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Anesthetic and Obstetric Considerations of Moyamoya Disease in Pregnancy after Intracranial Bypass Grafting: A Case Report and Literature Review

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Abstract

Moyamoya disease (MD) is a chronic idiopathic vasculopathy characterized by bilateral stenosis of the distal internal carotid arteries (ICA's) with subsequent formation of abnormal collateral vessels. The underlying etiology is unknown [1]. Patients with MD are predisposed to intracranial hemorrhage or cerebral ischemia given the fragile nature of the collateral circulation. Although there is no curative treatment for MD, there are surgical palliative procedures that can augment cerebral blood flow to areas of impaired perfusion and circumvent the abnormal collateral circulation. Medical management exists for patients who are not surgical candidates. Because the incidence of MD peaks during the second and third decades of life, it can potentially affect women of childbearing age [2]. However, the optimal anesthetic and obstetric management of the parturient with MD remains controversial. We present a case of a primigravida with a history of MD who underwent intracranial bypass grafting prior to conception and subsequently had a successful cesarean delivery that was complicated by transient ischemic attacks (TIA's) in the postpartum period.

Keywords

Moyamoya Disease, Pregnancy, Transient Ischemic Attack, Anesthesia

1. Case Report

A 28-year-old female with a past medical history of mild intermittent asthma

was referred to a neurologist by her primary care physician for repeated episodes of "tingling" and numbness in both of her hands and feet. She reported these episodes were present only when she exercised. They lasted for a few seconds and spontaneously resolved. She was diagnosed with MD by magnetic resonance angiogram (MRA), which showed evidence of bilateral ICA stenosis with associated collateral blood flow. She subsequently underwent bilateral superior temporal artery-middle cerebral artery (STA-MCA) bypass grafting. She did not experience any further neurologic symptoms.

Her first pregnancy occurred three years after the revascularization surgery. She was being treated with Aspirin 325 mg daily for stroke prophylaxis which was discontinued by her obstetrician one week prior to her hospital admission. She presented to the hospital at 39 weeks for a scheduled cesarean section. Her pregnancy had been unremarkable. She had a normal neurological exam on admission. Her admission lab work, which included a comprehensive metabolic panel (CMP) and a complete blood count (CBC), was unremarkable. Her vital signs on presentation were remarkable for an elevated blood pressure which remained less than 140/90 throughout her admission. She had a pre-eclampsia work-up done which resulted negative. She underwent an uneventful cesarean section under spinal anesthesia with subsequent delivery of a healthy female infant with Apgar scores of 8 and 9 at 1 and 5 minutes, respectively. Total blood loss after the procedure was estimated to be 700 mL. Twenty-four hours after delivery, the patient began to complain of "numbness and tingling" to her left cheek, lips, and tongue. The episode lasted approximately five minutes and subsided. One hour later, the patient reported feeling a similar sensation of left-sided facial numbness but this time lasting approximately three minutes. Upon evaluation by the Obstetrics team, the patient's neurological exam was unremarkable. She had no focal neurological signs or deficits. There was concern that she may be experiencing TIA's. Neurology service was then consulted. The on-call neurologist recommended that the patient undergo a brain MRI and MRA to rule out any new ischemic or hemorrhagic lesions. The brain MRI showed old lacunar infarcts in the right frontal deep white matter but did not show any evidence of acute ischemia or hemorrhagic lesions. The brain MRA showed stable obliteration of the distal supraclinoid and bilateral proximal anterior cerebral artery (ACA)/middle cerebral artery (MCA) consistent with the patient's history of MD (**Figure 1**). Given the imaging findings, the neurology service recommended restarting the patient's daily Aspirin, keeping her well hydrated, closely monitoring her blood pressure and avoiding episodes of hypotension, as well as neurological exam checks every four hours. She did not experience any further episodes of facial numbness or abnormal sensation. On post-operative day three, she was cleared by the neurology service given that her neurological condition remained stable. The rest of her post-operative course was unremarkable. She was discharged home the same day by the Obstetrics service.

