Anaesthetic management of a case of distal myopathy

Sir,

Congenital myopathies are a group of rare genetic disorders characterized by a defect in the contractile apparatus of the myocytes.[1] We describe the case of a patient with distal myopathy taken up for total abdominal hysterectomy under general anesthesia as patient refused for neuraxial blockade.

A 36-years-old female patient diagnosed to have abdominal leiomyosarcoma was planned for total abdominal hysterectomy under general anesthesia. On evaluation the patient gave a history of progressive weakness and thinning of distal aspect of both upper and lower limbs for the last 10 years. She had not sought any medical attention for these complaints.

On examination, she was thin built with a BMI of 20. She was haemodynamically stable and had an adequate airway. She had atrophy of distal hand muscles with bilateral claw hands. She also had bilateral foot drop with a high stepping gait [Figure 1a and b]. Deep tendon reflexes, sensory nervous system, cranial nerves and spine examination were normal. Her routine investigations, chest X-ray and ECG were normal. Electrophysiological studies were suggestive of distal myopathy with normal proximal muscles and sensory system.

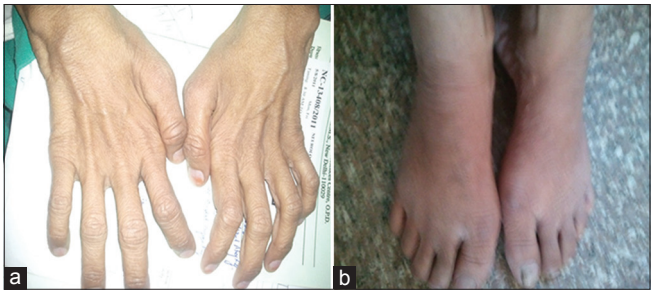

Figure 1: Distal myopathy (with clawing of hand and foot drop) (a) Clawing of hand. (b) Foot drop

Tab. alprazolam 0.25 mg was given the night before the surgery and adequate blood products were arranged. In the operation theatre, before taking the patient in, all vapourizers were removed from anaesthesia workstation. Soda lime, breathing circuit and face mask were changed. After securing peripheral intravenous line and attaching standard monitors, we started intravenous induction with midazolam 1 mg, fentanyl 60 μcg and 1% propofol 60 mg. After loss of verbal response, we started neuromuscular monitoring with train of four (TOF) and gave atracurium 15 mg. After obtaining no response to TOF, airway was secured with proseal laryngeal mask airway (LMA) of size 3 and post induction monitoring included capnography, nasopharyngeal temperature and TOF count. Maintenance of anesthesia was with with O_2/N_2O and intravenous propofol infusion at 30-50 μcg/kg/min. Top up doses of atracurium was guided intraoperatively with the TOF count of 3 to achieve adequate muscle relaxation. Haemodynamics and respiratory parameters were maintained within the normal limits throughout the procedure. Thirty minutes before the end of the surgery, diclofenac 1.5 mg/kg and ondansetron 10 μcg/kg were given. At the end of the surgery, neuromuscular blockade was reversed with normal doses of neostigmine and glycopyrrolate and when TOF ratio was ≥0.9, proseal LMA was removed. Post operative period was uneventful.

Anaesthetic management of patients with muscle diseases is challenging. In addition to unpredictable sporadic responses, such as rhabdomyolysis and metabolic stimulation, more predictable risks are associated with respiratory and bulbar muscle weakness, myocardial involvement, and difficult airway anatomy. To identify and minimize risk, a thorough preoperative workup is indispensable.[2,3] If regional anesthesia is planned, a preoperative assessment of peripheral sensory nerve dysfunction should be considered. Increased sensitivity to respiratory-depressant drugs should be anticipated and where possible, doses should be titrated to the desired effect. Even with the most careful anaesthetic management, postoperative ventilatory support may be required and, therefore, postoperative intensive care unit availability should be planned.

Baseline serum potassium and creatine kinase (CK) concentrations to assess muscle membrane integrity, family history, cardiac function test and other syndromic features must be sought. It is possible that high baseline values may be associated with increased risk of profound perioperative rhabdomyolysis, whereas the baseline value *per se* is required to differentiate perioperative rhabdomyolysis from preexisting muscle damage.[3]

All anesthetic techniques and drugs are associated with increased risk in patients with myopathies.[4,5] General anaesthesia performed as total intravenous anaesthesia or regional anaesthesia are equally safe. Inhalation agents especially halothane are not recommended because they may trigger a malignant hyperthermia (MH)-like syndrome.[6] Consideration should be given to the use of short-acting opioids to avoid the possibility of postoperative ventilation. Use of depolarizing neuromuscular blocking drugs should be generally discouraged in patients with neuromuscular diseases.[4,6] The depolarizing muscle relaxants might trigger MH, and prolonged depolarization leads to potassium release and calcium influx. Although theoretically reversal of neuromuscular block with anticholinesterases in patients with neuromuscular diseases may produce the similar situation, it was not observed in our case. So it can be recommended to use the normal dosage of reversal in such patients. The recent introduction of cyclodextrin reversal drugs (such as sugammadex) provides an attractive alternative in these circumstances. Succinylcholine may also cause acute profound rhabdomyolysis in patients susceptible to MH and those with myopathies. If neuromuscular blockade is required, a non-depolarizing neuromuscular blocking drug should be used, keeping in mind that patients with myopathies may show increased sensitivity to these drugs. On the basis of cardiac evaluation, there may be an indication for invasive monitoring and use of inotropes in patients with identified cardiac involvement.[5] Strength and range of motion can deteriorate rapidly after surgery and immobilization, and anaesthesia can unmask subclinical respiratory failure.[7] So, patients should be mobilized as soon as possible after surgery.[8]

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