Hypokalemic Familial Periodic Paralysis

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Anesthetic management of patients with hypokalemic familial periodic paralysis has been described. In all of these cases, the surgical team was aware of the diagnosis preoperatively. We describe the perioperative events of a patient with familial periodic paralysis that was undiagnosed at the time of surgery.

REPORT OF A CASE

The patient was a 25-year-old, 80-kg man who was admitted for surgical removal of an extruding wire mesh implant of the left eye and insertion of a new implant in the same eye. The left eye had previously been eviscerated and an implant inserted because of a traumatic injury. Preoperative history and physical evaluation by various personnel failed to reveal any significant findings other than a 10-pack/year history of cigarette smoking. The only past hospitalization was for the previous eye surgery which was done at a different hospital. Anesthetic details about the previous surgery were unknown. The patient stated he was not informed of any problems. A complete blood count, urinalysis, prothrombin time, partial thromboplastin time, creatinine, and SGOT were all within normal limits.

He was premedicated with 100 mg pentobarbital, 75 mg hydroxyzine, and 0.2 mg glycopyrrolate, iv. On arrival to the operating room he appeared calm and in no distress. He was then given 3 mg d-tubocurarine, iv, without adverse effects. Anesthesia was induced with 0.1 mg fentanyl and 400 mg thioental, iv. The trachea was intubated after administration of 120 mg, succinylcholine, iv. Anesthesia was maintained with 50% nitrous oxide and 0.5-1.0% inspired enflurane. Ventilation was controlled manually for the first 15 minutes of anesthesia, while the head was positioned, prepped and draped; the eye was then extensively examined, and the initial incisions made. After this, spontaneous ventilation returned and was assisted manually throughout surgery, which lasted one hour. ECG was monitored continuously and no abnormality or cardiac arrhythmias were noted. Asthmatic temperature remained at 36.5° C throughout the case. At the end of surgery, the trachea was extubated with no evidence of respiratory distress.

Within minutes of arriving in the recovery room, he became agitated and removed his nasal airway. He then appeared to relax. His color was good and ventilation appeared adequate. Within five minutes, he became responsive to verbal stimuli. Fifteen minutes later, he began complaining of weakness and could not move his arms and legs.

With more detailed examination, he was able to speak and breathe easily. He had good control of his head and neck, but was unable to move his arms and legs even with extreme effort. Deep tendon reflexes were absent. There were no pathologic reflexes. Sensation was intact.

With an PaO2 of 0.2, pH was 7.44, PaO2 91.0 mmHg, and PaCO2 46.5 mmHg. Serum electrolyte levels were all within normal limits, except for a potassium level of 3.1 mEq/l. As the patient became more alert, direct questioning revealed that this had happened to him in the past and that this also happened to his brother. These episodes of weakness spontaneously resolved. Episodes of weakness in the past, the present weakness of the extremities without sensory loss, and hypokalemia led to the presumptive diagnosis of hypokalemic familial periodic paralysis. A neurologist was consulted and his impression was the same. The family physician was contacted by telephone. He had been following the patient, his brother, and father for periodic paralysis. The father had not had an attack since age 40 years. The patient had been experiencing episodes of weakness since age 14 years. Weakness always involved only the extremities and never required hospitalization. During the past year, he experienced three episodes, all preceded by large meals and heavy alcohol intake. The anesthesiologist from the previous hospital was contacted and said he experienced no problems with the patient.

D5 in 0.25% saline with 40 mEq of potassium chloride per liter was administered at the rate of 120 ml/h. After four hours, the patient had regained some movement of his arms and legs. Since he was in no respiratory distress and muscular strength was returning, no further treatment was deemed necessary. The patient was returned to his room with one-on-one nursing care. The next morning he was stronger but could not ambulate without assistance. His potassium was 2.7 mEq/l. At this time the iv infusion was changed to 0.25% saline with 40 mEq potassium chloride per liter. He steadily improved and was ambulatory by mid afternoon. The next morning his potassium was 5.0 mEq/l. He was discharged on the second postoperative day with follow-up by his family physician. It was suggested he obtain a Med-Alert bracelet.

DISCUSSION

Case histories characterized by episodes of recurrent muscle weakness have been recognized since 1882.
Presently, at least four syndromes of this type are recognized. These syndromes include hypokalemic periodic paralysis, which is represented by our case; hyperthyroidism with periodic paralysis, which is also associated with hypokalemia; congenital paramyotonia of the tongue and hypothenar muscles; and adynamia episodica hereditaria which is clinically very similar to hypokalemic periodic paralysis, except that it is associated with hyperkalemia and generally occurs in a younger age group. Muscle weakness in the hypokalemic variety probably is triggered by a massive uptake of potassium into the muscle cells and a subsequent lowering of serum potassium. Drugs that decrease serum potassium can precipitate an attack of weakness. Currently, the drug of choice for prevention of attacks is acetazolamide which produces a metabolic acidosis, thereby increasing serum potassium. Spironolactone, a potassium-sparing diuretic, also is used.

Siler and Discavage described the administration of three separate anesthetics to a single patient with hypokalemic periodic paralysis. There were no major problems intraoperatively. Several hours after an emergency appendectomy, their patient experienced complete muscle paralysis and required endotracheal intubation and controlled ventilation. His serum potassium level was 2.0 mEq/l. Despite correction of the potassium to 4.0 mEq/l, the patient required controlled ventilation for 36 h. Horton described 21 anesthetic experiences with a family known to have hypokalemic paralysis. This family had no intraoperative, and at most, moderate postoperative problems with muscle weakness. There was one intraoperative death, but this was associated with hypertension and a ruptured cerebral aneurysm.

