
Intracerebral hemorrhage and Moyamoya disease in pregnancy

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Purpose: To present a case of Moyamoya disease with intracranial hemorrhage complicating pregnancy.

Clinical Features: A 36-yr-old parturient at 34 wk gestation presented with left hemiparesis, headache, nausea and vomiting. Subsequent deterioration in level of consciousness and the development of a dilated right pupil necessitated immediate intubation. Urgent non-contrast CT scan revealed a large right intracerebral hematoma with transtentorial herniation. The patient underwent simultaneous emergency Cesarean section and craniotomy. A postoperative angiogram revealed findings consistent with Moyamoya disease. The neonate survived but the patient developed severe cerebral edema and died eleven days postoperatively.

Conclusion: Adult patients with Moyamoya disease often present with intracranial hemorrhage which poses unique anesthetic challenges. We report a case of intracerebral hemorrhage during pregnancy, which is known to be associated with high morbidity and mortality. The anesthetic techniques are reviewed and discussed.

Objectif : Présenter un cas de maladie de Moyamoya accompagnée d'une hémorragie intracrânienne pendant la grossesse.

Éléments cliniques : Une parturiente de 36 ans, à 34 sem de grossesse, présentait une hémiparésie gauche, des céphalées, nausées et vomissements. Une détérioration subséquente du niveau de conscience et le développement d'une pupille droite dilatée ont nécessité une intubation immédiate. Une scanographie urgente, sans contraste, a révélé un gros hématome intracérébral droit et une hernie transtentorielle. La patiente a subi une césarienne d'urgence et une craniotomie simultanée. Un angiogramme postopératoire a mis en évidence des constatations caractéristiques de la maladie de Moyamoya. Le nouveau-né a survécu, mais la patiente a développé un œdème cérébral sévère et elle est décédée onze jours après la césarienne.

Conclusion : Les patients adultes atteints de la maladie de Moyamoya présentent souvent une hémorragie intracrânienne, donc des défis anesthésiques particuliers. Nous avons présenté un cas d'hémorragie intracérébrale pendant la grossesse, condition connue pour son taux élevé de morbidité et de mortalité. Les techniques anesthésiques ont été passées en revue et étudiées.

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MOYAMOYA disease is a rare cerebrovascular disorder characterised by bilateral steno-occlusion of the supraclinoid internal carotid artery, and formation of abnormal fine networks of collateral vessels at the base of the brain.¹ The disease was first described in Japanese patients but has subsequently been recognised in patients of all nationalities.² The incidence of Moyamoya disease is higher in female patients.³ Children often present in the first decade of life with clinical features of cerebral ischemia whereas adults often present in the third or fourth decade with features of intracranial hemorrhage.⁴

Case report

A 36-yr-old pregnant woman (gravida 3 para 0) at 34 wk gestation presented with a 48 hr history of headache, nausea and vomiting, and sudden onset of left hemiplegia. The patient was unaware of any major concurrent medical diseases. On examination, the patient was alert, had spontaneous eye opening and obeyed commands, but had confused speech (Glasgow Coma Score 14/15). Her blood pressure was 170/95 mmHg and other vital signs were stable. Further neurological examination revealed evidence of hypertonicity without clonus. Obstetric examination revealed a relaxed uterus appropriate for dates with a viable healthy fetus. Urinary examination showed evidence of trace protein. The patient was treated in the emergency room for presumed pre-eclampsia receiving intravenous magnesium sulphate (four grams followed by infusion 2 g·hr⁻¹) and small intravenous aliquots (2.5 mg) of hydralazine. Laboratory blood tests, which included hemoglobin, platelet count, coagulation profile and renal function were all within normal limits. The diastolic blood pressure remained stable between 90-95 mmHg as measured with intra-arterial monitoring.

Ninety minutes after presentation the patient suddenly became drowsy, poorly responsive to painful stimuli with left sided extensor posturing and abnormal flexion on the right, and then developed a dilated unreactive right pupil. The patient's trachea was intubated after she received 400 µg fentanyl, 100 mg thiopental and 120 mg succinylcholine. The blood pressure remained stable at 150/90 mmHg. Sedation was maintained with 2 mg midazolam and low-dose propofol infusion (50 µg·kg⁻¹·min⁻¹). Urgent non-contrast CT revealed a large right fronto-parietal intracerebral hemorrhage (ICH) centred at the putamen, considerable (2 cm) right-to-left midline shift and subfalcine and transtentorial herniation (Figure 1). The patient was transported to the operating room for emergent craniotomy and Cesarean section.

