Case Report

Acute life-threatening arrhythmias caused by severe hyperkalemia after induction of anesthesia in an infant with methylmalonic acidemia

Pei-Wen Chao a, Wen-Kuei Chang a, I-Wen Lai a, Chinsu Liu b, Kwok-Hon Chan a, Cheng-Ming Tsao a,

a Department of Anesthesiology, Taipei Veterans General Hospital and National Yang-Ming University School of Medicine, Taipei, Taiwan, ROC
b Division of Pediatric Surgery, Department of Surgery, Taipei Veterans General Hospital and National Yang-Ming University School of Medicine, Taipei, Taiwan, ROC

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Abstract

Methylmalonic acidemia (MMA) is a very rare genetic disease of metabolism that progressively leads to neurological and renal sequelae. This report describes an unusual case of a patient with MMA who developed severe hyperkalemia and severe dysrhythmia during anesthesia. A 13-month-old male infant with MMA underwent urgent insertion of a port-a-cath under general anesthesia. A life-threatening arrhythmia suddenly occurred, with severe hyperkalemia (up to 7.4 mmol/L), immediately following induction of anesthesia. Emergent resuscitation was successfully carried out, with a complete neurological recovery after 7 days after surgery. Although MMA is a rare complication, the possibility of severe hyperkalemia should be considered in the differential diagnosis of patients with MMA presenting with wide QRS complex tachycardia. The management and intraoperative complications of this disorder are reported here, and the available literature is reviewed.

Keywords: anesthesia; arrhythmia; hyperkalemia; methylmalonic acidemia; resuscitation

1. Introduction

Methylmalonic acidemia (MMA) is an autosomal recessive error of metabolism of methylmalonyl coenzyme A that occurs in about 1 per 100,000 newborns in Taiwan.1 Most cases of MMA are caused by a deficiency of methylmalonyl-CoA mutase, with patients developing progressive neurological and renal sequelae.2 Hyperkalemia (serum K+ level >5.5 mmol/L) after acute metabolic decompensation sometimes appears even in the phase of normal renal function.3 However, rare reports have been published on anesthesia and intraoperative complications associated with this disorder. Here, we describe a case of a male infant with MMA who suffered from sudden severe hyperkalemia and ventricular arrhythmia just after induction of anesthesia.

2. Case report

A 13-month-old boy, 12 kg in weight, with MMA was admitted because of progressive jaundice lasting for more than a week. The diagnosis of MMA was made after a series of newborn screening tests. The boy underwent a living donor liver transplantation (LDLT) in our hospital when he was 8 months old. After the transplantation, he took tacrolimus, lamivudine, and carnitene supplements for further treatment, and analysis showed amino acid levels within the normal range. Although his renal function was normal, persistent hyperkalemia (about 5.9–6.5 mmol/L) was noted during follow-up.

Anastomotic stricture of the biliary duct was suspected after admission, and percutaneous transhepatic biliary drainage...
(PTBD) was undertaken without incident under general anesthesia to relieve blockages in the bile duct. Unfortunately, the patient developed a postoperative fever and poor appetite with vomiting occurred for the first three days after PTBD, and a peripheral intravenous line could not easily be set up. Therefore, urgent port-a-cath implantation was suggested for long-term parenteral nutrition and fluid supply. Preoperative laboratory investigations showed serum levels of creatinine 94.6 μmol/L, urea nitrogen 23.2 mmol/L, K⁺ 5.8 mmol/L, Na⁺ 135 mmol/L, and C-reactive protein 489 nmol/L.

Upon the patient’s arrival in the operating room, electrocardiography (ECG), pulse oximetry, and automatic noninvasive blood pressure (BP) readings were monitored continuously. A preoperative ECG showed sinus tachycardia with peaked T-waves (Fig. 1A), the BP was 85/42 mmHg, and pulse oximetry gave a value of 100% saturation. We initially set up a peripheral intravenous line over the patient’s frontal area under inhalation anesthesia of sevoflurane and nitrous oxide with an FiO₂ of 0.4. Atropine 0.1 mg and cisatracurium 3 mg were then added to facilitate intubation with an uncuffed endotracheal tube. General anesthesia was maintained with sevoflurane 1.5—2.0% and nitrous oxide 60% in oxygen, with positive-pressure ventilation to keep the end-tidal carbon dioxide concentration at 30—35 mmHg.

About 5 minutes after intubation, wide QRS complex ventricular tachycardia (VT) suddenly appeared, with significant hemodynamic changes (Fig. 1B). The patient’s BP could not be measured using the pressure cuff. Two 10 mg doses of lidocaine were given, but the heart rhythm soon changed to low-voltage electrical activity (Fig. 1C). Meanwhile, emergent resuscitation was performed using cardiac massage and epinephrine. We then immediately set up an intra-arterial catheter in the radial artery, with an arterial BP reading of around 46/27 mmHg. The arterial blood gases showed pH 7.37, PaO₂ 450.7 mmHg, PaCO₂ 27.3 mmHg, base excess −9.5 mmol/L, and serum K⁺ 6.9 mmol/L.

