Anesthetic management of a patient with moyamoya disease: A case report

CPT LISA A. PETTY, CRNA, MHS, USA, AN*
Aurora, Colorado
*Winner of 1992 Student Writing Contest

A previously healthy 4-year-old female presented with an 18-month history of frequent headaches and seizures. Magnetic resonance imaging (MRI) and angiography revealed severe stenosis of the left proximal intracranial carotid artery, with bilateral development of moyamoya vessels, left greater than right. A diagnosis of moyamoya disease was made, and the patient was scheduled for surgical correction consisting of an encephalo-duro-arterio-synangiosis.

Moyamoya disease is a rare, occlusive cerebrovascular disorder characterized by bilateral stenosis of the internal carotid arteries and their branches. While its etiology is currently uncertain, recent studies indicate that focal arteritis, secondary to an antigen-antibody reaction, leads to the development of the stenosis.

Anesthetic management of patients with moyamoya disease focuses on maintenance of adequate cerebral blood flow, normalization of intracranial pressure, and avoidance of both cerebral vasoconstriction and vasodilation. Several anesthetic techniques have been successfully employed; one such method is presented.

Key words: Anesthetic management, encephalo-duro-arterio-synangiosis, intracranial carotid artery stenosis, moyamoya disease.

Introduction

Moyamoya disease is a rare, occlusive cerebrovascular disorder characterized by bilateral stenosis of the intracranial portion of the internal carotid artery, often with extension into the anterior cerebral artery and middle cerebral artery. This occlusive process results in an enlargement of the vascular network at the base of the brain and appears on angiography as a hazy shadow resembling a puff of smoke. Moyamoya in Japanese means something hazy, such as a puff of cigarette smoke drifting in the air.

Symptoms, including transient ischemic attacks and seizures, occur as a result of decreased regional cerebral blood flow beyond the stenotic lesion. Medical management centers on the control of symptoms until surgical intervention can be performed. The goal of surgery is to restore adequate cerebral blood flow to ischemic areas of the brain. Various techniques, both anastomotic and nonanastomotic, have been employed.

The following is a report of the anesthetic management of a child who presented with bilateral moyamoya occlusion, followed by a discussion of the disease, surgical intervention, and anesthetic considerations.

Case study

A 4-year-old, 10-kg female presented with an 18-month history of frequent headaches and seizures, manifesting as both grand mal and focal involvement of the left arm and face. At the time of admission the seizures were effectively controlled
with carbamazepine, and the child exhibited no residual hemiparesis.

Pregnancy had been complicated by pre-eclampsia and toxemia, and the patient was born prematurely at 34 weeks, weighing 3 pounds, 11 ounces. The child had also experienced two severe apneic episodes in the newborn nursery. She was developmentally delayed, estimated to be approximately 1 year behind cognitively and 1½ years behind in motor skills. Cerebral angiography and magnetic resonance imaging (MRI) revealed severe left proximal internal carotid artery stenosis with prominent thalamo-perforating arteries (moyamoya vessels). The right internal carotid artery was patent, but early moyamoya vessels were evident, as indicated by mild prominence of the thalamo-perforating arteries. The patient was kept on oral carbamazepine and scheduled for left encephaloduro-arterio-synangiosis.

A review of systems was unremarkable, except for the presence of Goldenhar’s syndrome (oculoauriculo-vertebral dysplasia), which did not significantly affect the anesthetic management. The child’s mother was of Japanese descent, and the father was Caucasian. At the preoperative interview, the child was very interactive and displayed no neurologic deficits or clinical evidence of intracranial hypertension. Her prior surgeries included strabismus repair and bilateral myringotomy with tube placement. A preoperative complete blood count and urinalysis were within normal limits. The patient was classified as ASA physical status II based upon her intracranial pathology. Two units of packed red blood cells were available.

Premedication consisted of 5 mL of oral carbamazepine, and the patient was transported to the operating room. Preinduction monitors were placed, including an oscillometric blood pressure cuff, precordial stethoscope, lead II electrocardiogram, pulse oximeter, and mass spectrometer. Anesthesia was induced with halothane, titrated up to 2% in a 3:1 nitrous oxide-oxygen mixture. Twenty-gauge intravenous (IV) lines were inserted in the dorsum of the right hand and the left wrist, and infusions of isolyte solution through pediatric buretrols were started. Atropine 0.2 mg was administered intravenously, and vecuronium 1.0 mg was given to facilitate tracheal intubation. Prior to intubation with a 4.5-mm oral RAEE® endotracheal tube, lidocaine 20 mg IV was also administered. Preinduction blood pressure was 94/50 mmHg, and pulse was 130 beats per minute (BPM). Vital signs during induction and intubation were stable, with a minimal response to intubation. An esophageal stethoscope and urinary catheter were then inserted, and a full-body Bair Hugger® warming blanket was placed. All IV fluids were warmed and administered at a rate of 4 mL/kg/hr, adjusted according to cardiovascular indices and urine output. Glucose-containing solutions were avoided, because elevated serum glucose levels have been associated with an adverse neurologic outcome in the face of cerebral ischemia.

