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Approach to A Pregnant Woman with Moyamoya Syndrome

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ABSTRACT

Many female patients with the moyamoya disease have normal fertility functions, including those diagnosed before they enter their childbearing years. Moya moya syndrome (MMS) is a rare cerebrovascular disorder caused by segmentary stenosis and occlusions of the small vessels around the circle of Willis and distal branches of carotid arteries. The name of "moyamoya" means "puff of smoke" in Japanese and describes the look of the tangle of tiny vessels formed to compensate for the blockage. This disorder may presented transient paresis, headache, epilepsy, dementia, aphasia, ataxia, and subarachnoid hemorrhage. Although MMS is considered a disease of unknown etiology, infectious and genetic causes have been blamed. However, there has been no comprehensive review of the management of pregnancy and delivery associated with Moyamoya disease. The purpose of this report is to describe a pregnant woman diagnosed with moyamoya disease and to review the literature on other such cases.

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Introduction

Moyamoya Syndrome (MMS) is a rare cerebrovascular disease characterized by segmental narrowing and occlusion of the anastomoses in the end branches of the carotid arteries. The disease presents with symptoms such as transient or recurrent hemiparesis, headache, convulsion, nystagmus, dementia, aphasia, ataxia, and subarachnoid hemorrhage. Moyamoya disease (MMD) is a diagnosis of cerebrovascular disorder with obstructive lesions of the circle of Willis. To date, both environmental and genetic factors have played a role in the pathogenesis of MMD. Allelic variations in RNF213 are known to pose a risk of MMD; however, the functional roles of RNF213 remain mostly elusive. Moyamoya disease (MMD) represents a specific intracranial vascular disorder characterized by progressive obstructive lesions of internal carotid arteries and branches within the circle of Willis. To compensate for the reduced blood flow in the affected brain area, Moyamoya's fine vascular network, a Japanese word for "smoke puffs," develops as arterial stenosis progresses [1, 2]. Previous studies have shown that environmental factors, including chickenpox zoster virus infection, contribute to the development of MMD [3, 4]. On the other hand, population-based studies indicate that the incidence of MMD is higher in oriental populations than Caucasians, suggesting that some genetic origins may pose a risk for the development of vascular lesions [5].

Clinical Presentation and Intervention

In this article, we present a 23-year-old primipara pregnant woman with moyamoya disease who gave birth by cesarean section under general anesthesia at her 36th gestational week. Our case developed headache and vomiting caused by intraventricular bleeding from the moyamoya veins in the sixth month of pregnancy. Superficial temporal artery-middle cerebral artery bypass surgery was performed in the sixth month of pregnancy to prevent intracranial rebleeding or ischemic cerebrovascular attacks during labor and delivery. The patient was successfully delivered by cesarean due to fetal distress in the full term without any complications. The baby was born with a weight of 2400 g, and his Apgar score was 7/8. Umbilical arterial pH values were between 7.27 and 7.39. The patient was intubated with a 7 Fr tube by ventilating for 1 minute with a mixture of 40% air and 60% O2. In induction, 200 mg propofol and 0.6 mg / kg i.v rocuronium bromide were administered. The operation took 35 minutes. The patient was awakened by the administration of 1 mg/kg sugammadex (i.v). The patient was discharged without any problem after being followed up in the intensive care unit for two days postoperatively.

Discussion

Pregnancy is associated with an increased risk of cerebrovascular events in patients with moyamoya disease. In more than 20 births with moyamoya disease, the first intracranial hemorrhages have been reported, or symptoms may include impaired consciousness, generalized seizure, hemiparesis, involuntary movement in the limb, visual defect, dysarthria, and hypertension as signs of postpartum cerebral ischemia with pregnancy [4-8]. These are not the exclusive features of moyamoya disease, so other cardiovascular disorders and eclampsia should be excluded. Emergency cesarean operation is sometimes considered due to deterioration of the maternal condition.

In contrast, a relatively low risk of cerebrovascular events has been reported in parents diagnosed with moyamoya disease before pregnancy. There is a risk during pregnancy and the postpartum period and even after revascularization. Because the surgical procedure is not curative, palliative treatment to reduce cerebral ischemia, the effect of surgery on bleeding, and pathological changes of the disease are essential in the clinical course [1, 2]. Neurological deterioration may occur during pregnancy and puerperium. We must maintain normotension in patients known to have Moyamoya disease because hypertension can cause intracranial events. Both hypocapnia and hypotension compromise cerebral blood flow. Hypercapnia can also create an intracranial stealing effect by altering the cerebral blood flow in the area affected by moyamoya. There are many case reports about childbirth in parents with moyamoya disease.

In 1998, Komiyama and colleagues comprehensively reviewed the literature in which they listed 25 cesarean cases. Both general and neuraxial anesthesia have been used successfully. Moyamoya disease has been known for 40 years and is most common in East Asia, especially in Japan [4, 9]. The number of sufferers in Japan was approximately 6000 in 1996 (4.7 per 100 000 population). However, the disease has been reported worldwide and deserves the attention of medical professionals outside East Asia. Since the disease is more prevalent in the female population (male: female = 1:1.8), one affected patient of childbearing age is not uncommon. However, there are no guidelines for obstetric management of patients with moyamoya disease. The optimal anesthetic management for cesarean section is not yet to be established [10, 11].

The benefits of neuraxial are easy to evaluate neurological changes, and neuraxial anesthesia usually prevents hypertension from tracheal intubation and extubation. Deficiencies are more likely to be hypotension, and the patient may develop hyperventilation and hypertension due to it. The benefits of general anesthesia enable us to control blood pressure better and we can comfortably avoid hyper or hypoventilation.

In the literature, 30 patients diagnosed with moyamoya disease before pregnancy and 23 symptomatic patients diagnosed with moyamoya disease for the first time during pregnancy have been reported. Our patient was a pregnant who was followed up under the control of neurosurgery and was followed up without symptoms. The patient's nasolabial grooves were asymmetrical, and there was a distal phalanx deficiency in the fingers. She was diagnosed at the age of 10 and had no seizures or syncope complaints.

Conclusion

There is no evidence that pregnancy increases the risk of cerebrovascular accidents or reduces the risk of bypass surgery. The poor prognosis of the patient or newborn is mostly due to cerebral hemorrhage, not cerebral ischemia. It is essential to control blood pressure and avoid toxemia, especially during pregnancy. Cesarean or vaginal delivery can be done safely. Any anesthetic method can be used if particular attention is paid to preventing hypocapnia, hypotension, and hypertension. Oral contraceptives should be avoided.

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