
Case Report

Crisis Management for Cesarean Section in a Parturient with Moyamoya Disease Under Spinal Anesthesia

Chia-Hsun You, Cheng-Yuan Lu, Ping-Heng Tan

Moyamoya disease is a cerebrovascular disease associated with stenosis of brain vessels and development of weak collateral circulation which may cause ischemia and hemorrhagic stroke. Parturients with moyamoya disease are at risk because labor pain may induce hypertension and brain hemorrhage. Elective cesarean section has been recommended for these patients, but the optimal anesthesia management is controversial. We report a 37-year-old pregnant woman with moyamoya disease who underwent cesarean section under spinal anesthesia. She had a massive drop of blood pressure from 157/101 to 90/50 mmHg during surgery. There were no complications in the parturient or baby. In conclusion, we reported a parturient with multiple systemic disease, who experienced hemodynamic instability after anesthesia. Despite the absence of complication, precautions should be taken against hemodynamic fluctuations for patients with moyamoya disease undergoing anesthesia.

Key words: moyamoya disease, cesarean section, neuraxial anesthesia

Introduction

Moyamoya disease is a cerebrovascular disease characterized by progressive stenosis of the intracranial internal carotid arteries and their proximal branches, which can cause strokes in the patients.¹ To compensate for the stenosis, collateral circulation develops around blocked major vessels. However, the collateral vessels are usually

small, weak, and prone to hemorrhage, aneurysm and thrombosis. The haziness and smoke-like appearance of the abnormal vessels on angiography gave rise to the term “moyamoya” in Japanese. The incidence of this disease peaks in two age groups: children about 5 years old and adults in the third to fourth decade of life. There are nearly twice as many female patients as male. It is most common in East Asia, especially in Japan. The overall incidence is about 0.35 per 100,000 people in

From the ¹Department of Anesthesiology, E-Da Hospital, I-Shou University, Kaohsiung, Taiwan.

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Address reprint request and correspondence to: Ping-Heng Tan, Department of Anesthesiology, E-Da Hospital/ I-Shou University, Kaohsiung City 82445, Taiwan.

Tel: +886-7-3477902, Fax: +886-7-3423348, E-mail: tanphphd@hotmail.com

Japan.² Major symptoms result from two mechanisms, brain ischemia (i.e., stroke and seizure) and compensatory vessels (i.e., hemorrhage from fragile collateral vessels and headache from dilated transdural collaterals).¹ Diagnosis of this disease is usually made by angiography (i.e., computed tomographic angiography, and magnetic resonance angiography) to identify intracranial internal carotid arteries.

Elective cesarean section has been recommended for parturients with moyamoya disease because labor pain could exacerbate hypertension which may lead to hemorrhage of collateral vessels and cause alteration in carbon dioxide tension leading to cerebral ischemia.³⁻⁵ In this article, we reported a parturient with preeclampsia and moyamoya disease who was admitted for cesarean section.

Case Report

A 37-year-old woman gravida 3, para 0, spontaneous abortion 2, was admitted at 35 weeks and 6 days of gestation with preeclampsia, and was scheduled for a cesarean section. She had a history of multiple systemic diseases, including type 2 diabetes mellitus, hypertension, and heart failure. In addition, she had brain ischemia when she was 28 years old when moyamoya disease was diagnosed. She was regularly followed up in the cardiovascular, neurology, and metabolism outpatient departments. Before this admission, she received transcranial Doppler ultrasound study which showed bilateral middle cerebral artery stenosis, absence of the right anterior cerebral artery, and left anterior cerebral artery stenosis. Preoperative physical examination showed mildly decreased muscle power of grade II in bilateral lower extremities. She received oxygen 2L/min via nasal cannula because of dyspnea. Complete blood and electrolyte data showed anemia (hemoglobin: 7.5 gm/dL, hematocrit: 24.1%). The platelet count (294000 /uL), coagulation function (prothrombin time:

9.1 sec, activated partial thromboplastin time: 25.9 sec, international normalized ratio: 0.9) and electrolyte data were in the normal ranges. No elevated liver enzyme or low platelet levels were noted. Chest radiography performed due to dyspnea showed a normal heart size without definite active lung lesion. Cardiac echocardiography revealed a normal sized left ventricle with an ejection-fraction of about 53.6%, left ventricle diastolic dysfunction, and mild tricuspid regurgitation.