Anesthesia was maintained with isoflurane (0.6-1.5%) in a 1:1 air-oxygen mixture, incremental fentanyl (total 55 μg), and vecuronium bromide (total 8 mg). Ventilation was manually controlled utilizing a Bain® coaxial circuit to maintain an end-tidal carbon dioxide concentration of 35-45 mmHg. Surgery was performed in the supine position, with the head turned to the right. Electroencephalographic monitoring was utilized, with no evidence of cerebral compromise throughout surgery.

The surgical procedure was uncomplicated and lasted 4 hours and 40 minutes. The patient remained hemodynamically stable throughout the procedure, with a blood pressure range of 80-100/40-50 mmHg and a heart rate of 100-110 BPM. Urine output was maintained at approximately 1.8 mL/kg/hr, and operative blood loss was estimated to be 35 mL. After closure of the dura mater, residual neuromuscular blockade was antagonized by the administration of neostigmine, 0.8 mg, and glycopyrrolate, 0.15 mg IV. The patient was extubated in the operating room and taken to the surgical intensive care unit (SICU).

Upon arrival at the SICU, the patient was awake and alert, with blood pressure 137/64 mmHg, heart rate 133 BPM, respiratory rate 28 breaths per minute, temperature 99.2°F, and oxygen saturation 99%, while breathing humidified blowby oxygen. The postoperative course was uneventful, and the patient was transferred out of the SICU the following day and discharged on the fourth postoperative day.

Discussion

Pathophysiology of moyamoya disease. Many cases of moyamoya disease have been reported since its first description by Shimizu and Takeuchi in 1955. It occurs predominantly in the Japanese population, most often in children, and more frequently in females. Approximately half of affected patients present before the age of 10 years, some as young as 6 months. Most often, the disease presents as bilateral stenosis of the internal carotid artery; however, cases of unilateral involvement in both children and adults have been reported.

Symptoms occur as a result of ischemia from cerebrovascular insufficiency, when developing collaterals fail to compensate for the progressive narrowing of the internal carotid artery. In children,
the disease commonly manifests as transient ischemic attacks, leading to neurologic sequelae, while adults usually present with subarachnoid or intracerebral hemorrhage from developing aneurysms in small arteries.

Hemiplegia can alternate between the right and left side of the body, and sensory deficits, as well as mental disorders, can exist. Irreversible neurologic deficits are more likely to develop when the patient is less than 4 years old at the time of disease onset. Headaches and seizures often occur as a result of crying and hyperventilation.

Although a definite cause for moyamoya disease has not been found, studies have indicated that a generalized immunological arteritis may play a role. There is a high correlation between moyamoya disease and preexisting infections above the neck, such as tonsillitis, sinusitis, otitis media, and boils. The infection is thought to stimulate an antigen-antibody reaction, which results in the formation of immune complexes that circulate systemically.

Antigens at the site of inflammation stimulate nearby sympathetic nerves with retrograde stimulation of the superior cervical ganglia. This results in enhanced permeability in those vessels innervated by the ganglia, allowing immune complexes to enter the vessel wall. Ultimately, inflammatory changes lead to stenosis of the arterial wall. On autopsy, histological examination frequently reveals an eccentrically thickened tunica intima around the carotid fork and proximal portions of the anterior cerebral artery and the middle cerebral artery. The internal elastic lamina appears folded and tortuous; however, the tunica media and adventitia often remain intact.

Cerebral angiography provides the definitive diagnosis for moyamoya disease. It generally reveals stenosis or occlusion of the internal carotid artery at its bifurcation, and the anterior cerebral artery and the middle cerebral artery cannot be visualized. Several investigators have also reported the use of a computed tomographic (CT) scan with contrast and an MRI.

In general, the CT scan findings are relatively nonspecific, and in approximately half of ischemic cases, normal CT scans have been reported. The superior anatomic resolution achieved with MRI has made it a more efficient means of detecting ischemic brain infarctions, as well as identifying vascular luminal changes and moyamoya vessels necessary to diagnose the disease.

