

General Anesthesia for Elective Cesarean Section in a Patient with Moyamoya Disease

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Abstract

Moyamoya disease (MMD) is defined by vaso-occlusive changes at the internal carotid and cerebral arteries, leading to proliferation of an abnormal vascular network at the base of the brain. The disease is manifested by ischemic, hemorrhagic strokes or transient ischemic attacks. Pregnant and puerperal MMD patients may be at a higher risk of developing new strokes as well as during the peripartum period. The aim of the current study is to report a case of an obstetric MMD patient.

A 22 year-old term female patient was diagnosed with MMD after having a stroke at 2 years of age, resulting in hemiparesis and paresthesia of the left upper and left lower limbs. Intravenous general anesthesia was performed with continuous remifentanyl and propofol infusions. Hemodynamic alterations did not occur before childbirth. After childbirth, hypotension occurred and was reversed with metaraminol.

We chose general anesthesia for better control of hypotension, in addition to prevention of hyperventilation, which produces hypocapnia and decreases cerebral blood flow. Disadvantages of general anesthesia include hemodynamic response to airway manipulation leading to alterations in cerebral blood flow and perfusion, difficulty in diagnosing neurologic alterations intraoperatively and impossibility of skin-to-skin contact at birth.

Keywords: Anesthesia; Anesthesiology; Moyamoya disease; Obstetrics; Cesarean section

Introduction

Moyamoya disease (MMD) is characterized by vaso-occlusive changes [1], ranging from progressive stenosis to total occlusion of the distal portion of the internal carotid and middle or anterior cerebral arteries, leading to proliferation of an abnormal vascular network at the base of the brain. Its etiology remains unknown. However, familial MMD suggests a low penetrance autosomal dominant mode of transmission. The disease predominates in females at a 2:1 ratio. The prevalence of MMD is higher in Japan, where the incidence and prevalence are 0.35 and 3.14 cases per 100,000 individuals, respectively. Mortality is around 10% in adults and 4.3% in children. Genes responsible for the disease are located in chromosomes 3p24.2-26, 6q25 and 17q25. Most recently, 8q23 was discovered. Moyamoya disease is associated with other conditions including thyrotoxicosis, sickle cell anemia, Down syndrome, coarctation of the aorta and hypertension [2].

The disease is initially manifested by ischemic cerebrovascular accidents (CVA) (loss of consciousness, dizziness, headaches and infarction), hemorrhagic or ischemic strokes or transient ischemic attacks. Pediatric patients typically present with ischemic symptoms in early episodes, while adults exhibit hemorrhagic symptoms.

Medical treatment to reverse the primary disease process is not currently available. Decreasing the risk for strokes remains the mainstay of treatment. Management consists of bypass surgery to

supply ischemic areas with increased blood flow in children and adults. However, the effectiveness of surgery and long-term outcomes are still uncertain, particularly in patients exhibiting hemorrhagic episodes. Guidelines for management of pregnancy and childbirth in MMD patients have still not been developed. To avoid the risk of stroke, over 70% of pregnant women undergo cesarean section.

Case Report

A 22 year-old term primigravida black female was admitted with a diagnosis of MMD after having a stroke at 2 years of life, resulting in hemiparesis and paresthesia of the left upper and left lower limbs without other alterations. Neurosurgical approach was required and she remained without any further symptoms throughout pregnancy. AAS 100 mg was used during pregnancy and interrupted 4 days before the procedure. Cesarean section was indicated due to maternal disease.

Physical examination

HR 76 beats/min, BP: 130 × 90 mmHg, W: 76 kg, H: 157 cm, Hb 12, Ht 37, Platelets 343 thousand, normal cardiopulmonary auscultation. The left upper and left lower limbs showed grade II strength. Intravenous (IV) general anesthesia was indicated without anesthetic premedication.

Volume expansion was performed before induction with 1000 ml of Ringer's lactate solution IV. Preoxygenation, rapid sequence induction and Sellick maneuver were performed for endotracheal intubation, using remifentanyl in pumps that were not target controlled, propofol 150 mg IV and succinylcholine 60 mg IV. Surgery was performed under total intravenous general anesthesia with continuous

remifentanil (0.1 mcg.kg.min⁻¹) + propofol (110 mcg.kg.min⁻¹) infusions. Ranitidine 50 mg and metoclopramide IV were given. Ketoprofen 100 mg IV was administered for analgesia.

Patient monitoring

Cardioscope, pulse oximetry, blood gas analyzer, and invasive arterial blood pressure. CO₂ was controlled with capnography and arterial blood gas measurements.

There was no hemodynamic alteration until birth. It took 3 min from the skin incision to delivery, and the APGAR was 8/9. Surgical procedure lasted 45 min and the patient was extubated in the operating room. Hypotension occurred after childbirth and was reversed with metaraminol 0.5 mg IV.

Discussion

In the gestational period of patients without comorbid conditions, there is a higher risk of stroke (ischemic or hemorrhagic). However, there is no evidence that MMD patients have an increased risk of stroke during pregnancy. It is most important to prevent new events related to MMD, which may progress to maternal death or severe neurologic symptoms. Hypercoagulability, venous stasis and endothelial lesion, common during pregnancy, may contribute to the occurrence of stroke. Therefore, pregnant MMD patients have more vulnerable cerebral vessels and may be at a higher risk than pregnant women without the disease.