Figure 1. MRA obtained during the patient's admission. The red arrow indicates the high-grade stenosis of the patient's right supraclinoid internal carotid artery (ICA) consistent with the patient's clinical history of MD. The stenosis of the left ICA is not clearly visualized in this MRA image.

2. Discussion

Moyamoya disease (MD), an idiopathic progressive occlusive disorder of the distal internal carotid arteries (ICA's), creates perturbations in cerebral blood flow which can potentially precipitate ischemic or hemorrhagic strokes, transient ischemic attacks (TIA's), or seizures. These alterations in cerebral perfusion can be exacerbated by the physiologic changes seen in pregnancy, making medical management of the parturient with MD particularly challenging. Despite prior literature addressing MD in pregnancy, there are currently no clear guidelines or standard recommendations as to the anesthetic and obstetric management in the parturient. Nevertheless, a clear understanding of the pathophysiology of MD and its complications can lead to safe and successful pregnancy outcomes.

Establishing a baseline neurological exam in the MD patient prior to the administration of any anesthetic is imperative. We believe that a full, thorough neurological assessment is essential to evaluate for any neurological deficit. A neurology consult may be needed to obtain a formal evaluation of the patient's neurological status and determine the need to obtain imaging. Post-delivery, vigilant monitoring of the patient's neurologic status is warranted, and the patient may require neurologic follow up. A brain MRI is needed if the patient reports any new symptoms or if a new neurological deficit is noted on physical exam.

The core of MD anesthetic management consists of maintaining the fine balance between cerebral blood flow (CBF) and oxygen consumption (*i.e.* $CMRO_2$) in order to minimize the risk of cerebrovascular events [3]. The autoregulatory mechanisms of CBF can be challenged and stressed via the physiologic hypervolemia, hyperventilation, and fluctuations in blood pressure seen during pregnancy. Despite these changes, there are strategies that can be implemented to maintain

a stable CBF—avoiding hypotension and hypertension, maintaining euvolemia and normothermia, and minimizing pain and hyperventilation. Blood pressure control is vital in the parturient with MD in order to minimize the risk of a cerebrovascular event. Any decrease in CBF could increase the risk of cerebral ischemia to areas of the brain that already have compromised blood flow from stenotic intracranial ICA's. Conversely, uncontrolled hypertension, such as that seen in severe pre-eclampsia, can lead to elevated cerebral perfusion pressure and potential rupture of the fragile collateral vessels, leading to a hemorrhagic stroke. Prior studies advocate for strict blood pressure control in the parturient with MD [4]. Calcium channel blockers and magnesium sulfate have been proposed as the best agents to control hypertension in MD because of their vasodilatory effects [5]. Maintaining euvolemia with oral and intravenous fluids can reduce the risk of cerebral ischemia from hypotension and hypovolemia [3]. Mild hypothermia as well as hyperthermia have been shown to induce arterial vasoconstriction, potentially compromising CBF [6] [7]. Hyperthermia can also increase $CMRO_2$, leading to a CBF-$CMRO_2$ imbalance which could precipitate cerebral ischemia [8]. Thus, it is advised to closely monitor the patient's temperature and any temperature disturbance be promptly addressed. Pain control is important for all parturients, even more so for those with MD. Pain and crying during labor can lead to hyperventilation and hypocapnia, increasing the risk for cerebral ischemia [5]. Thus, satisfactory pain relief should be strived for in all patients with MD.

