Section: Miscellaneous

Case Report

ANAESTHETIC MANAGEMENT OF A PATIENT WITH PROXIMAL MYOTONIC MYOPATHY AND DIFFICULT AIRWAY UNDERGOING BRANCHIAL CYST EXCISION

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ABSTRACT

Proximal Myotonic Myopathy (PROMM) presents unique anaesthetic challenges due to myotonia, multisystem involvement and sensitivity to anaesthetic agents.[1,2] We report the successful management of a 41-year-old male with PROMM and a predicted difficult airway undergoing branchial cyst excision. Awake intubation using a video bronchoscope and maintenance with total intravenous anaesthesia (TIVA) allowed safe perioperative management. Avoidance of triggering agents and preservation of spontaneous ventilation were key to the favourable outcome.

Keywords: PROMM, TIVA

INTRODUCTION

Proximal Myotonic Myopathy, also known as Myotonic Dystrophy Type 2, is a multisystem disorder characterized by myotonia, proximal muscle weakness and increased sensitivity to sedatives and anaesthetic drugs.[2] Anaesthetic considerations include the risk of myotonic contractures, respiratory compromise, cardiac conduction abnormalities and altered pharmacodynamics. Difficult airway scenarios further complicate management and require careful planning.

Case Report

A 41-year-old male with a known diagnosis of PROMM, hypertension and hypothyroidism on regular treatment was scheduled for elective excision of a branchial cyst.

Preoperative airway assessment revealed restricted mouth opening: one finger breadth, Mallampati Grade IV and had an anticipated difficult airway.

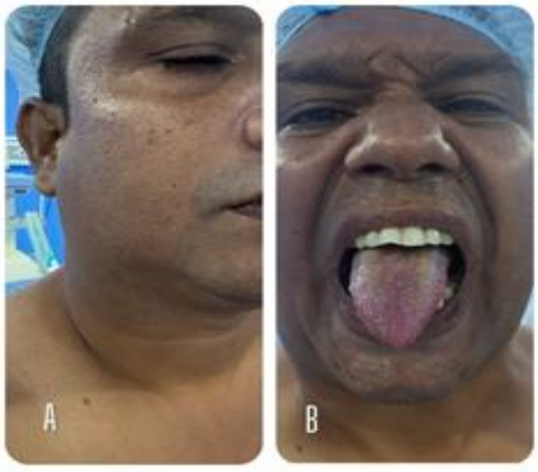

Figure 1: A. Patient picture showing the branchial cyst B. Patient picture depicting restricted mouth opening

He was diagnosed to have myotonic dystrophy at a tertiary care hospital on evaluation and follow up assessment for his muscle weakness since childhood. Specifically, he had weakness on getting up from bed, walking and climbing up the stairs. He wasn't found to have any respiratory, cardiac illnesses presumably associated with PROMM. Diagnosed with hypothyroidism, he is on oral

Thyroxine 50 mcg once daily for five years. Systemic examination was unremarkable and baseline investigations including pulmonary function tests and echocardiogram were within normal limits. Computed tomographic study was performed to study the extent of the branchial cyst and compression on the adjacent vital structures.

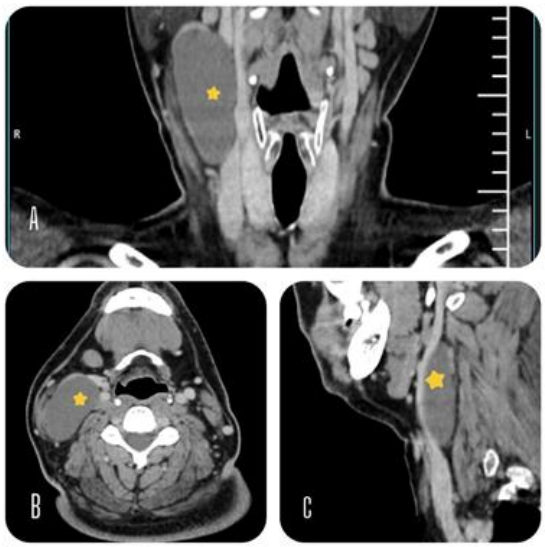

Figure 2: Computed Tomography of the neck depicting the branchial cyst in A. Sagittal section B. Axial section C. Coronal section

