Anaesthetists' Nightmare: Masseter Spasm After Induction in an Undiagnosed Case of Myotonia Congenita

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SUMMARY

We report an undiagnosed case of myotonia congenita in a 24 year old previously healthy primigravida, who developed life threatening masseter spasm following a standard dose of intravenous suxamethonium for induction of anaesthesia. Neither the patient nor the anaesthetist was aware of the diagnosis before this potentially lethal complication occurred.

KEY WORDS:
Masseter spasm, Suxamethonium, Undiagnosed myotonia congenita

INTRODUCTION

Myotonia Congenita (MC) is a rare genetic disorder characterized by failure of immediate muscle relaxation after voluntary contraction or mechanical stimulation. Two forms have been described: the autosomal dominantly inherited disease described in 1876 by Julius Thomsen who himself suffered from the disease, as did twenty other members of his family over four generations[1] ; and the recessively inherited disease described by Becker in 1950's. Becker's disease has a later onset; symptoms are progressive and more severe[2].

CASE REPORT

The patient was a 24 year old Malay primigravida at 40th week of gestation, who had no known previous medical illness. She was admitted to the Maternity Hospital in established labour. In view of her short stature of 139cm, and weight of 55kg, she was posted for emergency lower segment caesarean section, indicated by the cephalo-pelvic disproportion. She had no history of exposure to anaesthetic agents before. Pre-operative assessment did not reveal any features to suggest a difficult intubation.

During anaesthesia, vital signs were monitored closely. After adequate preoxygenation with 100% Oxygen for 4 minutes, rapid sequence induction was performed using intravenous thiopentone 225mg and suxamethonium 100mg. No obvious fasciculation was noted. Soon after that (around one minute later) when the anaesthetist tried to open the patient's mouth for intubation, patient developed severe masseter spasm. It was totally impossible for oral intubation. The patient desaturated rapidly and her saturation dropped to 50% despite mask ventilation with 100% oxygen. After several attempts, her mouth was finally forced open to insert an oropharyngeal airway. Following that she was manually ventilated until the oxygen saturation rose to more than 90%. Subsequently she was intubated with endotracheal tube, guided by a gum elastic bougie. The masseter spasm lasted for 3 minutes. Clinically, she was not warm or febrile. Urine was noted to be clear.

To proceed with surgery and aid with mechanical ventilation, she was paralysed with intravenous atracurium 30mg, a non-depolarizing muscle relaxant. Anaesthesia was maintained with 50% oxygen, 50% nitrous oxide and isoflurane. End-tidal carbon dioxide was maintained between 35 to 40 mmHg. Saturation was maintained at 97-100% throughout. The baby was born with a good APGAR score.

At the end of surgery, anaesthetic agents were discontinued and the patient began to breathe. Residual muscle paralysis was reversed with neostigmine and atropine. Despite being fully awake and obeying commands, her respiratory effort was unsatisfactory. Thus, a second dose of anticholinesterase was administered. However, end-tidal carbon dioxide progressively increased to 80 mmHg, associated with a desaturation to 92%. There was an initial suspicion of malignant hyperthermia due to the masseter spasm. However, she was not febrile, and there was no rise in carbon dioxide intraoperatively.

The arterial blood gas done at this time showed feature of respiratory acidosis possibly due to the poor recovery from the muscle relaxant. PH: 7.33, pCO2: 68.8 mm Hg, pO2: 288.2 mm Hg, HCO3: 25.9 mmol/L, BE: 3.3, oxygen saturation: 99%, FiO2:100%.

A thorough physical examination at the end of surgery revealed deltoid and gastrocnemius hypertrophy. She was transferred to the Intensive Care Unit (ICU) for ventilatory support in view of her poor breathing effort and the possibility of a myopathy. The subsequent arterial blood gas done in the ICU showed normal results.

In the ICU she was successfully extubated within the next 24 hours. She was then referred for a neurological opinion. On further enquiry by the neurologist, she explained that she had stiffness of muscles particularly upon releasing her fingers after handgrip. The symptoms were more prominent over the hands and feet. She had frequent falls when she was younger, especially when she tried to walk after a period of rest. However, she did not notice any amelioration of the symptoms after continuous muscle activities and cold weather worsened her symptoms. There was no similar

This article was accepted: 14 November 2008
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Fig. 1: Prominent deltoid hypertrophy.