Before the operation, the patient's blood pressure was 157/101 mmHg, heart rate 101 beats/min, and SpO₂ 99% on room air. To avoid hemodynamic fluctuation during tracheal intubation and the possibility of delayed extubation, general anesthesia was not considered. Initially, the anesthesiologist planned to perform combined epidural and spinal anesthesia. However, she had taken clopidogrel the day before the operation, so epidural anesthesia was not considered. Spinal anesthesia was performed by inserting a 26# spinal needle between the lumbar spine 3rd and 4th interspaces and 9 mg 0.5% heavy bupivacaine was injected intrathecally. The anesthesia level was up to T8. After spinal anesthesia, the patient's blood pressure dropped to about 80/50 mmHg, and fluid resuscitation with lactated Ringer's solution 500 mL and ephedrine 8 mg were given. The blood pressure remained around 90/50 mmHg for the next hour. Because of the low blood pressure, metabolic acidosis and electrolyte imbalance were possible due to inadequate tissue perfusion. Arterial blood gas analysis showed: pH 7.42, PaO₂ 255.5 mmHg, PaCO₂ 29.5 mmHg, HCO₃⁻ 18.7 mM/L, BE -5 mEq/L, hemoglobin 7.6 gm/dL, hematocrit 22%, Na 135 mEq/L, and K 3.7 mEq/L. Under the impression of anemia, two units packed red blood cells were transfused, and the blood pressure was increased to 140/70 mmHg. There were no postoperative complications for the patient or the newborn.

Discussion

The characteristics of moyamoya disease include occlusion of the intracranial arteries and formation of fragile collateral vessels to compensate for the occlusion. Both tend to result in cerebrovascular accident. Parturients with moyamoya disease have increased risks of cerebrovascular events, especially during the peripartum period. Previous research reported that some parturients with moyamoya disease experienced their first intracranial hemorrhage or cerebral ischemia during pregnancy or the postpartum period.² Elective cesarean section is suggested for these patients to prevent multiple physiological stimulations such as labor pain and exertion during normal spontaneous delivery. Maintenance of stable blood pressure and awareness of the patient's consciousness level are important during cesarean section. While substantially elevated blood pressure increases the risk of rupture of fragile brain collateral vessels, a decreased blood pressure may predispose the brain to ischemia. Although general anesthesia and neuroaxial anesthesia have both been reported in patients with moyamoya disease undergoing operations,^{5,6} the optimal anesthesia management remains controversial. In one previous report, general anesthesia was performed successfully.⁷ However, general anesthesia may cause blood pressure fluctuation associated with intubation/extubation, hypercarbia/hypocarbia, postoperative pain, nausea and vomiting. Unstable blood pressure may endanger the weak cerebrovasculature.⁵ In addition, general anesthesia in cesarean section may increase the risk of maternal aspiration and the possibility of delayed extubation. Neuroaxial anesthesia has been suggested to offer stable hemodynamics with adequate preanesthetic volume loading. In addition, the anesthesiologist can closely observe the consciousness level as the patient is awake.² However, spinal anesthesia could induce a substantial decrease in blood

pressure, which may increase the risk of brain ischemia.⁵ Epidural anesthesia, on the other hand, has the same advantages as spinal anesthesia without notable adverse hemodynamic impact so that the patient's cerebral blood flow and intracranial pressure could be maintained. A combination of spinal and epidural anesthesia has the advantage of neuroaxial anesthesia and abolishes the blood pressure effect of spinal anesthesia.

We reported a parturient with preeclampsia, multiple systemic diseases and moyamoya disease undergoing cesarean section. Considering that hemodynamic instability during tracheal intubation may increase the risk to the fragile brain vasculature and the possibility of delayed extubation, we initially planned to perform combined epidural and spinal anesthesia. However, since she took clopidogrel until the day before the operation, epidural anesthesia was not suitable because impaired coagulation function may induce epidural hematoma. As a result, spinal anesthesia was performed, causing a blood pressure drop by 30% compared with baseline. After ephedrine injection, fluid challenge and infusion of packed red blood cells, her blood pressure returned to baseline level. Fortunately, no new onset neurologic deficit was noted and the baby was born without complications. A previous report indicated that moyamoya disease may be associated with deteriorating neurologic deficits⁸ and intracerebral hemorrhage in the peripartum period.⁹

In this report, we described a case of a parturient with preeclampsia and moyamoya disease receiving cesarean section with blood pressure crisis after spinal anesthesia. A thorough preanesthetic evaluation including the patient's medical history, complete hemogram with evaluation of coagulation function, serum electrolyte levels, and associated examinations are needed. Preanesthesia preparation such as volume loading and premedication are also important. During operation, mainte-

nance of stable vital signs and close monitoring consciousness level are critical for avoiding new onset neurologic deficits. Postoperatively, adequate analgesics are also needed because pain may induce elevation of blood pressure which is also a risk for cerebral vascular accident.

In conclusion, we described a parturient with moyamoya disease who received a cesarean section under spinal anesthesia. Although the blood pressure drop during anesthesia did not cause any complication, we have to keep in mind that good preoperative preparation and maintenance of stable hemodynamics during surgery are the first priorities, especially in patients with moyamoya disease.

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