Given the underlying myopathy, the anaesthetic plan would need to be tailored to avoid agents known to precipitate myotonia or malignant hyperthermia. Succinylcholine was avoided due to its association with exaggerated myotonic responses and risk of life-threatening complications including malignant hyperthermia.[3] Similarly, inhalational anaesthetic agents, anticholinesterases (neostigmine), anticholinergics and muscle relaxants were deliberately avoided to prevent triggering myotonic contractures.[4,5]

Anaesthetic Management

After securing IV access with 18G IV cannula, standard ASA monitors were applied. After recording the baseline vitals, airway blocks were performed after adequately educating the patient about the procedure. Short acting benzodiazepine Midazolam 1 mg, antiemetic Ondansetron 8 mg were administered prior to airway anaesthesia.

Airway Strategy

Awake intubation was planned due to anticipated difficulty and the need to avoid muscle relaxants. Avoiding airway manipulation under deep anaesthesia without secured airway is crucial, as myotonia can impair ventilation and intubation.

Airway preparation was meticulously performed using a multimodal topical and regional anaesthesia approach. Nasal passages were prepared with lignocaine adrenaline soaked gauze packing along with oxymetazoline drops in both nostrils to achieve adequate vasoconstriction. Oropharyngeal anaesthesia was achieved using 10% lignocaine spray, followed by 2% lignocaine nebulization for deeper airway topicalization. This was complemented with targeted airway blocks, including bilateral superior laryngeal nerve blocks and a transtracheal recurrent laryngeal nerve block, ensuring optimal conditions for awake airway instrumentation.

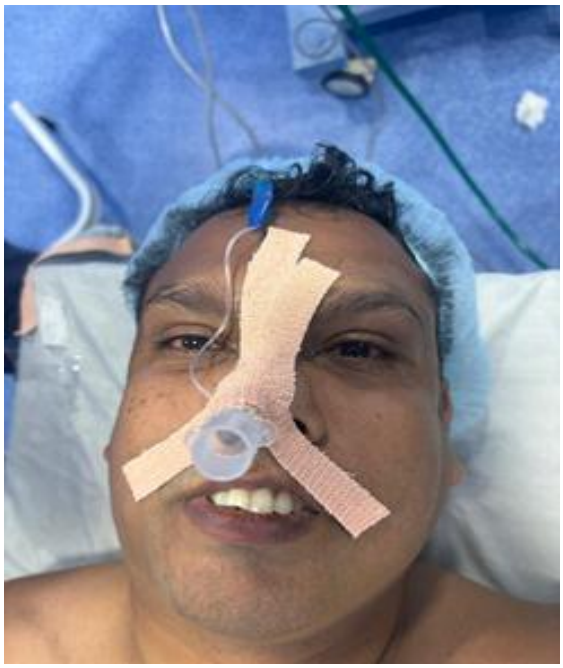

Figure 3: Awake patient with nasotracheal tube

Figure 4: A. Patient with extended neck position for the surgical excision of the cyst.

B. Intraoperative patient picture with the branchial cyst of size 8 x 4 centimetres

Airway was secured by a video bronchoscopy guided nasotracheal intubation with 7.0 mm ID cuffed tracheal tube. After confirmation of tube placement with capnography, anaesthesia was induced with Fentanyl 2mcg/kg and Propofol 2 mg/kg. The patient was positioned with head in

maximal extension for better surgical exposure. Anaesthesia was maintained using Total Intravenous Anaesthesia (TIVA) with propofol infusion at 6mg/kg delivered by a syringe pump, supplemented with nitrous oxide and oxygen at 2 litres each thro a closed circuit. Enough plane was anaesthesia was planned to keep the patient apneic and was ventilated in the volume control mode with a tidal volume of 8ml/ kg and respiratory rate of 16/ minute.

