A patient with known malignant hyperthermia susceptibility and latex allergy for robotic-assisted laparoscopic vaginal hysterectomy

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Abstract
In patients with known susceptibility to malignant hyperthermia, the necessity of avoiding triggering agents can complicate general anesthesia. Robotic-assisted laparoscopic procedures are associated with elevated carbon dioxide levels further complicating diagnosis of MH in susceptible individuals undergoing such procedures. In the following case, a patient with known malignant hyperthermia susceptibility and concurrent latex allergy presents for robotically assisted laparoscopic vaginal hysterectomy. The management of this patient is discussed.

IMPLICATIONS
In the malignant hyperthermia susceptible patient, diagnosis of MH under general anesthesia may be complicated by elevated carbon dioxide levels resulting from peritoneal insufflation during laparoscopy. Careful monitoring of end-tidal CO$_2$ and blood gas measurements coupled with communication with the surgeon is essential to ensure patient safety.

INTRODUCTION
The classic early signs of MH include tachycardia, tachypnea if the patient is allowed to breathe spontaneously, and markedly increased carbon dioxide production related to the hypermetabolic state. During robotic assisted laparoscopic dissection, carbon dioxide levels can increase significantly in any patient due to peritoneal insufflation. In the MH susceptible patient undergoing robotic surgery it may be difficult to determine the origin of elevated end tidal carbon dioxide levels. Frequent sampling of arterial blood gas measurements allows for the establishment of trends, is a more accurate indication of CO$_2$ levels, and provides pH data as well. As the case progresses, it is often necessary to reduce insufflation pressures to avoid excess CO$_2$ retention, which can be hard to reduce via increased ventilation in the steep Trendelenburg position. If MH is left untreated, a mixed respiratory and metabolic acidosis will develop, increased oxygen consumption may lead to hypoxia, and hyperthermia, muscle rigidity, and rhabdomyolysis, will occur. It is therefore essential to make the diagnosis as early as possible and begin treatment immediately.

CASE REPORT
A 38 year old, 72 kg Caucasian female with cervical dysplasia presented for robotic-assisted laparoscopic vaginal hysterectomy. Her past medical history was significant for idiopathic thrombocytopenic purpura (ITP) for which she had received multiple platelet transfusions, plasmapheresis and a splenectomy, which was performed ten years prior. She had received a general anesthetic for the splenectomy during which she developed intraoperative hyperthermia, muscle rigidity and metabolic acidosis requiring an extended stay in the intensive care unit. This episode was suggestive of malignant hyperthermia and she was referred for muscle biopsy but refused testing since her father had been tested following an episode of malignant hyperthermia he had sustained and was already known to be susceptible. It is unclear why she received a triggering anesthetic given her family history of malignant hyperthermia. Her past surgical history was also significant for an uneventful tonsillectomy performed as a child under general anesthesia, two cesarean section deliveries performed under spinal anesthesia and multiple orthopedic procedures secondary to injuries received as a result of a motor vehicle accident. All of the orthopedic procedures were performed after her episode of malignant hyperthermia and were conducted using regional techniques. At the time of her presentation for this surgery she reported taking no medications. She reported an allergy
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to latex, penicillin and sulfa drugs, all of which presented with anaphylaxis, and a severe rash associated with furosemide administration. As well, she reported a history of severe muscle cramping associated with ingestion of ibuprofen, percocet, darvocet, percodan, stadol and codine. She did not smoke, drink or use recreational drugs. Her family history was significant for malignant hyperthermia susceptibility as outlined above.

On physical exam the patient stood 163 cm tall and weighed 73 kg. Her pre-operative vitals were all within normal limits. She was found to have a Malimpati class 2 airway, a thyromental distance of 6.5 cm and a mouth opening greater than 5 cm. She reported no loose, chipped, missing or broken teeth and she had no limitations on her neck range of motion. Her pulmonary and cardiac exams were both unremarkable. A pre-operative complete blood count had been drawn and was remarkable only for a hematocrit of 34%.

Given her known susceptibility to malignant hyperthermia and need for general anesthesia, a total intravenous anesthesia (TIVA) technique was planned, using an infusion of propofol and remifentanil. After removing the vaporizer from the anesthesia machine and replacing the circuit and CO₂ absorber cartridge, the system was flushed with 100% oxygen at 10 liters per minute flow for 5 minutes. Additionally, a completely latex free set-up was used.

