

Anaesthetic Management for Caesarean Section in a Patient with Moyamoya Disease

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INTRODUCTION

Moyamoya disease (MMD) is a rare, progressive occlusive cerebrovascular disorder characterised by stenosis of the distal internal carotid arteries and other major vessels at the base of the brain.

This leads to formation of a network of new, small, fragile collateral vessels that compensate, appearing as a "puff of smoke" on angiography (1). These fragile vessels carry an increased risk of rupture, leading to haemorrhagic strokes, ischaemic strokes, or transient ischaemic attacks.

In the parturient, the physiological stresses of pregnancy and labour - specifically hypercoagulability and fluctuations in PaCO₂ - increase the risk of both ischaemic and haemorrhagic stroke.

Anaesthetic management aims to maintain a delicate balance between cerebral oxygen delivery and demand by strictly avoiding hypotension, hypocapnia, and pain-induced sympathetic surges.

The primary anaesthetic challenge is maintaining cerebral perfusion pressure (CPP) against the backdrop of pregnancy-induced haemodynamic shifts.

While general anaesthesia and epidural techniques are frequently discussed, spinal anaesthesia provides a rapid, definitive block. However, it also carries a risk of sudden hypotension.

We describe the successful management of an elective caesarean section using a spinal anaesthesia approach.



CASE REPORT

A 30-year-old primigravida with a known diagnosis of MMD presented at 38 weeks of gestation for an elective caesarean section.

At age 14, she had a stroke while sleeping, leaving weakness in the left arm and left leg. Following several angiograms at that time, she was diagnosed with MMD. Subsequent corrective bilateral cranial bypass surgery allowed a good recovery.

Following a pre-operative assessment and multidisciplinary team discussion, a spinal anaesthetic was planned. Pre-operative management included aggressive intravenous hydration in the operating theatre, standard AAGBI monitoring, and insertion of an invasive arterial line for strict BP monitoring.

The intrathecal injection was performed at the L3-4 intervertebral space, and a solution of 8 mg of 0.5% hyperbaric bupivacaine mixed with 400 mcg diamorphine was administered using a Sprotte needle. BP was maintained by titrating a metaraminol infusion, targeting within 10% of baseline.

Normoxia was achieved with a combination of room air and supplemental oxygen.

There were no surgical complications, and no neurological events occurred. Post-operative care was provided in a HDU, with a focus on strict normocapnia, avoidance of hyperventilation, and intravenous hydration for 24 hours. She was discharged uneventfully, with no change in her neurological status.



