

Postpartum Angiopathy Associated with Reversible Borderzone Ischemia

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Abstract

A 32-year-old woman developed a headache, seizures, and stupor on postpartum day 8. An initial diagnosis of possible encephalitis was made considering the presence of fever, neck stiffness, and abnormal CSF findings. MRI demonstrated hyperintense signals consistent with bilateral borderzone areas. MRA showed severe proximal narrowing of anterior, middle, and posterior cerebral arteries bilaterally. The patient recovered completely over 2 weeks, and repeated MRI and MRA scans were normal. Reversible vasoconstrictions have been known to occur during puerperium, and the clinical symptoms of our patient resembled such cases. Prior reports attributed these cases of postpartum angiopathy to capillary leakage and edema resulting in leukoencephalopathy. Our case suggests reversible borderzone ischemia as an additional pathological process.

Key words: leukoencephalopathy, eclampsia, vasoconstriction

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Introduction

Reversible vasoconstriction syndrome has been associated with migraines, vasoactive drug use, hypertensive encephalopathy, eclampsia, immunosuppressive drug therapy, or organ transplantation (1). Several cases have been reported in which the patient developed postpartum reversible vasoconstriction, which is now known as “postpartum angiopathy” (2).

Postpartum angiopathy is characterized by sudden onset headache, confusion, seizures, and focal neurological deficits after delivery. Patients generally display edematous lesions mainly distributed in the bilateral parieto-occipital white matter. However, abnormal cerebrospinal fluid (CSF) findings have not been reported before. Here, we report a patient with postpartum angiopathy associated with severe proximal narrowing of the major cerebral arteries, and reversible borderzone ischemia.

Case Report

A 32 year-old woman had an uncomplicated pregnancy and delivered her first baby at 40 weeks of gestation. She had no history of migraines. Furthermore, no signs of

edema, proteinuria, hypertension, or neurological complaints had been observed during the pregnancy. Her antepartum and intrapartum blood pressure ranged from 120/70 to 130/80 mmHg, and she was normotensive at least to postpartum day 6.

On postpartum day 4, she developed a severe throbbing headache, which continued but improved with analgesics. On postpartum day 8, her headache worsened upon awakening. Two hours later, she found herself unable to write, wash clothes and subsequently panicked at her inability to remember how to do simple activities of daily living. In fact, her husband found her at home crying next to the door, unable to unlock it from the inside to let him in. That night, the patient experienced a generalized tonic clonic seizure, and was transferred to our hospital immediately.

On arrival, she was confused, disorientated and somnolent. Her speech was slow and difficult to understand; however, she was able to express complaints of visual hallucinations consisting of red ants or green balls. Her blood pressure was 176/122 mmHg, and body temperature was 37.9°C. A general physical examination revealed normal results, and she did not have any peripheral edema or changes in the retinal arterioles and optic discs. A general physical examination was significant only for mild nuchal rigidity. There were no signs of peripheral edema or fundoscopic changes.

