General Anaesthesia in the Management of Myotonic Dystrophy Patient Posted for Tympanoplasty

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INTRODUCTION

Myotonic dystrophy is a genetic disorder in which there is progressive weakness and loss of muscle mass which often contract but are unable to relax. Anaesthetic management of myotonic dystrophy (DM) can be challenging with complications like loss of airway secondary to medication-induced respiratory depression, aspiration, hypoventilation and sudden death due to cardiac conduction abnormalities. In the light of these complications, one should consider "regional anaesthesia as a viable alternative if the surgical procedure is really necessary". However, in surgeries which require general anaesthesia one should avoid triggering factors like hypothermia, opioids, inhalational agents and succinylcholine. Severity of the patient disease must be elucidated on preanesthetic evaluation. PONV prophylaxis, rapid sequence intubation, short acting muscle relaxant, ECG and temperature monitoring are the essence of successful anaesthetic management.

Myotonica dystrophy (DM) is one of the variants of muscular dystrophy with a prevalence of 1 in 8000.[1] It is characterized by a compromised synthesis or regeneration of contractile proteins, affecting multiple organ systems and may present with progressive musculoskeletal weakness, cardiac dysrhythmia, hypoventilation, hyperthyroidism, and cognitive-behavioural disorders.[2,] Patients with DM can exhibit exquisite sensitivity to sedatives, neuromuscular blocking agents and volatile anaesthetics resulting in potential perioperative complications.[2,3] Thus meticulous preoperative assessment and proper anaesthetic planning are prerequisite for successful anaesthetic management. We report a case of myotonica dystrophy type 1 (DM1) with CSOM posted for tympanoplasty under general anaesthesia.

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PRESENTATION OF CASE

A 17 year old male patient with complaints of left ear discharge attended ENT OPD and was diagnosed as CSOM with myotonica dystrophy. He had undergone bilateral cataract surgery for congenital cataract in the past. In the family, two of his siblings are having similar disorder. Preanesthetic evaluation was done 4 days prior to surgery by a multi-specialty team. Patient was moderately built and nourished, conscious and oriented. There was no facial expression that is minimal muscle power impairment (scale 2) as assessed by muscular impairment rating scale. Vitals were PR 66 bmp, BP-110/60 mmHg and RR-15 cpm and cardiorespiratory system was normal. Airway examination- Mallampati class 2 with high arched palate and spine normal. Preop Hb 10.4 g%, platelet counts 2.13 lakhs, TC- 4600 cells/mm, RFT, FBS, serum electrolyte and ECG within normal limit. 2D ECHO shows bicuspid aortic valve and EF of 60%. Patient accepted as ASA1. A night prior to surgery PONV prophylaxis given with Tab ondansetron and Tab pantoprazole 40 mg and advised fasting for 8 hours before surgery.

DISCUSSION OF MANAGMENT

Planned for general anaesthesia with IPPV. In OR, ASA standard monitors connected, and vitals were PR-68 bpm, BP-118/70 mmHg, SpO_2-99% in room air and RR-15 cpm. 20G IV canula in left URL and Ringer lactate infusion started. Patient preoxygenated and induced with inj. glycopyrrolate 0.2 mg, inj. midazolam 1 mg, inj. fentanyl 50 mcg, inj. propofol 80 mg intravenously. Modified rapid sequence intubation done with ET tube (cuffed) 8 no after administration of inj. atracurium 20 mg IV. Anaesthesia maintained with O_2: N_2O, sevoflurane % 1 and with atracurium infusion. Temperature and neuromuscular monitoring done and external defibrillatory pads were applied. The operating room's ambient temperature was increased, a forced-air warming blanket and warm IV fluids were used to prevent hypothermia. Tympanoplasty procedure done and duration was three and half hour. Throughout the procedures vitals and ECG were normal. At the end of surgery, patient was extubated once train of four ratio was 0.9. He was conscious, responding to verbal commands and SpO_2 was 100% in room air. 1200 ml iv fluid and analgesic paracetamol 800 mg iv given intraoperatively. Intra op urine output was 300 ml. Patient was shifted to SICU for monitoring of vitals and respiration. Post op analgesia achieved injection paracetamol intravenously 20 mg/kg 12th hourly. After 24 hours, patient shifted to ward.

DISCUSSION

Muscular dystrophies are a set of more than 30 genetic diseases characterized by muscle weakness in varying gradient they belong to the same group of diseases, they have very different characteristics in their clinic. Although al presentation and in their genetic origin. The incidence of these diseases varies around 1 in 10,000 to 100,000 live births.[4] Although these are very rare diseases, they pose huge anaesthetic challenges.

Systems	Effects	
Musculoskeletal	(i) Myopathy, atrophy, myalgias (ii) Myotonia: triggers include stress and cold as well as specific medications	(i) DM1: tends to affect facial muscles e.g. distal muscles (ii) DM2: tends to affect proximal muscles e.g. hip flexors
Cardiac	(i) Arrhythmias (a) AV block, bundle branch block most common (b) Atrial flutter and fibrillation	(i) Cardiomyopathy: Hypertrophy, dilation, systolic dysfunction
Nervous system	(i) Cognitive impairment (ii) Mental retardation more common in DM1	(i) Axonal sensorimotor polyneuropathy (ii) Sensorineural hearing loss
Gastrointestinal	(i) Dysphagia (ii) GERD (iii) IBS-like symptom	(i) Gallstones
Eye	(i) Cataracts (ii) Proptosis	
Pulmonary	(i) OSA (ii) Hypersomnia/excessive daytime somnolence (iii) Increased risk of aspiration pneumonia	(i) Respiratory muscle weakness (ii) Increased sensitivity to respiratory depressants
Endocrine	(i) Primary Hypogonadism (ii) Diabetes, Insulin resistance (iii) Hyperthyroidism	(i) Hyperparathyroidism (ii) Hyperhidrosis (iii) Male pattern baldness
Cancer	(i) Increased risk for cancers of endometrium, brain, ovary, colon, and skin	
Reproductive	(i) Low sperm count secondary to hypogonadism	(i) Higher risk of miscarriage, preterm labour

