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Case Report Open Access

Two Patients with Moyamoya Disease Underwent Emergency Cesarean Delivery Resulted in Opposite Outcomes

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Abstract

Moyamoya disease is a rare cerebrovascular disorder, characterized by bilateral stenosis or occlusion of arteries at the base of the brain and formation of an abnormal network of collateral vessels, called moyamoya vessels, associated with cerebral hemorrhage and infarction. No appropriate method of delivery, anesthesia, and perioperative management of pregnant patients with moyamoya disease has yet been established. We report 2 cases of pregnant patients with moyamoya disease who underwent emergency cesarean delivery under general anesthesia. The postoperative courses were different, which might help us establish a protocol for the standard management of this disease. Case 1: A 33-year-old primipara was diagnosed as having moyamoya disease when she was 4-year-old. She underwent emergency cesarean delivery under general anesthesia at 37 weeks' gestation due to early rupture of the membranes. The perioperative period was uneventful. The patient was discharged without aggravation of neurological symptoms. Case 2: A 35-year-old multipara with no medical history experienced sudden nausea and headache disturbance at 30 weeks' gestation. Brain CT showed intracranial hemorrhage, and the patient was diagnosed moyamoya disease by cerebral angiography. The patient recovered after intraventricular drainage but lost consciousness at 32 weeks' gestation due to repeated hemorrhage. Cesarean delivery, aneurysm excision, direct hematoma evacuation, and external decompression were performed shortly thereafter under general anesthesia. The intracranial pressure continued to increase, and the patient died on the ninth postoperative day. In pregnant patients with moyamoya disease, careful management during the perinatal period is needed to decide the timing of emergency cesarean delivery, which affects prognosis.

Keywords: Moyamoya disease; Pregnancy; Emergency cesarean delivery

Introduction

Occlusive disease of the circle of Willis, known as moyamoya disease, is a relatively rare progressive cerebrovascular disorder, characterized by blocked arteries at the base of the brain and formation of hazy collateral vessels, called moyamoya vessels [1,2]. It is prevalent in Asian countries, especially in Japan [1,3], with 0.4–0.6 cases per 100,000 population [4,5], and it is more prevalent in women than in men. Therefore, we should establish a protocol for the management or anesthesia of pregnant patients with moyamoya disease who undergo cesarean delivery. However, no protocol for the standard management and anesthesia of emergency cesarean delivery in patients with moyamoya disease has been established yet, and moreover, the pathology varies according to each patient. Here, we report the case of 2 pregnant patients with moyamoya disease who underwent emergency cesarean delivery under general anesthesia. The postoperative courses were different, which might be helpful for the future management of pregnant patients with moyamoya disease.

Patient (case 1) or patient's family (Case 2) consents have been obtained.

Case Report

Case 1

A 33-year-old primipara, weighing 67 kg, underwent cesarean delivery at 38 weeks' gestation. The patient was diagnosed as having moyamoya disease when she was 4 years old. She had undergone surgery 6 times, including revascularization. When she was 19 years old, a cerebral aneurysm and subcortical infarct in the frontal lobe was detected. Dysarthria and weakness of the left upper and right lower extremities was noted, and she had been taking 200 mg/day ticlopidine, 15 mg/day clotiazepam, 0.6 mg/day clonazepam, and 15 mg/day nicergoline. The gestation period was uneventful, but at 37 weeks' gestation, the patient had diarrhea and vomiting and was transported to a nearby hospital. Vital signs were normal, and diarrhea and nausea disappeared upon arrival at the hospital. The gynecological examination revealed no abnormal findings, but analysis of the amniotic fluid indicated early rupture of the membranes. The patient was closely monitored overnight, and on the next day, cesarean delivery was performed under general anesthesia. No aggravation of neurological symptoms was noticed, and the fetal heart rate (HR), which was assessed by cardiotocography (CTG), was normal.