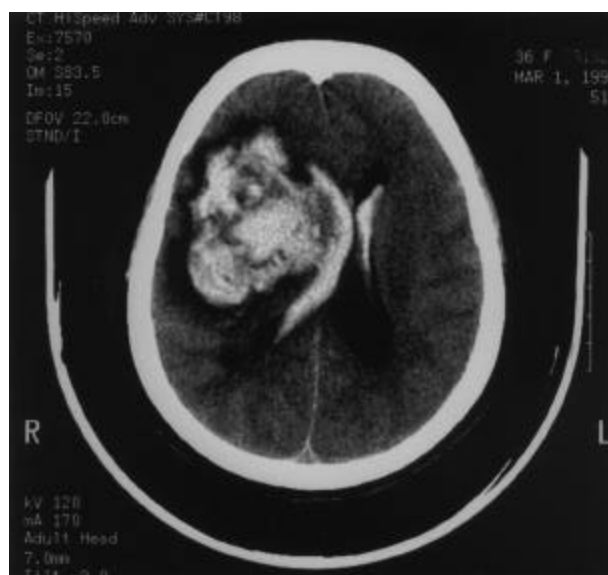


FIGURE 1 Non-contrast CT Brain showing a large right intracerebral haematoma with subfalcine herniation.

The patient was positioned supine with a left lateral tilt. Ventilation was maintained to achieve an arterial level of carbon dioxide in the normal range (31-33 mmHg) for pregnancy. A balanced anesthetic was administered which included 30 µg sufentanil, oxygen-nitrous oxide mixture (ratio 1:2), isoflurane (end-tidal < 0.7%) and rocuronium. An osmotic agent (50 g mannitol) was administered in an attempt to reduce cerebral parenchymal size. Simultaneous craniotomy and Cesarean section were performed.

A premature 2.65 kg female neonate was delivered via lower uterine segment Cesarean section. The neonate was immediately dried, and vigorously rubbed, and the oropharynx was suctioned. The one-minute Apgar score was 4. Bag and mask ventilation was provided for 30 sec with immediate improvement. The 5-min and 10-min Apgar scores were 8 and 10 respectively. The subsequent neonatal course was uneventful.

At craniotomy, clot evacuation was extremely difficult due to arterial bleeding. Estimated blood loss was six litres over a period of 180 min. This necessitated large blood and factor replacement via a rapid infuser device. Core temperature during the case was 35.0-35.8°C.

Postoperatively the patient was transferred to the intensive care unit with trachea intubated and lungs ventilated. The patient developed severe cerebral edema poorly responsive to medical therapy and cerebrospinal ventricular drainage. Four-vessel cerebral angiography revealed extensive arterial pathology con-

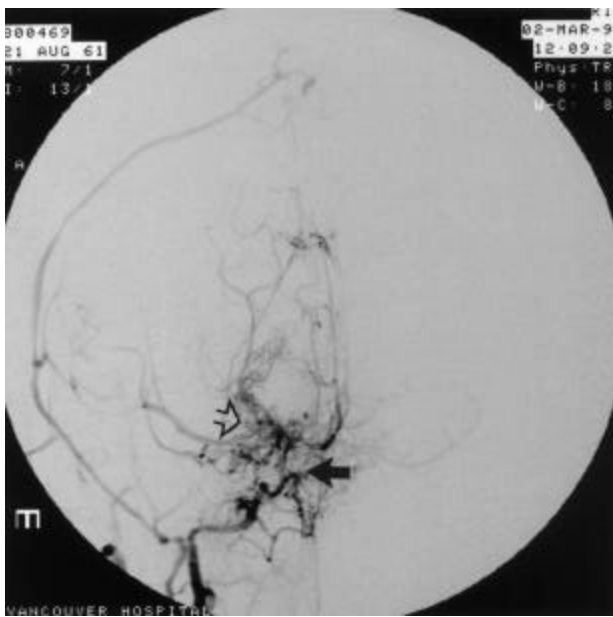


FIGURE 2 Right common carotid angiogram showing occlusion of the supraclinoid internal carotid artery (solid arrow) and extensive development of collateral vessels (open arrow).

sistent with Moyamoya disease (Figure 2). The patient died eleven days postoperatively.

Discussion

Moyamoya is a Japanese word that translates to something hazy like a puff of cigarette smoke drifting in the air.⁵ This “puff of smoke” appearance on cerebral angiography is due to the fine networks of vessels that form as a result of stenoses and occlusions of both internal carotid arteries and their terminal branches. Moyamoya patients are prone to intracranial hemorrhage because this fragile network of collaterals may be disrupted under increased hemodynamic stress, or alternatively because defined saccular aneurysms rupture in the circle of Willis (particularly the posterior circulation).⁶

Our patient presented with considerable neurological symptoms in the third trimester of pregnancy. Abnormal neurology during pregnancy can be due to a variety of etiologies, however it is most critical to distinguish between pregnancy-induced hypertension and intracranial hemorrhage.⁷ Our patient had unknown Moyamoya disease and suffered an intracranial hemorrhage, but transient hypertension and proteinuria made the precise diagnosis of hemorrhage unclear until non-contrast CT was performed.

