

Anesthesia in a child with Kinsbourne syndrome: Does anesthesia technique matters?

ABSTRACT

Kinsbourne syndrome is a rare neurological paraneoplastic syndrome associated with neuroblastic tumors. There are very few literatures on its anesthetic management and interaction with anesthetic agents. The epileptogenic potential of certain anesthetic agents such as ketamine, etomidate, and meperidine might trigger opsoclonus and myoclonus and have an impact on the long-term neurological outcome. The objective of this case report is to discuss the safety of anesthetic agents and their relationship in a patient with Kinsbourne syndrome. We discuss our experience in the anesthetic management of a child with Kinsbourne syndrome with ganglioneuroblastoma in the thoracic paravertebral space.

Key words: General anesthesia, kinsbourne syndrome

Introduction

Opsoclonus–myoclonus syndrome (OMS) also known as dancing eye syndrome or Kinsbourne^[1] disease as first described by Kinsbourne in 1963 is an extremely rare paraneoplastic syndrome associated with neuroblastic tumors.^[2] Incidence has been reported to be 0.03-0.18 cases per million total population.^[2] About 2-4% of neuroblastoma patients develop OMS^[3] and at least in about half of the patient with OMS, an underlying neuroblastoma will be found.^[2,4,5] It presents with opsoclonus, myoclonus, ataxia, and behavioral abnormalities such as sleep disturbances, irritability, developmental delays, decreased social interaction, lethargy, mutism, and visual disturbances.^[6] Anesthesia management of OMS is sparsely reported in literature and the objective of this case report is to discuss the safety of anesthetic agents in Kinsbourne syndrome and our experience in the anesthetic management of such a child with ganglioneuroblastoma.

Case Report

An 18-month-old male child weighing 14 kg presented with opsoclonus, myoclonus, and ataxia since 11-month-old. Magnetic resonance imaging of thorax and abdomen revealed a soft tissue mass in the left paravertebral region (D6-D10) and computed tomography-guided biopsy showed ganglioneuroblastoma, and the diagnosis of OMS was confirmed. He was started on adrenocorticotrophic hormone (ACTH) therapy and developed cushingoid features. After the neurological symptoms decreased, he was posted for surgical excision of the mass 7 months after the onset of OMS. During the preoperative examination, the child was active and playful with cushingoid features. Opsoclonus with occasional myoclonus and ataxia on walking were evident. Airway examination showed normal mouth opening, large tongue, and restricted neck extension with buffalo hump. Cardiopulmonary examination and laboratory parameters were unremarkable.

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NISA N, TALAWAR P, VASUDEVAN B

Department of Anesthesia, Critical Care and Pain Medicine, All India Institute of Medical Sciences, New Delhi, India

Address for correspondence: Dr. Neisevilie Nisa, Room No. 603, Masjid Moth Resident Doctors Hostel, All India Institute of Medical Sciences, New Delhi, India. E-mail: neisevilie@gmail.com

On the day of the surgery, the child was premedicated with syrup midazolam and inhalation induction was done with oxygen, nitrous oxide, and sevoflurane. Routine American Society of Anesthesiologists monitors were attached and after attaining adequate depth, two peripheral intravenous lines were secured. Fentanyl 30 mcg and atracurium 8 mg were given intravenously, and airway was secured with endotracheal tube of 4.5 mm internal diameter. Right radial artery was cannulated for monitoring invasive blood pressure, blood sugar, and arterial blood gas analysis. Caudal anesthesia was given with 15 ml of 0.125% preservative-free bupivacaine and 750 mcg morphine. Intraoperative analgesia was supplemented with intravenous paracetamol 250 mg and fentanyl boluses of 10 mcg based on the hemodynamic response. Blood loss was around 80 ml and replaced with 300 ml of ringer acetate. At the end of surgery, the patient was warm, hemodynamically stable, neuromuscular block was reversed, trachea was extubated, and the child was shifted to Postanesthesia Care Unit. Opsoclonus was present during inhalational induction, but disappeared once adequate depth was attained and there was no episode of opsoclonus or myoclonic jerk during the perioperative period. Caudal morphine was adequate for postoperative analgesia in the first 24 h along with intravenous paracetamol 250 mg every 6 h and later, fentanyl and paracetamol was given intravenously targeting a face, leg, activity, cry, consolability scale of <2. The postoperative period was uneventful, and the child was discharged 5 days later continuing on intramuscular ACTH and follow-up for the neurological symptoms was advised. The patient was doing well, but the neurological symptoms were still persistent with decreasing frequency at the time of writing this report.

Discussion

Currently, there are no data available supporting that surgical resection of neuroblastoma improves the neurological outcome of OMS,^[3] but prompt and potent immunosuppression with steroids, ACTH, immunoglobulins (IgGs), plasmapheresis, azathioprine, rituximab, mycophenolate, and cyclophosphamide are necessary to avoid the poor neurological prognosis.^[6] An autoimmune response between antibody produced against tumor antigen cross-reacting with central nervous system structures, mainly cerebellum and brain stem has been postulated as the main pathophysiology,^[6,7] and several antibodies such as anti-Hu, anti-Ri, and anti-Yo have been described and newer antibodies such as anti-neurofilament, anti-purkinje, and IgG autoantibodies have been isolated.^[6] An international consensus has been reached that three out of four criteria are required to diagnose the classical syndrome:

1. Myoclonus,
2. Opsoclonus,
3. Ataxia, and
4. Neuroblastoma.^[4]

The effect of anesthesia on OMS has not been studied in detail due to the rarity of the disease and three case reports on anesthesia management have been reported till date.^[8-10] Burrows and Seeman^[8] successfully administered anesthesia in a child with a kidney tumor and Kinsbourne syndrome using morphine, pancuronium, and nitrous oxide. They reported worsening of myoclonus and opsoclonus when intramuscular ketamine was used in the same patient for sedation during myelogram. Maranhão *et al.*^[9] used opioids, inhalational agents, nondepolarizing muscle relaxants (NDMRs), neostigmine, and ketorolac in a similar child without worsening of the symptoms. Lee *et al.*^[10] used propofol and remifentanyl for total intravenous anesthesia and noticed that opsoclonus and myoclonus disappeared once the target concentration was achieved.

In our patient, ACTH therapy was started 3 months before surgery and opsoclonus and myoclonus were fairly controlled during the preoperative evaluation and disappeared after induction. Whether anesthetic agents with epileptogenic potential really triggers the symptoms of OMS is controversial because an autoimmune reaction is the most acceptable explanation for the symptoms and not due to lowering of seizure threshold and so future studies are warranted for a better understanding of their interaction with anesthetic agents and their effect on the long-term neurological outcome.

We used a balanced anesthesia with inhalational induction supplemented by fentanyl, atracurium, paracetamol, and caudal block. We did not consider ketamine in our patient because of a previous report suggesting of worsening of symptoms.^[8] Regional anesthesia such as caudal block can minimize the requirement of inhalational and intravenous agents in children with OMS and provide good postoperative analgesia and should be considered, whenever feasible.

Conclusion

From our experience and the current literature available, we recommend that the use of inhalational agents, opioid, NDMR, nonsteroidal anti-inflammatory drugs, anticholinergics, and anticholinesterase are safe in patient with Kinsbourne syndrome and drugs such as etomidate, meperidine, and ketamine that can aggravate or trigger the symptoms should be avoided.

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Conflicts of interest

There are no conflicts of interest.

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