The patient was monitored electrocardiogram (ECG), non-invasive blood pressure (NIBP), pulse oximeter oxygen saturation (SpO_2), end-tidal CO_2 ($EtCO_2$), and bispectral index (BIS) during the operation. The initial blood pressure in the operation room was 112/64 mmHg, and other vital signs were stable. Oxygen, 200 mg thiamylal, 200μg fentanyl, and 100 mg suxamethonium were given following administration of 0.5 mg vecuronium for precurarization, and a tracheal tube was inserted. During induction and intubation, no significant alterations in hemodynamics were noted. Anesthesia was maintained with 0.5%

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sevoflurane until delivery and with target-controlled infusion (TCI) of propofol at a rate to achieve a concentration at the target site of 3μg/ml after delivery. Fentanyl (50μg) was administered when needed. During the operation, the ventilation setting was strictly adjusted to keep $EtCO_2$ at 38–40 mmHg. An arterial blood sample was taken once, and its analysis showed that pH was 7.334 and $PaCO_2$ was 41.5 mmHg. The patient was extubated shortly after the operation and discharged from the operation room (Figure 1). The operation time was 52 min, and anesthesia time was 1 h 25 min. Sufficient intravenous fentanyl administrations were used for postoperative analgesia to prevent hepervetilation until postoperative day (POD) 1. No aggravation of neurological symptoms was noted on PODs. The patient often became unsettled because of difficulties in child care due to paralysis, but started taking medicine and meals on POD 2. She was discharged with her child on POD 8.

Case 2

A 35-year-old multipara had uneventfully given birth to the first child via normal vaginal delivery 5 years ago. She weighed 55 kg at 30 weeks' gestation, and she had been transported to the emergency hospital due to nausea, headache, and sudden loss of consciousness. Head CT revealed intraventricular hemorrhage in the bilateral ventriculus lateralis and ventriculus quartus (Figure 2A); intraventricular drainage was performed immediately. Bilateral internal carotid artery stenosis and collateral vessels in the region of the middle cerebral artery was seen on cerebral angiography. Therefore, the patient was diagnosed as having intracranial hemorrhage caused by moyamoya disease. Consciousness was regained after the operation at the intensive care unit, and the patient was transported to our hospital for perinatal management of cerebrovascular complications. On admission, her initial blood pressure was 107/80 mmHg and HR was 73 bpm. Slight weakness of the right upper and lower extremity was noted. Fetal growth and results of the nonstress test (NST) were normal. Since admission, her blood pressure had been high and unstable, although we tried to control it by administering diltiazem. Cesarean delivery was scheduled at 34–38 weeks' gestation. However, at 32 weeks' gestation, the patient suddenly lost consciousness again and was intubated. Urgent CT of the head revealed hemorrhage with extravasation (Figure 2B) in left lateral ventricle and fourth ventricle. Emergency caesarean delivery, excision of aneurysm at left trigone of the lateral ventricle, direct hematoma evacuation, and external decompression were performed under general anesthesia. The initial blood pressure in the operation room was 150/54 mmHg, and anisocoria was noticed. The patient was monitored ECG, NIBP, SpO_2, $EtCO_2$, BIS, and an artery cannula was inserted via the left radial artery. After the infant was delivered, we inserted a central venous catheter via the right internal jugular vein. Anesthesia was maintained with 2% sevoflurane until the infant was delivered and then switched to TCI of propofol at 2–3 μg/ml. The operation time was 9 h 15 min, anesthesia time was 9 h 45 min, blood loss was 1980 ml, total volume of infusion was 4100 ml, and total volume of transfusion was 1080 ml (Figure 3). The head CT performed on the next day revealed hydrocephalus, and intraventricular drainage was performed immediately. On POD 3, the intracranial pressure (ICP) increased to over 20 mmHg. External decompression was performed, and therapeutic hypothermia was induced. The ICP increased to over 50 mmHg on POD 6, and epinephrine was continuously administered for hypotension, but the patient died on POD 9. The patient's child weighed 1852 g; it was delivered with Apgar scores of 2 (1 min) and 6 (5 min). The infant was intubated and temporally managed with mechanical ventilation but later extubated and discharged uneventfully.

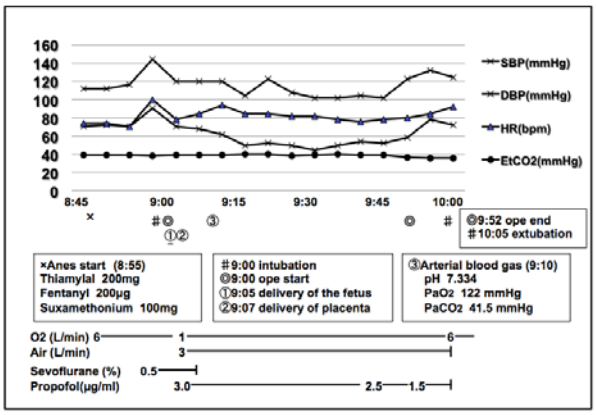

Figure 1: Procedure of anesthesia for case 1. SBP; systolic blood pressure, DBP; diastolic blood pressure, HR; heart rate, $EtCO_2$; end-tidal CO_2, PaO_2; partial pressure of oxygen in arterial blood, $PaCO_2$; partial pressure of carbon dioxide in arterial blood.

