Article published online: 2023-04-24



# Peripartum Cerebral Angiopathy

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J Neuroanaesthesiol Crit Care 2023;10:56–59.

## Abstract

Peripartum cerebral angiopathy (PCA) is a rare cerebrovascular disorder of unclear etiology that typically occurs in the first postpartum week. The aim of this case series is to present seven cases of PCA and the associated outcomes. Two of the cases were typical of a reversible cerebral vasoconstriction syndrome (RCVS) and had no long-term sequelae. Five of the cases involved intraparenchymal hemorrhage in the absence of an underlying vascular lesion, with three of the five cases having associated cerebral vasoconstriction. None of the patients had a documented history of preeclampsia or hypertension during pregnancy. It has been suggested that there is pathophysiological overlap between PCA and eclampsia and RCVS. There is limited evidence in the literature in this regard and, in most cases, treatment has been supportive only. As a condition, PCA may be underrecognized in the neurocritical care setting. Increased awareness of this condition, its manifestations, and options for management may optimize outcomes.

**Keywords**
- peripartum
- postpartum angiopathy
- reversible cerebral vasoconstriction syndrome
- cerebral intraparenchymal hemorrhage

## Introduction

Peripartum cerebral angiopathy (PCA) is a rare cerebrovascular disorder that typically occurs after pregnancy and delivery. Usually, there is segmental narrowing of large- and medium-sized intracranial arteries. Its manifestations range from headaches to ischemic and hemorrhagic strokes.

Here we present seven cases of PCA that demonstrate variable presentations and outcomes that occur with this poorly understood condition (►**Table 1**).

## Patient 1

A 36-year-old primigravid patient developed a headache and neurological deterioration during the first stage of labor. She underwent an emergency cesarean section under general anesthesia. Head computed tomography (CT) demonstrated a right basal ganglia intraparenchymal hematoma (IPH) with intraventricular extension. CT angiogram (CTA) showed narrowing of the right middle cerebral artery (MCA). She underwent a frontotemporoparietal craniectomy with evacuation of IPH. On day 2, cerebral digital subtraction angiography (DSA) demonstrated no underlying vascular abnormality; however, there was widespread narrowing of intracranial vessels. Six months later, DSA showed full reversal of previously detected vasoconstriction. She had residual left-sided hemiparesis.

## Patient 2

A 33-year-old female patient 12 days post-partum developed a sudden severe headache. Head CT and cerebrospinal fluid (CSF) sample were normal. DSA revealed widespread multifocal intracranial arterial narrowing. Inflammatory markers and a vasculitis screen (antineutrophilic cytoplasmic antibody (ANCA), ANCA-proteinase 3 (PR3), ANCA-myeloperoxidase (MPO), Rhesus (Rh) factor, complement C3, C4, connective tissue disease (CTD) screen, and antinuclear antibodies (ANA) were normal. MRI/MRA showed focal fluid-attenuated inversion recovery (FLAIR) hyperintensity in bilateral occipital sulci.

**article published online**
April 24, 2023

**DOI** https://doi.org/10.1055/s-0042-1757912.
**ISSN** 2348-0548.



Thieme Medical and Scientific Publishers Pvt. Ltd., A-12, 2nd Floor, Sector 2, Noida-201301 UP, India