The surgery lasted approximately two hours and remained haemodynamically stable throughout the procedure. A key intraoperative challenge was achieving optimal surgical exposure requiring neck extension, which necessitated maintaining an adequate depth of anaesthesia without compromising ventilation.

At the conclusion of surgery, the patient was allowed to emerge fully from anaesthesia. Extubation was performed on table once the patient was fully awake, pain free, with adequate respiratory effort and intact airway reflexes.

Throughout the tenure in the operation theatre, succinylcholine, muscle relaxants, anticholinesterases, anticholinergics and inhalation agents were avoided for fear of myotonic trigger and malignant hyperthermia.[5] The postoperative period was uneventful, with no evidence of myotonic episodes, respiratory compromise or residual weakness.

DISCUSSION

PROMM patients are highly sensitive to anaesthetic agents, particularly depolarizing muscle relaxants and volatile agents. Succinylcholine is contraindicated due to the risk of severe myotonic contractures and hyperkalaemia. Succinylcholine should be strictly avoided in patients with PROMM (proximal myotonic myopathy) because it can trigger severe myotonic crises, exaggerated muscle contractures, and life-threatening airway or jaw rigidity.[3] Severe masseter spasm and rigidity can make opening the airway extremely difficult which can be detrimental in a patient who already has a restricted mouth opening. Patients with underlying myopathies carry an increased susceptibility to massive potassium release and cardiac complications.

The role of volatile anaesthetics in triggering malignant hyperthermia in myotonic disorders remains controversial; however, cautious avoidance is often recommended. Additionally, volatile anaesthetics such as sevoflurane do not reliably prevent or relieve myotonic contractures.[6] Shivering, surgical stimulation or certain drugs can trigger clinical myotonia. Halogenated agents like sevoflurane are traditionally approached with caution in patients with uncharacterized myopathies.

Non-depolarizing muscle relaxants are generally considered much safer than depolarizing agents like succinylcholine in patients with proximal myotonic myopathy (PROMM), though they still require substantial dose reductions and close monitoring.[7] But these drugs are better avoided in view of potential danger of myotonia while using anticholinesterase and anticholinergic drugs for reversal of residual neuromuscular blockade.

Anticholinesterase agents like neostigmine increase acetylcholine at the neuromuscular junction, which can potentially provoke paradoxical muscle cramping, sustained muscle contraction (myotonia), or worsening weakness in patients with myotonic disorders.[8] If neostigmine is administered to reverse non-depolarizing neuromuscular blockade, it is standard practice to co-administer an anticholinergic agent (such as glycopyrrolate or atropine) to mitigate severe muscarinic side effects like bradycardia, though this does not eliminate muscle-specific myotonic risks. Quantitative neuromuscular monitoring (such as train-of-four monitoring) and prolonged post-procedure cardiorespiratory observation are critical while using muscle relaxants. Rocuronium is widely favoured for its rapid onset and reversibility with Sugammadex in patients with PROMM.[9]

Awake intubation remains the safest approach in the presence of a difficult airway, especially when muscle relaxants are contraindicated. TIVA using propofol provides stable anaesthesia while minimizing the risk of triggering myotonia associated with usage of muscle relaxants. Preservation of spontaneous ventilation while intubation is crucial in such patients.

Reversal agents such as neostigmine may exacerbate myotonia and their avoidance further supports a non–muscle relaxant technique.[8] Careful intraoperative monitoring including temperature and End Tidal Carbon dioxide, gentle handling, maintaining normothermia and vigilance in monitoring are essential to prevent myotonic episodes and catastrophic malignant hyperthermia

CONCLUSION

Patients with Proximal Myotonic Myopathy present significant anaesthetic challenges, particularly when associated with a difficult airway. Awake intubation combined with TIVA and strict avoidance of myotonia-triggering agents provides a safe and effective anaesthetic strategy.[10] Thorough preoperative assessment, meticulous planning, and vigilant perioperative monitoring are key to achieving favourable outcomes.

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