Anaesthetic management of a child with Lesch Nyhan syndrome

Sir,

Lesch-Nyhan (LN) syndrome, is a X-linked-recessive-disorder affecting predominantly males, caused by deficiency of hypoxanthine-guanine-p hosphoribosyl-transferase (HGPRT) an important enzyme in the uric acid metabolism. The syndrome is characterised by self-aggression, self-mutilation, choreoathetosis and intellectual abnormalities.[1] Severe deficiency of HGPRT in Grade-4 LN syndrome leads to the accumulation of uric acid crystals in different organs and is characterised by acute generalised dystonia, behaviour alterations such as self-aggression, seizures and cognitive impairment and therefore presents with multiple anaesthetic challenges.[1,2]

We report case of a 3-year-old boy, 8.5 kg, diagnosed clinically with Grade-4 LN-syndrome having cerebral palsy, dystonic movement, history of self-biting and posted for dental wiring under general anaesthesia in a tertiary care hospital in New Delhi. Preoperative evaluation revealed malnourishment, multiple injuries in upper and lower lips due to self-biting leading to lip scarring, injuries in fingers and hand. Developmental delay was present; could not hold neck or sit with support. Airway examination could not be done as the child was irritable. Blood investigations including uric acid levels were within normal limits.

ECG, NIBP, Pulse oximeter and skin temperature probe were attached for monitoring the vital parameters. 24-gauge IV cannula was secured in the right-lower limb due to difficult cannulations in the upper extremities because of mutilation. Injection atropine 0.1 mg and injection fentanyl 15 μg, injection metoclopramide 1 mg i.v were given as premedication. Child did not allow preoxygenation. Induction of anaesthesia was done by IV propofol 20 mg.

After assuring adequate ventilation with mask attached with 100% oxygen, injection rocuronium 8 mg iv was given to facilitate tracheal intubation. A 3.5-mm ID non-cuffed endotracheal tube was used to intubate the trachea nasally assisted by direct laryngoscopy, capnography, ETO_2 and was fixed at 13.5 cm. Bilateral equal air entry in the chest was ensured by placing a precordial stethoscope. Anaesthesia was maintained by intermittent positive pressure ventilation with JR circuit providing mixture of oxygen, air and isoflurane to maintain a MAC value of 0.9 to 1%. Padding of the extremities and pressure points were done. Intraoperatively vitals were stable. Injection paracetamol 100 mg IV infusion was given for pain relief. Injection dexamethasone 1 mg IV was given to prevent airway edema postoperatively as well as to decrease nausea vomiting. Surgery lasted for an hour, top-up doses of rocuronium were not required. At the end of the surgery spontaneous efforts returned, residual neuromuscular block was reversed using injection neostigmine 0.4 mg and injection glycopyrrolate 0.08 mg IV. Trachea was extubated after patient was awake with adequate motor power. Child was transferred to the HDU (high dependency unit) with oxygen mask in propped-up position; patient was monitored closely for apnea, bradycardia and aspiration. Remaining hospital stay was uneventful.

Encountering a grade-4 LN-syndrome patient for procedures under general anaesthesia is rare. It is important to divert the history-taking and examination to assess onset, severity, organ impairment including CNS (seizure), liver, renal dysfunction and airway abnormalities. For anaesthetic management, in addition to routine preparations special precautions were taken in this case (1) preparation of difficult paediatric airway cart and tracheostomy; (2) pre-warming the operating room, administering warm IV fluids, keeping over-head warmers ready to prevent hypothermia (3) padding of pressure points to prevent injuries.

Choice of anaesthetic drugs is critical in these children due to the challenges related to specific pharmacokinetic alterations in the disease. Injection atropine was used as a premedication to prevent sudden bradycardia and metoclopramide was used as a prokinetic agent to prevent pulmonary aspiration.[3] Despite previous reports of pain-insensitivity, it is currently indicated that mutilation is not due to lack of sensation, but is more of an obsessive-compulsive behaviour, with the child experiencing pain similar to other normal children.[4] Therefore, we used fentanyl as a premedication for pain-relief. Propofol was used as the induction agent as it has anti-emetic and uricosuric properties.[5] This was used under careful monitoring for bradycardia. Metabolism of barbiturates, ketamine, etomidate remains unchanged in these patients and can also be used safely.[3] Since mask ventilation was possible,

8 mg rocuronium was given to secure airway early, to avoid pulmonary aspiration. Rocuronium is excreted in the biliary system prevents renal impairment and is safe to use. Succinylcholine is reported to cause abnormal potassium influx and best avoided.[3] Although laudanosine-induced-seizures are unlikely in such short-duration surgical procedures, yet care must be taken while using atracurium in this group of seizure-prone patients.[3,6] NSAIDS were avoided to prevent any renal dysfunction. IV dexamethasone injection and avoidance of nitrous oxide helped to prevent nausea, vomiting and probable pulmonary aspiration.

Though the procedures may not be novel, but the careful set of precautions specific to this case and the judicious use of drugs under vigilant monitoring prevented any complications. The key points are: (1) Preoperative-anaesthetic evaluation with attention to oral anatomy, airway, renal function (2) using anaesthetics with low-risk of seizure; uricosuric properties and renal-safe (3) monitoring to prevent pressure injuries, sudden bradycardia, asphyxia, pulmonary aspiration and sudden death both intra and post-operatively.

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.

Shraya Banerjee, Sunil K Sinha, Preeti G Varshney, Pragya Ahuja

Department of Anaesthesia, Lady Hardinge Medical College and Smt. SK Hospital, New Delhi, India

Address for correspondence:
Dr. Shraya Banerjee,
A128, Second Floor, Malviya Nagar, New Delhi - 110 017, India.
E-mail: drshreyalhmc@gmail.com

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