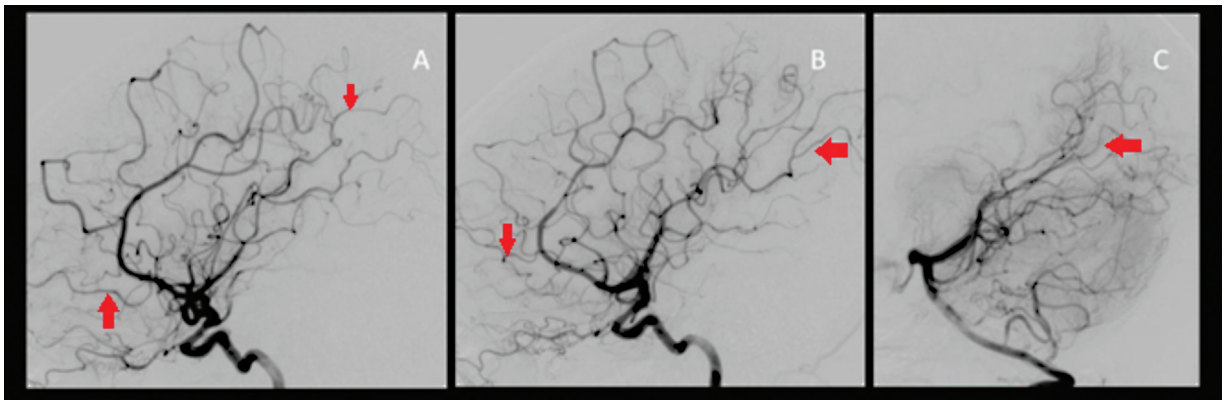


**Fig. 1** Cerebral angiogram images from patient 3. Right ICA (**A**) and left ICA (**B**) and L vertebral artery (**C**) digital subtraction angiography show bilateral peripheral ACA, MCA, and PCA stenoses. Stenoses highlighted with red arrows.

Nimodipine therapy was initiated. Two months later, the patient had repeated mild headaches. Cerebral angiogram demonstrated full resolution of the intracranial vasospasm.

## Patient 3

A 38-year-old female patient 5 days postpartum developed a sudden thunderclap headache. LP was positive for xanthochromia. CTA and venogram were normal. DSA showed multiple focal areas of mild vasoconstriction throughout the intracranial circulation (►**Fig. 1**). Inflammatory markers and a vasculitis screen (ANCA, RH factor, C3, C4 and CTD, ANA, MPO, PR3) were normal. She was commenced on nimodipine therapy to manage what was presumed to be RCVS. DSA performed 4 months later showed complete resolution of intracranial vasospasm.

## Patient 4

At 5 days post-partum, a 37-year-old woman developed a headache, right-sided weakness, and a facial droop. Head CT/CTA demonstrated a left basal ganglia IPH with no underlying vascular malformation. Deterioration in her level of consciousness prompted repeat imaging, which showed extension of her IPH. She underwent an emergency left frontotemporoparietal decompressive craniectomy with evacuation of hematoma. Cerebral angiogram demonstrated thinner caliber of her left MCA and branches but no underlying vascular malformation. After the rehabilitation period, the patient continued to have residual right-sided hemiparesis and dysphasia.

## Patient 5

A 36-year-old woman presented with a severe headache 10 days postpartum. Head CT/CTA were initially normal. Six hours later, she developed right-sided hemiparesis and her level of consciousness dropped. A second head CT demonstrated a large left posterior frontal IPH. Both CTA and venogram were normal. DSA demonstrated subtle peripheral branch narrowing of both middle cerebral arteries. The IPH was conservatively managed. Hematological investigations (factors 2, 5, 7, 8, 9, 10, 13, and von Willebrand factor) were all normal, as was a vasculitis screen. Three months later, she had made a full neurological recovery.

## Patient 6

A 36-year-old woman presented with a new-onset severe headache and leg weakness. She was 5 days postpartum and was normotensive. Head CT demonstrated a right basal ganglia hemorrhage. She underwent a frontotemporoparietal decompressive craniectomy with evacuation of IPH. DSA was normal and no underlying malformation was identified. Six months later, she had a hemiparesis and cognitive difficulties.

## Patient 7

A 33-year-old woman was 7 days postpartum and developed a severe headache. Initial head CT/CTA demonstrated a left basal ganglia IPH and subtle narrowing of the left posterior cerebral artery. Following transfer to our institution, she deteriorated neurologically and required a frontotemporoparietal decompressive craniectomy with evacuation of IPH. Head CT later demonstrated a new area of hemorrhage in the right occipital lobe and a left occipital infarct. Three months later, the patient had dysphasia, cognitive difficulties, and hemiparesis.

