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Original Research Paper

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ANESTHETIC CHALLENGES IN A PATIENT WITH MYOTONIA CONGENITA- A CASE REPORT

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ABSTRACT **Background:** Myotonia congenita (MC) was first described as a skeletal muscle disorder by Thomsen in 1876. As a result of the mutation of the chloride channel gene (CLCN1), which is on the 17th chromosome, patients suffer from muscle contractility and fatigue. Autosomal dominance and autosomal recessive heredity gene may present in this disease. The prevalence of MC is 1/100000 all over the world. In this disease, muscle contraction, hypertrophy and contracture are present without muscle weakness. Dysphagia, aspiration and cardiomyopathy are frequent symptoms. The most important complication is malignant hyperthermia in these patients. **Case Report:** A 27-year-old male who is a known case of myotonia congenita and on Tablet Phenytoin for past 10 years posted for orthognathic surgery. TIVA (Total intravenous anesthesia) was planned for the patient. Precautions were taken to prevent fatal malignant hyperthermia **Conclusion:** For the conduct of anesthesia in a patient with myotonia congenita preoperative evaluation, laboratory tests, relevant consultations, anesthetic drugs to be used, normothermia and ICU follow-up in postoperative period are very important. If anesthesiologist can't obtain dantrolene natrium, TIVA could be a safe solution with the precautions that were present above.

KEYWORDS : Myotonia congenita, malignant hyperthermia, total intravenous anesthesia (TIVA)

INTRODUCTION

Myotonia congenita (MC) is an inherited disorder of skeletal muscle excitability. It is caused by mutations in the muscle chloride channel gene (CLCN1) and can be inherited either as an autosomal dominant (Thomsen's myotonia) or autosomal recessive

(Becker's myotonia) trait. Becker's myotonia is more common than Thomsen's myotonia and usually presents with more severe symptoms and at an earlier age. Symptomatically, the stiffness is most pronounced in the extremities, ameliorates with continuous activity (warm-up phenomenon) and worsens after a period of rest. Patients with MC are at high risk of developing a severe myotonic response with generalized muscle spasms whenever depolarizing muscle relaxants are used. The most important complication is malignant hyperthermia in these patients. We presented our anesthesia experience in a patient who suffer from MC in this article.

CASE REPORT

A 27-year-old male patient (American Society Anesthesiologists grade 2, body mass index: 23.5) presented with complaints of congenital maxillary and mandibular misalignment with anodontia and difficulty in using the muscle of upper and lower limbs since birth. The patient was diagnosed with MC at the age of 14 with complaints of difficulty in climbing stairs. Since then, he was followed up in neurology department with oral treatment of phenytoin 100 mg twice daily. There was no family history of Myotonia congenita or any other congenital disease and doesn't have a history of any type of previous anesthesia exposure. General examination: mandibular prognathism and maxillary retrognathia present. On examination of central nervous system all higher functions, cranial nerve examination were normal. Power is decreased in bilateral upper and lower limb-4/5, All other system examinations are normal. Airway examination- Mallampati 2, mouth opening-more than 3 finger breadth, thyromental distance- more than 2 finger breadth, neck movements were normal. Anesthesia considerations like malignant hyperthermia, delayed recovery were kept in mind and drugs were tailored according to it. Before the operation, anesthesia machine breathing circuits were changed, soda lime and filter were changed. volatile agents were removed from the workstation to avoid accidental usage. No premedication was applied to the patient. Electrocardiogram (ECG), noninvasive blood pressure, (SpO2), (EtCO2) and esophageal temperature were monitorized.

Bristol regimen was followed in order to attain plasma concentration of 3.5microgram/ml, induction was carried out with 2mcgs/kg of fentanyl,1.5mg/kg blouses of propofol, Inj.Atracurium 30mg IV given, intubation was done with 8size North Pole RAE tube simultaneously propofol infusion were started at the rate of 10mg/kg/hr for 10min followed by 8mg/kg/hr for 10minutes followed by 6mg/kg/hr thereafter.

