Anesthesia management of an infant with moyamoya disease posted for encephaloduroarteriomyosynosteosis procedure

Dear Sir,

Moyamoya disease is a chronic, usually bilateral, vasculopathy with progressive narrowing of the terminal intracranial portion of the internal carotid artery and circle of Willis leading to decreased blood supply to the brain resulting in chronic ischemia characterized by symptoms of transient ischemic attacks and stroke.[1,2] The usual presentation of moyamoya disease occurs in bimodal peaks, in the pediatric age group at 5–9 years of age and in adults at 35–39 years of age.[3] This disease has a rare occurrence in the infant population so nothing much is described in the literature about anesthetic management of moyamoya in infants.[4] We, hereby, discuss the anesthetic management of moyamoya disease in an infant posted for encephaloduroarteriomyosynosteosis procedure (EDAMS).

An 11-month-old female, weighing 6 kg, was posted for an EDAMS procedure for moyamoya disease. The parents gave a history of multiple episodes of sudden clonic movements of the right upper limb (lasting for 1–2 min) till the seventh month of life. Magnetic resonance imaging (MRI) showed severe attenuation of the bilateral supraclinoid internal carotid artery and middle cerebral artery with A1 segment of the bilateral anterior cerebral artery with evident multiple lacunar chronic infarcts, encephalomalacia, and gliosis in bilateral cerebral hemispheres with multiple thalamo-perforating and lenticulostriate collaterals suggestive of moyamoya disease [Figure 1]. Routine investigations including complete blood count, coagulation profile, serum electrolytes, and renal function tests were normal. Her 2D echocardiography was normal. On examination, she also had microcephaly with fused fontanelles and overriding of sutures. The power was 4/5 in all limbs with increased tone in the right upper limb and lower limb and deep tendon reflexes of the right side were hyperactive grade +3. She was on tablet aspirin (25 mg), syrup clonazepam, and syrup levetiracetam.

On the day of surgery, she was calm and sleeping due to the morning dose of syrup clonazepam. She was shifted to the operation theater while in a state of sleep and there was no requirement of added sedation. All monitors were attached with pulse oximetry, electrocardiography, noninvasive blood pressure (BP), and nasopharyngeal temperature probe. She was induced with inj. fentanyl 12 mcg, inj. lignocaine (to avoid intubation response), inj. propofol 15 mg, and inj. atracurium 3 mg. Maintenance was done with O_2/air (50:50) and sevoflurane to keep a minimal alveolar concentration (MAC 0.8). 4F triple lumen central line was inserted in right-sided internal jugular vein. Left radial artery was cannulated with 24-G arterial line for continuous BP monitoring. The vital parameters were maintained with mean arterial pressure (MAP) targeted above 55 mmHg. End-tidal carbon dioxide was maintained between 28 and 30 mmHg throughout the procedure. She was extubated smoothly at the end of the procedure and was shifted to neonatal intensive care unit for close observation.

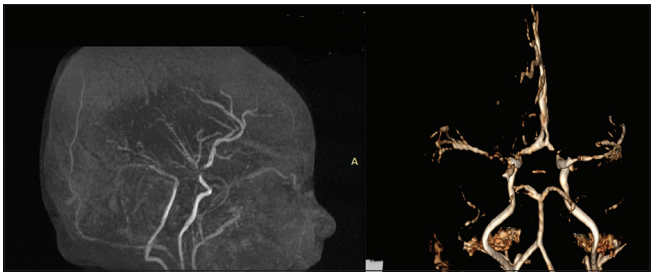

Figure 1: MRI image showing severe attenuation of bilateral supraclinoid internal carotid artery and middle cerebral artery

The goals of anesthesia in these surgeries included preservation of perfusion in areas prone to chronic ischemia by the maintenance of normothermia, normocarbia, normotension, prevention of hypoxia, and normal hematocrit.[5] Reduction in cerebral blood flow is less tolerated in pediatric patients than adults because children have a higher cerebral metabolic rate of oxygen consumption ($CMRO_2$), hence they develop ischemia even if the cerebral blood flow is marginally reduced.[6] Any factor that reduces the cerebral blood flow like reduction in MAP or rise in intracranial pressure (ICP) should be specifically avoided in these patients as blood flow is already compromised. In our patient, the margin was even lesser due to the presence of fused sutures and microcephaly. Rise in ICP at various stages was prevented by shifting the child to OT in a sedated state, using lignocaine IV for attenuation of intubation response, smooth extubation,

and good analgesia in the postoperative period. Invasive BP monitoring is required in such cases to continuously monitor for mean BP.

To conclude, avoidance of factors leading to hypoxia and hyperventilation (crying, agitation), maintenance of cerebral blood flow, and normothermia are the keys to successful anesthetic management of moyamoya disease in the infant population.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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