CASE REPORT

Anaesthetic Considerations for Interventional Neuroradiology in a Child with Moyamoya Disease - A Case Report

Gurnal P[1], Sivapurapu V[1], RanA L[2], Sharma P[1], Banik P[1]

[1]Department of Anaesthesia and Critical Care, All India Institute of Medical Sciences (AIIMS), Bilaspur, India.
[2]Department of Radiology, All India Institute of Medical Sciences (AIIMS), Bilaspur, India.

ABSTRACT **Introduction:** The Moyamoya disease is a chronic condition involving cerebrovascular system characterized by progressive stenosis and occlusion or narrowing of the terminal internal carotid arteries.

Case Presentation: Patient with severe cerebrovascular impairment with consequent cerebral ischemia and haemorrhage poses significant challenge due to compromised cerebral blood flow. The role of an anesthesiologist is particularly challenging in interventional neurological procedures such as revascularization as well as in digital subtraction angiography. These procedures require meticulous management of cerebral perfusion, hemodynamic stability and oxygenation to prevent ischemic complications or further deterioration in patients with pre-existing cerebral perfusion impairment.

Conclusion: We report a case of 11year male child with Moyamoya disease with right sided paresis who presented to us for digital subtraction angiography (DSA) under general anaesthesia.

Keywords: Moyamoya disease, Anaesthetic considerations, Interventional Neuroradiology.

Correspondence: Dr Gurnal Pooja, Assistant Professor, Department of Anaesthesiology and Critical Care, AIIMS Bilaspur (HP), India.
Pin Code:174001
Email ID: poojagurnal2@gmail.com

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INTRODUCTION

Moyamoya Disease (MMD) is a cerebrovascular arteriopathy of unknown cause, first described in 1957, characterized by progressive narrowing and eventual occlusion of the distal intracranial carotid arteries and branches i.e. middle cerebral artery and anterior cerebral artery and predisposing the patient to episodes of cerebrovascular insufficiency.[1,2] The collateral vessels develop from the leptomeninges, external and intracranial internal carotid artery to perfuse the ischemic brain beyond occlusion and the dilated basal collaterals from the intracranial internal carotid artery's supply various skull base structure creating a characteristic "puff of smoke" angiographic appearance.[3] The risk of cerebral ischemia is increased due to disrupted autoregulation of the cerebral blood flow with the precise blood pressure thresholds for autoregulation remaining undefined in paediatric population.[4,5] Despite diagnostic imaging with magnetic resonance imaging(MRI) or computed tomography(CT) suggesting MMD, digital subtraction angiography (DSA) is often required for definitive confirmation. The anaesthesiologist while providing anaesthesia to the patient with MMD during DSA must carefully manage the monitoring strategies, anaesthetic techniques and drug administration. We present the challenging management of a case of an 11-year-old child having cognitive decline, hemiparesis due to right middle cerebral artery territory infarct on MRI requiring confirmation of diagnosis by DSA.

CASE PRESENTATION

A 11-year-old male child weighing 19.5Kg presented with history of weakness of left upper and lower limb of body and slurring of speech for 15 days. There was a past history of generalized tonic-clonic seizures at 4 years of age for which he took tab sodium valproate for 3 years and stopped.

On examination, his vitals were stable. Systemic examination was unremarkable except decreased motor power on left lower and upper limb. All blood

investigations were within normal limits. Cerebrospinal fluid examination was normal. MRI brain was consistent with right-sided Middle cerebral artery and Anterior cerebral artery territory infarct. [Figure 2] Provisional diagnosis of MMD was made. The child was started on Tab Aspirin 75 mg OD and Tab sodium valproate 300mg BD and DSA was planned for confirming the diagnosis. No sedatives were advised in preoperative period. Child was taken for intervention to be done under general anaesthesia. After commencing electrocardiography, non-invasive blood pressure and pulse oximetry monitoring, child was induced with injection fentanyl 2 mcg /kg, injection propofol 2mg/kg, injection atracurium 0.5mg/kg intravenously and was intubated with a 5mm (internal diameter) endotracheal tube. Maintenance of anaesthesia was done with O_2, N_2O and isoflurane. Vitals remained unremarkable throughout the procedure. After the completion of one and half hour long procedure in the catheterization laboratory, child had tonic clonic seizures which were controlled with injection midazolam 1mg intravenously followed by injection propofol 20mg intravenously followed by Injection phenytoin infusion 500mg in 100ml normal saline. Child was transferred to the intensive care unit (ICU) for further monitoring and management. Child was kept sedated by means of injection fentanyl infusion2µ/kg/hour and put on ventilator overnight on synchronized intermittent mandatory ventilation (SIMV) mode. Patient was extubated the next day continuing with tab. sodium valproate. The child remained seizure free thereafter, was discharged in a stable condition and has been advised to undergo neurosurgical intervention.

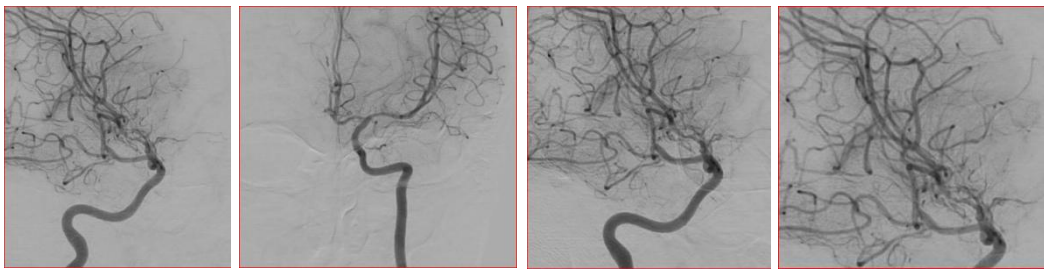

Figure 1. DSA shows presence of short segment stenosis in bilateral M1 segments with collaterals formation (Images A-D).

