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Anesthetic Management of a Patient with Autonomic Dysfunction, Hypokalemic Periodic Paralysis and Mast Cell Activation Syndrome Undergoing Bimaxillary Orthognathic Surgery: A Case Report

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Abstract

Autonomic-dysfunction refers to problems of the autonomic nervous system. Hypokalemic-periodic-paralysis results in episodes of muscle weakness and low blood potassium levels. Mast-cell-activation-syndrome is defined by the abnormal growth and accumulation of mast cells. Few clinical cases are found in the literature on the anesthetic management of each of these disorders, and there are no reported cases of adults with all three conditions undergoing surgery under general anesthesia. We report the case of a 36-year-old male with autonomic-dysfunction, hypokalemic-periodic-paralysis and mast-cell-activation-syndrome who safely underwent general anesthesia for bimaxillary orthognathic surgery.

Keywords: Autonomic dysfunction; Hypokalemic periodic paralysis; Mast cell activation syndrome; Malignant hyperthermia; Long QT syndrome

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Abbreviations

AD: Autonomic-Dysfunction; HKPP: Hypokalemic-Periodic-Paralysis; MCAS: Mast-Cell-Activation-Syndrome; OSAS: Obstructive-Sleep-Apnea-Syndrome

Introduction

Autonomic-Dysfunction (AD) or dysautonomia is a condition in which the immune system mistakenly attacks and damages certain parts of the autonomic nervous system. Hypokalemic-Periodic-Paralysis (HKPP) is a rare ion channel disorder caused by skeletal muscle ion channel mutations. Mast-Cell-Activation-Syndrome (MCAS) in turn is a clonal disease of mast cell progenitors in the bone marrow, characterized by the abnormal growth and accumulation of mast cells.

Few clinical cases are found in the literature on the anesthetic management of each of these disorders, and there are no reported cases of adults with all three conditions undergoing surgery under general anesthesia. We report the case of a 36-year-old male with AD, HKPP and MCAS who safely underwent general anesthesia for bimaxillary orthognathic surgery to correct Obstructive Sleep Apnea Syndrome (OSAS). Written informed consent has been obtained from the patient for the publication of this Case Report.

Case Presentation

A 36-year-old male (weight 70 kg, height 178 cm) with no known drug allergies, came to our center to undergo bimaxillary orthognathic surgery for the correction of OSAS. As part of his disease history, the patient provided a report from the Center for Complex Diseases (2500 Hospital Drive, Bldg, 4B Mountain View, CA 94040, USA) reflecting the diagnoses of OSAS, chronic rhinosinusitis, repetitive strain injury, chronic fatigue syndrome, circadian rhythm disturbance - AD, HKPP and MCAS. In this context, the patient was receiving regular treatment with intranasal budesonide, intranasal cromolyn sodium and nocturnal continuous positive airway pressure.

The patient was diagnosed in 2017 with AD, with abnormal elevation of anti-adrenergic antibodies, and suffered from postural orthostatic tachycardia syndrome. Control was currently afforded by proper hydration and regular exercise.

During that same year (2017) he was also diagnosed with HKPP. The patient reported to the emergency department due to repeated episodes of moderate hypokalemia (3.2 mmol/l) together with episodes of full-body muscle weakness. He required hospital admission and potassium replacement therapy. The electrocardiograms were normal. Finally, the genetic analysis confirmed the suspicion of HKPP, with the identification of CACNA1S and KCNJ2 gene mutations. Since 2018 the patient has been asymptomatic.

With regard to the surgical history, he had three surgeries under general anesthesia in recent years without incidents: Septoplasty in 2018, varicocelectomy in 2019 and palatal expansion surgery in 2020. In relation to this last operation, the patient provided an anesthetic report with the drugs used for general anesthesia at Stanford Health Care and University Healthcare Alliance (300 Pasteur Drive; Palo Alto, CA 94304, USA). Balanced general anesthesia was performed, and the following adjuvant drugs were administered: Midazolam, glycopyrrolate, fentanyl, propofol, lidocaine, sevoflurane, rocuronium, dexamethasone, cefazolin, ondansetron, Sugammadex, Ephedrine, Normosol® electrolyte solution, oxygen/air/nitrous oxide. There were no relevant incidents.

