Anaesthetic management of a case of Duchenne muscle dystrophy with Moyamoya disease

INTRODUCTION

Duchenne muscular dystrophy is an x linked recessive disorder caused by a mutation in the dystrophin gene located on chromosome Xp21.[1] Typical presentation is proximal muscle weakness. Patients develop respiratory distress and cardiomyopathy in advanced stage of the disease. The patients have associated difficult airway anatomy in the form of macroglossia and limited mobility of mandible and cervical spine. These patients are at an increased risk of developing extreme hyperthermia, rhabdomyolysis and hyperkalemic cardiac arrest when exposed to halogenated inhalational anaesthetics and depolarizing muscle relaxants.[2,3] Here, we describe the successful anaesthetic management of a case of moyamoya disease with Duchene muscular dystrophy who underwent encephaloduroangiomyosynangiosis (EDAMS) procedure under general anaesthesia.

CASE REPORT

An 8 years old male child, weighing 20 kg with diagnosis of moyamoya disease with Duchenne muscular dystrophy was scheduled for an EDAMS procedure. Magnetic resonance imaging showed right middle cerebral artery territory infarct with small foci of infarct in left frontal and high fronto-parietal white matter. There were also signs of left middle cerebral artery territory infarct and near total occlusion of right sphenoid internal carotid artery with multiple collateral vessels suggestive of moyamoya disease. The child had progressively increasing bilateral lower limb weakness and was diagnosed to have Duchenne muscular dystrophy at the age of 5 years. Patient was dysphasic and had right sided hemiparesis with right sided facial nerve palsy. Cardiovascular and respiratory systems were normal. Airway examination revealed a large tongue with Mallampati Grade 3 score. Prior to induction of anaesthesia, all the vaporizers were dismounted from the anaesthesia machine, fresh soda lime was filled into the soda lime canisters and the breathing circuits were changed. Then the anaesthesia machine was flushed at a fresh gas flow of 10 l/min for 30 min prior to remove any residual anaesthetic gases residue from prior use. It was ensured that there was absolutely no inhalational anaesthetic inside the anaesthesia machine and circuit as shown by gas monitoring. After attaching the routine monitors, child was induced with 40 mcg fentanyl and 40 mg propofol. Rocuronium 20 mg was given to facilitate endotracheal intubation. Intraoperative monitoring included electrocardiography (ECG), non-invasive blood pressure, invasive blood pressure, pulse oximetry, capnography, temperature and central venous pressure monitoring. Anaesthesia was maintained

with 60% nitrous oxide-oxygen-propofol-fentanyl and rocuronium. Patient was given positive pressure ventilation and to maintain the $ETCO_2$ between 32 and 34 mm Hg. Multiple episodes of haemodynamic fluctuations (systolic blood pressure variations from 90 mm Hg to 160 mm Hg and heart rate variations from 80 to 130 beats/min) were noticed during the dissection of subcutaneous tissue. These haemodynamic fluctuations were managed with adjustment of depth of anaesthesia and warning to the neurosurgeon. Apart from the haemodynamic variability, rest of the intraoperative period was unremarkable with no abnormality in cardiac rhythm, ETCO2, body temperature, serum electrolyte and urine output. The total duration of the procedure was 3 h. Propofol infusion was stopped 15 min prior to the end of surgery and muscle paralysis was reversed after surgery. Child was extubated when he was fully awake with good respiratory efforts.

The child was kept in the intensive care unit for 48 h where he made a successful recovery with no signs of rhabdomyolysis or hyperkalemia and shifted to ward for further care. He was discharged from the hospital on the 8th postoperative day.

DISCUSSION

The anaesthetic concerns in a child with Duchenne muscular dystrophy are the possibilities of difficult intubation, prolonged duration of neuromuscular block, possible need for postoperative ventilation and occurrence of rhabdomyolysis and cardiac arrhythmias when exposed to halogenated volatile anaesthetic agents and depolarizing muscle relaxants.

There are various reports of difficult intubation in patients with Duchenne muscular dystrophy.[4-6] In a retrospective study done by Breucking *et al.*, difficult intubation was experienced in eight of the 219 patients.[5] In another retrospective study by Muenster *et al.*, difficulty laryngoscopy was found in 4% of patients. The incidence was higher (7.5%) in older children.[6] Obesity, a large tongue, restricted mouth opening and limited mobility of cervical spine are thought to be causes of difficult intubation in this group of patients.[6]

Various studies have shown that the response of non depolarizing muscle relaxants is abnormal in patients of Duchenne muscular dystrophy. Delayed onsets of action and prolonged effect have been seen after the use of commonly used non depolarizing neuromuscular blocking agents.[7,8]

Due to the weakness of diaphragm, intercostal muscles, and the accessory muscles of respiration, various degree of restrictive pulmonary dysfunction may be present and pulmonary function tests should be done to assess the risk of perioperative respiratory complications and need for postoperative ventilation.[2] Normal preoperative ECG and echocardiogram findings do not exclude the possibility of severe cardiac complications in the perioperative period and cardiologist should be consulted in the preoperative period.[2,9]

There are various reports of acute onset hyperkalemic cardiac arrests, rhabdomyolysis, hyperthermia and tachycardia in patients of Duchenne muscular dystrophy following exposure of inhalational anaesthetic agents. Though only a small proportion of Duchenne muscular dystrophy patients develop rhabdomyolysis after inhalational anaesthesia, a total intravenous anaesthesia based anaesthesia is considered safest in these patients.[3] Nitrous oxide is generally considered safe in these patients. In a large series involving 232 cases, nitrous oxide in oxygen was used in 135 patients without any problem.[6]

Precautions must be taken to ensure that patient is not exposed to inhalational agents during surgery. This can be ensured by unmounting all the vaporizers from the anaesthesia machine, using fresh sodalime and breathing circuits by running the machine at a high fresh gas flow rate for 20—30 min.[10]

CONCLUSION

We describe the successful anaesthetic management of a case Duchenne muscular dystrophy who underwent EDAMS procedure under general anaesthesia for moyamoya disease. A careful preoperative evaluation and avoidance of anaesthetic agents which may trigger rhabdomyolysis and severe hyperkalemia are the keys to successful anaesthesia outcome in patients with Duchenne muscular dystrophy.

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