

## A Case of Postpartum Cerebral Angiopathy

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### Introduction

The incidence of ischemic stroke during pregnancy and postpartum is very low; however, it could be a serious event for mothers and infants. Once it does occur, many concerns arise about the safety of the mother and fetus in relation to common diagnostic tests and therapies. Brain scanning might reveal pathological results in spite of a normal neurological examination. With neurological examination and brain scanning, it may be possible to diagnose and treat severe complications that may otherwise result in maternal mortality [1].

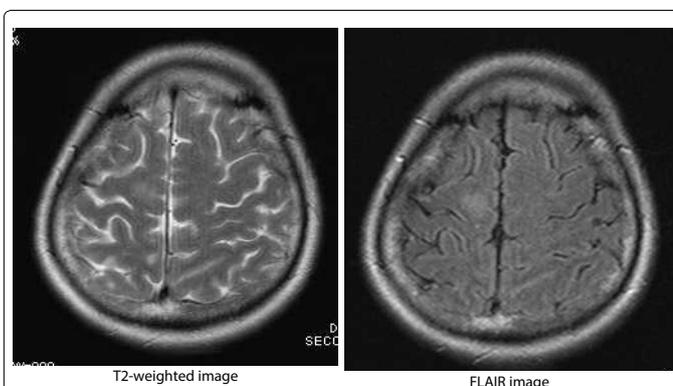
Postpartum Cerebral Angiopathy (PCA) is a reversible clinico-radiological syndrome, characterized by the acute onset of severe headache, focal neurologic deficits, and reversible cerebral segmental vasoconstriction [2-6]. It is diagnosed by angiography, which demonstrates multifocal segmental narrowing in large and medium-sized cerebral arteries, with a similar appearance to vasculitis [2-8]. The process is generally self-limiting; with the resolution of angiographic abnormalities within 4-12 weeks and typically complete resolution of symptoms [1,4]. However, owing to its association with both infarction and hemorrhage, PCA does carry a risk of morbidity and mortality [2,9].

Patients with PCA generally presented the acute onset of severe headache; however, we present a rare case of PCA involving presentation with paresis but without headache.

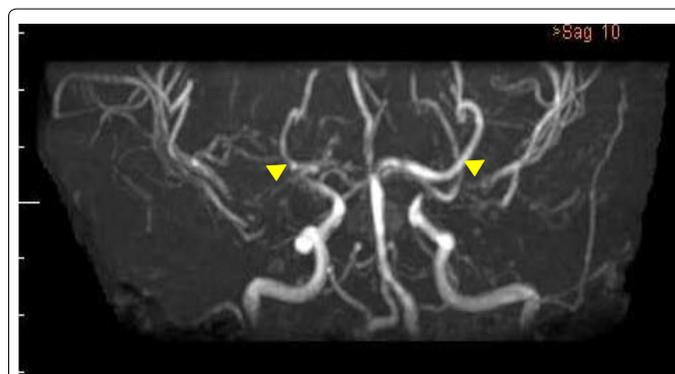
### Case Report

A 34-year-old woman, gravid 1, para 0, was admitted to our hospital for pregnancy-induced hypertension. At 39 weeks of gestation, her blood pressure was 158/86 mmHg, and she presented headache. She had no previous history of heart disease, migraine, diabetes mellitus, or collagen-vascular diseases. At bed rest, although her blood pressure normalized, her headache persisted. On the fourth admission day, a cerebral Computed Tomography scan (CT) was performed, with no significant finding. At 40 weeks of gestation, owing to prolonged labor following premature rupture of the fetal membrane, she underwent an uneventful Cesarean Section (CS). After the CS, her blood pressure