## Discussion

Peripartum cerebral angiopathy (PCA) typically occurs within the first postpartum week after a normal uneventful pregnancy and delivery. In this series, one patient became symptomatic in the first stage of labor, thus the term "peripartum" may be more appropriate than "postpartum."

In a case series of postpartum patients with acute neurological problems by Raps et al, three out of the four patients did not have features of proteinuria or edema required for the diagnosis of eclampsia.[1] Two of the patients had diffuse cerebral vasospasm on angiography. MRI abnormalities were present in all patients, most prominently in the parietooccipital regions. They termed this condition "delayed peripartum vasculopathy," suggesting it is a variant of

**Table 1** Summary of seven cases of peripartum cerebral angiopathy

| Patient no. | Age | Parity | Pregnancy complications | Days postpartum | Symptoms at presentation | Findings at initial imaging | Angiopathy Treatment | Outcome |
|---|---|---|---|---|---|---|---|---|
| 1 | 36 | 1 | Nil | Intrapartum | Low GCS | IPH (BG), right MCA vasoconstriction | Magnesium | Hemiparesis |
| 2 | 33 | 3 | Nil | 12 | Headache | Multifocal vasoconstriction | Nimodipine | Normal |
| 3 | 38 | 3 | Nil | 5 | Headache | Multifocal vasoconstriction | Nimodipine | Normal |
| 4 | 37 | 2 | Nil | 5 | Headache, hemiparesis | IPH | Nil | Hemiparesis, dysphasia |
| 5 | 36 | 3 | Nil | 10 | Headache | IPH, mild bilateral MCA vasoconstriction | Nil | Normal |
| 6 | 36 | 4 | Gestational diabetes | 5 | Headache, leg weakness | IPH (BG) | Nil | Hemiparesis, cognitive difficulties |
| 7 | 33 | 2 | Gestational diabetes, polyhydramnios | 7 | Headache | IPH, Left PCA narrowing | Nil | Hemiparesis, dysphasia |

Abbreviations: BG, basal ganglia; IPH, intraparenchymal hemorrhage; MCA, middle cerebral artery; PCA, posterior cerebral artery.

cerebral eclampsia. Reporting a fatal case of PCA, Williams et al suggested that if there is overlap between classic eclampsia, late postpartum eclampsia and PCA, it appears that PCA is limited to the cerebral vasculature, while eclampsia involves both the systemic and cerebral vasculature.[2]

RCVS has been defined as a group of conditions, characterized by reversible multifocal narrowing of the cerebral arteries heralded by severe headaches with or without associated neurologic deficits. Some case series of RCVS have included postpartum patients within the larger cohorts – 12/139, 1/77, 8/89, and 5/67 patients, respectively.[3–6]

Progesterone is the most abundant hormone in pregnancy and begins to fall during labor, with a precipitous decline after delivery.[7] Progesterone is a vasoactive hormone, causing vascular relaxation, an effect potentially mediated via L-type calcium channels. There are case reports of RCVS occurring during periods of decreased progesterone and estrogen levels for non-pregnancy related reasons.[8,9] It is plausible that the rapid change in hormone levels in the peripartum period acts as a precipitant to cerebral angiopathy in people who have a genetic predisposition or other risk factors yet to be elucidated.

Fugate et al reported 18 cases of PCA with half of the patients having residual neurological deficits or dying.[10] Initial arterial brain imaging was often normal, as was the case in this series. This suggests that PCA may begin as a small vessel vasculopathy with progression to involvement of medium and larger size vessels in some cases.

Treatment approaches to PCA have paralleled those for subarachnoid hemorrhage associated vasospasm, eclampsia, posterior reversible encephalopathy syndrome, and primary angiitis of the central nervous system.

## Conclusion

Peripartum cerebral angiopathy (PCA) is a rare syndrome of dysregulated cerebrovascular tone manifesting in its mildest form as a headache, with hemorrhagic and ischemic strokes representing its most severe presentation. The diagnosis of PCA should be considered in all women presenting with new neurological symptoms in the peripartum period.

**Conflict of Interest**
None declared.

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