Medical and surgical management. Management of moyamoya disease includes initial medical intervention and, ultimately, definitive surgical treatment. Medical management to control the course of the disease until the patient is old enough to tolerate surgery centers on vasodilation to improve regional blood flow, control of the inflammatory process, and management of neurologic symptoms. Agents employed include anticonvulsants, corticosteroids, calcium channel blockers, and antiplatelet agents, such as salicylates. Medical management alone is rarely sufficient to completely eradicate symptoms.

The goal of surgical intervention in moyamoya disease is to establish collateral blood flow with the intention of revascularizing previously ischemic areas of the brain. The most common procedure, encephalo-duro-arterio-synangiosis (EDAS), involves attaching a superficial scalp artery to either the dura or arachnoid mater in a nonanastomotic fashion. In a manner that is not completely understood, over a period of several months collateral circulation develops which eventually provides adequate regional blood flow and resolution of symptoms.

Given that moyamoya disease is rare and the number of patients treated surgically is relatively small, surgical intervention appears to be relatively successful. Matsushima and associates operated on 22 lesions (both bilateral and unilateral) in 16 patients. Follow-up angiography performed from 6 months to 1 year postoperatively revealed the development of adequate collateral circulation in 72% of these lesions. Suzuki and colleagues obtained follow-up angiograms over a 10-month period from 21 patients who had received surgical intervention using EDAS. Increases in hemispheric and cortical blood flow were evident in 60% of post-EDAS patients. The development of successful collateral flow was more significant in patients who had presented with transient ischemic attacks than in those who had suffered cerebral infarctions. Cases of moyamoya refractory to EDAS have also been reported.

Anesthetic management. The anesthetic management of patients with moyamoya disease focuses on maintenance of adequate cerebral blood flow, normalization of intracranial pressure, and the avoidance of both cerebral vasoconstriction and vasodilation. Little has been written on the management of these patients under general anesthesia.

In the preoperative period, the patient should be assessed for the presence of intracranial hypertension and evidence of cerebral ischemia. Anticonvulsant therapy should be continued, and a bleeding time is indicated for those patients who may have been treated with antiplatelet agents, such as aspirin.

During induction of anesthesia, two important goals are to maintain stable hemodynamics and
normocarbia in order to preserve cerebral blood flow. In addition, moyamoya disease may be complicated by the presence of cerebral aneurysms. Meticulous control of blood pressure is imperative, because hypertension may lead to aneurysmal rupture, while hypotension may cause ischemia.²

No anesthetic technique has been shown to be superior to another in the management of moyamoya disease; however, some considerations are important. Since many of these patients may have established hemiplegia, with its increased risk for hyperkalemia, the use of a nondepolarizing muscle relaxant is indicated.²

Vecuronium is an attractive alternative, because of its demonstrated cardiovascular stability. Nitrous oxide can cause cerebral vasodilation, and its use in neuroanesthesia is controversial.² Isoflurane has beneficial effects on cerebral hemodynamics. While it is a mild cerebral vasodilator, it greatly decreases cerebral metabolic oxygen requirements, serving to protect the brain from ischemia during hypotensive episodes.⁴

Although successful use of halothane has been reported,³⁰, ³¹ it is not considered the agent of choice for maintenance of anesthesia.² Halothane is a potent cerebral vasodilator and can greatly increase cerebral blood flow, which can induce a “steal” phenomenon, whereby blood flow is diverted away from ischemic tissue. In this case, halothane was used only for induction, because it tends to be more readily accepted by the pediatric patient.

Maintenance of normothermia is also important, because changes in body temperature have been implicated as a factor in precipitating transient ischemia in moyamoya disease patients.³⁰ In addition, monitoring of end-tidal carbon dioxide (ETCO₂) or arterial carbon dioxide (PCO₂) is essential.

It has been well-established that changes in carbon dioxide can profoundly affect cerebral circulation. Takeuchi and associates demonstrated that both hyperventilation and hyperventilation can jeopardize blood flow to the frontal region of the brain in patients with moyamoya disease.³⁵

Sumikawa and Nagai reported severe neurological deficits postoperatively in patients who were hyperventilated (PCO₂ 30-35 mmHg) intraoperatively and no neurologic complications when ETCO₂ was maintained between 40-50 mmHg.³⁰ Brown and Lam maintained normocapnia during anesthesia without postoperative neurologic sequelae. In the present case, ETCO₂ was maintained between 35-45 mmHg throughout the procedure; no neurological changes were noted postoperatively.²

Patients with moyamoya disease are prone to seizures in the immediate postoperative period. Continuation of anticonvulsant therapy is important, and vigilant nursing care is imperative.²

Moyamoya disease, while rare, presents the anesthetist with clinical challenges similar to those provided by other more common intracranial vascular lesions. Adherence to the principles of safe neuroanesthetic management is essential in providing for a positive patient outcome.

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