DISCUSSION

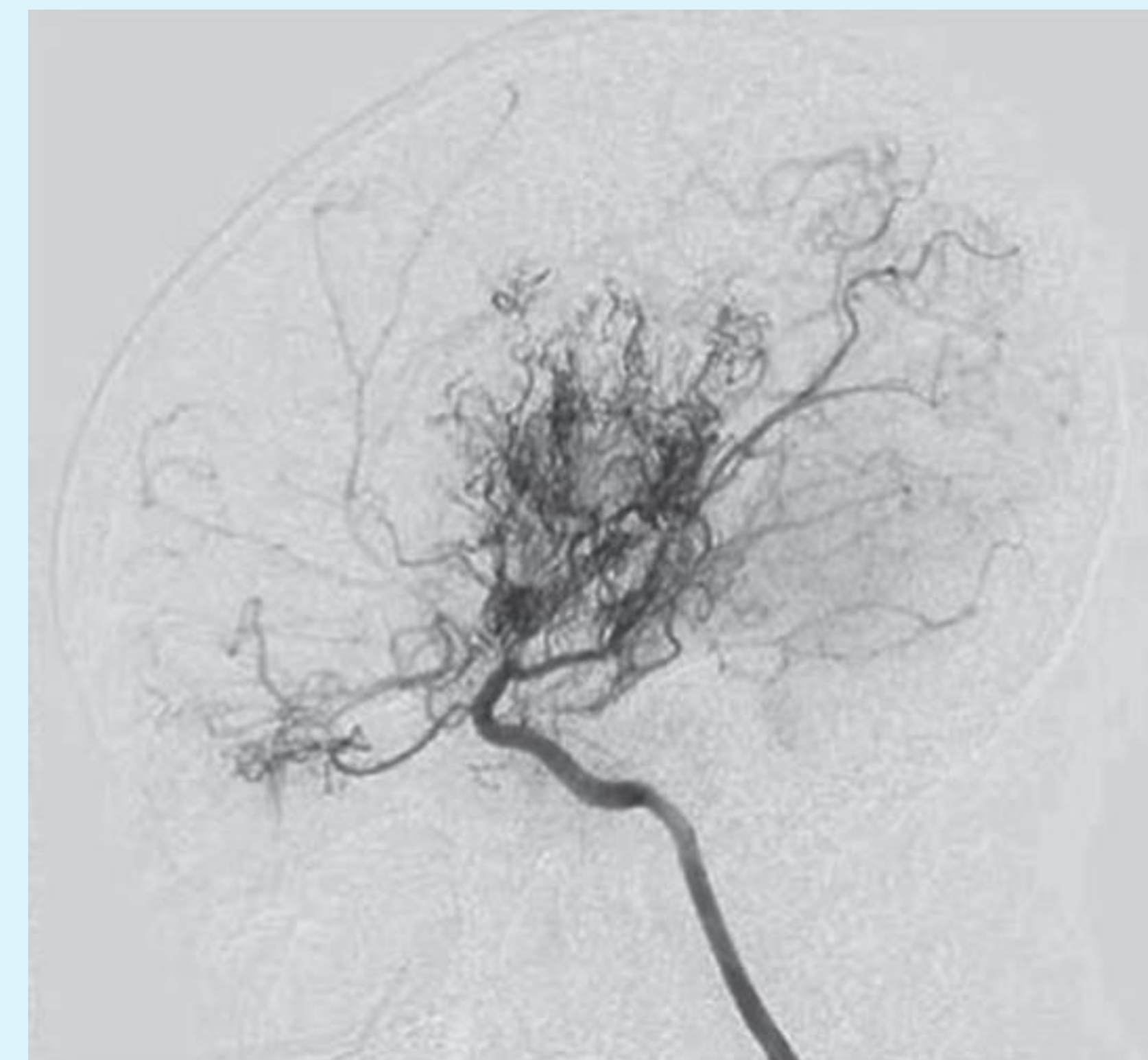
The most common clinical presentation of MMD is ischaemic, followed by haemorrhage, headaches, and seizures.

MMD has a complex pathophysiology with a range of associated genetic, molecular, and environmental factors. There is no treatment for the underlying cause, and antiplatelet therapy and surgical revascularisation procedures are the mainstay of management (1). The primary anaesthetic goal in MMD is the maintenance of CPP - this is achieved by adhering to the five normals: normotension, normoglycaemia, normocapnia, normoxia, and normothermia.

A regional technique was preferred to avoid the sympathetic stimulation of intubation during GA and allowed for continuous neurological monitoring of the awake patient.

The use of a metaraminol infusion is advantageous as it maintains MAP without increasing heart rate, thereby maintaining the same CMRO₂. This case demonstrates that a multidisciplinary approach, focusing on strict physiological targets, can result in a safe maternal and neonatal outcome.

Regional anaesthesia is beneficial for maintaining an awake patient for neurological monitoring and avoiding airway manipulation, but it carries a risk of sudden hypotension. GA is beneficial for controlling PaCO₂ and reducing cerebral oxygen demand but is associated with risk during induction and emergence.



**Carotid Angiography:
The "Puff of Smoke" Sign**

Lateral view, right carotid angiography showing Moyamoya vessels. The ACA branches are extremely narrowed.

*(Chapter 71, Surgical Treatment of MMD in Adults
(Neupsykey.com: Clinical search engine)*

