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CASE REPORT

Anesthetic management of an adult patient with childhood x-linked adrenoleukodystrophy

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ABSTRACT

X-linked adrenoleukodystrophy is a disorder of peroxisomal fatty acid beta-oxidation causing the accumulation of very long chain fatty acids (VLCFA) in tissues. The hallmarks of the disease are CNS demyelination and primary adrenal insufficiency. The anesthetic considerations include mental retardation, seizure disorder, impaired adrenocortical function, immunosuppression, risk of iatrogenic fractures, hypotonia and delayed awakening. The anesthetic plan should be case specific. Pre-operative sedation should be avoided because of hypotonia of pharyngeal muscles. Corticosteroids should be given intra-operatively. We anesthetized our case using titrated doses of short acting intravenous and inhalational agents. We secured the airway with i-gel. Few cases have been reported in literature. Still there exists no established anesthetic management plan for these patients. To the best of our knowledge, this is the first reported use of titrated doses of anesthetic agents under i-gel supraglottic airway device for a case of x-linked adrenoleukodystrophy posted for orchidectomy, and with a favorable outcome.

Key words: Adrenoleukodystrophy; Demyelination; Hypotonia; Orchidectomy; Seizure

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INTRODUCTION

Adrenoleukodystrophy (ALD), also known as Addison–Schilder disease or Siemerling–Creutzfeldt disease, is a rare inherited disorder that leads to progressive brain damage, adrenal gland failure and eventually death.[1,2] Patients with ALD are deficient in an essential protein necessary for breakdown of very-long-chain fatty acids (FAs) found in the normal diet. Resulting FA accumulation damages the myelin sheath, thus affecting the brain and adrenal gland.[3]

The gene responsible for the condition has been mapped to Xq28.[4] The prevalence of ALD is estimated to be between 1:20,000 and 1:50,000.[5] We present a case report of general anesthesia with i-gel of a patient with this disorder with good outcome.

CASE REPORT

A 26 year old, weighing approximately 40 kg male with x-linked ALD presented for right orchidectomy. His medical history was significant for advanced adrenoleukodystrophy. The parents gave history of onset of adrenoleukodystrophy at 9 y of age. There was progressive deterioration of neurological function leading to generalized spasticity and he was bed ridden for the last 15 y. He had a history of mental retardation and generalized seizures. The patient was not on any treatment except for anti-epileptic medication which was started a week before the surgery.

On examination, the patient was conscious of his surroundings and responded to verbal commands. His vitals and airway examination were within normal limits. The laboratory investigations (complete blood count, renal and liver function tests, coagulation profile), chest x-ray and ECG were within normal limits. The patient was adequately fasting as per the guidelines. Patient had a 22G intravenous cannula in situ.

Written, informed consent for general anesthesia was obtained from the parents.

In the operation theatre, after applying standard monitors (ECG, NIBP and SpO_2), inj. hydrocortisone 100 mg IV, inj. ondansetron (0.8 mg/kg) and inj. fentanyl (1 μg/kg) was administered.

Patient was induced with titrated doses of inj. propofol to achieve hypnosis (upto 2 mg/kg) and inj. atracurium (20 mg IV). After ventilating for 1 min and assessing adequate depth of anesthesia, i-gel No. 5 was inserted. Gastric tube was inserted via dedicated port. Anesthesia was maintained with O_2/ air/sevoflurane 0.8 MAC on volume control mode. TOF ratio was used to assess neuromuscular blockade (NMB) intraoperatively.

The vital signs remained stable throughout the surgery. Inj. paracetamol 1 gm IV was given for analgesia. Total duration of surgery was one hour. After completion of surgery, the incisional site was infiltrated with 10 ml 0.25% levobupivacaine by the surgeon. Post-surgery, the patient started breathing spontaneously within 5 min. The neuromuscular blockade was reversed. I-gel was removed once the patient was fully conscious. The patient was shifted to PACU and discharged home the next day.

DISCUSSION

Our case describes the anesthetic management for a bed-ridden adult patient with childhood x-linked ALD with seizure disorder. Despite being diagnosed with the disease 15 years back, he was not on any treatment. Antiepileptics were started recently, which were continued pre-operatively.

Anesthetic considerations include several factors like mental retardation, seizure disorder, hypotonia, liver dysfunction, gastro-esophageal reflux, abnormal adrenocortical function, altered pharmacological response, risk of iatrogenic fractures, etc.[6]

Current therapies for the disease include bone marrow transplantation, which can leave the patient immunosuppressed iatrogenically, and Lorenzo's oil, which can cause thrombocytopenia and cardiac dysfunction.[7,8] Anticonvulsant therapy for seizures can lead to hepatic enzyme induction and the need to modify anesthetic drug dosages.[9] We decided to use short-acting drugs and titrate the level of anesthesia.

Pre-operative sedation is an issue because it is unknown whether these patients are prone to respiratory depression. It is a good practice to avoid any sedative agents until the patient is under direct supervision.[10]

Chronically bedridden patients are prone to hyperkalemia if given succinylcholine, so it should be avoided.[9,10,11] The response to depolarizing muscle relaxants is unknown and clinical monitoring should guide administration.[6,9,10,12]

Another important consideration is patient positioning. These patients are susceptible to iatrogenic fracture during transfer.[10,13] We took utmost care during induction, positioning and shifting the patient to and from the bed.

Regional anesthesia techniques could be difficult for many reasons. These patients may have scoliosis, secondary to CNS involvement. Patients with mental retardation may be unable or unwilling to consent to a regional technique. Thrombocytopenia secondary to dietary manipulation could also limit the use of regional techniques.[7,10]

Pre-medication with inj hydrocortisone 100 mg was given keeping in view the associated adrenal deficiency.

We managed our patient with titrated doses of propofol with a low dose of non-depolarising agent - atracurium and sevoflurane based balanced anesthesia technique.[14] I-gel was used for airway management. Ryle's tube inserted by a side port provided for aspiration of gastric contents. However, our patient's stomach was empty.

Throughout the surgery, the patient was maintained with oxygen / air / sevoflurane 0.8 MAC. TOF was used to assess the NMB adequacy. Towards the end of surgery, the MAC was gradually decreased. Our patient woke up within five minutes of completion of surgery and NMB was reversed.

As advocated elsewhere in literature, the airway device should be removed once the patient is fully awake. The patient should be monitored for 24 h postoperatively, to recognize early postoperative problems such as respiratory failure due to hypotonia and precipitation of seizures.[10]

On the basis of review of literature and our experience, we would like to emphasize the importance of following: Premedication: avoid sedation; administer corticosteroids. Induction: short acting IV and inhalational agents; avoid succinylcholine, NMB monitoring guided NDMR administration. Maintenance: careful positioning, multimodal analgesia, short acting agents. Recovery: monitoring for at least 24 h postoperatively.

We shifted our patient to PACU in fully awake and pain-free state. The patient was discharged home the next day.

CONCLUSION

Anesthetic management of an adult patient with

childhood x-linked ADL is a rare occurrence. In our patient, a favorable outcome was achieved by the judicious and titrated use of anesthetic agents along with continuous monitoring and good analgesia. This avoided post-operative complications and facilitated early hospital discharge in a high risk patient with uncommon medical condition.

Conflict of interest: None declared by the authors

Authors' contribution:

JG: Concept, conduct of study work, literature search, manuscript writing and editing

DA: Concept, manuscript editing, guidance

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