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Multimodal Anesthesia for Glaucoma Surgery in a Child with Mitochondrial Disease and Malignant Hyperthermia

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Abstract

Mitochondrial diseases (MD) are characterized by impairments of mitochondrial function that precipitate metabolic acidosis. An 8-year-old MD female with prior anesthesia exposure complicated by metabolic acidosis, seizures and propofol infusion syndrome, presented for glaucoma surgery. We present a multimodal balanced anesthetic technique used to successfully manage this complex case.

Introduction

Mitochondrial disorders (MD) are a heterogeneous group of genetic disorders that impair mitochondrial integrity and result in deficient energy production. The disorder has an incidence of 1:5000 live births [1], and affects tissues with high-energy requirements such as the central nervous system, retina, heart and muscle [2]. Consequently, these patients have multiple co-morbidities that include cardiac, endocrine, and neurologic dysfunction [3-5]. Current evidence suggests that mitochondrial disease may be a contributing factor in the pathogenesis of glaucoma [6]. Anesthesia in this patient population may prove hazardous because the stress of surgery and fasting can induce marked metabolic aberrations, most commonly lactic acidosis [7].

Case Presentation

Institutional Review Board approval is not required for single case reports at Jackson Memorial Hospital and University of Miami. An 8 year old, 25kg Caucasian female with infantile glaucoma, mitochondrial disease, renal tubular acidosis type II and family history of malignant hyperthermia presented for bilateral glaucoma surgery. Of note, at age 6, eye surgery under GA had been complicated by severe post-operative metabolic acidosis, propofol infusion syndrome (PRIS) and prolonged two week PICU stay. At that time, genetic investigation revealed an abnormality of the 2-oxoglutarate-dehydrogenase enzyme.

A multidisciplinary group comprising physicians from genetics, neurology, cardiology, nephrology and anesthesiology was convened and recommended new studies in order to identify the gene deficit. Also, the group recommended preoperative nutritional supplementation and electrolyte correction. Her ECG demonstrated prolongation of the QT interval (426ms) and the 2-D cardiac echo was normal. Current medications included trileptal, brimonidine and lantaprost ophthalmic solutions, coenzyme Q10 and a multivitamin supplement.

Our anesthesia plan centered on a balanced technique utilizing intravenous agents for sedation and hypnosis in tandem with extraconal ophthalmic block for analgesia. A vapor-free operating room was prepared. Monitoring included standard ASA monitors and bispectral analysis (BIS) (Model 185-0151) with the target range set at 40-60. In the holding suite the child was sedated with midazolam. Then, in the OR, infusions of remifentanil (0.1mcg/kg/min) and dexmedetomidine (1mcg/kg/hr) were started. When BIS levels fell below 60 we administered cisatracurium (4mg) and performed endotracheal intubation. A left radial intra-arterial catheter was placed for blood gas and metabolic panel

analysis. Prior to surgery, a periorbital block using 2% lidocaine was performed.

Serial metabolic analysis demonstrated stable acid/base status with marginal elevations in serum lactic acid (1.3 to 1.5mg/dL). Surgery was completed in 110 minutes and passed uneventfully. At this point the neuromuscular block was antagonized, infusions were discontinued and the child was extubated and transferred to PICU for overnight observation. She was discharged from the hospital on postoperative day 1.

Discussion

Mitochondrial disease (MD) are a divergent group of more than 100 genetic aberrations in which defects of the organelle impair oxidative phosphorylation and the production of energy, resulting in cell injury and metabolic acidosis. It is estimated that MD afflicts 1:5000 children. Symptoms typically manifest in tissues that are wholly dependent on mitochondria as a source of energy [8].

The anesthetic management of children with MD is complex because studies indicate that volatile inhalational agents depress mitochondrial function [9,10]. Additionally, severe co morbidities such as organ failure are potential risks for lactic acidosis. Further anesthesia considerations include sensitivity to neuromuscular blocking agents, altered homeostasis and impairment of mitochondrial integrity triggered by several intravenous agents. There are no controlled clinical trials to assess the effects of anesthetic agents and their correlation with intra- and postoperative complications [3]. Currently, available data stems from case reports, expert opinion and retrospective chart reviews [10, 11].

A recent rat model study demonstrated that ketamine produces alterations in the mitochondrial respiratory chain complex 1 [12]. Other investigations implicate etomidate and barbiturates as inhibitors of the same complex [13]. Moreover, evidence suggests that propofol impairs mitochondrial function, and that propofol infusions are associated with PRIS because this agent is highly lipophilic and readily diffuses across cell membranes. Therefore, propofol should only be administered as a single bolus [14,15]. On the other hand, dexmedetomidine, a selective α2 agonist, is known to have beneficial effects on the mitochondrial membrane [16]. Although the association between MD and malignant hyperthermia (MH) remains unproven, the Malignant Hyperthermia Association of the United States (MHAUS) continues to recommend avoidance of volatile anesthetic agents and caution in using succinylcholine [17- 19].

In this patient we were concerned about the development of lactic acidosis, malignant hyperthermia and PRIS. At the same time, we aimed to optimize intraoperative surgical conditions and avoid patient movement. In light of these considerations; we adopted a balanced intravenous anesthetic technique that eliminated the need for a volatile inhalational agent. Thus, we integrated the hypnotic effect of dexmedetomidine and the analgesic property of an ultra-short acting opioid (remifentanil) with a non-depolarizing neuromuscular blocking agent having negligible renal elimination (cisatracurium) and also performed an extraconal eye block (2% lidocaine). We selected lidocaine because bupivacaine inhibits carnitine-acylcarnitine translocase and has been reported to precipitate ventricular dysrhythmias [20]. The eye block was quintessential because it reduced anesthetic requirements and assured a smooth transition between the operating room (OR) and post anesthesia care unit (PACU).

In summary, the anesthesia care for MD children is challenging. This case report highlights two important points; [1] the value of a multidisciplinary team to optimize preoperative status, and [2] the benefit of judicious selection of anesthetic technique in order to avoid complications, such as lactic acidosis, MH or PRIS. Finally, this case underscores the benefit of regional anesthesia in reducing anesthetic requirements and facilitating an uneventful transition from the operating room to post anesthesia care unit.

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