrence of RCVS and PRES is rare and has been reported in the following: postpartum and post-transfusion states, intracranial hypotension, licorice toxicity, hemolytic uremic syndrome, bilateral carotid paraganglioma resection, oral contraceptive pill usage, intravenous immune globulin therapy in Guillain-Barre syndrome and heart transplantation.[7,11-21] The pathogenesis of RCVS and PRES is poorly understood, however, autonomic dysregulation, oxidative stress, and genetic predisposition have been postulated.[1]

Angiographic analysis has shown more severe vasoconstriction in distal versus proximal segments in all lesion types (ICH, cSAH, infarction, vasogenic edema).[24] Early distal vasoconstriction was associated with lobar ICH and cSAH and delayed proximal vasoconstriction with infarction. It has also been reported that early vasogenic cerebral edema was related to small vessel dysfunction with acute disruption of the blood-brain barrier.[25] The pathophysiology of hemorrhagic complications underlying in RCVS or PRES remains uncertain. However, alteration in cerebral vascular tone leading to vasoconstriction is recognized to be a key pathophysiologic mechanism underlying the development of RCVS.[26]

RCVS typically occurs one to three weeks after deliver but has been described up to 6 weeks post-partum.[26] Our case presented with RCVS 50 days after delivery and extends the known time period for recognition of this condition post-partum.

In summary, RCVS is considered a rare syndrome, but the growing use of vasoactive drugs, postpartum, combined with the more frequent use of non-invasive neurovascular imaging increase the incidence. RCVS often occurs in association with PRES and cSAH, and those conditions are likely to share a common pathophysiology. However, concurrence of PRES and cSAH is rare. Our case also illustrates the possibility of the diagnosis of RCVS even when initial vascular imaging is normal. Early diagnosis and treatment is crucial for a better prognosis.

DISCLOSURE

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Conflict of interest: None

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