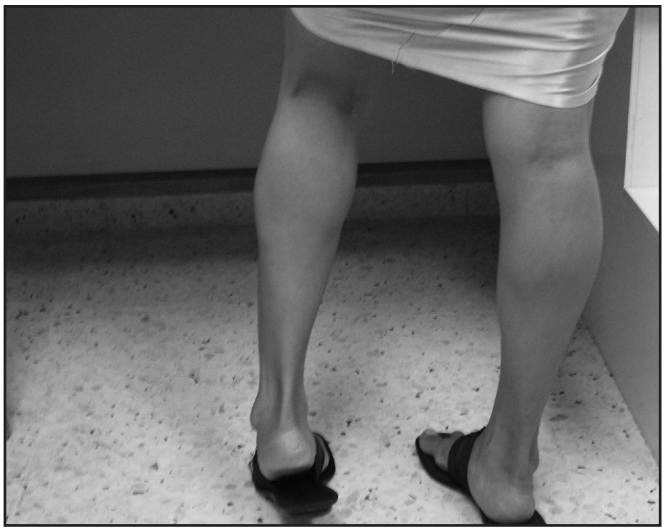

Fig. 2: Calves hypertrophy

problem among her siblings and there was no consanguinity between her parents. She had been married for a year to her factory–operator husband. She was a housewife.

The patient underwent a thorough neurological examination after delivery. Both grip and percussion myotonia were present. Her chest X-Ray showed an abnormally shaped thoracic cage with a restrictive pattern. Blood investigations showed elevated creatinine kinase – 933 IU/L and slightly lowish cholinesterase – 3006 (normal: 3930-10800). Electromyography showed typical spontaneous 'dive-bomber' myotonic discharges and the myopathic patterns of brief low amplitude polyphasic motor unit potentials and early recruitment. A diagnosis of myotonia congenita, most likely Becker's disease, was established in view of the fact that there was no similar problem among the family members. At a 3-months' follow up visit, she was clinically stable, with no significant progression of symptoms.

DISCUSSION

We describe a patient who developed life-threatening muscular spasm following the administration of suxamethonium at induction and later neostigmine at reversal. Subsequent evaluation revealed typical features of myotonia congenita and diagnosis was further confirmed by an electromyogram.

In an acute situation when patients develop masseter rigidity during induction of anaesthesia, differential diagnosis like malignant hyperthermia, pseudocholinesterase deficiency and myotonia should be considered [4].

Masseter muscle spasm after administration of suxamethonium should raise the suspicion of malignant hyperthermia(MH) as it presages clinical MH in up to 30% of cases. Even in the absence of clinical MH, myoglobinuria in individuals with masseter spasm is common post-operatively. In the case of an emergency surgical procedure, the anesthetic should be converted to a "non-trigger" technique and the patient observed for an early signs of MH. The modalities that should be monitored closely include end tidal carbon dioxide, arterial blood gas, electrolytes, coagulation, core temperature, creatinine kinase levels, myoglobin level and urine output. Dantrolene and supportive treatment must be made ready.

Myotonia congenita (MC) is a nondystrophic muscular disorder caused by mutation of the gene encoding the chloride channel of skeletal muscle. Proximal muscular hypertrophy is a prominent feature. Myotonia manifests as persistent contraction of a muscle after cessation of voluntary contraction. Transient weakness is noticed on quick movement and is ameliorated by exercise, the so called "warm-up" phenomenon. It is not cold sensitive and is not associated with abnormal potassium homeostasis[2].

The basic pathology of this condition is a reduced chloride conductance that fails to buffer the after-potential and triggers new premature action potentials. Patients with myotonia congenital have altered response to anaesthetic drugs [2]. Suxamethonium is contraindicated as it precipitates intense myotonic contraction and trismus which prevents opening of the mouth for intubation. Neostigmine may aggravate muscle contraction by facilitating depolarization of the neuromuscular junction[3].

Short acting non-depolarising muscle relaxant seems to behave normally in myotonic patients and is therefore recommended[1]. Intraoperative maintenance of body temperature and postoperative avoidance of shivering is important as cold may induce myotonia in certain type of myotonic conditions[3].

It is important to exclude malignant hyperthermia (MH) in a patient who develops severe masseter spasm following administration of induction agents. MH is characterized by

increased body temperature and rigidity during anaesthesia. It is triggered by volatile anaesthetics and depolarizing muscle relaxants. The cause is most often a mutation involving the sarcoplasmic reticulum (SR) calcium release channel (ryanodine receptor: RYR1 gene) but genetic heterogeneity exists. During a MH episode, the SR calcium channels open persistently and the resulting calcium influx causes sustained muscle contraction, hyperthermia and increased metabolism. This may subsequently lead to complications like metabolic acidosis, increased oxygen consumption, increased carbon dioxide production, increased temperature and haemodynamic instability[4].

Predisposition to MH is confirmed in three muscular disorders apart from true MH itself: central core disease, Evans myopathy and King Denborough syndrome (short stature, mental retardation and musculoskeletal abnormalities)[4].

Apart from MH, pseudocholinesterase deficiency should be suspected when paralysis of respiratory and other skeletal muscles fail to spontaneously resolve after suxamethonium is administered. It is an inherited enzyme abnormality that results in abnormally slow metabolic degradation of exogenous choline ester drugs such as suxamethonium[4].

In conclusion, 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. If this is suspected, depolarizing muscle relaxants should be avoided during future anaesthesia. 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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