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Case Report

CASE REPORT

A headache-free reversible cerebral vasoconstriction syndrome (RCVS) with symptomatic brain stem ischemia at late pregnancy as a rare manifestation of RCVS resolved with termination of pregnancy by semi-urgent cesarean section

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Abstract

A 32-year-old pregnant woman in her 39th week of pregnancy presented at the emergency room complaining of sudden-onset dizziness with gaze disturbance and was admitted to our hospital. Her past medical history included hypertension, diabetes mellitus and infarction in the right medulla oblongata 18 months prior to this event. Magnetic resonance (MR) angiography showed multiple irregular stenosis of the intracranial arterial system. Although MR images revealed no fresh ischemic or hemorrhagic lesions, she was diagnosed with reversible cerebral vasoconstriction syndrome (RVCS) associated with pregnancy. Cesarean section immediately resolved the headache-free ischemic RCVS. The postpartum course of the patient was uneventful as well as that of her baby. Follow-up MR angiography showed improvement of intracranial vasoconstriction and follow-up MR imaging showed improvement of a left medial pontine ischemic lesion on diffusion-weighted image. This report describes a rare manifestation of pregnancy-related RCVS.

INTRODUCTION

This report describes a case of pregnant-related reversible cerebral vasoconstriction syndrome (RCVS), as a variant of reversible cerebral vasoconstriction syndrome (RCVS), which resolved immediately after semi-urgent cesarean section (C-section) as a means of termination of pregnancy despite a fine ischemic

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lesion in the left medial pons. To the best of our knowledge, this is the first description of brain stem ischemia/infarction caused by pregnancy-relating reversible vasoconstriction of the central nervous system (CNS). Many cases of RCVS have been published [1–5] since Calabrese and colleagues proposed the name RCVS and a set of diagnostic criteria to regroup all similar cases that had been reported in the 1970s. Subsequently, RCVS was published under several other names such as 'isolated benign cerebral vasculitis' [6, 7], 'acute benign cerebral angiopathy' [8], 'reversible cerebral segmental vasoconstriction' [9, 10], 'Call or Call-Fleming syndrome' [10], 'CNS pseudovasculitis' [11], 'benign angiopathy of the CNS' [12, 13], 'postpartum angiopathy' [14], 'migraine angiitis' [15], 'migrainous vasospasm' [7], 'primary thunderclap headache' [16], 'cerebral vasculopathy' [17, 18] and 'vasospasm in fatal migrainous infarction' [19]. Among them, headache-free RCVS, which causes brain stem ischemia like our case, has not been reported. Through this first case presentation of pregnancy-related headache-free RCVS with symptomatic brain stem ischemia, as a rare pattern of RCVS, we provide clinicians with clues for clinical decision making for progressive brain stem ischemia in late pregnancy.

