Olgu Sunumu

MASSETER SPASM IN AN UNDIAGNOSED CASE OF LIPID STORAGE MYOPATHY

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Summary:

Masseter muscle spasm; it is important to perform necessary interventions in time with regards to providing safe airway and probability of development of malignant hyperthermia. Patients who develop masseter rigidity during anaesthesia may have an underlying myotonic disorder. In this case, we report an undiagnosed case of lipid stroge myopathy in a patient, who developed life masseter spasm following a routine standard induction of anaesthesia.

Key Words: Masseter spasm, malign hyperthermia, lipid storage diseases, general anesthesia

Tanısız Lipid Depo Hastalığı Olgusunda Massater Kas Spazmı

Özet:

Masseter kas spazmı; güvenli havayolu sağlanması ve malign hipertermi gelişebilmesi açısından önemlidir. Anestezi esnasında masseter spazmı gelişen hastada altta yatan myotonik hastalık olabilir. Bu olguda; standart anestezi indüksiyonunu takiben masseter kas spazmı gelişen tanısı olmayan lipid depo hastalığı olan hastayı sunduk.

Anahtar Kelimeler: Masseter spasm, malign hipertermi, lipid depo hastalığı, genel anestezi

Introduction:

Although masseter muscle spasm is rarely seen, it may cause severe complications. Failure to achieve mouth opening, failure to achieve direct laryngoscopy and endotracheal intubation, failure to achieve mask ventilation with any airway apparatus in the patient after induction may appear as a life-threatening condition[1].

After receving the consent of the patient, we report an undiagnosed case of lipid stroge myopathy in a 40 year old male patient, who developed life threatening masseter spasm following a routine standard induction of anaesthesia. Neither the patient nor the anesthesiologists were aware of the disease before this potentially lethal complication occurred.

Case: A 40-year old male patient (height 170 cm, weight, 80 kg, ASA I) was scheduled for

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nasal septoplasty surgery in the Ear Nose Throat Clinic (ENT). He had no general anaesthesia before. During anaesthesia, vital signs were monitored closely. After preoxygenation, standart anaesthesia induction was performed. with fentanil 100 µg, propofol 200mg and rocuronium 50 mg. Ventilation via face mask could not be performed. Then the anaesthetist tried to open the patient's mouth, but the mandible resisted by massive spasm of both masseteric muscles. Mouth opening was completely impossible at this time and all attempts to insert a laryngoscope blade or an alternative airway management device failed. Manual bag ventilation via face mask was impossible. The patient was gradually desaturated (80%). An emergent tracheotomy was decided. Tracheotomy was performed by ENT surgeon approximately in 2 minutes without any complication. Following the tracheotomy, the patient was ventilated effectively and saturation rose to 100 % and heart rate returned to normal. Moderete rigidity of upper and lower extremity muscle was noted. His core temparature measured with eusophageal probe was 37.2 0C. Urine was clear. Surgery was cancelled and the patient was transfered to the intensive care unit for ventilatory support. In the ICU he was successfully weaning within the next 24 hours. He was conscious, fully orientated. He hadn't any neurological deficit. Routine laboratory evaluations analysis all were normal. Creatin kinase levels were 320 UI-1 (at 1st hr), 438 UI-1(at 4th hr), 348 UI-1 (at 8th hr), 332 UI-1(at 12thhr), 165 UI-1 (at 24th hr), 154 UI-1 (at 48th hr) and 120 UI-1 (at 72th hr). No myoglobulinuria was detected by dipstick urine analysis. Computorized tomography showed that the mandibule and temporomandibular joints were normal. The patient was decanulated at the 3rd day. Muscle biopsy from right deltoid muscle was done regarding any possible myopathic disease. The histological diagnosis was ''lipid storage myopathy". After the patient was stable he was transferred to neurology clinic.