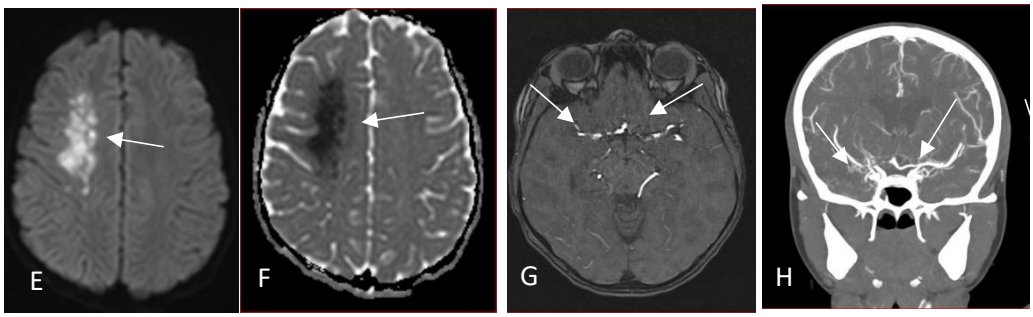

Figure 2. MRI (E-DWI, F-ADC and G-MRA) and CT angiographic (H) images of the patient showing presence of occlusive changes in bilateral (Right >Left) as shown by white arrows. The MR images E and F shows presence of infarct in right MCA territory region of brain.

DISCUSSION

There are no definitive guidelines on anaesthetic management of MMD patients. Goal remains to prevent any ischaemic episodes perioperatively. Maintaining normovolemia, normocapnia, normothermia are the mainstays of management. Some studies have recommended total intravenous anaesthesia(TIVA) for revascularization procedures on the basis of regional blood flow because inhaled anaesthesia can provoke intracerebral steal in MMD. However, Adachi et al.[6] in their study found no significant difference in postoperative complications in both types of anaesthesia. Hypocapnia can induce brain ischemia and brain infarction, while hypercapnia can induce vasodilation and hyper-perfusion of the fragile vessels in the brain, which might cause brain haemorrhage. In patients with MMD, both vasoconstriction and vasodilation should be avoided. That is why keeping normocapnia is quite important.[7] Recovery period after neurosurgical procedures remains a time of potential danger to patients given the high incidence of postoperative complications independent of the anaesthetic strategy.[8] Hypocapnia-induced seizure and neurological worsening has been reported in a child with MMD during postoperative period.[9] It is difficult to prove the association between hypocapnia and seizure in our patient as $ETCO_2$ was maintained within normal limits, i.e., 35-40mmHg throughout the procedure.

Niu et al.[10] analysed MMD patients to investigate the causes of early postoperative seizures after revascularization procedures. They found that cerebral infarction before surgery and new infarction after surgery were the risk factors for seizures. One mechanism is extravasation of blood vessels. Iron

released from haemoglobin reacts with hydrogen peroxide in surrounding tissue to produce free radicals which leads to seizures. Exposure to free radicals leads to formation of epileptic foci. Other mechanism is ion imbalance due to ischaemia or hypoxia. In our patient, there was bilateral stenosis in M1 segment of middle cerebral artery (MCA) and infarct was present in the middle cerebral artery region which was a risk factor.

Choi et al.[11] found association of seizure with hypoperfusion. Hyperperfusion of brain tissue after reperfusion surgery can itself cause transient neurological dysfunction and seizures.[12] Regarding routine use of antiseizure prophylaxis, there are no definite guidelines but it may be considered if there are known risk factors like prior history of seizures, infarction or aneurysm in middle cerebral artey.[13] Our patient was on sodium valproate therapy which continued till day of intervention and continued after the procedure during ICU stay and at the time of discharge from hospital also.

Maintenance of normotension is another recommendation to prevent ischaemic insult. However, hypotension was not associated with ischaemic complications in a study by Iwama et al.[14] Recently, autoregulation monitoring with infra-red spectroscopy derived indices was used for finding optimal blood pressure range in children with MMD. Optimal blood pressure with best autoregulation was found to be around 60-80mmHg.[15] We had maintained normotension throughout the procedure, though invasive blood pressure monitoring should have been done to have continuous monitoring of blood pressure instead of intermittent non-invasive monitoring.

There may be some role of EEG monitoring during neurosurgical intervention in MMD patients as it can predict and possibly prevent perioperative ischaemic events.[16] These cases become particularly challenging as they are performed outside operation theatre requiring meticulous preparation and vigilance so that any complication arising could be handled appropriately. We had well equipped catheterization laboratory at par with operation theatre so we were able to manage the case even when complication arose in the form of seizure at the end of the procedure.

CONCLUSION

A good understanding of the pathophysiology of MMD favours a well-planned perioperative strategy for successful management of MMD patients. The crux of the management in MMD remains maintenance of normotension, normocapnia, normovolemia and normothermia throughout the procedures. Because there is high propensity of seizures and neurological worsening during intervention and after the procedures, patients should be monitored in well-equipped intensive care units / recovery units instead of the wards so that, patient can be managed in a timely manner when such complications arise.

Conflict of interest: None.

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