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CLINICAL VIGNETTE

Anesthetic Considerations of an Obstetric Patient with Moyamoya Disease

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Introduction

Moyamoya disease (MMD) is a rare cerebrovascular disorder that is characterized by progressive stenosis, usually bilaterally, of the terminal portion of the internal carotid arteries (ICA) and anterior and middle cerebral arteries (ACA, MCA).¹ This stenosis results in the formation of dilated and fragile collateral arteries (moyamoya vessels) at the base of the brain that can form aneurysms and cause intracranial hemorrhage (ICH) or cerebral ischemia including transient ischemic attacks, and ischemic stroke.^{2,3} Other clinical manifestations can include epilepsy, headaches, altered mental status, and weakness.¹ The prevalence of cerebrovascular events in patients with MMD is approximately 50-75%.¹

The term moyamoya means “puff of smoke” in Japanese and alludes to the appearance of these collateral vessels on angiography.² Patients who have the classical angiographic appearance of moyamoya, without any other risk factors are known to have the disease while those with any of the associated conditions (i.e. Down’s syndrome, neurofibromatosis type 1, sickle cell) are said to have the syndrome.⁴

Although the incidence is highest in East Asian countries such as Japan and Korea, MMD appears throughout the world with an incidence of 0.086 per 100,000 in the United States.^{1,3} Since MMD occurs more frequently in younger people and in women of reproductive age, it is not uncommon for MMD to be diagnosed during pregnancy or postpartum.^{1,3} Furthermore, the physiological changes during pregnancy may present potential challenges to the anesthetic care of patients with MMD. We present a patient with suspected MMD and superimposed preeclampsia who presented for induction of labor at 36 weeks and 2 days.

Case

A 39-year-old female, at the time G5P2022, presented at 36 weeks and 2 days for induction of labor for superimposed preeclampsia with severe features. Her pregnancy was also complicated by a history of a cerebrovascular accident with residual right sided weakness one year prior which was suspected to be secondary to Moyamoya disease, diet-controlled gestational diabetes, chronic hypertension, advanced maternal age, and a body mass index of 36. Other past medical history included depression which was stable at the time of presentation. Her medications included daily baby aspirin, oral labetalol, subcutaneous enoxaprin, and prenatal vitamins. The

patient had no known drug allergies. Her surgical history was significant for an appendectomy at age 10.

Upon admission to Labor and Delivery, her enoxaprin was held and vital signs included: temperature 36.7 C, BP 142/73 mmHg, RR 15/min, and O2 saturation of 99% on room air. The patient was alert, oriented and in no acute distress. Airway exam, revealed a short and thick neck with normal range of motion, midline trachea, adequate mouth opening, Mallampati III classification. The rest of the physical exam was unremarkable.

The patient had requested for a labor epidural, which was placed without difficulty and she had an uneventful normal spontaneous vaginal delivery and maintained a systolic blood pressure below 140 mmHg throughout the delivery process.

Discussion

The physiological changes during pregnancy may present potential challenges to the anesthetic care of pregnant patients with MMD.

Physiology of Pregnancy and MMD

Pregnancy is a pro-thrombotic state due to increased estrogen, clotting factors, and venous stasis.⁵ For women with MMD, these physiological changes can be triggers for ischemia. The incidence of stroke in pregnancy may be increased by the ischemia induced by MMD, wherein emboli form in stenotic arteries and where fragile collaterals may bleed.¹

Increased blood volume during pregnancy may also increase the risk of ICH amongst women with MMD and this increased blood flow may also lead to an increase in the development of fragile collateral vessels.^{1,5}

The patient in this case presented with superimposed preeclampsia with severe features which is not uncommon in women with MMD.¹ Blood pressure management is therefore critical during pregnancy since high cerebral perfusion pressures may precipitate ICH. For our patient, was recommended to remain normotensive throughout her pregnancy and beyond.

Hypocapnia during labor due to hyperventilation and pain could also precipitate ICH.¹ Therefore, adequate pain control during labor is critical for MMD patients.

Despite these concerns, there is currently no definitive evidence of increased risk of cerebrovascular events in pregnancy amongst MMD patients.¹ However, some suggest that ischemic events, especially TIAs, are more common than hemorrhagic events in women with MMD, and that most of these patients have good outcomes.⁶ Others suggest that poor outcomes in pregnant women with MMD are typically due to the less common hemorrhagic events versus the more common ischemic events.⁷⁻⁹ Given the lack of definite evidence, MMD patients should be monitored vigilantly throughout pregnancy using a multidisciplinary care team approach.

Obstetric Anesthetic Considerations

Although no standardized protocols exist for the anesthetic management during delivery of patients with MMD, most strategies center around preventing hypertensive episodes and the associated sequela of neurological injury, particularly ICH.³

Since there are no guidelines establishing a preferred mode of delivery for these patients, until recently, most patients with MMD have had elective cesarean sections to avoid the increases in systemic and intracranial pressures associated with pain and pushing during vaginal delivery.^{1,8,9} For MMD patients electing a cesarean section, an epidural or combined spinal-epidural may be preferred over spinal analgesia alone due to the ability for the anesthesiologist to titrate medications to desired levels.^{1,8} These regional techniques also avoid the hypertension associated with intubation and extubation/emergence with general anesthesia. Since patients are generally alert and awake when using regional techniques, this also allows the anesthesiologist to continuously monitor for any changes to mental status which can be important neurological signs, especially when caring for a laboring patient with MMD. Despite these benefits, it is important to consider risks associated with cesarean deliveries such as blood loss, longer hospital stays, infections, blood transfusions, and deep vein thrombosis.³

Although cesarean deliveries have been most common, there are reports of good outcomes after vaginal delivery for pregnant patients with MMD, such as the patient in our case.^{8,10} However, for these deliveries, analgesia with an epidural is critically important to reduce the risk of ischemic and hemorrhagic events.¹ Epidurals decrease endogenous catecholamine levels associated with the onset of labor and aid in maintaining normotension and normocapnia.¹¹ Normocapnia is important to prevent periods of decreased cerebral blood flow. Although the patient in this case had two prior deliveries without anesthesia or epidural analgesia, she elected epidural analgesia for this delivery. Since MMD peaks in the second and third decade of life, it is possible that this patient did not develop MDD until after her two previous deliveries.

Overall, hyperventilation, hypotension, hypertension, alkalosis, hypocapnia and hypercapnia should be avoided in MMD.¹ Anxiolytics can be considered to prevent hyperventilation and an assisted second stage can also be considered to prevent increases in systemic and intracranial pressures with valsalva.¹

Conclusion

Although Moyamoya disease is a rare cerebrovascular condition, it is more prevalent amongst women of reproductive age. Anesthesiologists should gain familiarity of this condition and its associated anesthetic challenges. Misdiagnosing MMD patients can have catastrophic consequences for mother and fetus and it is important to consider MMD amongst patients of all ethnicities. In this case, our patient with MMD and superimposed preeclampsia had a safe induction of labor and vaginal delivery with an epidural catheter.

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