was stable. Two days after the CS, she presented left crural paresis without headache; however, her blood pressure was 130/64 mmHg and her body temperature was 36.5°C. Neurological and general physical examinations were normal, excluding left crural paresis 4 hours after onset. Brain Magnetic Resonance Imaging (MRI) was performed, which revealed T2/Fluid Attenuated Inversion Recovery (FLAIR) hyperintensity in the right frontal-parietal lobe (Figure 1). It also revealed an acute phase of cerebral infarction. Blood tests, including those for immunologic lupus erythematosus preparation, such as antinuclear antibodies, the erythrocyte sedimentation rate, and antiphospholipid antibodies, were all normal. Treatment with edarabon as a free radical scavenger and oral aspirin was started immediately (day 1). Magnetic Resonance Angiography (MRA) on day 2 showed an occlusion of the bilateral proximal middle cerebral arteries (Figure 2). By day 6, her paresis had almost disappeared. On day 8, MRA showed improvement of occlusion of the bilateral proximal middle cerebral arteries and new, irregular, multiple narrowings of the bilateral internal carotid arteries (Figure 3). Cerebral arteriography, performed on day 9, showed a marked narrowing of the bilateral middle cerebral and anterior cerebral arteries, and irregular, multiple narrowings of the bilateral internal carotid arteries. These findings ruled out a spasm of the cerebral arteries. A month later, although MRA showed multiple narrowings remaining of the internal carotid arteries, her paresis had completely resolved. Four months later, MRA showed no abnormal findings, and she is currently well without headache or paresis.



**Figure 1:** MRI 4 hours after onset  
Both T2-weighted and FLAIR imaging showed a hyperintense area (arrows) in the right frontal-parietal lobe. Findings are consistent with acute ischemic stroke.



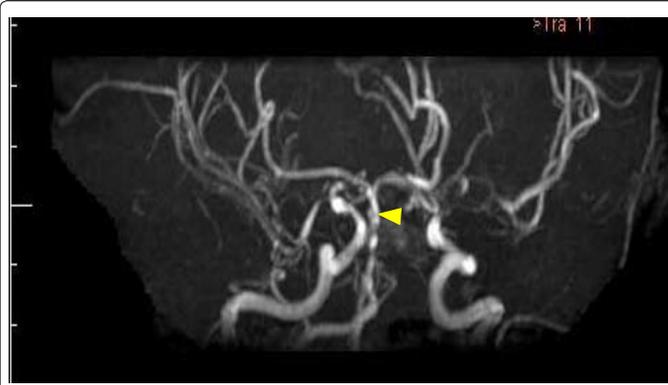
**Figure 2:** MRA 2 days after onset  
MRA performed 2 days after onset: occlusion of the bilateral proximal middle cerebral arteries (arrows).

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**Figure 3:** MRA 8 days after onset  
MRA performed 8 days after onset: an improvement of occlusion of the bilateral proximal middle cerebral arteries, and new, irregular, multiple narrowings of the bilateral internal carotid arteries (arrow).

## Discussion

Stroke related to pregnancy is associated with significant morbidity and mortality. The American Maternal Mortality Collaborative placed cerebrovascular disease as the fifth most common cause of maternal death during 1980-1985 [10]. Moreover, pregnancy and the postpartum period are associated with an increased risk of stroke and cerebral hemorrhage [2,9]. Evaluations of both the incidence and risk of stroke in pregnancy have varied markedly, and diverse inclusion criteria have been proposed as a result of population-based studies [8,11-13]. One study using data from 46 hospitals in the Baltimore-Washington DC area concluded that the risk of ischemic stroke and Intracerebral Hemorrhage (ICH) increased in the postpartum period, but not during pregnancy, with a relative risk of 8.7 and 28.3 for ICH [11]. Considering this study along with other population-based studies, the incidence of stroke is from 4 to 11 cases per 100 000 deliveries [8,11-13].

Non-contrast head CT is usually the first line in imaging because of its speed and wide-spread availability. Although safety considerations for fetus are an important concern, fetal radiation doses for head CT in which the fetus is not directly imaged are minimal and need not figure in the risk-benefit analysis to perform the examination. One of the most important advantages of MRI is machine parameters on the signal intensities. Digital subtraction angiography is widely accepted as the gold standard for evaluation of vascular pathology including vascular malformations, Moyamoya disease. However, poor delineation of hemorrhage is a disadvantage of MRI. Therefore, we use CT instead of MRI as the first examination.