CASE REPORT

A 32-year-old pregnant woman in the 39th week of her first pregnancy presented at the emergency room and was admitted to our hospital complaining of dizziness with gaze disturbance. She had a medical history of hypertension, diabetes mellitus (DM) and infarction in the right medulla oblongata 18 months prior to this event (Fig. 1). When she suffered from the medullary infarction, she was diagnosed with DM and hypertension (HT). Diabetes nutrition therapy was recommended for her. Since then, she has taken antihypertensives (amlodipine besylate 2.5 mg/day, telmisartan 20 mg/day), statin (rosuvastatin calcium 2.5 mg/day) and antiplatelets (cilostazol 200 mg/day or aspirin 100 mg/day). After her pregnancy was revealed, insulin therapy was introduced to control her plasma glucose more appropriately. Antiplatelets were continued until the 28th week of pregnancy. On arrival, she was in the 39th week of pregnancy. The temperature was 37.2°C, the pulse was 80 beats/min, and the respirations were 20/min. The blood pressure (BP) was 170/92 mmHg. The pupils were 3 mm in diameter and briskly reactive to light. The neck was supple. She denied having a headache or neck pain. She complained of diplopia on the right lateral gaze. Fine horizontal nystagmus on right gaze made apparent by looking to the right was observed in both eyes. She was mostly closing her eyes and did not sufficiently gaze because of vertigo. Movements of her extremities were intact and not ataxic. The remainder of the physical examination was normal. The hematocrit, hemoglobin level, platelet count, and results of liver- and renal-function tests were normal or nearly normal, as were blood levels of total protein, albumin, globulin and calcium. After admission, she suffered from progressive vertigo with horizontal nystagmus gaze and could not keep her eyes open. Magnetic resonance (MR) angiography showed multiple irregular stenosis of the intracranial arterial system (Fig. 2). Although diffusion-weighted images (DWIs) revealed no fresh ischemic lesions, her brain stem symptoms, such as vertigo, nystagmus and gaze disturbance, were aggravated. Although her BP was controlled under 160 mmHg with continuous intravenous nicardipine, BP control gradually became difficult and required more dose of intravenous nicardipine, indicating progression of brain stem ischemia and reactive HT. Movements of her extremities were intact and not ataxic at that time. She did not complain of headaches during the late pregnancy. We diagnosed her as suffering from RCVS associated with pregnancy of the latter period, with pre-eclampsia and decided to terminate the pregnancy with semi-urgent C-section for curbing further brain stem ischemia to infarction. The C-section under lumbar anesthesia at 39 weeks of pregnancy immediately resolved the brain stem ischemic symptomized by diplopia, vertigo and gaze disturbance. The newborn's birth weight was 3197 g with Apgar scores of 8/9 (1 min/5 min). On the third postoperative day, fluid attenuation inversion recovery and DWI showed left medial pontine ischemia (Fig. 3). Heparin was continued until the third postoperative day. Heparin was substituted for peroral aspirin (100 mg/day). Her postoperative clinical course was uneventful as well as that of her baby. Follow-up MR angiography (Fig. 2) showed improvement of intracranial vasoconstriction despite a fine ischemic scar in the left medial pons on MR imaging (Fig. 3D, upper). She and her baby left our hospital without sequelae.

DISCUSSION

Despite recent available reviews of RCVS [20–28] or posterior reversible encephalopathy syndrome during pregnancy and

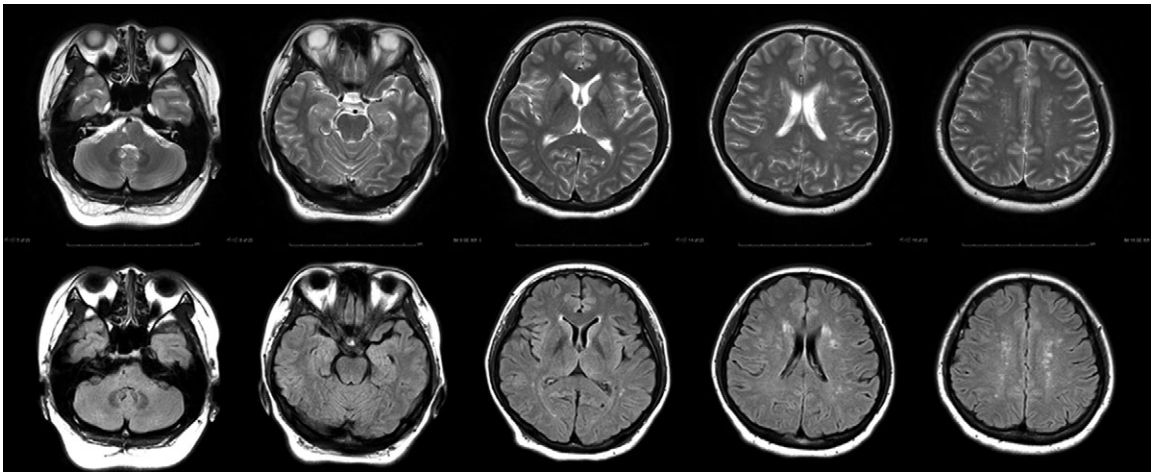

Figure 1: Pre-pregnant axial sections of T2-weighted image (upper row) and fluid attenuation inversion recovery (lower row) show a small infarction scar in the right medulla oblongata and mild leukoaraiosis in the cerebral white matter.

