Anesthetic management of a patient with Kearns-Sayre syndrome

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Introduction

Herein, we describe the interesting case of a 13-year-old female with Kearns-Sayre syndrome (KSS) who underwent a successful T3-L3 posterior spinal instrumentation and fusion (PSIF). KSS is a rare mitochondrial myopathy with only 226 cases reported in published literature as of 1992. It is the result of deletions in mitochondrial DNA and manifests as a triad of: 1) chronic progressive external ophthalmoplegia 2) bilateral pigmentary retinopathy 3) cardiac conduction abnormalities.

The operative presentation of a child with KSS is rarely encountered by anesthesiologists. As such, anesthetic management of a child with this disorder has seldom been reported.

Case Presentation

In addition to the classical triad, this teenager had bilateral sensorineural hearing loss, progressive atrioventricular (AV) conduction abnormality with right bundle branch block, diabetes mellitus type I, hypothyroidism, and attention deficit hyperactivity disorder. She was found to have a 70-degree scoliosis and was scheduled for a T3 to L3 PSIF with allograft.

Preoperative management

The 33.8-kg teenager underwent placement of an intravenous catheter (IV) upon admission and:
- received her last oral dose of Levocarnitine 1000mg at 2000
- maintained on insulin regimen with frequent blood glucose checks
- magnet was prepared in the operating room in the event of a need to convert the pacemaker to magnet mode
- 4 units of packed red blood cells confirmed to be available
- received 200mg of Gabapentin orally on the morning of surgery, 1.5mg of IV Midazolam

Intraoperative management

General anesthesia was then induced:
- 2mg/kg of Ketamine, 3mcg/kg of Fentanyl, 1mcg/kg of Dexmedetomidine
- no muscle relaxant or inhalation agents were used
- trachea was intubated with a 5.5 cuffed oral endotracheal tube
- second IV and a 22 Gauge left radial arterial line were inserted
- patient was then placed prone on a padded head rest
- Anesthesia maintained with 5-10mcg/kg/min of Ketamine, 5-30mcg/kg/hr of Fentanyl, and 0.6-1mcg/kg/hr of Dexmedetomidine.

Postoperative management

The patient remained intubated and was transferred to the intensive care unit
- estabulted that evening and transferred to the floor on postoperative day (POD) 1
- Oral Levocarnitine, Vitamin B50 complex, and Levothyroxine was subsequently restarted

For pain relief, her regimen included:
- 352mg of oral Acetaminophen every 4 hours
- 15mg of IV Ketorolac every 6 hours for 72 hours
- Hydromorphone patient-controlled analgesia (PCA) pump

On POD 3, she was taking:
- oral hydromorphone
- oral ibuprofen for analgesia
- 1.5mg of IV Midazolam

Discussion

The peroperative management of this patient is complex. KSS patients have:
- impaired respiratory response to hypoxemia
- opioids should be used with caution
- potential to induce respiratory acidosis in addition to the underlying metabolic acidosis
- dysfunctional aerobic metabolism
- any increase in the basic metabolic rate should be prevented
- shivering, hypoxia, ketoacidosis, and fasting in such patients may also exacerbate the lactic acidosis

Critical to peroperative management of this patient are also the steps needed to avoid medications shown to inhibit complex I of the electron transport chain:
- barbiturates, propofol, and volatile anesthetics

For this patient and her surgical procedure, we were able to provide:
- total intravenous anesthesia
- without utilizing medications that would inhibit the electron transport chain or interfere with neuromonitoring.

We tailored our care to:
- prevent hypothermia, respiratory acidosis, and ketoacidosis

The decision to proceed with surgical procedures in KSS children is challenging. However, as this case demonstrates, with multidisciplinary collaboration and placing considerable attention on providing a smooth and safe anesthetic along with good perioperative anesthesia, we are able to optimize chances for a successful peroperative outcome.

REFERENCES: