Moyamoya disease in pregnancy and delivery planning: A case series and literature review

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Abstract

Moyamoya disease is a rare condition characterized by stenosis or occlusion of the intracranial internal carotid arteries leading to the development of fragile collateral vessels. Disruption of these vessels can lead to both ischemic and hemorrhagic strokes. As such, these patients are sensitive to changes in intracranial pressure and pose a challenge in pregnancy and delivery planning. Two cases of a parturients with moyamoya disease are presented, and the literature regarding safe method of delivery in similar patients is reviewed. The available evidence suggests that adequate anesthesia, and maintenance of hemodynamic status, takes precedence over the exact method of delivery in these patients. Cesarean delivery under epidural anesthesia can be a safe option for parturients with moyamoya disease, but an uncomplicated vaginal delivery, most often assisted with either vacuum or forceps, has also been frequently reported.

Keywords

Neurology, cardiovascular, maternal-fetal medicine, high-risk pregnancy, physiology

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Introduction

Moyamoya disease (MMD) is a rare condition characterized by stenosis or occlusion of the intracranial internal carotid arteries leading to the development of collateral vessels. The image created on angiography of this collateral vasculature gives this condition its name, meaning “hazy like a puff of smoke” in Japanese. The incidence of this condition is highest in Japan with an incidence rate of 0.54 per 100,000 person-years. It is almost twice as frequent in women as in men.^{1–4}

Presenting symptoms can include ischemic stroke or transient ischemic attack (TIA) (50–75%), intracranial hemorrhage (10–40%), and less commonly seizures, headache, choreiform movements, and cognitive or psychiatric changes.⁵ Treatment includes supportive care, and in ischemic cases, anticoagulation in the form of anti-platelet therapy, low molecular weight heparin, as well as revascularization surgery.^{5,6}

MMD can become symptomatic during pregnancy due to physiological changes associated with the gravid state and thoughtful management, especially surrounding labor and delivery, is essential. A large survey study in Japan reported rates of neurological events during pregnancy, delivery, and the puerperium in patients with known MMD of 5.1%.⁷ As a strategy for minimizing MMD complications in pregnancy and the puerperium, Lee et al.⁸ describe the use of cerebral hemodynamic status assessment via single-photon emission computed tomography (SPECT) imaging prior to conception in order to identify patients most at risk of cerebrovascular events during pregnancy and up to 7 months postpartum. In identifying these high risk patients, Lee et al.⁸ postulate that they may benefit from pre-conception revascularization surgery. However, there are no definitive guidelines available regarding recommendations for delivery, and peri-delivery anesthesia for gravidas with MMD. Here, we present two cases of MMD in pregnancy followed by a review of the literature focusing on delivery management.

Case 1

A 34-year-old gravida 2 para 1 Caucasian woman initially presented with bilateral hand tingling, choreiform movements of the arms, and

mania, at the age of 22 years. An MRI diagnosed ischemic strokes in bilateral hemispheres, and angiogram was diagnostic of MMD. One year later, internal carotid stenosis progressed, prompting revascularization surgery in the form of bilateral encephaloduroarteriosynangiosis. This procedure accomplishes indirect re-vascularization via transposition of an external carotid artery branch to the leptomeninges overlying the affected area of brain.⁹ Her symptoms resolved post-operatively.

During her first pregnancy, at age 30 years, she delivered via uncomplicated planned primary cesarean section at term with epidural anesthesia. Post operatively, she experienced transient left-sided vision loss with no evidence of acute changes on MRI. She remained asymptomatic following the delivery with no residual deficits.

During her second pregnancy, at 36 weeks and 5 days gestational age, she experienced hand tingling. She underwent an uncomplicated repeat cesarean section at 37 weeks gestational age under epidural anesthesia. Intra-operatively, her blood pressure was maintained between 100 and 120 mmHg systolic and 60 and 70 mmHg diastolic. Estimated blood loss was 800 ml. She delivered a male infant weighing 3629 g with Apgar scores of 8 and 9 at 1 and 5 min, respectively. Post-operatively, on day 0, she experienced an episode of left eye ptosis. MRI and MRA showed no acute changes (Figures 1 and 2) and her symptoms resolved spontaneously.

Case 2

A 30-year-old gravid 10 para 1 with a history of developmental delay and chronic hypertension was evaluated after a rapid decline in her IQ was noted from 91 at age 19 to 54 at age 29. An MRI showed features

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Figure 1. There is focal encephalomalacia and gliosis in the anterior limb of the left internal capsule and left caudate nucleus head (thick arrow) consistent with old ischemic infarction.

Figure 2. 3-D time-of-flight MR angiography of the circle of Willis shows high-grade stenosis of both the left and right supraclinoid carotid arteries (thin arrows) compatible with the clinical history of MMD.

consistent with MMD with complete occlusion of the left internal cerebral artery (ICA) at the proximal supraclinoid, and occlusion of the right ICA distal to the origin of the posterior communicating artery. Additionally, left cerebral atrophy was noted. She was 30 weeks pregnant at the time. Subsequently, during pregnancy, she developed a seizure disorder necessitating treatment with levetiracetam.

At 37 weeks and 5 days gestational age, she developed gestational hypertension and underwent induction of labor. She received cervical ripening with a cervical foley and two doses of misoprostol. She received epidural anesthesia and progressed to fully dilated with pitocin augmentation. Passive descent took place for 2 h until the fetal vertex reached 3+ station. She then delivered a 2660 g infant with

Apgars of 8 and 9 via vacuum-assisted vaginal delivery without valsalva. She had an uncomplicated postpartum course.