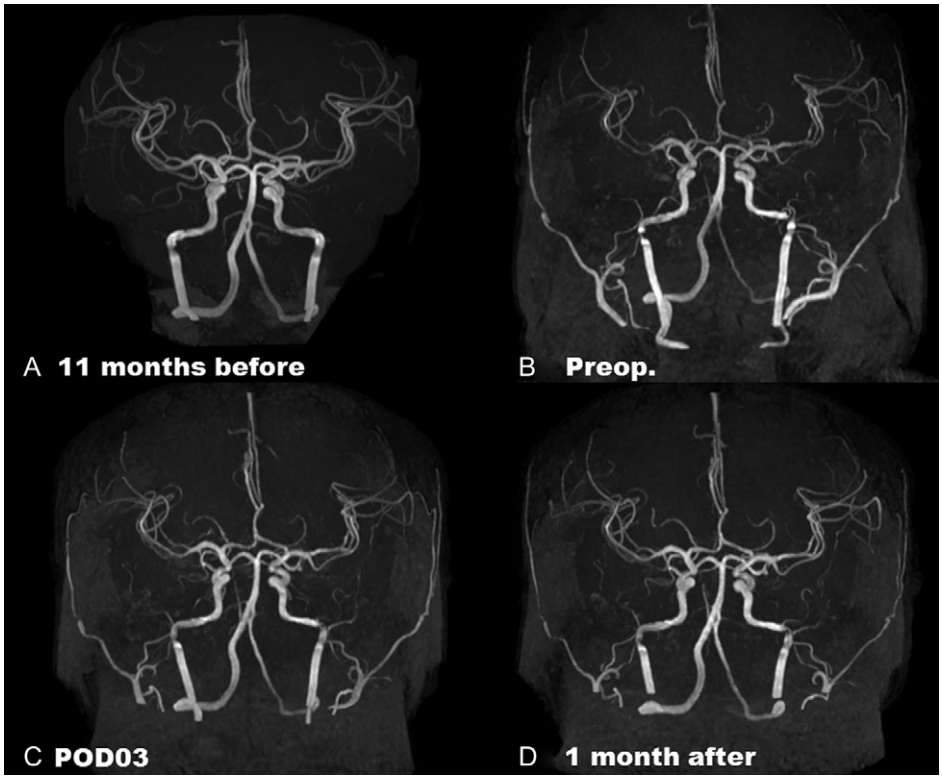

Figure 2: In comparison with the examination 11 months before admission (**A**), magnetic resonance angiographies show emergence (**B**) and disappearance (**C** and **D**) of multiple stenotic changes especially in the middle cerebral arteries and in the left vertebral artery.

