CASE REPORT

Moyamoya disease, revascularisation surgery and anaesthetic considerations

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SUMMARY

We report a case of a rare disease that requires a unique anaesthetic approach for a unique surgical procedure. Anaesthesiologists need to be familiar with different pathophysiological aspects of this disease and its perioperative management.

BACKGROUND

Moyamoya disease (MMD) is a progressive cerebral arteriopathy occluding the distal portion of the internal carotid arteries (ICAs) and/or the proximal portions of the anterior cerebral arteries and middle cerebral arteries (MCAs).^{1 2} The clinical presentation of MMD includes ischaemic or haemorrhagic stroke, transient ischaemic attack (TIA), seizure, headache and cognitive decline.³ The aetiology of MMD is unknown and medical treatment (including antiplatelet agents, anticoagulation and calcium channel blockers) has a very limited success rate.^{2 3} Surgical revascularisation has been shown to be effective in preventing ischaemic attacks in MMD.⁴ Perioperative management of these patients is challenging to anaesthesiologists and avoidance of hypotension, hyperthermia, hypocarbia or hypercarbia is crucial to maintain adequate cerebral blood flow and reduce the risk of stroke.^{3 5} The authors describe the anaesthetic management of a 49-year-old woman with MMD who presented for revascularisation surgery.

CASE PRESENTATION

A 49-year-old Hispanic female (weight 63 kg, height 155 cm, body mass index 26) with MMD was scheduled for a right-sided superficial temporal artery (STA) to MCA bypass surgery in our institution. The patient's medical history was significant for MMD diagnosed at the age of 45 and multiple episodes of TIA/infarction ever since with residual weakness in bilateral upper extremities (left>right), worsening cognitive impairment, speech disturbance and depression. She had two previous abdominal surgeries without any incident.

On the day of surgery, an intra-arterial catheter for blood pressure monitoring was inserted pre-induction; she received 1 mg intravenous midazolam and was transferred to the operating room (OR). On arrival to the OR, ASA standard monitors were applied and initial vital signs were recorded as follows: blood pressure (129/64 mm Hg), heart rate (88 bpm), respiratory rate (16 breaths/min) and oxygen saturation (95% on room air). Following preoxygenation, general anaesthesia was induced using fentanyl, propofol,

rocuronium and the trachea was intubated. Maintenance of anaesthesia consisted of sevoflurane in oxygen and air, and an infusion of remifentanyl/propofol. Sevoflurane level was maintained at 0.5 minimum alveolar concentration (MAC) throughout the entire case. Somatosensory evoked potentials (SSEP) and motor evoked potentials (MEP) were used to monitor the integrity of sensory and motor tracts function. The patient underwent a left-sided superficial temporal artery to middle cerebral artery bypass using a microscopic technique. SSEPs and MEPs remained unchanged compared with the patient's baseline. Oxygen saturation was kept at 99–100% and partial pressure of carbon dioxide (PaCO₂) was monitored with intermittent arterial blood gas tests and was maintained around 40 mm Hg; minute ventilation was adjusted accordingly to achieve this goal. At the end of the case, muscle relaxant was reversed and the patient's trachea was extubated without any incidence. Her neurological examination was unchanged and she was transferred to the intensive care unit.

DISCUSSION

First described in the Japanese population, MMD has been observed in other ethnicities with variable incidences.³ MMD has a bimodal age distribution with one peak in the first decade and the other in the fourth decade of life. The signs and symptoms of MMD vary depending on the age of patients.³ Children usually present with ischaemia/infarction in regions supplied by ICA and MCA resulting in hemiparesis, dysarthria, aphasia and cognitive impairment. Adults present with subarachnoid, parenchymal or intraventricular haemorrhage secondary to rupture of collateral arteries.^{3 5 6} Cerebral angiography is the gold standard for the diagnosis of MMD.⁶

Surgical revascularisation has been shown to be a successful method to improve the regional cerebral blood flow and reduce the risk of stroke.^{3 4} Direct revascularisation (superficial temporal artery to middle cerebral artery bypass), indirect bypasses (encephaloduroarteriosynangiosis, encephaloduroarteriomyosynangiosis, pial synangiosis) or a combination of direct and indirect procedures have been used with various outcomes.⁴

Essentially, direct bypass surgery is considered if an immediate increase of blood flow to ischaemic areas is required. Perioperative course can be complicated by ischaemic infarction, haemorrhage or infection. The principle in anaesthetic management of MMD is to maintain a balance between cerebral

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blood flow and oxygen consumption.¹ Particularly described in paediatric cases, crying and hyperventilation result in low arterial PaCO₂ and may induce ischaemia due to cerebral vasoconstriction.^{3–7} Hypercapnia is also detrimental as cerebral vasculature is maximally dilated in ischaemic areas and high PaCO₂ levels and vasodilation will further impair perfusion to already hypoperfused areas.⁸ There are no clear guidelines in the literature about anaesthetic management of revascularisation surgery in MMD. However, considering cerebral haemodynamics of MMD, avoidance of hypotension, hyperventilation and hypercarbia seems to be a reasonable approach in maintenance of CBF.⁹ Mean arterial pressure (MAP) should be kept close to the patient's baseline and PaCO₂ be maintained around 40 mm Hg. Insertion of arterial catheter before induction of anaesthesia is preferred for blood pressure monitoring during induction.^{1–7}

Intravenous induction of anaesthesia with propofol or etomidate is routinely performed followed by a non-depolarising muscle relaxant without histamine releasing properties.^{10–11} Intravenous opioids and lidocaine help to attenuate the sympathetic response to direct laryngoscopy and tracheal intubation.¹¹ The monitoring besides American Society of Anaesthesiologists (ASA) standard monitors may include electroencephalography (EEG), transcranial Doppler, somatosensory evoked potentials (SSEP) or near infrared spectroscopy (NIRS) according to the institutional experience.^{1–12}

Sato *et al* compared TIVA with volatile anaesthetics for maintenance of anaesthesia and concluded that the regional cortical blood flow might be decreased with volatile agents.¹³ Adachi *et al*¹⁴ showed an improved outcome using TIVA, whereas other studies have not shown a significant difference between TIVA and volatile agents.¹⁵ We used SSEP during the surgery and opted to use total intravenous anaesthesia (TIVA) with propofol and remifentanyl infusion combined with low doses of sevoflurane (0.5 MAC) to avoid interference with neurological monitoring.

There is no consensus supporting a desirable haematocrit level in patients with MMD. However, a haematocrit greater than 30% seems to be sufficient to minimise the risk of perioperative ischaemic events.¹⁶

Postoperative care of MMD patients undergoing revascularisation surgery preferably will take place in the intensive care unit.¹⁷ They need to be closely monitored for any haemodynamic and neurological changes as postoperative ischaemic events have been attributed to haemodynamic swings.^{1–7–17} Intravenous fluid administration to maintain normovolemia and starting aspirin on the first postoperative day have been recommended.¹²

In conclusion, there are increasing numbers of patients with MMD requiring anaesthesia either for diagnostic studies or revascularisation surgery. Therefore, it is important that anaesthesiologists practicing in tertiary care centres make themselves familiar with the pathophysiology of MMD, the surgical techniques and their anaesthetic implications.

Learning points

- Unique pathophysiology of moyamoya disease mandates anaesthesiologists to formulate an individualised anaesthetic plan for these patients.
- Perioperative management of moyamoya disease has a direct impact on the outcome of surgery.
- Perioperative hyperventilation/hypoventilation and hypotension should be avoided to reduce the risk of further neurological impairment.

Contributors All authors contributed to patient care and preparation of this manuscript equally.

Competing interests None.

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