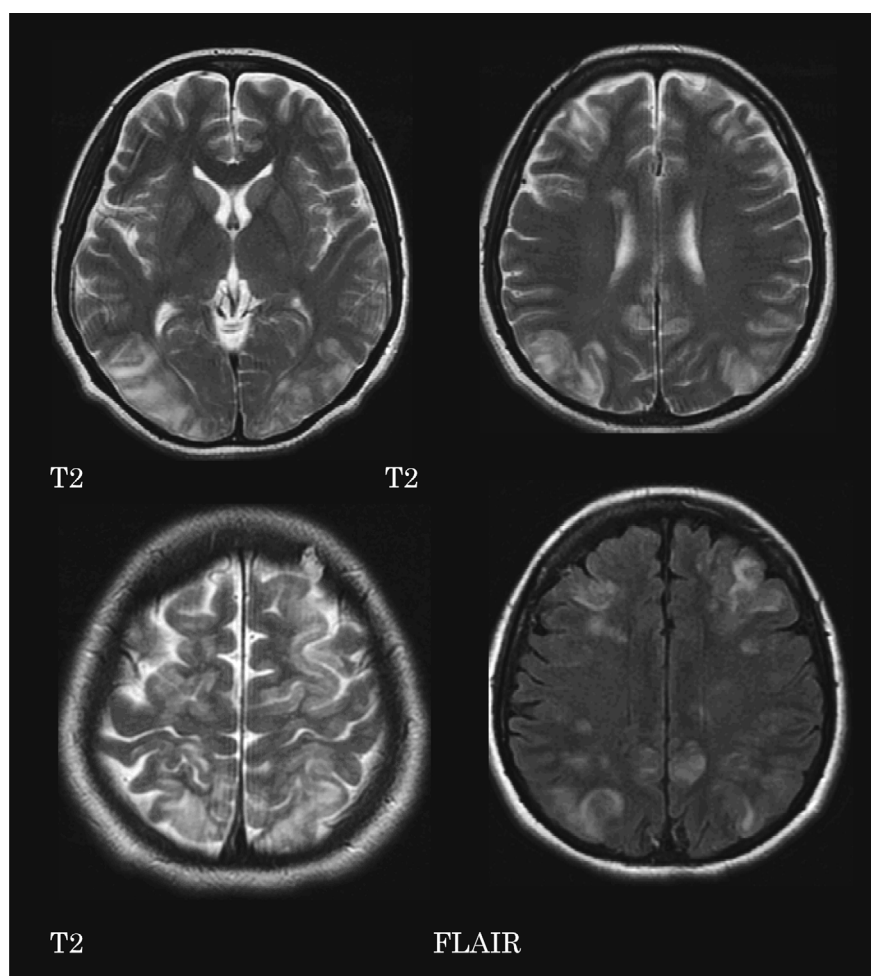


Figure 1. Brain imaging on arrival. T2-weighted MRI demonstrated hyperintense lesions in the bilateral parieto-occipital lobes and the frontal cortices and subcortices. Diffusion-weighted images (DW) showed hyperintensities in the cortices consistent with borderzone arterial territories.

Strength, sensation and deep tendon reflexes were all intact. The patient had two more generalized tonic clonic seizures in our emergency room.

Laboratory results revealed leukocytosis of 17,100/mm³, and a normal platelet count. Serum chemistry showed an elevation of lactic dehydrogenase (LDH, 374 IU/L), creatinine kinase (CK, 1,488 IU/L), and C-reactive protein (CRP, 2.71 mg/dl). Her urinalysis revealed proteinuria (>3+). Electrolytes, liver and renal function test, rheumatoid factor, antinuclear antibodies were all normal. Echocardiogram and electroencephalography were both unremarkable. Lumbar puncture showed clear colorless cerebrospinal fluid (CSF) with an opening pressure of 16 cm H₂O. CSF contents showed protein elevated to 128 mg/dl, glucose normal at 77 mg/dl, and a leukocyte count of 1/μL. Antiviral antibodies or herpes simplex virus DNA in the CSF were negative.

A head CT scan revealed hypodense lesions in both occipital lobes. An MRI demonstrated hyperintense lesions in the bilateral parieto-occipital lobes and the frontal cortices and subcortices (Fig. 1). Diffusion-weighted images showed hyperintensities in the cortices consistent with borderzone arterial territories (Fig. 1). The apparent diffusion coefficient

(ADC) in the lesions showed no obvious changes.

An initial diagnosis of possible encephalitis was made considering the presence of fever, neck stiffness, and abnormal CSF findings. She was treated with acyclovir, glycerol, phenytoin and analgesics. On the following day, the patient's blood pressure fell to 110/72 mmHg without antihypertensive drugs, and she had no more seizures but still suffered from confusion and headaches. MR angiography (MRA) performed 4 days later showed severe proximal narrowing of anterior, middle, and posterior cerebral arteries bilaterally (Fig. 2).

The remainder of her hospital course was uneventful, and the proteinuria noted to be 3+ on admission decreased to 1+ upon discharge. A repeat CSF study performed 1 week after admission was normal. Her headache and the clinical deficits completely disappeared over the following 2 weeks. One month later, a follow up MRI (Fig. 3) and MRA (Fig. 4) were normal.

Discussion

Reversible vasoconstriction syndrome was first angiographically demonstrated in 1988 by Call and Fleming (2).

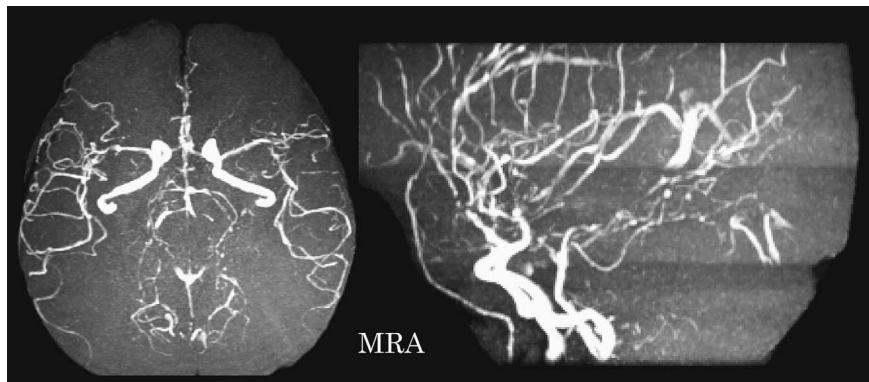


Figure 2. MR angiography (MRA) performed 4 days later. Severe proximal narrowing of anterior cerebral arteries (ACA), middle cerebral arteries (MCA), and posterior cerebral arteries (PCA) is seen bilaterally.