The present case report describes the decision making carried out by the anesthetic-surgical team for bimaxillary osteotomy surgery under general anesthesia in an adult male with three infrequent disease conditions involving severe and potentially fatal risks for the patient.

Dysautonomia or autonomic-dysfunction is a condition in which the immune system mistakenly attacks and damages certain parts of the autonomic nervous system. The symptoms may include severe orthostatic hypotension, fainting, constipation, fixed and dilated pupils, urinary retention, and dry mouth and eyes. The underlying cause of AD is poorly understood. The main recommendations for the surgical management of patients with AD focus on two aspects. Firstly, in relation to airway management, these individuals should be regarded as having a full stomach due to their gastroparesis and intestinal motility alterations, and thus rapid sequence anesthetic induction should be performed. Secondly, at hemodynamic level, proper perioperative hydration is to be maintained, considering that these patients are very labile to modifications in position and ventilation, with paradoxical responses to physiological and pharmacological changes [1].

Hypokalemic-periodic-paralysis is a rare ion channel disorder caused by skeletal muscle ion channel mutations (CACNA1S, SCN4A, and KCNJ18 are the genes involved). Patients characteristically present with sudden onset of generalized or focal flaccid paralysis associated to hypokalemia, which persists for several hours before resolving spontaneously. Anesthesiologists must be aware of this condition, especially as regards mutations of the CACNA1 and KNCJ genes, associated with malignant hyperthermia and long QT syndrome, respectively. The main recommendations for the management of patients with HKPP include strict control of plasma potassium concentration, the avoidance of large glucose and salt loads, the maintenance of body temperature and acid-base balance, and careful use of neuromuscular blocking agents [2,3]. In addition, in those patients with CACNA and KNCJ gene alterations susceptible to malignant hyperthermia and long QT syndrome, a number of anesthetic considerations should be added. In effect, patients susceptible to malignant hyperthermia have genetic abnormalities in skeletal muscle receptors that allow an excessive accumulation of myoplasmic calcium in the presence of certain anesthetic triggers. Trigger-free general anesthesia, avoiding volatile anesthetics and succinylcholine, is mandatory [4-6]. In case of administering inhaled anesthetics, the goal is to keep the inspired volatile anesthetic concentration at 5 ppm or less. This threshold is commonly accepted as "safe" regardless of country-specific legal regulations [7]. Likewise, in patients susceptible to long QT syndrome, careful anesthetic management is needed because of the risk of Torsades de Pointes and malignant arrhythmias, and premedication with midazolam, sedoanalgesia with morphine or fentanyl, induction and maintenance of anesthesia with thiopental or propofol Total Intravenous Anesthesia (TIVA) avoiding halogenated volatile anesthetics and ketamine, is recommended [8].

Mast-cell activation-syndrome is a clonal disease of mast cell progenitors in the bone marrow, characterized by the abnormal growth and accumulation of mast cells. Patients with this condition are at risk of suffering severe allergic reactions in the form of anaphylaxis, due to the potential of releasing large amounts of histamine into the bloodstream [9]. The primary goal of anesthetic management in patients with MCAS is to prevent mast cell degranulation using anti-anxiety premedication, maintaining a steady temperature, with careful positioning of the patient, the avoidance of histamine-releasing drugs, the prevention of nausea and vomiting, aggressive treatment of pain - which is a potent mast cell degranulator (including the use of acceptable forms of opioids such as fentanyl, remifentanil) - and the use of H1 and H2 receptor antagonists to maintain mast cell stability [10-12].