Discussion:

Lipid storage myopathy (LSM) is pathologically characterized by prominent lipid accumulation in muscle fibers due to lipid dysmetabolism[2]. Lipid storage myopathy is mainly seen with clinical findings such as hypotonia, weakness and it has subtypes as primary carnitine deficiency, multiple Acyl-COA dehydrogenase and neutral lipid storage[3]. Definitive diagnosis can be made by muscle biopsy and genetic tests for subgroup in the direction of clinical findings. Definitive diagnosis in our patient was made by muscle biopsy and EMG findings supported the diagnosis[4]. Genetic subdiagnosis could not be provided since clinical symptoms were not observed clearly in our patient. Presence of a lot of odds numbers in genetic tests uses clinical symptoms as base for probable subgroup in order to reduce the number of tests to applicable number. Otherwise, tests to be performed reach to the numbers not complying with practice. Therefore, lipid storage myopathy subgroup was not described.

Masseter muscle spasm is a life-threatening condition in the myopathic patients which may occur with administration of the anesthetic agents and accompany malignant hyperthermia[5]. As there may be an underlying myopathy diagnosis, there may also be no previous clinically significant finding. In this case; myopathy should be investigated, history and diagnosis methods should be used. Although our patient has no known myopathy diagnosis; during the detailed history we learned that a previous tooth extraction procedure was performed with difficulty due to difficulty in opening of the mouth because of spasm. This information shows the importance of the detailed history during preoperative evaluation while it may be a clue for myopathy.

While masseter muscle spasm may occur together with malignant hyperthermia development, it can also be seen alone[6,7]. It may be observed as a myotonic reaction that may occur during the administration of anesthesia in the individuals with neuromuscular disease. It is an important clinical picture regarding either the maintenance of the airway provided safely or the observation of malignant hyperthermia together with the spasm. Underlying neuromuscular disease in the patient developing masseter muscle spasm may be diagnosed previously or may not be diagnosed due to not observation of clinical symptom. When Looi et al.[8] investigated

masseter muscle spasm case developed after induction of anesthesia with thiopental and suxamethonium, the authors reached to underlying and previously undiagnosed myotonia congenita diagnosis.

Probability of accompanying of malignant hyperthermia to the picture gains a different point of view to the case. We did not find any finding specific to malignant hyperthermia in our patient. This condition may depend on not maintaining anesthesia by us. Operation could be postponed since it was septal deviation surgery. Caution should be exercised regarding malignant hyperthermia in a case in which the patient must have been operated and dantrolene should be accessible. The patient and his relatives were informed written and verbally about this subject.

Another point that we want to emphasize in presentation of our case is the accessibility of invasive airway devices like cricotomy in difficult airway cases 'who can not be intubated, ventilated' similarly. Since we were in otorhinolaryngology operating room we had emergency tracheostomy equipment and the procedure was performed successfully in a short time by the surgical team. With the occasion of this case; requirement of presence of emergency cricotomy equipment in all operating rooms which can be used by the anesthetist was emphasized once more importantly.

In a similar case, Wolfgang et al.[9] presented masseter muscle spasm developed in their patient after propofol administration during anesthesia induction. Differently from our patient, mask ventilation could be provided in the patient in whom endotracheal intubation could not be performed due to failure to achieve mouth opening. When the spontaneous breathing of the patient returned, endotracheal intubation was performed with the blind nasal intubation method using a Portex endotracheal tube guided by breath sounds and the operation was maintained. Operation was completed uneventfully with careful follow-up regarding malignant hyperthermia. The authors emphasized that blind-nasal, breath-sound-guided endotracheal intubation was as efficient as video laryngoscopy in difficult airway.

In masseter muscle spasm; it is important to perform necessary interventions in time with regards to providing safe airway and probability of development of malignant hyperthermia. Based on this case, we wanted to emphasize the importance of the availability of invasive difficult airway devices like cricotomy set in anesthesia. Patients who develop masseter rigidity during induction of anaesthesia may have an underlying myotonic disorder. A careful preoperative history with questions directed at muscular symptoms and family history are vital. On top of that, patient must be counseled regarding the disease and its significant anaesthetic implication, that is the dangerous and deadly nature of malignant hyperthermia syndrome which may follow masseter spasm. The family members must be investigated for the disease as well.

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