Highly Suspicious Malignant Hyperthermia: A Case Report

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Abstract

Malignant hyperthermia (MH) crisis could be fatal during general anesthesia. We report a case of a 34-year-old male who was scheduled to undergo bilateral tonsillectomy with diagnosis of left tonsil mass, chronic tonsillitis and obstructive sleep apnea-hypopnea syndrome (OSAHS). The patient inhaled 3% sevoflurane for 5 minutes mask ventilation with bag was well for his pre-oxygenation procedure. During induction of anesthesia, the patient developed masseter muscle rigidity (MMR) rapidly following administration of succinylcholine, but laryngoscopy was still available. Low-flow sevoflurane was inhaled after the intubation. The patient suffered from hypercapnia (55mmHg), slightly increase of temperature, sinus tachycardia, combined metabolic-respiratory acidosis during intraoperative period. The patient was temporary diagnosed with malignant hyperthermia and treated accordingly. Further the patient was carefully monitored and investigated to exclude other possible conditions in postoperative period. The patient was highly suspected MH according to the DNA analysis. He was discharged from the hospital without any complication and discomfort.

ABBREVIATIONS

MH: Malignant Hyperthermia; MMR: Masseter Muscle Rigidity; CKP: Creatine Phosphokinase; EtCO₂: End-tidal Carbon dioxide Pressure; SR: Sarcoplasmic Reticulum; AST: Aspartate Aminotrans Ferase; OSAHS: Obstructive Sleep Apnea-hypopnea Syndrome; T: Temperature; HR: Heart Rate; RYR1: Ryanodine Receptor

INTRODUCTION

Malignant hyperthermia (MH) is a hyper metabolic, lethal syndrome triggered by anesthetic drugs in those genetically susceptible individuals [1]. The Pathophysiology of MH is related to altered sarcoplasmic reticulum (SR) calcium release channel. Two genes’ mutation has been reported associated with MH: ryanodine receptor subtype 1(RyR1) and CACNA1S [2]. During MH episode the SR calcium channels open persistently and responding calcium influx causes sustained muscle contraction and increased metabolism of oxygen and glucose resulting in acidosis, rigidity and hyperkalemia. Sustained muscle contraction expands large number of adenosine triphosphate. Consumption of cellular adenosine triphosphate stores finally leads to rhabdomyolysis and release of the contents of cells (e.g., potassium, creatine phosphokinase, and myoglobin) [3,4]. Increased metabolism accounts for hyperthermia and a high level of end tidal CO₂ (EtCO₂) out of proportion to clinical setting indicates MH during anesthesia because persistent metabolism lead to increased oxygen consumption, hypoxia, progressive lactate acidosis, excessive production of CO₂ and increased body temperature [5].

We here present a case showing all the clinical signs and symptoms of malignant hyperthermia after administration of succinylcholine and sevo sevoflurane in a patient who was scheduled to undergo tonsillectomy.

CASE PRESENTATION

A 34-year-old and 85 kilograms male who was scheduled to receive bilateral tonsillectomy under general anesthesia was diagnosed with left tonsil mass, chronic tonsillitis and obstructive sleep apnea-hypopnea syndrome (OSAHS). This patient has suffered sleep apnea-hypopnea at night for half a year, 1 to 2 times/month, no dizzy or sick at the next morning.

According to pre-anesthetic physical examination, there was no significant finding except for bilateral tonsil enlargement (1F°). The mouth opening of this patient was 4.5cm with a 5.5cm thyromental distance and no abnormality was found in the extension of the atlanto-occipital joint. The Mallampati grade was 1F°.

Routine adjunctive investigations such as complete blood count, coagulation parameters, liver and kidney function and ECG were all normal and no allergic history was found. The result of polysomnogram indicated apnea recurrent 5 times during
temperature was 37.0. On the fifth day after surgery, the whole muscle soreness remitted partly, urine was faint yellow, and alkalize urine; fluid infusion and diuretic to ensure urine volume micturition or urodynia and osphyalgia. Nephrologists suggested His urine was tawny without urgent urination, frequent especially when he swallowed, the whole body muscle soreness.  