The maternal mortality associated with intracranial hemorrhage, irrespective of the underlying etiology is

30-50%.^{7,8} Cesarean section is frequently performed in conjunction with craniotomy because intracranial hemorrhage often occurs in near-term pregnancy. Optimally Cesarean section is performed as soon after induction of anesthesia as possible to avoid 1. prolonged exposure to pharmacologic agents, 2. hemodynamic fluxes, and 3. changes in temperature, that may have unfavourable effects on the fetus. In our case simultaneous Cesarean section and craniotomy were performed because the patient had clinical and radiological evidence of brainstem herniation necessitating immediate clot evacuation.

Pregnant patients with known Moyamoya disease are vulnerable to both cerebral ischemia and intracranial hemorrhage. Komiyama *et al.*⁷ reviewed 30 pregnant patients known to have Moyamoya disease. Four patients had transient cerebral ischemia during their pregnancies, but all made full recoveries. One patient had an intraventricular hemorrhage at 30 wk gestation with subsequent poor neurologic outcome.

The optimal mode of delivery for patients with known Moyamoya disease has traditionally been Cesarean section. Cesarean section aims to prevent hyperventilation-induced cerebral ischemia and hypertension during labour. However, significant circulatory changes can occur at laparotomy and at delivery. There are six case reports of patients with known Moyamoya disease who have had vaginal deliveries without neurologic sequelae.⁸ Vaginal delivery under either epidural or spinal anesthesia is appropriate if the birth canal is favourable and delivery proceeds rapidly.⁸

The principle aim of anesthesia is the preservation of maternal and fetal well-being. It is critical to maintain maternal cerebral perfusion. Control of ventilation is of particular importance in optimising cerebral hemodynamics. Hyperventilation causes moyamoya vessels to vasoconstrict. This is known to cause diffuse slowing of the electroencephalogram, significant decreases in regional cortical blood flow, and clinical cerebral ischemia.^{9,10} Hypoventilation also leads to reductions in cortical blood flow.¹¹ The presumed mechanism is that hypercapnia results in steal from maximally dilated moyamoya vessels due to cerebral vasodilatation of normal carbon dioxide reactive regions. It would seem most appropriate, therefore, to maintain normocapnia whenever possible to promote maximal cerebral perfusion in Moyamoya patients.¹² Hypothermia is known to encourage vasospasm in moyamoya vessels, so maintenance of normothermia during anesthesia is also important in optimising cerebral perfusion.¹³

Elective Cesarean section can be performed under general anesthesia or major regional block. Neuraxial blockade is more frequently utilised because it avoids

the specific hazards associated with general anesthesia, and allows continuous monitoring of cerebral function.⁸ It is important to titrate neuraxial blockade to avoid hypotension and subsequent cerebral hypoperfusion. Patient reassurance can provide anxiolysis and thereby prevent hyperventilation and potential cerebral ischemia.

General anesthesia allows optimal control of ventilation but may be hazardous due to the risks of hypertension at intubation, gastric aspiration and neonatal depression. Traditionally, the choice of anesthetic agents has consisted of a combination of potent volatile anesthetic, nitrous oxide and intravenous narcotics. Recent evidence offers support for continuous propofol intravenous anesthesia in Moyamoya disease. Sato *et al.* found that regional cortical blood flow diminished under isoflurane-nitrous oxide general anesthesia compared with total intravenous anesthesia using propofol.¹⁴ Volatile anesthetics are known to have cerebral vasodilating properties and it postulated that a steal phenomenon analogous to hypercapnia occurs in Moyamoya disease. Propofol anesthesia has been successfully used for Cesarean section in a patient with known Moyamoya disease.¹⁵ Continuous propofol infusion reduces the hypertensive response to laryngoscopy and intubation^{15,16} which is important in Moyamoya patients prone to intracranial hemorrhage. Neonatal status as assessed by Apgar scores, cord acid base status and the neurologic adaptive capacity scores are similar when using intravenous propofol *vs* thiamylal-isoflurane anesthesia.¹⁷ Therefore, propofol anesthesia appears to be a safe alternative to "balanced anesthesia" for Cesarean section, and may offer specific benefits in Moyamoya patients.

Vaginal delivery can be performed under spinal or epidural analgesia. Epidural anesthesia has been shown to prevent hyperventilation and hypocapnia associated with labour.¹⁸ In Moyamoya patients neuraxial blockade can be used to help reduce hyperventilation and hypertension during the first and second stages of labour.

Conclusion

Intracranial hemorrhage can be confused with pregnancy-induced hypertension because transient hypertension and proteinuria can complicate ICH. The specific diagnosis of ICH can only be made after non-contrast CT scan has been performed.

In pregnant patients with known Moyamoya disease the principle aim of anesthesia is to maintain normoventilation, normothermia and normotension during delivery.

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