Successful anaesthetic management of a patient with prior history of malignant hyperthermia for corrective scoliosis surgery

INTRODUCTION

Malignant hyperthermia is a pharmacogenetic condition triggered by the administration of volatile anaesthetic and/or depolarizing muscle relaxants causing the sustained release of calcium ions from the sarcoplasmic reticulum, thereby leading to hypermetabolism and rhabdomyolysis and multiorgan failure. In the absence of timely recognition and intervention, it can prove to be a fatal complication in susceptible individuals. In most cases, it is caused by a defect in the ryanodine (RYR1) receptor.[1]

Various studies have established an increased incidence of malignant hyperthermia in patients with congenital myopathies.[2,3] Congenital myopathy with type 1 fibre predominance is a non-progressive myopathy affecting the skeletal muscles and majorly causing proximal muscle weakness in both upper and lower limbs. Even in the absence of underlying disorders, moderate to severe degree of scoliosis tends to have a restrictive effect on the respiratory mechanics. Decreased chest wall compliance, impaired lung growth and decreased respiratory muscle strength contribute to decreased lung volumes in these individuals, thereby leading to respiratory compromise.

CASE REPORT

A 14-year-old girl was scheduled for correction of idiopathic infantile right thoracolumbar scoliosis. The surgery was previously attempted at a different hospital 2 years before. But, because she developed features suggestive of malignant hyperthermia, the procedure was abandoned. She had apparently developed some features during the 3 h of intra-operative period; tachycardia, muscle rigidity, hypercarbia, dark-coloured urine and hyperthermia (108^0 F). On laboratory evaluation, she was found to have respiratory acidosis and elevated CK levels, which settled over the next 3 days, for which she was treated symptomatically and discharged. Muscle

biopsy performed at a later date showed congenital myopathy with type 1 fibre predominance.

Her past history revealed delayed gross motor milestones, recurrent respiratory tract infections and progressive spine deformity with back pain. With a history of easy fatigability and dyspnoea on mild exertion and a breath holding time of 8-10 s, she presented a picture of gross respiratory compromise, which was confirmed by pulmonary function tests that revealed severe restrictive defect [forced expiratory volume in 1 second (FEV1) - 25% predicted; forced vital capacity (FVC) - 28% predicted, FEV1/FVC - 83% predicted] and not responding to bronchodilators. Other than being underweight, her general physical and airway examinations were normal. All laboratory investigations (complete blood picture, blood urea, serum creatinine, serum electrolytes, coagulation profile, liver function tests) and two-dimensional echocardiogram were normal. X-ray chest showed right thoracic scoliosis with left lumbar compensation, with a Cobb's angle of 90 degrees and normal lung fields. Baseline arterial blood gases (ABG) and CK were normal and urine tested negative for myoglobinuria.

Informed consent was taken after counselling the family members regarding all the potential complications – recurrence of malignant hyperthermia, rhabdomyolysis, increased blood loss, haemodynamic fluctuations, delayed recovery, post-operative ventilator support and intensive care unit (ICU) stay.

On the day of the surgery, the anaesthesia workstation was flushed with 100% oxygen after removing the vaporizers. Fresh soda lime and anaesthetic circuit were used. Dantrolene, all other resuscitation drugs and ice packs were kept ready. Pre-operative vitals and arterial blood sample on room air were normal. She was wheeled into the operating theatre and anaesthesia was induced with inj fentanyl 2 mcg/kg and inj propofol 2 mg/kg. After confirming adequate ventilation, inj atracurium 0.5 mg/kg was administered and intubated with an oral cuffed endotracheal tube 6.5 and connected to the ventilator. The right internal jugular vein and the left radial artery were cannulated. She was then turned prone and the pressure points were adequately padded. Oral airway was placed to prevent tongue bite during monitored evoked potential (MEP) stimulus.

Apart from standard monitoring, neuromuscular monitoring was also performed using MEP. Anaesthesia was maintained using oxygen – air mixture 40:60 and propofol (125-150 mcg/kg/min) and fentanyl (1-1.5 mcg/kg/h). No further muscle relaxants were administered. The procedure lasted for nearly 7 h and was uneventful. MEPs were adequate during the procedure. Intra-operative blood loss was approximately 1500 mL and four units of whole blood were transfused. Intra-operative urine output was 1 mL/kg/h. At the end of surgery, the patient was turned supine and muscle relaxation was reversed. Extubation was performed as she was fully conscious, obeying commands and had stable respiratory parameters.

Post-operatively, her ABG revealed respiratory acidosis. As she was conscious and co-operative, intermittent bilevel positive airway pressure (BiPAP) support was given for 2 days post-operatively. She was shifted out of the ICU on the 4th post-operative day and discharged on the 6th post-operative day.

DISCUSSION

Corrective scoliosis surgery poses a challenge to the anaesthesiologist as it requires us to have a perfect understanding of the patient's diagnosis and co-morbidities and the physiological derangements that may be induced by anaesthesia and the surgical procedure.[1] In this case, the presence of congenital myopathy and severe restrictive lung disease with a prior history of malignant hyperthermia further compounded the challenge.

Published evidence suggesting mutations in RYR1 gene in patients with congenital myopathy with fibre-type disproportion exist.[2] Because RYR1 mutation is most commonly implicated in malignant hyperthermia-susceptible patients, we decided to be well equipped to be able to avert, recognize and treat any recurring episode as per the Malignant Hyperthermia Association Of United States (MHAUS) protocol.

Total intravenous anaesthesia (TIVA) using propofol and fentanyl infusions with complete avoidance of inhalational agents was successfully used with no undue haemodynamic effects. Other than the loading dose to facilitate intubation, no further doses of muscle relaxants were administered to facilitate intra-operative MEP monitoring and prevent delayed recovery and further deterioration in respiratory function.[4,5]

Pulmonary complications are the principal causes of post-operative morbidity and mortality in corrective

scoliosis surgeries. Although surgical intervention corrects the spinal curvature, its effect on lung volumes and arterial oxygenation only becomes apparent after two or more years.[6] Owing to patient's poor pulmonary function, post-operative respiratory compromise was anticipated. As the patient was fully conscious and co-operative, it was decided to give a trial of BiPAP to avoid reintubation and its subsequent complications. The effectiveness of NIPPV in hypercapnic respiratory failure secondary to chest wall defects and neuromuscular disorders is well established.[7,8] In the present case, it took 3 days for the respiratory acidosis to subside and for the patient to be clinically fit to be shifted out of the ICU.

CONCLUSION

A thorough pre-anaesthetic assessment, high degree of vigilance and strictly adhering to the protocol goes a long way in averting an episode of malignant hyperthermia. Dantrolene has to be made available as increasing cases of malignant hyperthermia are being reported in India. Intra-operatively, TIVA with avoidance of muscle relaxants can be effective in corrective scoliosis surgery, especially in the presence of MEP monitoring. BiPAP can be effectively used in averting endotracheal intubation in post-operative respiratory acidosis.

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