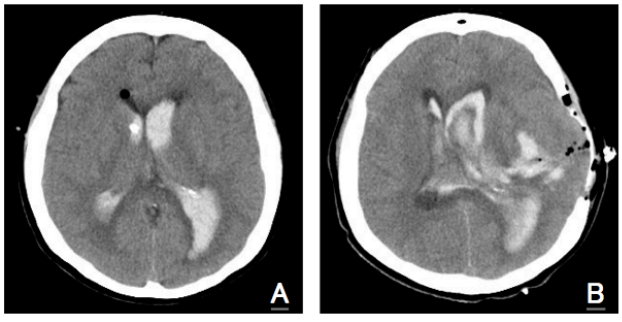

Figure 2: Brain computed tomography (CT) of Case 2. At 30 w 4 d of gestation (A) shows bilateral intraventricular hemorrhage. It extended to the intraventricular foramen. At 32 w 0 d of gestation (B) shows intracerebral hemorrhage with extravasation.

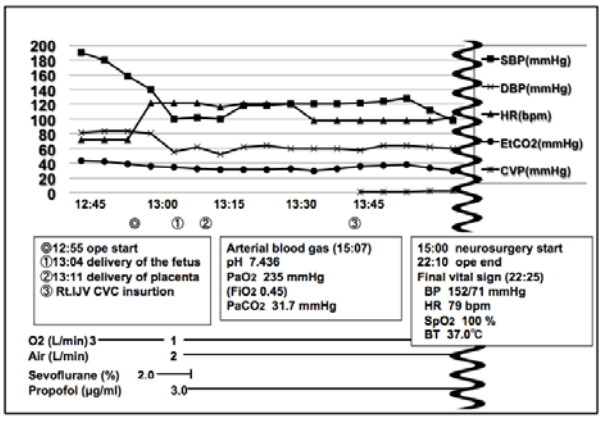

Figure 3: Procedure of anesthesia for case 2. SBP; systolic blood pressure, DBP; diastolic blood pressure, HR; heart rate, $EtCO_2$; end-tidal CO_2, CVP; central vein pressure, IJV; intrajugular vein, CVC; central venous catheter, PaO_2; partial pressure of oxygen in arterial blood, $PaCO_2$; partial pressure of carbon dioxide in arterial blood, FiO_2; fraction of inspired oxygen, SpO_2; pulse oximeter oxygen saturation, BT; body temperature.

