Anesthetic management of a case of neurodegeneration brain iron accumulation (Hallervorden–Spatz disease)

Madam,

Neurodegeneration brain iron accumulation (NBIA) is characterized by dementia, dystonia, dysphagia, dysarthria, muscular rigidity, and parkinsonism-like extrapyramidal features.\[1\]-\[3\] The anesthetic concerns include difficult airway management, increased risk of gastric aspiration, dehydration, and postoperative respiratory and renal insufficiency.\[2\],\[3\] We describe the successful management of such a patient who came for dental extraction.

A 26-year-old, 32 kg, female, a known case of NBIA presented with ulcerations of lower lip and oral cavity wounds. On examination, the patient was bed-ridden, mute, not following any command, had impaired cognition, and dystonic posture of head, trunk, and limbs. There was generalized wasting of muscle, flexion deformity of knees, and equinovalovar deformity of both feet. The patient had spastic quadriaparesis with lower limb involvement more than the upper limb. Teeth were irregular and protruding. Patient also had torticollis and was not able to open her mouth.

Investigations such as complete blood count, kidney function, liver function, urine analysis, blood sugar, serum electrolytes, X-ray chest, and electrocardiogram (ECG) were within normal limits. Magnetic resonance imaging (MRI) of the brain showed bilateral T2-weighted hypointense lesion in the bilateral basal ganglia. Her treatment consists of limb physiotherapy, tablet baclofen to decrease rigidity, tablet pacitane for Parkinson-like symptoms, and syrup diphenhydramine to decrease secretions and for sedation. She was accepted for dental extraction under ASA grade III, and her attendants were explained about the high risk involved in the surgery and the possibility of postoperative elective ventilation. All the medications were continued till the morning of the surgery, along with syrup ranitidine 150 mg, syrup ondansetron 2 tablespoons for aspiration prophylaxis.

In the operation theatre, the patient was positioned supine in her natural posture with adequate padding at pressure points and monitors were attached. Her baseline pulse, blood pressure, respiratory rate, \( \text{SpO}_2 \) on room air were 96/min, 138/90, 22/min, and 98%, respectively. She was premedicated with Inj. glycopyrrolate 0.2 mg. Nasal drops 0.1% xylometazoline were instilled in both nares as we planned nasal intubation. No sedative/narcotic was given. Airway management cart and tracheotony was kept ready before inducing the patient. She was induced with 4% sevoflurane in oxygen-nitrous oxide mixture in a ratio of 1:1. Mask ventilation was done and airway was maintained easily. Mouth opening and oromandibular rigidity improved as the depth of anesthesia increased. Nasal intubation was done with high volume, low pressure, cuffed, armored endotracheal tube no. 6.5 followed by oropharyngeal packing. Anesthesia was maintained with sevoflurane 1–2% in oxygen-nitrous oxide mixture in a ratio of 1:1 and neuromuscular blockade was avoided. The dental surgeon was asked to give local anesthesia with 2% lignocaine with adrenaline for the teeth required to be extracted. Inj. diclofenac sodium 50 mg was given as a pre-emptive analgesia. Intraoperative monitoring of NIBP, HR, ECG, \( \text{SpO}_2 \), and EtCO\(_2\) was done which remained within normal limits throughout the procedure. Surgery was uneventful. After thorough suctioning of oropharynx, pack was removed and sevoflurane and nitrous oxide was switched off. Oxygenation with 100% \( \text{O}_2 \) was started and the patient was allowed to breathe spontaneously. Extubation was done after ensuring adequate tidal volume and regular respiration. Patient was shifted to the postanesthesia recovery room with oxygen via facemask. In the recovery room, patient was monitored for 2 h and then shifted to the ward. No episodes of vomiting and airway obstruction were noticed postoperatively and she was discharged next day from hospital.

Preanaesthetic assessment is very important in such patients. It is difficult to assess respiratory and cardiovascular systems due to abnormal posture, hyper-rigidity, and mental retardation. Difficulty in feeding makes the patients malnourished. There is a risk of aspiration and chest infection leading to compromised gas exchange. Patients are usually on long-term benzodiazepines, so it can be used for premedication on the day of the surgery along with antiemetic as prophylaxis for vomiting.\[2\],\[6\]

Involuntary movements can make intravenous cannulation difficult. Mouth opening is limited due to muscular rigidity of jaw muscles. Scoliosis, joint rigidity, and muscle contracture may fix the cervical spine. This can make endotracheal intubation difficult. Facility for emergency tracheotomy should be at hand. Awake intubation can be difficult due to lack of cooperation from patient and a noxious stimulus may aggravate dystonia.\[2\],\[6\]

Induction agents such as thiopentone and propofol have been used safely by Keegan et al. and Balas et al.\[2\],\[3\],\[6\]
Succinylcholine should be avoided due to immobility, leading to possibility of hyperkalemic cardiac arrest. Nondepolarizing muscle relaxants and inhalation agents such as halothane and sevoflurane can be used safely. In our case, rigidity decreased as the depth of anesthesia increased with sevoflurane and nasal intubation could be easily done. Regional anesthesia can be difficult due to deranged mental state, scoliosis, and rigidity.[2‑6]

Postoperatively, extubation should be done only when all extubation criteria are met. Longer observation and oxygen therapy may be required in the postoperative period because re-intubation may be required due to muscular rigidity and respiratory disability. Risk of postoperative aspiration and compromised gas exchange should be kept in mind.[2‑6]

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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