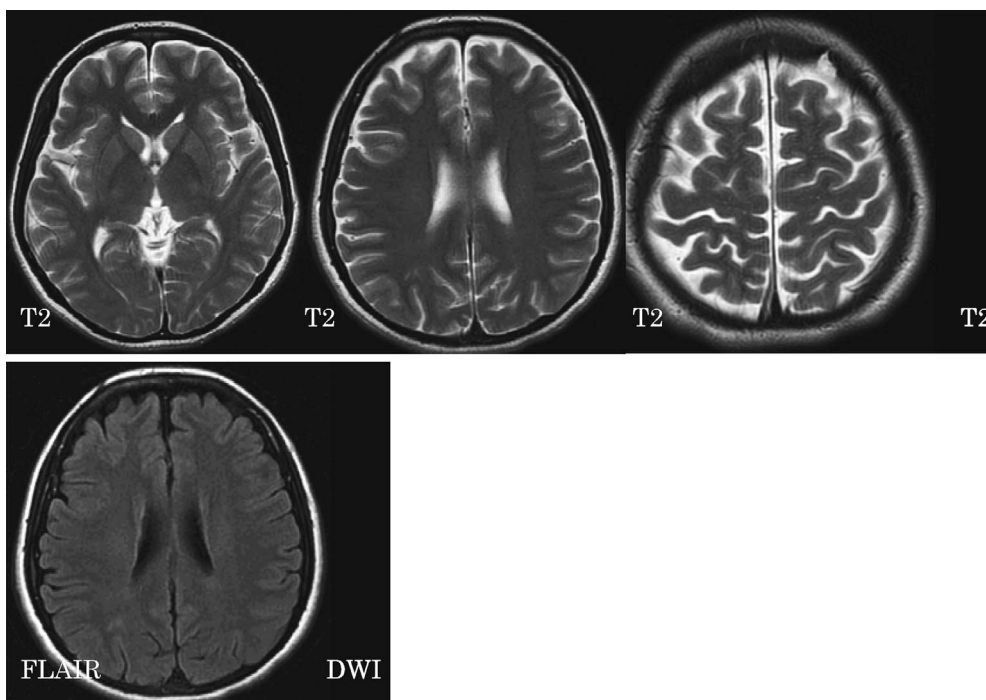


Figure 3. Follow-up MRI and MRA at 1 month. All images were normal.

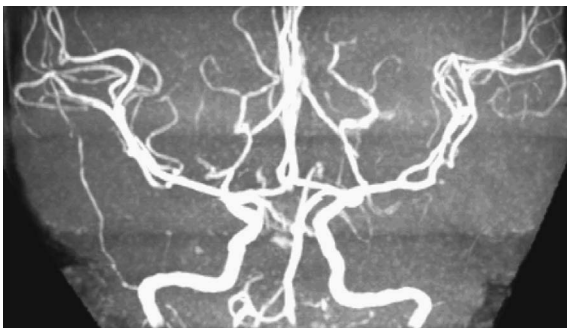


Figure 4. A follow up MRA at 1 month. All follow-up images were normal.

Their paper described transient, fully reversible vasoconstrictions and dilatations predominantly involving arteries around

the circle of Willis in four patients with severe headaches and motor and sensory deficits. Reversible vasoconstriction syndrome has subsequently been found to be associated with conditions like migraine, vasoactive drug use, pregnancy and the puerperium (1).

The symptoms of reversible vasoconstriction syndrome are quite similar to that of eclampsia, a disorder defined by the development of convulsion and/or coma during pregnancy and the postpartum period in severely hypertensive women (3). The presence of generalized edema, hypertension, proteinuria, and convulsions are well known as the signs of pending eclampsia. However, women who develop eclampsia may exhibit a much wider spectrum of signs with no proteinuria, no edema, and only minimal hypertension (3). The angiographic changes during eclampsia are yet to be fully characterized.

Patients with postpartum angiopathy (PPA) are reported to develop symptoms similar to reversible vasoconstriction syndrome, such as severe headaches with an abrupt onset, seizures, or focal neurological deficits. While eclampsia is often thought to occur within 48 hours of delivery, the time frame for PPA is much longer, with onset up to two weeks after delivery (1). The neurological symptoms of PPA similarly remit within days to weeks. The symptoms of the present patient resembled such cases.

Prior reports attributed the case of postpartum angiopathy to capillary leakage and edema resulting in leukoencephalopathy (4). However, in the present case, MRI demonstrated hyperintense lesions in the cortices and the subcortices, which were consistent with borderzone ischemia. Consider-

ing these findings, we speculate that proximal artery vasospasm and subsequent endothelial injury to small vessels of the cortices due to borderzone ischemia caused leakage of fluid across a transiently abnormal blood-brain barrier, and thus resulted in elevated protein levels in the CSF. We are unable to explain the precise mechanism why our case is different from usual cases of PPA (leukoencephalopathy and capillary leak).

It is possible that all of these disorders represent different manifestations of similar underlying processes. Our patient had aspects of reversible vasoconstriction syndrome, eclampsia, and PPA. The proximal artery vasospasm may cause borderzone ischemia by a hemodynamic mechanism.

References

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