After application of a non-invasive blood pressure cuff, pulse oximetry, and electrocardiogram leads, a 20 g intravenous was placed in the left hand. An uneventful induction was performed with propofol, followed by vecuronium and intubation with a 7.5 endotracheal tube. Once the endotracheal tube had been secured and its position in the trachea verified, a bispectral index (BIS) monitor was applied and an infusion of propofol and remifentanil was begun. A 20 gauge arterial catheter was placed in the right radial artery for intraoperative blood gas measurements and a second 18 gauge intravenous line was placed in the right antecubital fossa. The patient was ventilated with a mixture of 80% oxygen in air using the pressure-regulated volume guarantee mode and 5 centimeters of positive end expiratory pressure was applied as the patient would be placed in steep Trendelenburg during the robotic portion of the procedure.

Temperature was monitored using both esophageal and bladder probes and serial blood gas measurements were obtained throughout the anesthetic. The baseline arterial blood gas measurement performed after arterial line placement showed a pH of 7.43 and a CO₂ of 33 mmHg, at which time the end tidal CO₂ was 30 mmHg with an esophageal temperature of 36.2 degrees centigrade. Blood gas measurements were then repeated at one hour intervals. The second arterial blood gas measurement was obtained 15 minutes after peritoneal insufflation and showed a pH of 7.37 and a CO₂ of 32 mmHg, at which time the end tidal CO₂ was 35 mmHg and the esophageal temperature was 36.7 degrees centigrade. Over the following hour the end tidal CO₂ increased progressively, requiring an increase in ventilatory rate and, after discussion with the surgeon, a decrease in the peritoneal insufflation pressure. The third arterial blood gas measurement was obtained 75 minutes after peritoneal insufflation and showed a pH of 7.36 and a CO₂ of 39 mmHg, at which time the end tidal CO₂ was 35 mmHg and the esophageal temperature was 37.1 degrees centigrade. The procedure was completed in three and one half hours, after which the patient was allowed to wake up and transferred to the post-anesthesia care unit for observation. She was subsequently transferred to the general ward and discharged with no anesthetic sequelae.

DISCUSSION

Malignant Hyperthermia (MH) is a genetic disorder, which in most cases is caused by a defect in the ryanodine receptor. The RYR-1 gene, which codes for the receptor, is located on chromosome 19q13.1. Over 90 mutations have been identified in this gene and 28 are known to be causal for MH. These mutations in the skeletal muscle RYR-1 gene can render affected individuals susceptible to a hypermetabolic state when exposed to triggering agents such as the volatile anesthetics and depolarizing muscle relaxants. The inheritance of MH is considered to be autosomal dominant with variable penetrance, and the discovery of more than one genetic locus in some families indicates that it is a heterogenetic disorder. This hypermetabolic state results from a dramatic release of calcium from the sarcoplasmic reticulum and is characterized by hyperthermia, muscle rigidity and severe metabolic acidosis which can lead to death if untreated. It is possible that as many as 1 in 2,000 individuals in the general population may be MH susceptible, however the incidence of documented MH episodes is considerably lower and estimates range from 1 in 15,000 to 1 in 60,000 administered anesthetics. Since the volatile anesthetics are known triggers of MH, the anesthesia machine needs to be prepared before use on an
MH susceptible individual. This can be achieved by removal or sealing of the vaporizers, changing the absorber cartridge, replacement of the fresh gas outlet hose, and use of a disposable circuit. The machine should then be flushed with a flow of 10 L/min for at least 5 minutes. It is possible that, the volatile agent's concentration may again increase once the flow rate is reduced, and some of the newer machines require a constant high flow to minimize volatile agent concentrations, so the gas analyzer must be checked after flushing to ensure absent levels of volatile anesthetic agent.  

As well as avoiding the volatile agents, depolarizing neuromuscular blocking agents such as succnylcholine should not be used. Providing that triggering agents are avoided, it is not necessary to pre-treat susceptible patients with dantrolene, but a fully stocked MH cart with enough dantrolene to treat the patient based on weight should be readily available in the operating room.

While latex allergy has been associated with MH susceptibility in patients with Lobstein's syndrome, there is no known association with Idiopathic Thrombocytopenic Purpura and it is likely that in this case, the coexisting pathologies are unrelated and simply further complicate the anesthetic management of this at-risk patient. Her history of severe muscle cramping associated with ingestion of ibuprophen, percocet, darvocet, percodan, stadol and codine is suggestive of a non-specific myopathy which may or may not be related to her MH susceptibility, but which did present further complications for postoperative pain management. Awake triggering of MH has been associated with exercise and heat stroke in humans, and it has been suggested that pain itself and the associated physical stress may be a trigger for MH. In any case, adequate postoperative pain control is mandatory, and this patient's history leaves few options. The prospect of an epidural for postoperative pain control was discussed with both the patient and the surgeon, but declined by the patient as only moderate postoperative pain was expected. Fentanyl was administered intraoperatively and the surgical trocar insertion sites were infiltrated with 0.25% bupivicaine. In the post-anesthesia care unit the patient reported moderate discomfort which was adequately treated with Tylenol.

References
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