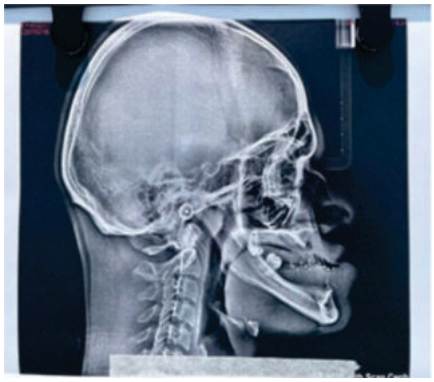

Figure 1: X-ray Picture Of Head And Neck (lateral View)

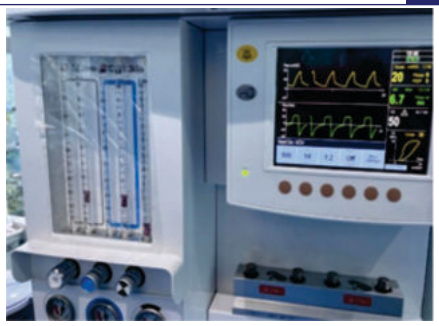

Figure 2: Picture Showing A Ventilator Setting For The Patient.

Patient was ventilated with 6 L/min 50% oxygen/nitrous mixture. During the operation, 2000 mL isotonic saline was infused. In the operation period patient vital parameters and hemodynamic status were stable and esophageal temperature was 35.9-36.2 °C, (ETCO2) values were 35-38 mmHg. During the operation blood gas analysis values were within normal limits (Ph: 7.35, PCO2:35, P02: 190, lactate: 0.8, K: 3.6). Operation was finished after 180 minutes. Inj.ondensteron 4mg IV was given. TIVA was stopped once the surgery is over. After 10 to 15 minutes spontaneous ventilation was achieved, once the patient achieves the extubation criteria, extubation was carried out after reversing the patient with Inj.neostigmine 2.5mg and inj.glycopyrollate 0.5mg IV.The patient was transferred to the intensive care unit (ICU) for postoperative complications like delayed acute onset of apnea. Postoperative period was uneventful. Adequate analgesia was given with the drugs such as paracetamol 1gm IV and weak opioid tramadol 100mg iv was administered.

DISCUSSION

MC first described by A. J. Thomsen in 1876 in his own family, Therefore, this autosomal dominate myotonia is named as Thomsen's disease. It begins at the childhood and affects upper extremities than lower. Temporary muscle rigidity diminishes with rest. The prognosis of this disease is good. In our patient similar symptoms (fatigue without muscle weakness, muscle rigidity which diminishes with rest and responds medical treatment) were present. The disorder had been diagnosed at 14 years old and had responded well to drug therapy with phenytoin. In another type of myotonia is named by Becker, lower extremities are mainly affected. In this form, symptoms are more severe than to be in the Thomsen type. In pulmonary and neuromuscular diseases such as MCs, regional anesthesia should be preferred instead of general anesthesia. If general anesthesia is performed, short-acting drugs such as propofol and remifentanil can be used. It should also be kept in mind that masseter muscle spasm may develop during the induction of anesthesia in myotonic disease. When the inhalation anesthetics were used, shivering may occur, and myotonia may increase. We administered TIVA with propofol, instead of inhalation anesthetics. In our anesthetic plan, we have used only reduced dose of muscle relaxants and subsequent doses were not given. This type of neuromuscular diseases can cause malignant hyperthermia during anesthesia because of increased muscle activity. We took some precautions against possibility of malignant hyperthermia.

Triggers for MH	Safe drug for MH
Ether	Propofol
Halothene	Ketamine
Enflurane	Etomidate
Isoflurane	Benzodiazepines
Sevoflurane	Barbiturates
Desflurane	Opiods
Succinylcholine	Nitrous oxide
	Non depolarising muscle relaxants
	Local anesthestics

Figure 3: Triggers and safe drugs for MH (Malignant hyperthermia)

Dantrolene sodium is the drug of choice for malignant hyperthermia. Dose required will be 2.5mg/kg IV every 5 minutes until symptoms subsides; may required upto 30mg/kg, but Dantrolene sodium were not available in our institution and hence maximum precautions were taken to prevent malignant hyperthermia like we changed the ventilation circuits, soda lime and filter, and we chose the anesthetic drugs that did not trigger malignant hyperthermia as we mentioned above in figure 3. Since pain could be induced muscle rigidity, postoperative pain control is very important in these patients. Therefore, we administered paracetamol and tramadol intravenous intraoperative and postoperative period to our patient. It was reported in the literature that cold environment, tremors, hyperkalemia, and mechanical or electrical stimulation may trigger myotonia in these patients, and acidosis may occur.For this reason, arterial blood gas analysis was followed and normothermia was provided by heat monitoring in our patient. Thanks to these precautions, acidosis was not observed. Prior to anesthesia of patients with CM, preoperative evaluation, laboratory tests, relevant consultations, anesthetic drugs to be used, normothermia and ICU follow-up in postoperative period are also important. If anesthesiologist can't obtain dantrolene natrium, TIVA could be a safe solution with the precautions that were present above.

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