At 11:40 am, an ABG done showed: pH 7.199, PaCO\textsubscript{2} 6.13 mmHg, PaO\textsubscript{2} 113.5 mmHg, HCO\textsubscript{3} - 23.4 mEq/L, BE -5.69 mmol/L, SO\textsubscript{4} 98%, K\textsuperscript{+} 4.45 mmol/L, Ca\textsuperscript{2+} 1.073 mmHg. Surgery ended at 12:00 with temperature 37 and EtCO\textsubscript{2} 45mmHg. The patient recovered at 01:05 pm with temperature 37.7 and EtCO\textsubscript{2} 56mmHg. The patient received cholecystectomy under general anesthesia and no significant complication was found. The surgery day is On October 30, 2014

The patient was back to the ward after careful observation in PACU. First day after surgery the patient's highest temperature was 37.1. He complained about pharyngeal pain and discomfort especially when he swallowed, the whole body muscle soreness. His urine was tawny without urgent urination, frequent micturition or urodynia and osphyalgia. Nephrologists suggested alkalize urine; fluid infusion and diuretic to ensure urine volume reach 300ml/h. On the third day after surgery, the whole body muscle soreness remitted partly, urine was faint yellow, and temperature was 37.0. On the fifth day after surgery, the whole body muscle soreness remitted completely, urine also was faint yellow, temperature was 37.2. Serum myoglobin and creatine phosphokinase rose severely in 24 hours post operation then gradually reduced to normal. Renal function, coagulation function and electrolyte were normal during the postoperative period. 11 days after surgery the patient discharged without any complication or discomfort.

In this case, we found multiple heterozygous mutations by DNA analysis in the patient's RYR1 Figure (1a-1d). The consequence of his younger sister was negative, but his parents' gene mutation was found in RYR1 which also related to MH (Table 2).

### DISCUSSION

Clinical presentations of MH are various from slight or moderate symptoms to fatal MH crises cause by severe skeletal muscle hyper metabolism and rhabdomyolysis [6]. However, it was an atypical MH in this case. The clinical manifestations were characterized by masseter muscle rigidity (MMR) following intravenous succinylcholine, moderate rise of EtCO\textsubscript{2} and slightly increase of temperature. It is very interesting that the patient did not suffer from severe hyperpyrexia at all. Some other cases also reported no significant changes in body temperature, particularly when appropriate treatment was started early [7,8]. The moderate clinical presentation due to the early suspicion of MH and rapidly appropriate treatment. The severity of MH also depended on the dose of triggering agents, such as volatile anesthetics or succinylcholine, which was given to the patient [9]. A small amount of sevoflurane was inhaled during the beginning of the surgery in this case.

A clinical grading scale considered a useful tool for detection of MH [10]. Among seven criteria of this score there are four criteria, i.e. masseter spasm following succinylcholine (15 points), EtCO\textsubscript{2} >55mmHg with appropriately controlled ventilation (15 points), Arterial pH <7.25 (10 points) and inappropriate sinus tachycardia(3 point) were present in this case, leading to a total score of 43 corresponding to MH rank 5, classified as 'MH Very likely'[11].

Compared with the invasive contracture test, DNA analysis is becoming more and more important over the last decades [12,13]. As we known, there are three mammalian isoforms of the ryanodine receptor: RYR1, expressed in skeletal muscle; RYR2, predominant in heart muscle; RYR3, found in skeletal and smooth

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muscle [14,15]. Many mutations in the RYR1 gene have been connected with a susceptible to MH [16]. Multiple mutations in RYR1 which are associated with MH, were found in the patient's and his parents' DNA [17]. So it's very essential to ask the patient questions about individual or family history of MH in every pre-anesthetic interview.

The early suspicion of MH and rapidly appropriate treatment lead to favorable prognosis of a MH crisis [6]. In this case, MH was suspected by masseter muscle rigidity (MMR), high level of EtCO₂, and slightly increase in temperature then immediate treatment (including stopping the trigger agents, removing the volatile anesthesia from the anesthesia machine hyperventilating with 100% oxygen at maximum fresh gas flow, turning to total intravenous anesthesia) was initiated [18]. Dantrolene as a specific medicine for crisis of MH can decrease the mortality rate prominently [19,20]. In this case, the favorable prognosis without administration of dantrolene was due to early diagnosis and treatment.
Without severe Hemodynamic changes and pulmonary complications, after operation, the patient was recovery and exubated successfully in the operation room. Patient was carefully monitored and in investigatd in postoperative period and a severe rise in creatine phosphokinase and myoglobin level was recorded at 24 hours. Most patients were killed by acute renal failure cause by high level of myoglobin during the post-operation. Thus, fluid infusion and diuretic were employed to prevent acute renal failure in our case [8].

In conclusion, the prognosis of a MH crisis depends on close observation and early diagnosis of MH. Rapidly appropriate treatment including initial and symptomatic treatment is crucially important as well. Masseter muscle rigidity (MMR) following trigger agents (succinylcholine and volatile anesthetic), increase in temperature and EtCO$_2$ should be suspicious MH. Treatment including initial and symptomatic treatment is crucially important as well. Masseter muscle rigidity (MMR) following trigger agents (succinylcholine and volatile anesthetic), increase in temperature and EtCO$_2$ should be suspicious MH.

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REFERENCES