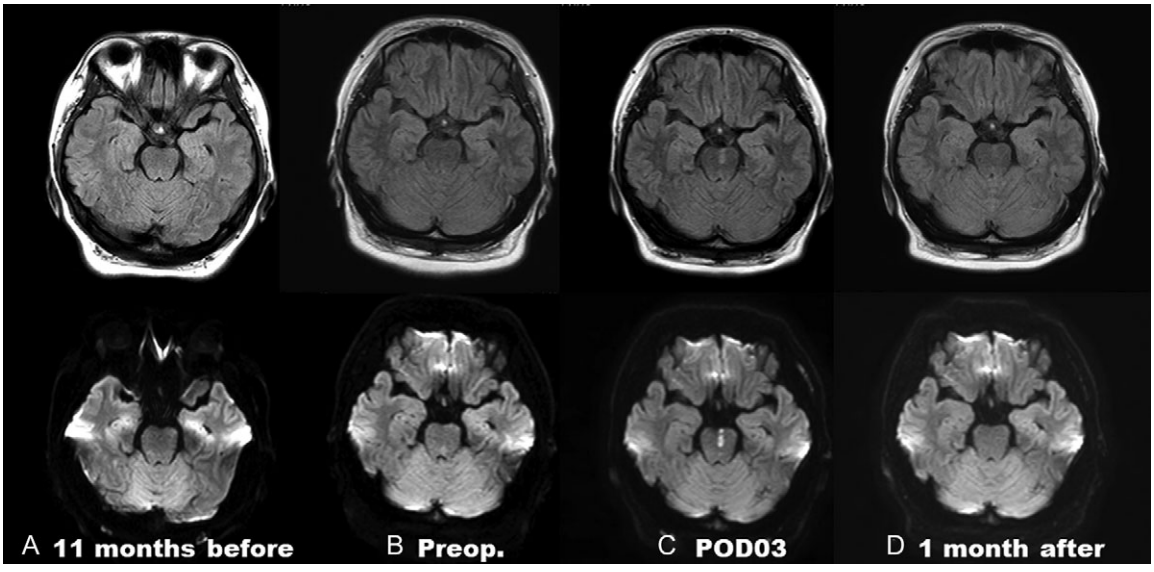

Figure 3: Serial magnetic resonance images (**A–D**) show emergence (**B** and **C**) and disappearance (**C** and **D**) of left medial pontine ischemic lesion. Upper: FLAIR, lower: DWI.

postpartum [29], no case reports of brain stem ischemia due to RCVS are available as of early 2018.

To the best of our knowledge, this report describes the first case of symptoms of pregnant-related RCVS with symptomatic brain stem ischemia resolved immediately after semi-urgent C-section, which indicates that semi-urgent C-section can improve RCVS with symptomatic brain stem ischemia in late pregnancy.

Stroke is a rare complication of the peripartum period, with low incidence (34.2 per 100 000 deliveries [30, 31]) but potentially devastating consequences. Previous studies estimated that peripartum women are at a 3-fold increased risk of stroke

[31–35], with the postpartum period conferring the highest risk of both ischemic and hemorrhagic stroke [35, 36].

The estimated incidence of pregnancy-associated stroke was 10.2 per 100 000 deliveries in Japan [37]. Hemorrhagic strokes were observed in 73.5% of the cases, ischemic strokes in 24.5%, and mixed type in 2.0% [37]. Among the ischemic strokes, 75.7% were arterial and 24.3% were venous infarctions. The most frequent cause of arterial infarctions was RCVS [38].

In healthy adults, global CBF is maintained at ~50 mL/100 g brain tissue per minute at cerebral perfusion pressures (CPP) between ~60 and 160 mmHg [39, 40]. In settings of acute hypertension when arterial pressure may rise above the cerebral blood flow (CBF) autoregulatory range, such as in some cases of pre-eclampsia, the increased intravascular pressures can overcome the myogenic vasoconstriction of arteries and arterioles, causing them to lose their ability to provide vascular resistance [40–43]. The resulting loss of autoregulation and hyperperfusion can lead to endothelial damage, edema and risk of brain injury [42–46]. In our case, hyperperfusion symptoms, such as white matter lesions or cognitive changes, were not observed clinically and radiologically. MR angiography was useful to diagnose the RCVS. Although hypertensive encephalopathy and cerebral infarction have been reported in a few individual case reports [47], brain stem ischemia/infarction has not been described clinically and/or radiologically to the best of our knowledge.

Taking past medical history of the patient into consideration, her pontine ischemia/infarction is due to not only RCVS but also cerebrovascular risk factors, such as hypertension, DM and the previous medullar infarction. Although her brain stem ischemic symptoms were transient, she clearly had a small pontine infarction.

In this case, we attribute the pontine infarction to headache-free RCVS. RCVS in our case may overlap pre-eclampsia in a broad sense. they are not mutually exclusive and can occur together. The most frequent mode of delivery for women suffering from pre-eclampsia is elective C-section; however, neonates delivered by emergency C-section did not show an adverse neonatal outcome [48]. Through our practical experience, timely C-section is an effective and safe treatment of RCVS with progressive ischemic stroke at late pregnancy.

Clinicians should keep in mind the possibility of RCVS if they encounter sudden-onset brain stem symptoms in a pregnant woman despite lack of headaches, because early intervention can lead to favorable outcome in RCVS during pregnancy.

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CONFLICT OF INTEREST STATEMENT

The author declared no potential conflicts of interest with respect to the research, authorship and/or publication of this article.

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ETHICAL APPROVAL

Informed consent was obtained from the patient after discharge, in order to anonymously use the medical data for scientific purpose and all information reported generally without mentioning patient's name.

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