Cerebral autosomal dominant arteriopathy with subcortical infarct and leukoencephalopathy: A rare syndrome raising anesthetic concerns!

Sir,

We report the case of a 22-year-old man, weighing 53 kg, with a probable diagnosis of Cerebral autosomal dominant arteriopathy with subcortical infarct and leukoencephalopathy (CADASIL), made by a neurologist who was scheduled for a brain biopsy for the confirmation of diagnosis. Earlier, he presented in the neurology outpatient department with a history of seizures since 4 years, imbalance while walking, change in behavior and personality, and forgetfulness since 2 years, as well as change in speech since 6 months. His history was significant for delayed mental milestones and normal motor milestones. Neurological examination was significant for increased muscle tone in all limbs, exaggerated deep tendon reflexes, and dorsi flexed plantar response. Magnetic resonance imaging of the brain revealed bilateral cerebellar and cerebral atrophic changes, with bilateral cerebral white matter hyperintensities.

We reviewed anesthetic literature for the management of CADASIL-type arteriopathy. An isolated report by Dieu and Veyckemans[1] described anesthetic management in a 30-year-old female patient with CADASIL, who underwent laparoscopy. Our patient received intramuscular glycopyrrolate 0.2 mg as premedication one hour prior to surgery. General anesthesia was induced with propofol 80 mg and fentanyl 100 μg intravenously. Tracheal intubation was facilitated with rocuronium 1 mg/kg. Anesthesia was maintained with isoflurane and nitrous oxide in oxygen. Intraoperatively, the mean blood pressure was kept around 80 mm Hg and normocapnia was maintained. Intraoperative course was uneventful and the trachea was extubated at the end of surgery after reversal of the neuromuscular block. A week later, the brain biopsy report confirmed the diagnosis.

The CADASIL syndrome is a rare inherited neurological condition resulting from non-atherosclerotic and non-amyloidotic microangiopathy. The clinical features of this syndrome include migraine, cognitive problems, seizures, psychiatric problems, and dementia accompanied by difficulty in walking, urinary incontinence, and pseudobulbar syndromes. The anesthetic considerations are similar to patients with cerebrovascular disease, which include maintenance of normocapnia and mean arterial blood pressure within the autoregulation range. Considering altered cerebral physiology in patients with CADASIL syndrome, Dieu and Veyckemans[1] suggest use of isoflurane for maintenance of anesthesia, norepinephrine for hypotension, and nimodipine for hypertensive episodes. Our patient had an uneventful intraoperative and postoperative course, thereby suggesting appropriate anesthetic management.

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Failure in properly checking equipment is a factor in many critical incidents. Proper checking of equipment can help prevent equipment-related morbidity and mortality, improve preventive maintenance, and educate the anesthesia provider about equipment. Unfortunately, failure to perform a proper check before use is common. We encountered an unusual foreign body in the breathing circuit and felt the need to re-emphasize the importance of the cockpit drill before induction of anesthesia.

There are numerous reports of obstruction of anesthesia breathing circuits by different foreign bodies. However, we did not come across literature reporting breathing circuit obstruction caused by a cap of urobag tubing. While checking the anesthesia machine and breathing circuit, it was noticed that intra-circuit pressure did not decrease on opening the end of elbow connector to the atmosphere. On thorough inspection, no source of obstruction could be found, but on turning the circuit upside down and removing the elbow connector, a foreign body, which was, in fact, a cap of catheter end of urobag tubing, was noticed. It was in the Y-piece of breathing circuit with the open end abutting the bifurcation and blind end in the straight limb [Figure 1]. On initial inspection, the foreign body appeared as a continuation of the elbow connector because of the same color. It took some time to find the exact site of obstruction, which could have been sufficient to cause hypoxia. The exact reason for this unusual occurrence could not be determined. It was the first case on that day; moreover, on the previous day, three cases were managed in the same operation theatre uneventfully using the same circuit. One hypothesis is that nontechnical staff of the operation theatre, while cleaning at the end of the previous day, might have misconnected the urobag cap thinking it to be a part of the breathing circuit. A foreign body lodged in the elbow connector (a glass ampoule fragment after it was used to open a propofol ampoule) causing an impossible ventilation has been reported earlier.

Although, the exact cause of this unusual incident could not be concluded, but its timely detection definitely avoided a potential catastrophe. We recommend that readers thoroughly follow the pre-use checkout guidelines to prevent critical incidents and improve patient safety. Lastly, the nontechnical staff of the operation theatre should be educated and trained about isolation and disposal methods of the items such as cap of urobag tubing or similar items, which have the potential to cause circuit obstruction.

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