Severe hyperkalemia and metabolic acidosis with respiratory compensation were diagnosed. Therefore, calcium chloride, insulin, sodium bicarbonate, and furosemide were administered to treat the hyperkalemia. About 5 minutes later, wide QRS complex VT resumed at a rate of 233 beats per minute (bpm; Fig. 1D), and the serum K⁺ level was recorded as 7.4 mmol/L. The frequency of VT progressively decreased, the ECG showed sinus tachycardia (Fig. 1E), and the serum K⁺ level decreased to 5.7 mmol/L. The systolic BP recovered to around 80 mmHg. During this episode, the patient’s oxygen saturation remained stable at 94—100%. However, levels of serum creatinine of 196.2 μmol/L and urea nitrogen of 42.8 mmol/L were found. Thereafter, the port-a-cath implantation went smoothly under general anesthesia.

After the operation, the boy was transferred to the pediatric intensive care unit for further evaluation and medical treatment. His recovery was uneventful and his ECG normal. No cardiovascular or neurological complications developed, and he was discharged. He continued to do well during the 6 months of follow-up.

3. Discussion

In considering what led to such extreme potassium shifts in our patient, the potential causes might be divided into two types: chronic and acute. Chronic hyperkalemia usually develops with ineffective potassium excretion, resulting from renal insufficiency, a defect in tubular secretion, and/or medications. In this case, the patient’s serum creatinine level was within normal limits before the operation, suggesting that there was no significant renal insufficiency. Some case studies have also shown persistent hyperkalemia in children with MMA, even though their creatinine clearance was low.²,³ Pela et al reported two patients with MMA with normal glomerular filtration who displayed hyperkalemia and metabolic acidosis; type IV renal tubular acidosis was diagnosed.³ Therefore, it is supposed that hyperkalemia can be caused by acute metabolic decompensation even in the phase of normal renal function.

In addition, it is known that tacrolimus, an immunosuppressant, may lead to calcineurin inhibitor nephrotoxicity and renal tubular acidosis via unknown mechanisms.⁶,⁷ This infant received tacrolimus after LDLT to attenuate the risk of organ rejection. Therefore, it is likely that the tacrolimus might have aggravated the complications of hyperkalemia and metabolic crisis in our case.

The acceptable value for serum K⁺ prior to surgery in order to avoid the life-threatening risk of hyperkalemia has been reported to be 6.0—6.2 mmol/L,⁸ although one newborn with

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Fig. 1. Electrocardiograph changes: (A) normal sinus rhythm with peaked T-wave at 131 beats/min (bpm); (B) wide QRS complex ventricular tachycardia at 146 bpm; (C) pulseless electricity activity; (D) wide QRS complex ventricular tachycardia at 130 bpm; (E) sinus tachycardia at 208 bpm.
a higher serum K⁺ level (>8 mmol/L) had no significant sequelae under general anesthesia.9 Potential causes of perioperative hyperkalemia might be attributed to an excessive potassium load and transmembrane potassium ion redistribution caused by acidosis and drugs.10 Our patient did not receive a massive blood transfusion, potassium supplements, succinylcholine, or beta-blockers, all of which may elevate serum K⁺ levels.

However, children with MMA are at increased risk for metabolic decompensation, particularly with infections, trauma, surgery, anesthesia, and psychosocial stress. Therefore, acidosis, hyperglycemia, and increased levels of stress hormones may develop after induction of anesthesia, and serum K⁺ concentration then becomes transiently elevated. In addition, a rapid deterioration in renal function caused by inadequate fluid intake before operation might have further aggravated the elevation in serum K⁺ concentration in our case. Thus, it is better to keep within the normal limits of potassium level before induction of anesthesia, especially in patients with MMA.

Progressive changes in the ECG manifest as serum K⁺ level increases: a peaked T wave, prolonged PR interval, widened QRS complexes, ventricular fibrillation, and asystole.10 To treat wide QRS VT with a pulse, synchronized electrical cardioversion and/or antiarrhythmic drugs are immediately used if hemodynamic conditions are unstable, but defibrillation should be used if there is no pulse.11 Lidocaine is a class Ib antiarrhythmic that decreases ectopic firing points in the heart to suppress sustained ventricular arrhythmia. However, McLean et al reported that lidocaine precipitated a profound conduction disturbance and asystole in two patients with hyperkalemia.12 Hyperkalemia-induced resting membrane depolarization can inactivate sodium channels and then markedly increase lidocaine binding, potentiating its effects on blocking conduction.13 Thus, it is probable that the patient’s rhythm degenerated to low-voltage electric activity due to lidocaine use.

There is some evidence to suggest a reduced efficacy of antiarrhythmic therapy in the presence of severe hyperkalemia.14,15 Therefore, in the presence of severe hyperkalemia in anesthesia, the serum K⁺ concentration must be directly reduced using calcium to stabilize the myocardial membrane, sodium bicarbonate and glucose plus insulin given to shift K⁺ into the cells, and furosemide administered to effectively promote potassium excretion, rather than following the treatment algorithms for wide QRS complex VT.16

In conclusion, patients with MMA are potentially at risk for perioperative hyperkalemia caused by acute metabolic acidosis and renal insufficiency. Although this is a rare complication, the possibility of severe hyperkalemia should be considered in the differential diagnosis of patients with MMA presenting with wide QRS complex tachycardia.

References