

Moyamoya-disease-related ischemic stroke in the postpartum period

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Abstract

Stroke during pregnancy or the puerperium is an extremely rare yet serious cause of perinatal morbidity and mortality. Moyamoya disease, a cerebrovascular occlusive pathology with a female predominance, may become symptomatic for the first time in association with pregnancy. A 36-year-old woman with postpartum pre-eclampsia suddenly developed hemiparesis in the left arm with dysarthria after the initiation of antihypertensive measures. Cranial magnetic resonance imaging and angiography revealed acute ischemic lesions in the right hemisphere along with the steno-occlusive lesions of bilateral terminal portions of the internal carotid artery, indicating Moyamoya disease. With anti-platelet medication, the patient recovered gradually and was diagnosed as having Moyamoya disease using conventional angiography 3 months postpartum. In this case, the fluctuations in blood pressure in association with pre-eclampsia appear to have exacerbated the clinical symptoms of Moyamoya disease. As such, Moyamoya disease should be considered as an underlying disease of ischemic stroke associated with pregnancy.

Key words: ischemic stroke, Moyamoya disease, pre-eclampsia, pregnancy.

Introduction

Moyamoya disease is a rare cerebrovascular occlusive disease with a female predominance. Because of fluctuations in blood pressure associated with pregnancy, Moyamoya disease may become symptomatic for the first time during pregnancy and the puerperium.¹ Intracranial hemorrhage of sudden onset predominates in patients diagnosed with Moyamoya disease for the first time in association with pregnancy. To the best of our knowledge, there are few case reports regarding the diagnosis of Moyamoya disease following an ischemic attack during pregnancy and the puerperium.^{2–4} We herein present a case of postpartum ischemic stroke in a pre-eclamptic woman, who was subsequently diagnosed as having Moyamoya disease.

Case Report

A 36-year-old second parous Japanese woman presented at 31 weeks' gestation with complaints of decreased fetal movement, epigastric pain with nausea and edema for 2 days. Mild elevated blood pressures of 140–150/70–80 mmHg were apparent after attending our prenatal clinic. Upon arrival, her blood pressure was 180–190/100–110 mmHg with proteinuria of 100 mg/dl and the ultrasound evaluation revealed an appropriate-for-date fetus without movement. Because of the non-reassuring fetal heart rate pattern and severe pre-eclampsia, emergency cesarean was performed and placental abruption was found. A depressed female baby weighing 1436 g was resuscitated and treated at the neonatal intensive care unit. The woman's

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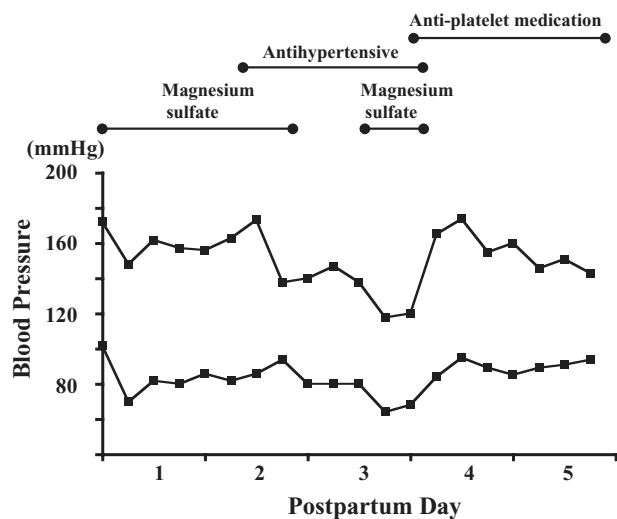


Figure 1 Changes in blood pressure during the postpartum period. On postpartum day 3, blood pressure was controlled, however, hemiparesis in the left arm with dysarthria developed.

medical history was unremarkable and her previous pregnancy was uncomplicated except for cesarean delivery because of cephalopelvic disproportion.