Normotension and normocapnia are mandatory. Blood vessels of pregnant women are vulnerable and a rapid increase in blood pressure may cause intracranial bleeding. In contrast, an abrupt decrease in blood pressure may provoke venous spasm in MMD patients, progressing with ischemic attacks. Abrupt changes in arterial blood pressure are more frequently observed during labor, although not in the case described here. During anesthesia, a balance should be maintained between the metabolic oxygen consumption rate and cerebral flow to prevent neurologic morbidity [2]. Therefore, the delivery route is important, taking into account the risks involved in patients harboring the disease. Hypothermia may precipitate vasospasm, thus maintenance of normothermia during anesthesia is fundamental to optimize cerebral perfusion [3].

More than 70% of pregnant women with MMD undergo cesarean section. This is the delivery route of choice to prevent intracranial hemorrhage due to hypertension during labor. In contrast, acute hypotension caused by anesthetic induction and postoperative hypercoagulation may lead to ischemic cerebral events. Cesarean section is more appropriate for patients with a higher risk of intracranial bleeding than cerebral ischemia, such as those who experienced previous bleeding episodes or did not have bypass surgery.

General anesthesia and spinal block have been described as successful for the performance of cesarean section. However, both techniques may cause sudden hypotension, which may progress to ischemic events in MMD patients [4]. Arterial hypertension produced by tracheal intubation during general anesthesia may cause intracranial hemorrhage. However, direct laryngoscopy of short duration associated with lidocaine administered in the laryngotracheal region, as performed in the case described, reduces pressor changes. Control of ventilation is important to prevent hypocapnia due to hyperventilation, which may cause dizziness, fainting and even complicate labor. General anesthesia in patients with severe anxiety

may be beneficial for the prevention of hypocapnia. In contrast, general anesthesia may produce low neonatal APGAR score at birth due to fetal circulation of drugs administered to the mother. There is also a risk of broncoaspiration during anesthetic induction [3]. Anesthesia with propofol has been successfully used for cesarean deliveries in MMD patients. Continuous propofol infusion reduces hypertensive response to laryngoscopy and intubation in the prevention of hemorrhagic stroke.

Vaginal delivery should be considered in patients at low risk for intracranial hemorrhage, such as those who underwent bypass surgery without a history of bleeding. Spinal anesthesia may reduce endogenous catecholamine levels with labor pain relief, resulting in decreased systemic vascular resistance. Furthermore, pain relief can prevent hypocapnia caused by hyperventilation.

Disadvantages of general anesthesia are potential hypertension during laryngoscopy and extubation, leading to intracranial hypertension, difficulty in diagnosing neurologic alterations in the intraoperative period and impossibility of skin-to-skin contact at birth.

There are no curative or remissive treatments for MMD. Initial supportive care is based on the symptoms and findings at each presentation for maintenance of cerebrovascular function [3]. Antithrombotic agents are used in patients with ischemic symptoms. Antiplatelet agents are more commonly used than anticoagulants due to the known risk of hemorrhagic stroke in these patients. Calcium channel blockers were employed in some studies in an attempt to reverse ischemic symptoms. Anticonvulsants have shown results in patients with seizures. Corticosteroids have been used, despite the lack of scientific confirmation [4].

Hemorrhagic cases may require drainage and removal of the hematoma, depending on size and location. In ischemic cases, treatment is aimed at increasing blood flow to hypoperfused areas by creating collateral circulation. Surgery is performed in an attempt to reduce the formation of abnormal blood vessels and decrease the risk of hemorrhagic stroke produced by hemodynamic oscillations in vessels affected by MMD. Early neurosurgical intervention is made to revascularize directly, indirectly or both. Direct procedures are more commonly used in adults, while indirect procedures are used in children. Controlled randomized studies of recurrent infarction comparing different revascularization procedures with each other or with drug treatment have not been published.

Diverse surgical procedures have been used such as superficial temporal artery-middle cerebral artery (STA-MCA) anastomosis, encephaloduroarteriosynangiosis (EDAS) and encephalo duro arterio myo-synangiosis (EDAMS). Prognosis is more favorable after the STA-MCA surgery with prompt increase in the blood supply [2]. Bypass surgery, commonly performed in the treatment of MMD, reduced not only the incidence of ischemic attacks, but also decreased bleeding episodes in patients initially presenting with ischemic symptoms. Cases with hemorrhagic stroke as the initial manifestation are considered more severe than those with ischemic strokes [5]. Multiple strokes or transient ischemic attacks (TIAs) can provoke mental deterioration and may be fatal due to hemorrhagic stroke, particularly at term or during labor.

Conclusion

Hemorrhagic stroke may be confused with pregnancy-induced hypertension since transient hypertension and proteinuria may

complicate a hemorrhagic stroke. Non-contrast computed tomography or magnetic resonance imaging may help differentiate between both conditions. We chose general anesthesia for better control of hypotension (common in spinal anesthesia, leading to hypoperfusion of vaso-occlusive areas). In addition, the technique could prevent hyperventilation secondary to maternal anxiety, which causes hypocapnia and decreases cerebral blood flow. In this case, no new event related to MMD in the peripartum period occurred.

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