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CASE REPORT

Malignant Hyperthermia in the Dental Setting: A Case Report

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Abstract

Understanding rare diseases, such as Malignant hyperthermia (MH), will help provide optimal care to these patients, especially in dental settings. As MH is a sporadic, inherited disorder, dental clinicians must have sufficient knowledge and information about such a rare disease to manage a patient with such a condition. This article will describe a case with MH, highlighting the patient's or parents' concerns about using local anesthetic agents in dental treatment.

Keywords: Malignant hyperthermia; CACNA1S; RYR1; Whole exome sequencing; Inhalation agents

Introduction

MH can be defined as a hypermetabolic response to potent inhalation agents such as halothane, sevoflurane, desflurane, the depolarizing muscle relaxant succinylcholine, and rarely, in humans, to stresses such as vigorous exercise and heat.[1] This condition is rare and could be life threatening.

According to the current literature, the ryanodine receptor RYR1 plays an important role in the pathophysiology of MH and the voltage-dependent calcium channel Cav1.1 located in the T- tubular membrane of myocytes.[2] Concerning age and gender, the average age of MH patient were found to be children of 12 years or younger, with a higher tendency of affecting males.[3,4]

Pediatric dental patients are usually managed safely in the dental chair using topical and local anesthetic agents before dental treatment. In some occasions where the pediatric patient is considered anxious, relative analgesia (conscious sedation) or general anesthesia is indicated.

Topical anesthesia is a valuable tool in treating pediatric patients, as it will reduce the pain of local anesthesia injection, which will relieve the patient of his fear and anxiety, ensuring a good experience for the patient and the parent.[5]

Local anesthesia might be required for some dental treatment. According to current consensus, local anesthesia is safe to use in MH patients. 2% lidocaine with epinephrine 1:100,000 is thought to be the ideal agent for pediatric patients as it has a low chance of toxicity.[6]

Case Report

A 9-year-old male patient of Arabic ethnicity attended the Dental and Maxillofacial Centre (DMFC) at the Royal Medical Services (RMS) and was advised extractions of primary canines.

After assessment by a pedodontist and orthodontist, the patient`s parent was concerned about the technique of dental extractions to be performed as the patient is being diagnosed with MH.

The patient had a medical report of a recently diagnosed recurrent rhabdomyolysis due to MH (type 5 susceptibility) from King Hamad University Hospital (KHUH), where whole exome sequencing (WES) genetic test was performed and a variation of the CACNA1S gene was found. According to the family history of the child and parent, no other family member had been diagnosed with MH.

Before dental extraction was carried out, it was clearly explained to the patient and parent that the use of the topical anesthetic gel (20% Benzocaine, iolite, Dharma-Research, Inc-USA) and the local anesthetic agent (Lidocaine HCL 2% and Epinephrine 1:100000, Octacaine, Novocol Pharmaceutical of Canada Inc.) is safe to use according to the literature.[5] On the same dental visit, two primary canines were extracted without complications. The patient was recalled for follow-up after 4 weeks, and neither the patient nor the patient's parents reported any signs or symptoms of MH.

Discussion

MH is a pharmacogenetic disorder that manifests as a hypermetabolic response to potent inhalation agents.[7] In the past, local anesthesia was considered to be a triggering agent for MH, but these days, local anesthesia is considered safe to use as well as inhalation sedation agent nitrous oxide, which is the only safe inhalation sedation agent.[8,9]

According to the UK guidelines for the use of general anesthesia in pediatric dentistry,[10] the patient cooperation level must be determined along with the perceived anxiety, how the child responded to similar procedures, the procedure's complexity, and the child's medical condition.[10] It’s essential to formulate a complete treatment plan to avoid the repeat of general anesthesia.

Multiple procedures are more suitable to be done under general anesthesia, such as acute swelling requiring removal of an infected tooth, symptomatic teeth in more than 1 quadrant, surgical extraction of 1 or more teeth, examination of teeth for special needs kids, and the most commonly observed are severe pulpitis and acute infections.[10]

Sedation might be suitable for treating multiple patients, but general anesthesia might be required for comprehensive dental treatment. Fortunately for patients with MH, several safe drugs can be used, such as propofol, ketamine, opioids, and non-depolarizing muscle relaxants.[1]

The two genes that have been definitively associated with MH causative mutations are RYR1 and CACNA1S.[5] Diagnosing this condition can be difficult as most patients of MH will not have any symptoms without them being exposed to an anesthetic or specific diagnosis test.[7]

The key clinical features for MH will include an unexplained elevation of expired carbon dioxide despite increased minute ventilation, muscle rigidity, rhabdomyolysis, hyperthermia, tachycardia, acidosis, and hyperkalemia.[7]

Masseter muscle rigidity (MMR) can be used as an indicator to predict MH in a patient. Trismus after succinylcholine is one of the most common early signs of MH. Therefore, it can be used for early detection of MH. It was concluded that the MMR is associated with MH around 50% of the time.[11]

The golden standard for laboratory diagnostic methods involves an in vitro contracture test, which tests the muscle contracture in halothane or caffeine.[5]

If muscle contracture is observed in the presence of both chemicals, then the patient is susceptible to MH. Another diagnostic approach includes using the whole exome sequencing test to identify any mutations in the RYR1 and CACNA1S gens to determine the susceptibility of an individual to MH.[12]

The incidence of an MH episode is between 1:10000 and 1:250000. Even though MH can occur from the first exposure to the agent, it usually takes up to three times to finally show signs. MH is reported to

occur in any ethnicity but is more common in males than females, with a ratio of 2:1.[13]

In 1994, Larach et al. created a grading scale for MH to classify patient susceptibility from 1 to 6, with 1 being rarely and 6 being almost certain. The scale uses 6 processes, represented by clinical indicators, each given an assigned score. The processes include Rigidity, muscle breakdown, respiratory acidosis, temperature increase, cardiac involvement, and family history, and after calculating the raw score, a rank will be given from 1 to 6 to determine the chance of an MH episode occurring.[8,14]

Management of MH initially involves removing the trigger agent and then giving 100% oxygen for 2-3 minutes. Second, we will start giving intravenous dantrolene, an antidote that inhibits the excessive release of calcium into the muscles. Lastly, we will start body cooling since increased body temperature enhances calcium release into muscle cells.[15]

If the initial management was successful in the operating room and there was no contraindication to finishing the surgical procedure, the patient would need to be monitored for 24 hours in the ICU before release.[15]

Ideally, they should try to prevent an episode from occurring by taking a thorough anesthetic history for the patient and his family members; if there is suspicion that MH exists, the family members of the patient should not be exposed to a trigger agent, all of the patients that might undergo more than brief general anesthesia should have their body temperature monitored. We should avoid succinylcholine in patients under 12 or patients with any form of myotonia.[1]

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