The major emphasis in anesthetic management of affected patients is the prevention of episodes of muscle weakness. Guidelines for this have been suggested. The patient should be instructed not to overeat the night before surgery. Large carbohydrate meals are often followed by weakness. As glucose enters the cells, potassium does also. The most certain method of inducing an attack for diagnostic purposes is the administration of glucose and insulin. Preoperative serum electrolyte concentrations should be obtained. If the patient is receiving a diuretic and the potassium level is even slightly below normal, it should be corrected. The iv administration of dextrose and sodium should be limited as much as possible. Severe attacks of paralysis are usually preceded by sodium retention, and sodium loading has been shown to precipitate attacks. Hypothermia can precipitate attacks so careful temperature monitoring is required. ECG signs of hypokalemia are more pronounced than in normal persons with the same level of hypokalemia. Careful monitoring can detect early potassium changes. The relationship between muscle relaxants and postoperative weakness is unclear, so they should be avoided if possible.

Although the patient had been diagnosed as having hypokalemic familial periodic paralysis preoperatively, he did not relate this to any member of the hospital staff. If this history had been obtained, several of the guidelines which were not followed could have been. The patient received 5% dextrose in normal saline as his iv infusion. A solution such as 0.25 normal saline should have been used. Rectal or esophageal temperature, rather than axillary, should have been used, as they probably are better measures of core temperatures. A warming blanket should have been in place and intravenous solutions warmed. Neuromuscular blocking drugs could have been avoided. The postoperative hypokalemia, despite potassium replacement, may have been due to the iv infusion of dextrose.

Two important points are illustrated by this case. Our patient’s muscular function improved postoperatively even though his serum potassium level decreased. The patient described by Siler and Discavage did not improve even though his serum potassium was returned to within normal limits. This suggests there is no direct relationship between serum potassium levels and muscle weakness. Closely monitoring serum potassium levels may therefore be of limited value in predicting extent of muscular weakness.

Our patient, like others reported, appeared to experience no intraoperative disease-related problems. He did experience postoperative weakness. This is true of some, but not all, of the cases previously reported. We did not follow the above suggested guidelines; the other authors did. The common factor in all the reported cases of weakness is anesthesia and surgery. No matter how limited the operative procedure, anesthesia and surgery apparently does something, as yet undefined, to initiate an episode of weakness. We therefore feel that the above guidelines should be followed, but that adherence to these guidelines does not insure against the development of postoperative weakness. Prolonged postoperative monitoring is essential even in the presence of a benign intraoperative course and normal serum potassium levels.

References
Zomepirac is an orally administered non-narcotic analgesic, which is a prostaglandin synthetase inhibitor. Its analgesic properties appear to be similar to those of morphine.\(^1\,^2\) Our objective was to assess the efficacy of zomepirac as an orally administered analgesic premedication in short-stay patients, where an effective non-narcotic analgesic might be particularly useful for prophylaxis of postoperative pain. Codeine was chosen for comparison because it is often used for the treatment of postoperative pain.

**METHODS**

Institutional approval of the protocol was obtained and all patients gave written informed consent. Eighty-eight patients were studied, 40 of whom were scheduled for elective surgical removal of impacted wisdom teeth, and 48 for laparoscopic sterilization. All patients studied were A.S.A. I, aged between 18 and 45 years old (dental patients, 23.2 ± 1.08 years; laparoscopic patients, 31.2 ± 0.75 years), and weighed 45–85 kg (dental, 69.4 ± 1.72 kg; laparoscopic, 62.2 ± 1.20 kg). Patients who were taking medications that could confuse pain assessments were excluded from the study.

Patients were stratified by surgical procedure and randomized to one of two premedication regimens: 100 mg zomepirac or 60 mg codeine administered orally approximately 30 minutes prior to surgery. A double-blind design was assured by the use of drug capsules of identical size and appearance, labeled by number alone. All patients received a standardized general anesthetic; dental patients received thiopental for induction, succinylcholine to facilitate intubation of the trachea, and anesthesia was maintained with enfuran and 70% nitrous oxide with spontaneous respiration; patients undergoing laparoscopy received a similar anesthetic except that ventilation was controlled and muscle relaxation was achieved during maintenance with a succinylcholine infusion. The use of narcotics or local anesthetics was avoided.

All observations were made by one nurse observer. Pain intensity was assessed using a vertical 10-cm visual analogue scale\(^3\) (VAS), and a five-point ordinal scale\(^4\) comprising the following categories: none (0), mild (1), moderate (2), severe (3), and very severe (4), the numerical score being subsequently assigned to facilitate statistical analysis.

Following preoperative practice with the analogue and ordinal scales, pain assessments were made postoperatively at 1.5, 2, 3, 4, 5, and 6 hours following premedication, or until the patient was judged fit for discharge from hospital. Recovery was assessed as the interval from termination of anesthesia until the patients could first recall their date of birth ("recovery time"), and the patients were fit for discharge from the hospital as judged by a modified Fay\(^5\) scoring system ("street fitness time").

All patients received additional postoperative analgesic drugs on request after assessment by the nurse observer. The time of request was recorded. A standardized regimen was used which comprised 25 mg iv meperidine during the first postoperative hour, and 60