Postpartum, the patient received 48 h of intravenous magnesium sulfate as seizure prophylaxis (Fig. 1). The immediate postoperative course was uneventful with blood pressures of 150–160/70–80 mmHg. On postpartum day 2, her blood pressure again elevated to 170–180/90–100 mmHg and antihypertensive treatment using long-acting nifedipine was initiated. The following day, her blood pressure was controlled, ranging from 120–140/60–80 mmHg. However, hemiparesis in the left arm with dysarthria suddenly developed and magnesium sulfate was restarted for possible seizure. Intracranial hemorrhage was not found by immediate computed tomography. Cranial magnetic resonance imaging (MRI) illustrated acute ischemic lesions in the right hemisphere (Fig. 2). Cerebral edema suggesting posterior reversible encephalopathy syndrome was not demonstrated on MRI. Additionally, magnetic resonance angiography (MRA) revealed the stenocclusive lesions of bilateral terminal portions of the internal carotid artery, indicating Moyamoya disease (Fig. 3a). Magnesium sulfate and an antihypertensive were discontinued, and anti-platelet medication with aspirin was given. On postpartum day 4, her blood pressure showed a transient elevation to 170–180/90–100 mmHg but she manifested no additional neurological symptoms. Thereafter, her blood pressure

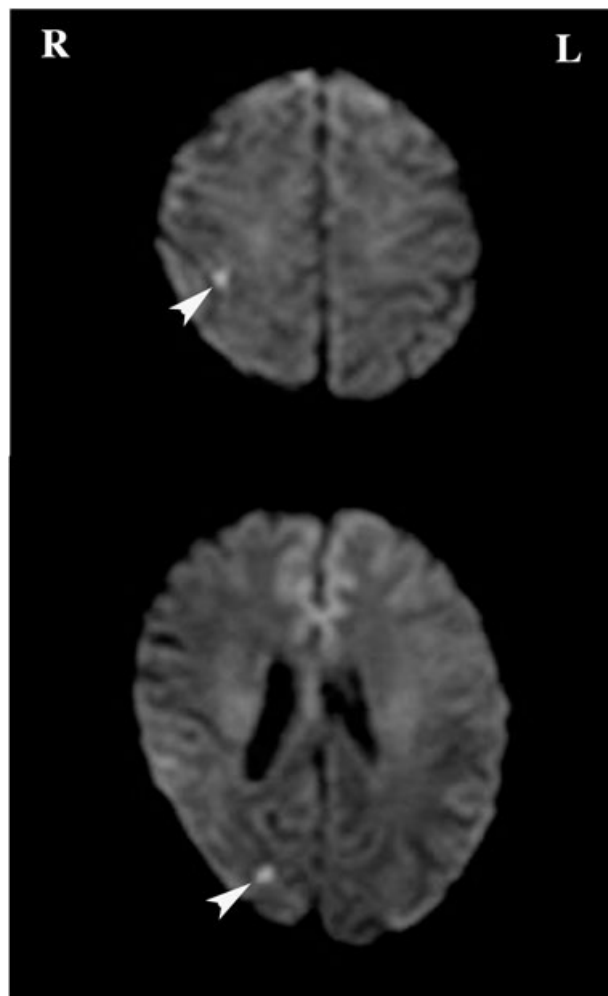


Figure 2 Brain axial diffusion-weighted magnetic resonance image showing acute ischemic lesions in the right hemisphere (arrowheads). R, right; L, left.

lowered gradually and remained mildly elevated but stable at 140–150/80–90 mmHg without antihypertensive measures (Fig. 1). She recovered gradually and the neurological status was normal 5 days after the onset of symptoms. Three months postpartum, Moyamoya disease was confirmed by conventional angiography (Fig. 3b,c).

Discussion

Moyamoya disease is characterized by stenosis or occlusion of the terminal portion of internal carotid arteries, along with abnormal capillary networks, known as Moyamoya vessels.⁵ Although a variety of genetic, environmental, and infectious causes have

