Malignant hyperthermia: case report

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Abstract

The incidence of malignant hyperthermia (MH) in children is much higher than in adults. The management of children during dental treatment may require general anesthesia. Better understanding of the disease helps to detect the susceptible cases, make an early diagnosis, give effective treatment, and minimize the mortality and morbidity.

The management of an acute hyperthermic crisis during surgery is reported. The crisis was aborted mainly by intravenous dantrolene sodium. The patient recovered completely.

Malignant hyperthermia; a disease of pharmaco-genetic origin, can have a fatal outcome. Increase in core body temperature, cardiac arrhythmias, muscle rigidity, metabolic acidosis, and elevated serum enzymes are characteristics. The disease usually is encountered during the administration of general anesthesia. Unless life saving measures are initiated during an episode, the patient's survival is doubtful. Dentists who administer general and local anesthesia and those who admit patients for treatment in hospital operating rooms should be familiar with the signs and treatment of this disease.

Case Report

S.G. is a 12-year-old, 30 kg boy scheduled for multiple restoration of dental caries under general anesthesia. At six months of age, he developed postvaccination meningo-encephalitis, leaving him blind and mentally retarded. No family history of intolerance to anesthesia was elicited. Review of systems showed no other significant abnormalities, and serum electrolyte, enzymes, routine blood cell counts, coagulation profile, and urinalysis were within normal limits. Pulse was 92/min, respiration 18/min, and blood pressure was 120/60 torr.

Meperidine, 25 mg, hydroxyzine, 25 mg and atropine, 0.4 mg were given intramuscularly one hour before the operation. Routine monitoring included precordial stethoscope, electrocardiogram (ECG), blood pressure (BP), and rectal temperature. A thermobanket was used. Anesthesia was induced with halothane by gravity, then by mask due to patient's apprehension. After the induction, succinylcholine, 60 mg, was given intravenously to facilitate endotracheal intubation. It was followed by generalized muscle fasciculation, difficulty in mouth opening and tachycardia (140–150/min). The trachea was intubated with some difficulty and anesthesia was maintained with halothane.

Tachycardia (140/min) persisted, and occasional premature ventricular contractions were noted shortly after the operation began. Rectal temperature remained unchanged, but surgeons complained of difficulty in opening the mouth. Fifteen minutes later the temperature rose from 36.0 to 38.8 °C, then to 42.2 °C in another 15 minutes with heart rates of 150–160/min.

The surgical procedure was stopped, all anesthetics were discontinued, and the patient was hyperventilated (minute volume 12 L) with 100% oxygen. Dantrolene sodium* (30 mg IV push) was given. Other immediate treatment included ice cold IV fluid infusion, cooling blanket, and crushed ice around the body. Arterial, CVP, and Foley catheters were inserted. Severe metabolic and respiratory acidosis (base excess—18 mEq/liter) with a pCO2 of 115 torr was noted and treated with sodium bicarbonate. Serum creatinine phosphokinase (CPK), potassium (K+) and calcium were 69,200 units, 4.6 mEq/liter and 8.7 mg/dl, respectively. The temperature was lowered to 38.8 °C in 45 minutes.

Approximately 110 minutes from the onset of rising temperature, the patient was able to open his eyes and was extubated off the ventilator.

Thereafter, dantrolene was given 30 mg IV every six hours for 2½ days in the ICU to prevent recurrence and the rectal temperature was monitored continuously. At the end of this period, all invasive monitoring lines were removed and cultures from the cannulae and urine showed negative growth. The patient was transferred to the pediatric ward on the fourth postoperative day. Oral a Dantrium, Norwich-Eaton Pharmaceuticals, Norwich, NY.
Dantrolene was continued (30 mg PO every six hours) for five more days. On the twelfth postoperative day the patient was discharged from the hospital with a CPK of 95 units.

Discussion

The incidence of MH is approximately 1/14,000 anesthetics in children and 1/52,000 in adults. The initial mortality rate of 80% has been reduced to 53% by increased awareness and early recognition and treatment. MH is a genetic disease of autosomal dominant inheritance with variable penetrance and is present in varying degrees in siblings of families inheriting the disease.

Families susceptible to MH in North America appear to be clustered in Wisconsin, Toronto, and Nebraska. There is usually little or no functional impairment of those MH-affected people and their families. Myopathy is detectable only by specific testing. Some affected people have obvious musculoskeletal abnormalities but diseases such as myotonia, polymyositis, and muscular dystrophy do not show the characteristic abnormalities found in muscle of MH-susceptible patients, and most of those afflicted do not develop MH.

MH can be triggered by several potent volatile anesthetic agents, but the onset is usually more abrupt when succinylcholine is used alone or in conjunction with volatile agents. The sarcoplasmic reticular membrane is probably abnormal, and depolarization results in greater than normal amounts of Ca\(^{2+}\) moving into the cytoplasm. Active reuptake of Ca\(^{2+}\) also is blocked, resulting in an inability of the muscle to relax. Various mechanisms of heat production in MH have been proposed. These include aerobic metabolism, glycolysis and hydrolysis of high-energy phosphates involving ion transport, and contraction-relaxation and neutralization of the hydrogen ion. Some authors have suggested that preoperative stress associated with inadequate sedation, or intraoperative stress with light anesthesia might contribute to the initiation of MH in the absence of triggering anesthetic agents in MH-susceptible subjects.

A careful medical history is most important in finding susceptible patients and is aided by recognition of abnormal CPK. Laboratory tests show contracture of biopsied skeletal muscle in response to low concentration of caffeine, and halothane and abnormal platelet ATP level.

The choice of drugs and preparation for susceptible patients include the use of vapor-free anesthetic machine and breathing system, nitrous oxide, thiopental and other barbiturates, narcotics, droperidol, and pancuronium. In order to reduce stress for the pediatric patient, it is important that he become familiar with his surroundings and be accompanied by a family member. The pediatric patient should be admitted at least 24 hours before surgery to accomplish the above and to administer dantrolene sodium orally at that time. Preoperative preparation includes oral dantrolene 4 mg/kg/day in three or four divided doses within 24 hours before general anesthesia, heavy premedication with tranquilizers (no phenothiazines), barbiturates and/or narcotics. Atropine should be given intravenously in small doses only as needed.

Dantrolene is a recent addition to the armamentarium for treating MH patients. Following successful animal studies, its use in humans proved successful. The suspected mode of action is dissociation of excitation-contraction coupling in skeletal muscle, and attenuation of calcium release from terminal cisternae of the sarcoplasmic reticulum.

Monitoring of MH-susceptible patients during surgery should include electrocardiogram, temperature, and other routine monitoring for anesthesia. After induction, an arterial line is inserted for direct BP recording and arterial blood gas determinations. A central venous pressure line is inserted for pressure measurements and venous sampling of electrolytes. The patient is catheterized so urine output can be measured.

Clinical diagnosis is based upon muscle rigidity, particularly masseter spasm, following the administration of succinylcholine or potent inhalation anesthetic agents, unexplained tachycardia or arrhythmias, rapid rise in body temperature (1°C/5 min), hyperkalemia, and acidosis. Suggested criteria for the diagnosis include a base excess of less than minus 5mEq/liter and a PaCO\(_2\) greater than 60 torr without reasonable explanation.

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References