Postpartum Cerebral Angiopathy (PCA) is one of several Reversible Vasoconstriction Syndromes (RCVS), characterized by reversible vasoconstriction of the cerebral arteries [2,4,5,14]. A retrospective and prospective study in France of 348,295 deliveries revealed 31 cases of stroke, and only 1 case was attributed to PCA [13]. Although the pathology of RCVS remains unknown, the prevailing hypothesis is of a transient disturbance in the control of cerebral vascular tone leading to segmental and multifocal arterial constriction and dilatation [4,5,8]. It is unclear whether vasospasm, vasculitis, or another mechanism or factor is the underlying cause of PCA, but the pathophysiology may be similar to the disease mechanism in eclampsia [4,8,14-16].

In pregnancy, two-thirds of PCA cases show an onset during the first week after delivery [5]. In 50-70% of cases, it is associated with the intake of vasoconstrictors, mostly ergot alkaloids, which are commonly used to treat postpartum hemorrhage [5,6]. Ducros et al. reported

that eclampsia/ preeclampsia, selective serotonin reuptake inhibitors, ergotamine tartrate, methergine, and bromocriptine are causes of RCVS [5]. In our case, an ergot alkaloid was administered to reduce postpartum hemorrhage; however, she had no previous history of collagen-vascular diseases, miscarriage, drug abuse, or bromocriptine treatment, which is indicative of an inflammatory process. The vasculitic process might cause cerebral ischemia.

Classically, PCA was considered to present thunderclap headache, vomiting, and an altered mental state with or without focal neurologic deficits within days of delivery. According to the current diagnostic criteria for RCVS proposed by the International Headache Society for "acute reversible cerebral angiopathy" and the criteria proposed in 2007 by Calabrese [5], it is very difficult to diagnose in the absence of headache. However, PCA without headache or low-level headache does exist [5]. Fugate et al. reported that two cases in 18 patients with PCA had no headache [3]. Our patient also did not present headache.

A diagnosis of PCA is made with angiography. It demonstrates multifocal segmental narrowing and dilatation (string of beads) in large and medium-sized cerebral arteries [2-8]. Ducros et al. reported that noninvasive angiography (MRA or CT angiography) was 80% sensitive in their series compared with the gold standard of catheter angiography [5]. Brain MRI may show areas of T2/FLAIR hyperintensity at any location, especially in watershed areas between vascular territories [8]. By definition, the process is generally self-limiting, with the resolution of angiographic abnormalities within 4-12 weeks and typically complete resolution of symptoms [2,5,14]. However, the first angiogram, including MRA and CT Angiography (CTA), regardless of the modality, may be normal if performed very early, within 4-5 days of the onset of symptoms; therefore, if the first MRA or CTA is normal, a second angiogram a few days later may be diagnostic [3,5].

In our case, she was diagnosed as PCA, without headache on the basis of the following: 1) cerebral angiography, including MRA, showed multi-segmental areas of narrowing and dilatation of arteries supplying the hemisphere, 2) exclusion of systemic inflammatory disorders such as infectious vasculitis and collagen-vascular diseases, and 3) resolution of both MRA abnormalities and paresis in only a few months.

Cerebral arterial vasoconstriction can be seen in the postpartum period. The obstetric neurologic literature has described a similar entity, given various labels [5,6,8,14], including PCA, postpartum angiopathy, postpartum angitis, and puerperal vasospasm. This syndrome is probably still underdiagnosed or goes unnoticed, in cases with the pure cephalalgic or paralytic forms. Therefore, it is important for obstetricians to recognize this particular disorder. We should suspect RCVS in patients with thunderclap headache, with or without other neurologic deficits. However, we should be also aware that PCA could occur only with neurologic deficits, differing from RCVS in non-pregnant cases.

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