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## CLINICAL INFORMATION

# Reversible cerebral vasoconstriction syndrome a rare cause of post-partum headache: an anesthetic overview



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### KEYWORDS

Reversible cerebral vasoconstriction syndrome;  
Postpartum headache;  
Young female

### Abstract

Reversible cerebral vasoconstriction syndrome is a cerebrovascular disorder leading to multifocal arterial constriction and dilation. Reversible cerebral vasoconstriction syndrome is possibly caused by transient deregulation of cerebral vascular tone. We report a rare case of a patient with chief complain of postpartum headache, was later diagnosed as a case of reversible cerebral vasoconstriction syndrome. A young full term primigravida with good uterine contraction admitted to labour room. Later she complained of leaking per vagina and on examination meconium stained liquor was noted. Caesarean delivery under spinal anesthesia was done and intra-operative period was uneventful. Both mother and baby were normal and shifted to post-operative ward and nursery respectively. In postoperative ward, mother complained of severe headache after 1 h and later developed seizure. Midazolam was given intravenously and was intubated and transferred to critical care unit for further investigation and management. Non contrast computerised tomography scan of brain showed right occipital intracerebral as well as subarachnoid bleed. CT angiography showed right vertebral artery narrowing without any other vascular malformation. Patient was managed in critical care unit for 2 days and then extubated and shifted to high dependency ward after a day observation and discharged 3 days later after a full uneventful recovery.

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### PALAVRAS-CHAVE

Síndrome de vasoconstrição cerebral reversível;  
Cefaleia pós-parto;  
Mulher jovem

### Síndrome de vasoconstrição cerebral reversível, uma causa rara de cefaleia pós-parto: visão da anestesia

### Resumo

A síndrome de vasoconstrição cerebral reversível é uma doença cerebrovascular que leva à constrição e dilatação arterial multifocal. A síndrome de vasoconstrição cerebral reversível é possivelmente causada pela desregulação transitória do tônus vascular cerebral.

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Relatamos um caso raro de uma paciente com queixa principal de cefaleia pós-parto, posteriormente diagnosticada como um caso de síndrome de vasoconstrição cerebral reversível. A jovem primigesta a termo apresentando boa contração uterina foi internada em sala de parto. Mais tarde, a parturiente queixou-se de perda de líquido pela vagina e, ao exame, líquido amniótico manchado foi observado. O parto cesariano sob raqui-anestesia foi realizado, e não houve intercorrência no período intraoperatório. Tanto a mãe quanto o bebê estavam normais e foram transferidos para a sala de recuperação pós-operatória e berçário, respectivamente. Na sala de recuperação, a mãe queixou-se de forte dor de cabeça após uma hora e depois desenvolveu convulsão. Midazolam foi administrado por via intravenosa, e a paciente foi intubada e transferida para uma unidade de terapia intensiva para posterior investigação e tratamento. A tomografia computadorizada sem contraste do cérebro mostrou hemorragia intracerebral occipital direita e subaracnoide. A angiotomografia mostrou estreitamento da artéria vertebral direita, sem qualquer outra malformação vascular. A paciente foi tratada em unidade de terapia intensiva por dois dias e, em seguida, foi extubada e transferida para a ala de alta dependência onde permaneceu um dia em observação, recebendo alta hospitalar três dias depois, após uma recuperação completa e sem intercorrências.

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## Introduction

Reversible cerebral vasoconstriction syndrome is defined as sudden thunderclap headache with reversible multifocal narrowing of cerebral arteries lasting 1–3 months with or without focal neurological symptoms.<sup>1</sup> Postpartum headache is mostly considered a benign symptom.<sup>2</sup> However, it may be a serious premonitory sign. RCVS is mostly found in women 20–50 years of age. Different names are given to this syndrome according to clinical context: Call-Fleming syndrome, benign angiopathy of the CNS, postpartum angiopathy, thunderclap headache with reversible vasospasm, migrainous vasospasm or angiitis, and drug-induced cerebral arteritis or angiopathy. The main clinical manifestation of RCVS are recurrent sudden-onset and severe (thunderclap) headaches over 1–3 weeks, often accompanied by nausea, vomiting, photophobia, confusion and blurred vision.<sup>3</sup> The syndrome is generally self-limited and has a low incidence of recurrence

## Case report

Our patient a full term primigravida, aged 26 years old reported to labour room of our hospital, with good uterine contractions and complained of decreased foetal movements. Her antenatal period was normal, however diagnosed for hypothyroidism and started on tablet thyroxin 50 µg once daily. Foetal wellbeing was assured after a bedside sonography and cardiotocography (CTG). She was planned for vaginal delivery. Later on patient complained of leaking per vagina and on examination meconium stained liquor was noted. Later CTG was done which was non-reactive and she was planned for an emergency caesarean delivery. Spinal block was performed in left lateral position at L4–5 intervertebral space. Injection bupivacaine heavy 10 mg was injected after aspiration of cerebrospinal fluid. Healthy baby was delivered. Surgery lasted for 1 h and all the vitals



Figure 1 Occipital subarachnoid bleed.