Discussion

MMD is a rare condition in the non-Japanese population. However, because of its potential catastrophic effects on parturients, understanding the effects on pregnancy and delivery are essential in managing these patients. The first case presented here is an example of a woman diagnosed with MMD as an adult after presenting with an ischemic stroke and later underwent revascularization surgery. During two pregnancies toward term, she experienced symptoms consistent with TIAs, though did not suffer any lasting neurologic sequelae.

These patients were cared for by a multidisciplinary team including obstetricians, maternal fetal medicine specialists, obstetric internists, anesthesiologists, and neurologists. This team worked to develop the safest delivery plan to optimize delivery outcome. Given the complexity of this condition, as well as its rare incidence, multidisciplinary approach to care was essential in ensuring evidence-based high quality care for these patients.

Decision for timing and method of delivery in MMD patients is based on reducing risks associated with hemodynamic instability. Key components to maintaining hemodynamic stability in the intrapartum period include minimizing pain, avoiding blood pressure fluctuations, minimizing valsalva, and judicious fluid management. Minimizing pain during labor reduces the risk of hyperventilation or crying, and subsequent hypocapnoea, and resultant cerebral ischemia.⁵ Hypertension and hypotension can lead to hemorrhagic or ischemic cerebrovascular events, respectively, in MMD patients. Neuraxial analgesia, valsalva, intubation for general anesthesia, and changes in fluid status can result in fluctuations of blood pressure and can thus increase the risk of cerebrovascular events in these patients.^{10,11} Additionally, close monitoring of neurological status is essential. Kato et al.¹¹ cite the benefit of neuraxial anesthesia over general anesthesia for cesarean delivery, because patients can verbally report any new symptoms intra-operatively. In summary, maintaining hemodynamic stability, minimizing pain, and reducing valsalva are factors that should be taken into account when planning delivery for a patient with MMD.

In the majority of case series of parturients with MMD, scheduled cesarean delivery predominates as the chosen delivery method. There are however reported cases of vaginal delivery, the majority performed with epidural anesthesia, and many are operative (assisted) deliveries (Table 1).^{7,11–16} No increase in risk of neurologic complications was noted in patients who underwent vaginal delivery in these case series.

In Case 1, cesarean delivery was chosen due to prior history of cesarean and patient choice. Literature review suggests that cesarean delivery under epidural anesthesia can be a safe option for parturients with MMD, but an uneventful vaginal delivery has also been frequently reported, as described in Case 2. The evidence guiding delivery management is limited to case series; but it would seem that the method of delivery is less important than avoiding hypocapnia, hypertension, and hypotension, to maintain cerebral blood flow homeostasis and prevent ischemia or hemorrhage. Thus, neuraxial anesthesia and close attention to fluid status is recommended in all cases.

In conclusion, in patients with MMD, cesarean delivery under epidural anesthesia remains a safe option. We recognize the added morbidity associated with cesarean delivery,¹⁷ and based upon literature review, note that vaginal delivery assisted with vacuum or forceps can be a viable option for some patients.

Table 1. Case series describing method of delivery and anesthesia type in parturients with moyamoya disease.

References	Cesarean sections, N (%)	Anesthesia type	Vaginal deliveries, N (%)	Anesthesia type	Complications	Notes
Takahashi ⁷	45 (76.3)	Not described	14 (23.7)	11 CSE, 3 none	No intra or postpartum neurologic complications. One antepartum intracerebral hemorrhage causing severe impairment of mother.	Survey of 132 medical centers in Japan over 5 years.
Jung ¹²	25 (92.5)	19 Regional ^a , 6 Gen	2 (7.5)	Not described	Five patients had neurologic symptoms that resolved spontaneously.	Two patients diagnosed postpartum with MMD. All patients had favorable delivery outcomes.
Fukushima et al. ¹³	18 (90)	2 Epi, 15 CSE, 1 Gen	2 (11)	1 Gen, 1 none ^b	Two patients developed TIAs after cesarean delivery.	Planned cesarean delivery was recommended.
Kato ¹¹	5 (100)	4 Epi, 1 CSE	0 (0)	N/A	No neurologic complications.	Used sedation in all cases to reduce anxiety induced hyperventilation.
Tanaka ¹⁴	7 (26)	Not described	20 (74)	20 Epi	No neurologic complications.	All vaginal deliveries were assisted with either vacuum or forceps. Cesarean deliveries were performed for obstetric indications not related to MMD.
Sato ¹⁵	4 (28.5)	Not described	10 (71.5)	10 Epi	One patient experienced TIAs after two subsequent vaginal deliveries.	60% of vaginal deliveries were assisted with either vacuum or forceps. Vaginal delivery was recommended for women with no recent ischemic symptoms.
Sei et al. ¹⁶	5 (71.4)	2 CSE, 3 Gen	2 (28.6)	2 Epi	No neurologic complications.	Vacuum-assisted vaginal deliveries were recommended to reduce vasa previa. General anesthesia chosen in two cases due to patient with severe anxiety and in one case for thrombocytopenia secondary to HELLP syndrome.

^aIncludes epidural and CSE.^bUnintentional un-anesthetized vaginal delivery due to precipitous labor.

Epi: epidural; CSE: combined spinal-epidural; Gen: general; MMD: moyamoya disease; VD: vaginal delivery; CS: cesarean section; HELLP: hemolysis, elevated liver enzymes, low platelets.

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