The optimal method of delivery for the parturient with MD has yet to be elucidated. No prospective trials have been conducted to assess the safest mode of delivery. A systematic review of case series in 2018 of pregnant patients with MD did not find any outcome benefits of cesarean delivery over vaginal delivery [2]. Prior case series have noted no increase in neurological complications in patients who underwent vaginal delivery [5] [9]. The ideal method of delivery for the parturient with MD remains unknown and should be individualized based on the patient's risk for cerebrovascular accidents during the peripartum and postpartum period. For most MD patients, scheduled cesarean delivery is the preferred method of delivery [10]. There is concern that the pushing and Valsalva maneuvers during labor and vaginal delivery can increase intracranial pressure and cause the fragile blood vessels of the collateral circulation to rupture, leading to a hemorrhagic stroke [10]. However, cesarean delivery also poses its own risks, with potential for blood loss, blood pressure fluctuations, and fluid shifts leading to an increased risk for brain hypoperfusion and ischemic stroke. There is also little to no data demonstrating the benefit of one anesthetic modality over another (*i.e.* general anesthesia vs neuraxial) for the pregnant patient with MD. Based on current literature, neuraxial anesthesia tends to be the preferred mode of anesthesia for these patients [11]. Neuraxial anesthesia allows the patient to remain awake and it gives the anesthetic provider an opportunity to monitor the patient's neurological status. Some literature suggests that cesarean section under epidural anesthesia is the safest option for delivery [11]. Epidural is preferred

over spinal anesthesia as it provides steadier blood pressure control. Spinal anesthesia can lead to more significant hypotension over epidural anesthesia and potentially precipitate a stroke secondary to intracranial hypoperfusion. An arterial line placement may be needed for closer blood pressure monitoring if large fluctuations in blood pressure are anticipated. Regardless, a careful anesthetic plan employing strict blood pressure control should be instituted for the patient with MD.

Pregnancy produces a physiologic state of hypercoagulability secondary to elevated estrogen levels and decreased fibrinolytic activity [12]. Pregnant women are at a 4- to 5-fold increased risk of thromboembolism compared to women that are not pregnant [13]. Patients with MD have an increased incidence of forming emboli at the sites of arterial stenosis, further increasing the baseline risk of thrombosis seen in pregnancy [5]. Thus, it is recommended to start patients with MD on long-term antiplatelet therapy to reduce the risk of ischemic symptoms [14]. Medications such as aspirin or cilostazol have been previously used for primary or secondary stroke prevention. However, no randomized trials on the use of these drugs by patients with MD exist [15]. Antiplatelet therapy should be avoided in patients with MD and a history of hemorrhagic strokes.

During pregnancy, women develop dilutional anemia due to increased plasma volume that is disproportionate to the rise of red blood cells [16]. Thus, treatment of anemia with packed red blood cell transfusions may be needed to increase the oxygen-carrying capacity of blood and ameliorate this risk. The hemoglobin levels decrease throughout pregnancy [16] and should return to normal within 1 week of childbirth. During a cesarean section, there is on average an 800 to 1000 ml blood loss which further causes a reduction in hemoglobin [17]. Anemia is a risk factor for ischemic stroke [18] and can further potentiate the possibility of this occurrence in the MD patient.

Although there is no definitive treatment for MD, there are surgical palliative procedures that can circumvent the stenotic arteries and increase blood supply to areas of the brain at risk for ischemia. This can be accomplished via extracranial-intracranial revascularization techniques (*i.e.* superficial temporal artery-middle cerebral artery bypass, EMS, EDAS, etc.). Surgical intervention is usually reserved for patients that have symptomatic MD or for asymptomatic patients with findings on neuroimaging showing severe limitations in resting blood flow [5]. Whether patients with known MD should undergo surgical palliation prior to conception is unclear. Some authors advocate for surgery prior to conception in patients at high risk of neurovascular complications based on single-photon emission computed tomography (SPECT) imaging [19]. However, the absolute benefit of surgery prior to conception has not been demonstrated [5].

3. Summary

In summary, we present the case of a parturient with MD status-post intracranial bypass grafting who experienced neurological symptoms in the postpartum period after an uneventful cesarean delivery. We further discuss treatment options

for these patients. Although the anesthetic and obstetric management of MD in pregnancy may prove to be challenging, a clear understanding of the disease and its treatment can lead to safe and successful deliveries in this unique patient population.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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