Discussion

Moyamoya disease is a rare cerebrovascular disorder of unknown cause, which is likely to be seen in Asian countries, especially in Japan. The source of the disease is bilateral stenosis or occlusion at the end of the internal carotid artery or at the origin of the anterior-mid cerebral artery. This leads to the growth of collateral vessels at the base of the brain and to formation of an abnormal network of vessels, called moyamoya vessels, which are associated with cerebral hemorrhage and infarction. In Japan, the incidence of moyamoya disease is reported to be 0.4–0.6 cases per 100,000 population, and the male:female ratio is 1:1.8.4,5 About 10% of patients have been reported to have hereditary factors [3,5]. Moyamoya disease has 2 peaks of incidence, one in childhood (around 5 years of age) and the other one in the third decade.5 In children, the main symptoms of moyamoya disease are transient ischemic attacks, which often occur during hyperventilation, while the major manifestation in adults is cerebral bleeding [3,6]. The main goal of medical or surgical therapy of moyamoya disease is to prevent repeated cerebral infarction or bleeding in the future, and bypass surgery of the vessels is effective to prevent re-infarction in patients with ischemic onset [6]. However, no effective treatment has been established to prevent re-bleeding in patients with hemorrhagic onset. Sometimes, pregnant patients with moyamoya disease who undergo cesarean delivery require management or anesthesia. Pregnant women with moyamoya disease are divided into 2 types: patients who had been diagnosed before they got pregnant, and patients who are diagnosed or identified for the first time because of cerebral apoplexy during the gestation period [4,6]. In general, patients who had already been diagnosed before pregnancy may be at low risk of neurological events; therefore, strict and cautions management results in most cases in an uneventful postoperative course [5,6]. On the other hand, patients who have been first diagnosed during their perinatal period, often have an onset of cerebral bleeding and, in general, are at high risk of mortality or neurological events [5,6]. There have been several reports of pregnant patients with moyamoya disease in Japan, but an appropriate method for delivery has not been established yet. Normal vaginal delivery is not recommended for pregnant patients with moyamoya disease because of the risk of hyperventilation and hypertension due to pain occurring in the second stage of delivery, leading to ischemia and hemorrhage. Cesarean delivery has been widely recommended, but is not necessarily superior to vaginal delivery because the incision and delivery often cause significant alteration in hemodynamics [2,5]. The optimal method of anesthesia in patients with moyamoya disease is still debated. Local anesthesia is preferred and used in many cases because the patients are awake. This allows the physician to observe the patient's neurological status, but it may cause hyperventilation and hypertension during operation [4,7]. General anesthesia is utilized for emergent operation due to the shorter induction time, moreover, is reported to be useful to prevent hyperventilation [8]. However, general anesthesia has risks of hypertension, hypotension, and aspiration during intubation and it may be difficult to notice some neurological symptoms during general anesthesia [4,9]. In addition, the use of inhalational halogenated anesthetics may cause an impairment of cerebral autoregulation [10]. This is a critical problem for the patients of moyamoya disease, however, sevoflurane does not affect cerebral autoregulation less than 1.5 MAC [11,12]. Therefore, we used low concentrations of sevoflurane as a sedative agent until the neonate was delivered.

Whichever method of delivery or anesthesia is used, the goal of the management of pregnant patients with moyamoya disease is to maintain normotension, normocapnia and normothermia to stabilize circulation and brain blood flow [4,13-16].

In case 1, general anesthesia was performed because the patient had been administered antiplatelet, ticlopidine until 9 days before operation. In addition, a neurosurgeon had been consulted who advised that general anesthesia would be better than local anesthesia to avoid rupture of the cerebral aneurysm due to straining. Crash induction with suxamethonium was performed to prevent aspiration in this case. Blood pressure and $EtCO_2$ were stable during the operation, and no deterioration of neurological symptoms was seen during the gestation period. However, there is no evidence that anti-platelet agents have prophylactic effects on re-infarction in the chronic period in patients with moyamoya disease with ischemic onset. Moreover, treatment with anti-platelet agents may increase the risk of cerebral hemorrhage when the patient becomes an adult. Therefore, the patient should have stopped ticlopidine treatment to decrease the risk of intracranial hemorrhage before getting pregnant.

In case 2, the patient had onset of intracranial bleeding in her gestation period and her blood pressure was unstable after intravascular drainage. Moreover, the patient had already intubated, therefore, the operation was performed with general anesthesia. It was very important to control the blood pressure in order to prevent re-bleeding, but no other effective therapy was available. The incidence of re-bleeding in patients with moyamoya disease with hemorrhagic onset is reported to be about 7%. The prophylactic effect of surgical therapy for re-bleeding is controversial. A prospective multicenter trial, investigating the prophylactic effect of direct revascularization in patients with moyamoya disease with hemorrhagic onset compared with medical therapy, is ongoing since January 2001. Our patient did not have any familial disposition. Therefore, it was difficult to diagnose the underlying disease. After the mother had been diagnosed with the disease, it might have been better to deliver the neonate as soon as the fetus had grown enough to be safe and to prevent the second cerebral hemorrhage. However, it was difficult to make the right decision.

In both cases, we maintained normotherimia (35.5-37.5 C) throughout the operation. It is crucial since in has been observed that in moyamoya disease patients, vasospasm likely to occur in the presence of hypothermia, whistle, hyperthermia also trigger ischemic events [17-19].

We report the case of 2 pregnant patients with moyamoya disease who underwent emergency cesarean delivery under general anesthesia, which resulted in opposite outcomes. In pregnant patients with moyamoya disease, careful management during the perinatal period are essential to decide the timing of emergency cesarean delivery, which affects prognosis.

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