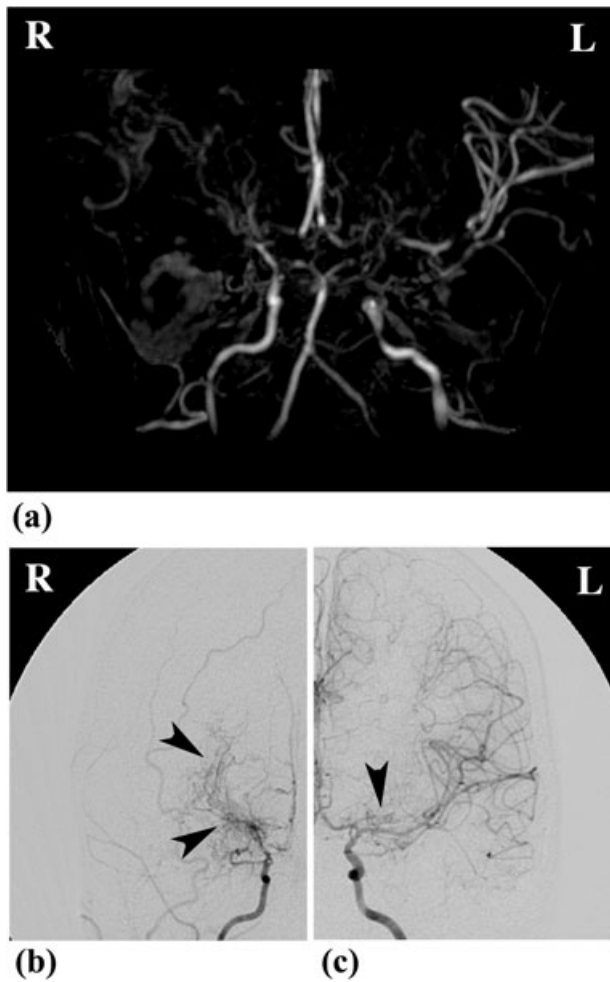


Figure 3 Radiological findings of stenotic lesions of the terminal portion of bilateral internal carotid arteries and Moyamoya vessels. (a) Antero-posterior view of magnetic resonance angiogram. Frontal angiogram of right common (b) and left internal (c) carotid artery. Note that the distal internal carotid artery is occluded and the middle cerebral artery is not visualized in the right hemisphere. The left distal internal carotid artery is seen as stenotic. Abnormal capillary networks known as Moyamoya vessels are clearly demonstrated by conventional angiography (arrowheads). R, right; L, left.

been postulated, the etiology of Moyamoya disease remains unclear.^{4,5} The diagnosis of Moyamoya disease is made based on angiographic features from MRI and MRA as well as conventional angiography.⁶ Its occurrence has been reported worldwide, although it was originally reported in Japan. Among the Japanese, it depicts a bimodal age distribution with a higher peak in 10–14 year olds and a lower peak in 30–40 year olds.⁷

Common manifestations in Japanese patients are transient ischemic attacks in children and cerebral hemorrhage in adults. Of interest, epidemiological studies from the USA have shown that ischemic attack is the predominant initial symptom in both adults and children, indicating racial differences in its clinical presentation.⁸

As the diagnosis of Moyamoya disease is based on its angiographic features, case detection may hinge on the performance of angiography. In this regard, the use of conventional angiography in the evaluation of ischemic stroke varies widely between individual health care providers. In our case, MRI revealed acute ischemic lesions without cerebral edema in the right hemisphere. The patient had no history of a cardioembolic source (i.e. cardiac structural abnormalities or arrhythmias). Although pre-eclampsia may be associated with the development of cerebral ischemia, the MRI results warranted further evaluation of underlying cerebrovascular disease, and then the use of MRA revealed vascular lesions indicative of Moyamoya disease.

Our patient showed ischemic symptoms when her blood pressure was lowest after the initiation of anti-hypertensive measures. Hemodynamic change during the early puerperium may place undue stress on the cerebral vessels.¹ Komiyama *et al.* reported that the development of pre-eclampsia may cause deterioration of clinical symptoms of Moyamoya disease.¹ Pre-eclampsia, a pregnancy-specific multi-organ disorder, is characterized by endothelial dysfunction.⁹ The development of neurological disorders in pre-eclampsia is thought to be related to vasoconstriction associated with endothelial damage, as well as impaired autoregulation of the cerebral vessels.⁹ Therefore, fluctuation of blood pressure and cerebral pathologies associated with pre-eclampsia could make patients with Moyamoya disease become symptomatic of this disease. In contrast to the previous pregnancy, our patient developed severe pre-eclampsia in the index pregnancy. Ischemic lesions were found in the right hemisphere which showed marked vascular changes associated with Moyamoya disease. The combination of acute reduction in blood pressure with an antihypertensive, pre-eclampsia, and underlying Moyamoya disease appeared to be associated with ischemic stroke in this case.

Stroke during pregnancy or the puerperium is an extremely rare yet serious cause of perinatal morbidity and mortality.¹⁰ Hypertension and ruptured cerebral aneurysm are considered as major causes of cerebral

stroke during pregnancy. Although Moyamoya disease is a rare cerebrovascular disorder, it should be considered as an underlying disease of ischemic stroke associated with pregnancy.

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