Table 1

Two genes have been identified as playing a role in the development of myotonica dystrophy (DM). ACTG expansion in DMPK gene results in type 1 (DM1), while an expansion in the ZNF gene results in type 2 (DM2) and severity of the disease appears to correlate with the expansion repeats. Patients may present with different symptomatology as it affects multiple organ system (table 1).[2,4,5] Hence multidisciplinary approach and assessment of the patient is critical to successful perioperative management. In our case the boy was 17-year-old with positive history of congenital cataract and generalized mild muscle impairment scale 2 (Table 2).

Grade	Description
1	No muscular involvement
2	Minimal signs Myotomy, mandibular and temporal weakness Facial weakness, weakness of flexor muscle of neck Lid ptosis, nasal voice, No distal weakness, except for digital flexor of the hand
3	Distal weakness No proximal weakness, except for extensor of elbow
4	Mild to moderate proximal weakness
5	Sever proximal weakness

Table 2

He was able to carry out day to day activity but became fatigued often with no facial expression. As patient is uncooperative, we planned for general anaesthesia and prepared for post op mechanical ventilation if needed. Myotonica dystrophy patients are prone to aspiration due to incompetence of the lower oesophageal sphincter and weakness of the laryngeal muscles. Thus, PONV prophylaxis and strict NPO are essential.[6,7] While RSI is recommended to prevent aspiration.[8] DM patient have increased sensitivity to sedatives, anxiolytics and analgesics. They are at high risk for compromised ventilatory drive, so sedative premedication prior to surgery was withheld.[2] Perioperatively titrated dose of injection midazolam and injection fentanyl administered.[9] In our case, induction was done with propofol as it is effective and safe although thiopentone and etomidate can also be used.[10,11,12] After adequate preoxygenation and maintaining cricoid pressure tracheal intubation was done with atracurium 20 mg.[13] Atracurium and rocuronium can be used safely in DM patients. The rapid breakdown of atracurium by "Hofmann" elimination leads to a predictable recovery, and avoids the use of neostigmine which may precipitate myotonia.[13,14,15]

The succinylcholine may lead to masseter spasm, laryngospasm and also exaggerated hyperkalaemia response seen in DM patient hence avoided.[16] Maintenance of anaesthesia done with oxygen, nitrous oxide and sevoflurane 1% with atracurium infusion with aid of neuromuscular monitor.[17] DM patient are no more susceptible to malignant hyperthermia than general population due to inhalational agents but may exuberate cardiomyopathies due to myocardial depressive effects.[8] Sevoflurane has cardiostable properties provides stable hemodynamic intraoperatively. At the end of surgery complete recovery from muscle relaxant was assessed by neuromuscular monitoring (TOF 0.9), extubated and maintained oxygen saturation in room air. Perioperative monitoring is very essential for the smooth recovery of the

patient.[2] Continues 12 lead ECG, SpO_2, NIBP and temperature monitoring done. These patients are prone for atrioventricular arrythmias and common cause of sudden deaths of 47% while respiratory complications lead to 29.2% of deaths. Patient with cardiac involvement may have implantable cardioverter and defibrillator otherwise defibrillator along with external pad should be available in perioperative setting.[18,19] Hence external defibrillator was made available. Myotonia may be precipitated by hypothermia, shivering so temperature monitoring is vital intraoperatively and it is achieved with help of warm blanket, warm IV fluids throughout the procedures.[2,8] Postoperative respiratory and cardiovascular complications are high due to prolonged sedative effect of opioids, neuromuscular blockade drugs hence observation for 24 hour is necessary in intensive care unit.[8] For post OP analgesia inj. paracetamol given 12th hourly to avoid opioid induced respiratory depression and random blood sugar level was 140 mg/dL. Post op was uneventful. Regional anaesthesia is safer than general anaesthesia in DM patient wherever possible.[8] Suggestions for Perioperative Management of Patients with DM by Myotonica Dystrophy Foundation.

1) Check preoperative blood sugar.
2) Keep patient warm. Use forced-air warming device and increase ambient temperature in OR.
3) Have defibrillator available in the operating room and defibrillator pads on patient.
4) Avoid succinylcholine and neostigmine.
5) Utilize continuous pulse oximetry and EKG monitoring.
6) Plan for possible prolonged postoperative stay

In a patient with myotonica dystrophy, one should consider regional anesthesia if the surgical procedure is really necessary. However, for surgeries which require general anesthesia one should avoid triggering factors like hypothermia, opioids, inhalational agent, succinylcholine and neostigmine. The severity of the patient disease must be elucidated during preanesthetic evaluation. PONV prophylaxis, rapid sequence intubation, short acting muscle relaxant, ECG, temperature and neuromuscular monitoring are the essence of successful anesthetic management.

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