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Non-Trigger Anesthesia Management in a Patient With Leigh's Syndrome Presenting for Dental Rehabilitation

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Abstract

Introduction: Usually presenting in infancy, Leigh's syndrome is an inherited condition often manifesting with seizures, ataxia, developmental delay, and dysarthria. The disorder is rare, appearing in approximately 1 in 40,000 live births. Consequently, providing these patients with a suitable plan by which to administer anesthetics remains problematic.
Case Presentation: We report a male patient with Leigh's syndrome and a family history suggestive of unknown hypotonia and malignant hyperthermia presenting for dental rehabilitation.
Conclusions: Dexmedetomidine with remifentanil can be used with no complication in this senerio.

Keywords: Leigh's Syndrome, Anesthesia, Encephalomyelopathy, Mitochondrial Disorders, Malignant Hyperthermia

1. Introduction

Leigh's syndrome is a sub-acute necrotizing encephalomyelopathy that was first reported in 1951 by Archibald Denis Leigh, a British neuropathologist (1). It is one disease of the family of disorders classified as 'mitochondrial myopathies' that include mitochondrial encephalopathy, lactic acidosis, and stroke-like episodes (MELAS); neurogenic muscle weakness, ataxia, and retinitis pigmentosa (NARP). Leigh's syndrome can be caused by mutations in mitochondrial DNA or by deficiencies of the enzyme pyruvate dehydrogenase. It is rare disorder with an estimated prevalence of approximately 1 in 40,000 live births (2).

We report our experience in non-trigger anesthesia management in a patient with Leigh's syndrome and a family history suggestive of unknown hypotonia and malignant hyperthermia who underwent dental rehabilitation. Our institutional review board does not require review for case reports when no identifying patient information is given. This report does not include any identifying information.

2. Case Presentation

A 19-year-old male scheduled for dental rehabilitation was diagnosed with Leigh's syndrome in infancy. At 3-months-of-age, the patient presented with staring/lethargic spells and loss of developmental milestones. Following a muscle biopsy, he was diagnosed with Leigh's syndrome after 2 months. His past surgical history included a gastrostomy and Nissen fundoplication and magnetic resonance imaging (MRI) in infancy. Family history was positive for a first cousin with unknown hypotonia and symptoms suggestive of malignant hyperthermia (masseter spasm) on exposure to anesthesia. Physical examination of the patient revealed a Mallampati class II airway; he was nonverbal and wheelchair-confined with secondary spasticity. He also had severe neuromuscular scoliosis with a pectus carinatum deformity noted. He weighed 36.8 kilograms. His medical history included known allergies to amoxicillin clavulanate, latex, and tape. His medication included levetiracetam, clonazepam, and tamsulosin. Vital signs were within normal limits and his respiratory examinations were unremarkable. Hematological workup and hermia, tricuspid insufficiency electrolytes were within normal limits. His last echocardiogram was 9 years previously showing mild tricuspid insufficiency. An echocardiogram on the day of surgery reported cardiac dextroposition due to severe pectus carinatum deformity, and normal left ventricular (LV) and right ventricular (RV) functions with a right ventricular systolic pressure (RVSP) of 24.1 mmHg.

The anesthetic plan included performing the procedure under general anesthesia using total intravenous anesthesia. The patient was held nil per os (NPO) for 6 hours. Midazolam (5 mg) was administered subcutaneously in the preoperative holding area. General anesthesia was in-

duced with 70% N_2O in O_2 following a 22-gauge peripheral intravenous line (PIV) placed in the left hand. A 1 μg/kg bolus of dexmedetomidine over 10 minutes was started. Fentanyl (1.5 μg/kg) was given to attenuate the hemodynamic response to intubation and a 6.5 mm endotracheal tube was placed and secured in position. On direct laryngoscopy, the airway was a Cormack and Lehane grade 3 view. Anesthesia was maintained with 1 L oxygen/2 L nitrous oxide and continuous infusions of dexmedetomidine (0.4 - 1.4 μg/kg/hour) and remifentanil (0.8 - 1.2 μg/kg). Infusions were titrated to hemodynamic response of surgery. Intraoperative end-tidal carbon dioxide concentration in the expired air (ETCO2) was maintained between 35 and 40 mmHg. An underbody blanket was used to maintain normothermia. The procedure lasted 1.15 hours and the total fluid administration included 600 mL of normal saline. Infusions stopped 10 minutes before the end of surgery. Following the procedure, the patient was extubated and transferred to the post-anesthesia care unit with close observation. The postoperative course was uncomplicated, and postoperatively no additional opioids were given. He returned to his baseline mental status, maintaining normal oxygen saturation on room air. The patient was discharged home with advice to take ibuprofen 400mg every 4 hours as needed.

3. Discussion

The rarity of patients with Leigh's Syndrome results in a paucity of information regarding anesthesia administration and management, particularly for those patients who exhibit mitochondrial disorders that involve multi organ systems. Anesthetic management depends on the assessment of these systems prior to any surgical procedure and close observation following surgery. Reports of complications using volatile agents in patients with mitochondrial defects are mixed. In a 2006 animal study, Bains and colleagues (3) suggested that volatile agents could becontra indicated in patients with mitochondrial defects. Conversely, a study by Morgan et al. (4) suggested no contraindications in patients with mitochondrial dysfunction; the study of 16 patients included 1 diagnosed with Leigh's Syndrome. Although malignant hyperthermia has not been reported with Leigh's syndrome, it may be best to avoid triggering agents as other myopathic conditions with susceptibility for malignant hyperthermia may be misdiagnosed as Leigh's syndrome. In this case specifically, the patient had a relative with malignant hyperthermia and this further limits the anesthetics that can be used. Additionally, close patient monitoring following surgery is important to avoid respiratory failure.

In our case, we chose to use dexmedetomidine with remifentanil for the induction and maintenance of anesthesia for its rapid recovery profile, with limited postoperative central nervous system and respiratory depression and safety in patients at risk for malignant hyperthermia. Dexmedetomidine and remifentanil infusion provided a good combination of sedation and analgesia which can be used in other procedures that require cardiac stability. We recommend avoiding circumstances that place a metabolic burden on these patients like prolonged fasting, hypoglycemia, postoperative nausea and vomiting, hypothermia with resulting shivering, prolonged tourniquets, acidosis, and hypovolemia.

Learning Points are included as:

1. Leigh's syndrome is an inherited condition often manifesting with seizures, ataxia, developmental delay, and dysarthria.

2. Leigh's syndrome is rare.

3. Although malignant hyperthermia has not been reported with Leigh's syndrome, it may be best to avoid triggering agents as other myopathic conditions with susceptibility for malignant hyperthermia may be misdiagnosed as Leigh's syndrome.

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