Lastly, the anesthetic plan in our patient with all three of these infrequent disease conditions, and who had recently received several potential risk medications without complications, was as follows: Preoperatively one hour before surgery, a peripheral venous line was placed for the administration of saline solution together with midazolam 2 mg, dexchlorpheniramine 5 mg, cefazolin 2 g and ondansetron 4 mg. Once in the operating room, a thermal blanket was placed, the patient was positioned correctly, noninvasive monitoring was arranged, and rapid-sequence anesthetic induction was performed with a Sellick maneuver together with propofol (2 $mg.kg^{-1}$, rocuronium (0.6 $mg.kg^{-1}$) and fentanyl (2 $\mu g.kg^{-1}$). Anesthetic maintenance was then carried out with propofol (Target-Controlled Infusion [TCI], Schnider-model effective concentration 2 $mcg.ml^{-1}$) and remifentanil (TCI Schnider-model effective concentration 2 $ng.ml^{-1}$) to achieve correct permissible perioperative hypotension with a mean arterial pressure of >60 mmHg. Intravenous paracetamol 1 g, dexamethasone 4 mg and tranexamic acid 15 $mg.kg^{-1}$ were administered as adjuvants. Arousal took place with prior muscle relaxation reversal using Sugammadex. The postoperative instructions were as follows: Liquid diet low in sugar, analgesia with paracetamol, and one delayed-release fentanyl transdermal patch (72 h), with local cold - facial mask circuit. The patient was discharged 18 h after surgery according to the protocol of our center, with no relevant medical-surgical complications.

References

1. Mustafa HI, Fessel JP, Barwise J, Shannon JR, Raj SR, Diedrich A, et al. Dysautonomia: Perioperative Implications. Anesthesiology. 2012;116:205-15.

2. Phuyal P, Nagalli S. Hypokalemic Periodic Paralysis. Treasure Island (FL): StatPearls Publishing. 2022.

3. Henshaw DS, Harstroem C, Forest D. Hypokalemic periodic paralysis masquerading as a compressive neuraxial lesion following lumbar epidural placement in a parturient: A case report. A A Pract. 2021;15(4)e01431.

4. Rosenberg H, Pollock N, Schiemann A, Bulger T, Stowell K. Malignant hyperthermia: A review. Orphanet J Rare Dis. 2015;10:93.

5. Lambert C, Blanloeil Y, Horber RK, Bérard L, Reyford H, Pinaud M. Malignant hyperthermia in a patient with hypokalemic periodic paralysis. Anesth Analg. 1994;79(5):1012-4.

6. Litman RS, Smith VI, Larach MG, Mayes L, Shukry M, Theroux MC, et al. Consensus statement of the malignant hyperthermia association of the United States on unresolved clinical questions concerning the management of patients with malignant hyperthermia. Anesth Analg. 2019;128(4):652-9.

7. Rüffert H, Bastian B, Bendixen D, Girard T, Heiderich S, Hellblom A, et al. Consensus guidelines on perioperative management of malignant hyperthermia suspected or susceptible patients from the European Malignant Hyperthermia Group. Br J Anaesth. 2021;126(1):120-30.

8. Fazio G, Vernuccio F, Grutta G, Lo Re G. Drugs to be avoided in patients with long QT syndrome: Focus on the anaesthesiological management. World J Cardiol. 2013;5(4):87-93.

9. American Academy of Allergy, Asthma & Immunology. Mast Cell Activation Syndrome (MCAS). 2022.

10. Dewachter P, Castells MC, Hepner DL, Mouton-Faivre C. Perioperative management of patients with mastocytosis. Anesthesiology. 2014;120(3):753-9.

11. Borgeat A, Ruetsch YA. Anesthesia in a patient with malignant systemic mastocytosis using a total intravenous anesthetic technique. Anesth Analg. 1998;86(2):442-4.

12. Becerra-Bolaños A, Muiño-Palomar V, Cabrera-Doreste S, Rodríguez-Pérez A. Sugammadex in systemic mastocytosis: Case report and a systematic review of literature. Anaesthesist. 2021.