ANAESTHESIA MANAGEMENT FOR A PATIENT WITH FAMILIAL HYPOKALEMIC PERIODIC PARALYSİS

Case Report

FAMILIAL HIPOKALEMIK PERİYODİK PARALİZİLİ HASTADA ANESTEZİ YAKLAŞIMI

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ABSTRACT

Familial hypokalemic periodic paralysis(FHPP),is a rare inherited autosomal dominant genetic disease characterized by attacks of severe muscle weakness and flaccid muscle paralysis. It is reported that anesthesia, stress , surgery, hypothermia, high carbohydrate intake, infections, low potassium levels, menstruation, pregnancy and exercise was induced the attacks in FHPP. In this case report, our anesthesia management and İntesive Care Unit (ICU) follow up in a craniotomy patient with FHPP has been previously described.

Keywords: Familial hypokalemic periodic paralysis , general anesthesia ,craniotomy.

ÖZET

Familial Hipokalemik Periyodik Paralizi (FHPP); ataklar şeklinde ciddi kas güçsüzlüğü ve flask kas paralizisi ile seyreden, genetik olarak otozomal dominant geçişli, nadir görülen bir hastalıktır. Anestezi, stress, cerrahi, hipotermi, fazla karbonhidrat alımı, enfeksiyonlar, düşük serum potasyum düzeyi, menstruasyon, doğum ve egzersizin FHPP'yi tetiklediği bildirilmiştir. Bu olgu sunumunda intrakranial kitle tanısıyla kraniotomi uygulanan hastadaki anestezi deneyimimizi ve yoğun bakım takibimizi sunmaktayız.

Anahtar kelimeler: Familial Hipokalemik Periyodik Paralizi, Genel Anestezi, Kraniotomi.Batık tırnak, enfeksiyon

INTRODUCTION

Familial hypokalemic periodic paralysis (FHPP), is a rare inherited autosomal dominant genetic disease characterized by attacks of severe muscle weakness and flaccid muscle paralysis. Anesthesia, emotional stress , surgery, hypothermia, high carbohydrate intake, infections, menstruation, pregnancy and exercise induced the attacks. General anesthesia, postoperative stress, dextrose containing intravenous solutions, hypokalemia and neuromusculer agents are related with the postoperative attacks . In this report, we described the anesthesia management and postoperatif ICU follow up in a craniotomy patient with FHPP.

CASE REPORT :

The patient was a 42 year-old-man (107 kg/180 cm) presenting for craniotomy of intracranial tumor located in the right ventricul. The patient said that attacks were triggered by emotional stress and exercise. İn his history he has no respiratory distress or oropharyngeal muscle paralysis, but he has bronchial asthma , sleep apnea, gastroesophageal reflux disease and numbness in the left arm and leg becouse of intracranial tumor. İn the operating room heart rate, noninvasive blood pressure , oxygen saturation and skin temperature were monitored.

Anesthesia was induced with intravenous thiopental 5 mg / kg and fentanyl 100 mcg. After the ventilation with a face mask that is comfortable iv cisatracurium 0.2 mg / kg was given and the patient was intubated. Anesthesia was maintained with propofol 100 mcg / kg / min and remifentanil 0.25 mcg / kg / min was achieved by infusion . The patient underwent invasive arterial and central venous pressure monitoring . Also the skin and nasopharyngeal temperature were monitored. Pre-operative evaluation of potassium (K+) levels were found to be normal and during the operation 50 mEq of potassium phosphate in 250 ml of normal saline 10 mEq / hour iv infusion administered . During the operation normal levels of K + concentration (3.65 to 4.33 mEq / dL) remained . Body temperature and EtCO2 was monitored. Operation time was 5 hours .

During the operation, the patient remained hemodynamically stable . Postoperative analgesia was provided with paracetamol 4x1 g (iv) and morphine HCl 1 mg (iv). The patient was extubated with no problem . During the ICU period patient was hemodinamically stable and Glasgow Coma Scale was 15. We checked the blood glucose, potasyum levels and potasyum phosphate 10 meq was replace in ICU. Visual Analogue Scale was 0-3 point. And the patient did not require additional analgesic and antiemetic therapy. There was no postoperative paralysis or muscle weakness observed. The patient was transferred to the service at the end of the 24 hour follow up in İCU and after the 48 hour service follow up the patient was discharged without any complication.

DISCUSSION

This case report is the first to describe the anesthetic management in craniotomy operation with FHPP.

FHPP is an autosomal dominant disease that is characterized by muscle weakness and flaccid muscle paralysis. (1-8). İn many cases perioperative period attacks are serious enough to require emergency intubation and mechanical ventilation may be remained. (3).İn a small number of cases respiratory arrest, and death have been reported. (9).

Anesthesia, hypothermia , postoperative stress, high carbohydrate intake, long-acting neuromuscular muscle relaxants and iv solutions containing dextrose are associated with postoperative attacks . (10). İn clinical management of this patient we must avoid the factors that may attack the FHPP and keep the potassium levels in normal rates.(3,4,8-10).

In FHPP patients depending on the patient's condition and the type of surgery, regional anesthesia techniques can also be applied to . Due to high-dose local anesthetic epidural or spinal anesthesia may be preferable to avoid muscle weakness . (11-14).

In patient with FHPP electrolyte levels are important and should be checked. Monitoring the serum potassium levels in a normal range should be maintain and if needed potassium replacement is important.(1-17).İn this case we measured the potassium levels closely (every 1 hour in operation and every 4 hour in İCU) and observed no clinical signs of muscle paralysis.

ECG monitoring to observe the arrhythmias is essential. During hypokalemic episodes patients with FHPP are

prone to develop ventricular arrhythmias. (16) İn our patient no arrhythmia was developed.

Hypothermia can precipitate the attacks so careful temperature monitoring is required. Malignant hyperthermia was also reported in many cases. Lambert et al. reported a patient with FHPP who suffered a hypermetabolic crisis consistent with malignant hyperthermia during an operation(10).Occasionally, general anaesthesia using volatile anaesthetics and succinylcholine results as safe agents in patients with FHPP without malignant hyperthermia(8-10).Malignant hyperthermia is a rare clinical progress in a patient with FHPP. For this reason we prefered total intravenous anaesthetic (TIVA) technique with propofol and remifentanil. The real benefit of this technique was a rapid and complete recovery of consciousness and muscle strength. Increased pain following termination of the short-acting narcotic may have triggered postoperative muscle weakness. However, combination of an opioid and paracetamol can allowed an uneventful recovery in our patient.

Since many patients report an increase in weakness following a large carbohydrate meal, patients should have a light supper the evening before surgery. The amount of glucose infused should be kept at a minimum, as glucose may promote the intracellular movement of potassium. Dextrose containing solutions administered during surgery should be avoided and normal saline (%0.9)should be prefferred.(1,8-17)

Preoperative to alley anxiety good premedication, avoidance of stress and postoperative adequate analgesia is vital in preventing attacks.

The role of neuromuscular blocking agents in FHPP is controversial. Postoperative muscle weakness has been described following the use of depolarising muscle relaxants(10).Therefore, avoidance of neuromuscular blocking agents in FHPP has been recommended_(4,8). However, different non-depolarising muscle relaxants have been used successfully during general anaesthesia for patients with FHPP (11-14).

In conclusion, we report the successful anaesthetic management and ICU follow up in a patient with FHPP who underwent craniotomy surgery. Avoiding the triggering factors of FHPP and keeping the potassium level in normal range are the most critical issues in the management of such patients.

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