are within normal limit intra-operatively. Later both mother and baby were shifted to postoperative ward and nursery respectively. One hour later patient started complaining of severe headache. Patient was thus examined and found to have blood pressure of 170/100 mm of Hg along with surgical site pain. She was suspected of post dural puncture headache (PDPH) or surgical pain. She was thus advised for tablet paracetamol, injection fentanyl 50 µg i.v. and intravenous fluids. Soon, patient developed two episodes of generalised tonic clonic seizure half an hour later. Her consciousness also deteriorated to a Glasgow Coma Scale (GCS) of 9/15. She was administered 5 mg of injection midazolam. Her trachea was intubated and shifted for NCCT scan. NCCT scan revealed right occipital intracerebral as well as subarachnoid bleed (Figs. 1 and 2). Patient was further shifted to critical care unit on ventilator. Injection phenytoin 100 mg thrice daily along with other supportive measures was started. Hypertension resolved after 9 h of episode by tablet Nifedipine 10 mg twice daily. Her blood and urine investigation did not reveal any aetiology. However, CT angiography revealed right vertebral artery



**Figure 2** Occipital intracerebral bleed.

narrowing without any other vascular malformation. Patient further had no episodes of hypertension or seizures next day. She was extubated after confirming a normal GCS. She was shifted to high dependency wards after a day observation and discharged 3 days later with full uneventful recovery.

## Discussion

Postpartum headache is described as a complaint of cephalic, neck or shoulder pain from placental delivery to six weeks postpartum. Goldszmidt et al. even reported a high incidence of 39% in first postpartum week. With increased concern of PDPH among obstetricians, anaesthetists' are commonly the first one called to review post caesarean patients complaining of headache.

International headache society classifies headache as primary and secondary.<sup>4</sup> Primary headaches includes causes without underlying pathology. Contrary to the common dictum primary headaches *i.e.* tension and migraine not the PDPH are the most common cause of postpartum headache. Further PDPH is not even the most common secondary cause of headache. Musculoskeletal headache (11–14%), pregnancy induced hypertension (8–24%) followed by PDPH (4–16%) accounts for most of the secondary postpartum headache.

A postpartum presenting with headache and sudden onset hypertension can put any anaesthetist in dilemma of varied possibilities. The possibilities could vary from simple post-operative pain to cerebrovascular accidents.

Our patient had sudden onset headache followed by transient hypertension, seizures, unconsciousness followed by later to complete recovery. Clinical and NCCT finding raised the suspicion of RCVS.

There have been many similar reports of RCVS.<sup>5</sup> Most of these patients had acute and transient onset of headache, hypertension followed by neurological deficits. Some cases reported intracerebral bleed, subarachnoid bleed and both.<sup>6</sup> Cerebral angiography of all these cases revealed classic picture of vasculitis with no increase in inflammatory markers.

Reversible cerebral vasoconstriction syndrome is a recently recognised syndrome marked by sudden onset headache, hypertension, seizure with or without neurological deficits. RCVS presents as a wide clinico-radiological spectrum varying from benign oedema, ischaemic or haemorrhagic picture. Various names including benign angiopathy of the central nervous system,

postpartum angiopathy and others have been proposed to describe the same clinical–radiological syndromes.<sup>7</sup> Finally, in 2007 a common term RCVS has been agreed on by a panel of experts.

RCVS as a cause of stroke is not well understood. Multiple theories have been prescribed for pathophysiology; most accepted being that of disrupted cerebral auto regulation. Increased blood pressure and loss of auto regulation in postpartum phase may cause the clinical picture. Experiments have shown that acute hypertension can produce areas of vasospasm and dilation.<sup>3</sup> Further rapid resolution of symptoms without an increase of inflammatory markers suggests a transient vasospasm rather than vasculitis.<sup>8</sup> It is although still unclear that cerebral vasoconstriction is a reaction to hypertensive episode or represents an independent primary process.

Although, it mostly follows complete improvement, at least one fatal case has been reported. Due to the rare incidence of RCVS, although, there is no true standard protocol for management. Case studies have reported successful treatments even without steroids mostly with favourable outcomes after treatment of hypertension.<sup>5</sup>

Headache could be a serious premonitory sign of RCVS. Although outcomes are mostly favourable, the consequences could be life threatening as well. The major complications of RCVS are non-aneurismal cortical surface Subarachnoid Haemorrhage (SAH), intracerebral haemorrhage, TIA or ischaemic stroke, seizures, and Posterior Reversible Encephalopathy Syndrome (PRES).<sup>9,10</sup> Women and patients with a history of migraines appear to be more at risk for intracranial haemorrhage in those with RCVS.<sup>11</sup> In fact, RCVS recurrence is reported to occur in approximately 5% of all RCVS cases.<sup>12</sup> Prevention of these complications necessitates active and early intervention for hypertension and other offending agents. Studies are thus required to determine threshold and strategies for management RCVS.

## Conclusion

A postpartum headache should be taken seriously. Reversible cerebral vasoconstriction syndrome may present as sudden onset headache, hypertension, seizure with or without neurological deficits. Its clinico-radiological spectrum varying from benign oedema, intraparenchymal haemorrhage, subarachnoid haemorrhage and ischaemic stroke. We should consider CT scan and angiogram for proper diagnosis and management. Outcome is mostly favourable in all reported cases till now even with supportive treatment.

## Conflicts of interest

The authors declare no conflicts of interest.

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