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Letter to Editor

Challenges and strategies for safe eye surgery in duchenne muscular dystrophy patients

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Dear Editor,

A 22-year-old male presented for phacoemulsification of right eye cataract. During preoperative evaluation, it was noted that patient had an abnormal facies comprising of a flat nasal bridge, prominent forehead and macroglossia (Figure 1). Airway examination revealed a Modified Mallampati grade of IV with limited neck extension and flexion, suggesting an anticipated difficult airway. The patient had a known diagnosis of Duchenne Muscular Dystrophy (DMD), based on clinical history and features. Progressive muscle weakness had led to joint contractures in both the elbow and knee, complicating positioning on the operating table. On inspection of spine, a thoracic scoliosis was noted. Functional capacity could not be assessed as patient's mobility was limited. An attempt at performing spirometry failed. A chest Xray was done to gauge severity of pulmonary limitation, which suggested left lung field to be small and atelectatic and right lung field hyperinflated in compensation. Auscultation of chest was clear with no added sounds. ECHO suggested normal LV systolic function of 55-60% with no valvular or other abnormality. All the blood investigations were normal. Patient had mild global developmental delay but with well performed perioperative counselling, required cooperation was expected.

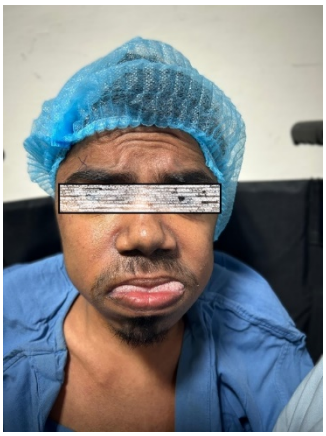

Figure 1: Wheel chair bound patient with macroglossia

DMD is a rare X-linked recessive inherited muscle disorder. Mutations in the dystrophin gene (chromosome Xp21) causes gradual degeneration and weakening of muscle fibres. Mortality typically occurs between 20-40 years of age due to respiratory muscle weakness or heart problems. Progressive involvement of musculoskeletal system, respiratory muscles and cardiac anomalies possess a significant challenge to anaesthetist during perioperative phase.[1]

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DMD can be associated with macroglossia, weak neck flexors and limited mobility of mandible and cervical spine leading to difficult intubation and difficulty laryngoscopy.[2] Cardiac issues including dilated cardiomyopathy and arrhythmias could lead to perioperative hemodynamic fluctuations. Respiratory muscle weakness, along with scoliosis, can lead to restrictive pulmonary impairment, complicating mechanical ventilation and increasing the risk of pulmonary complications.[2] Patients of DMD can have Intellectual disability in the form of ADHD and autism spectrum disorders, which may make them uncooperative for procedure or venous cannulation.

Patients with DMD in GA are at increased risk of developing extreme hyperthermia, rhabdomyolysis and hyperkalemic cardiac arrest when exposed to depolarizing muscle relaxants and halogenated inhalational anaesthetics and thus are contraindicated. The use of Trigger-free' anaesthetic and 'clean' anesthesia machine is advocated.[3,4] Further regional and local anesthesia offers significant advantage in terms of avoidance of anaesthetic drugs and reduction in postoperative pulmonary complications.

For ophthalmologic procedures, which can be performed under topical or local anesthesia, it is advisable to use Monitored Anesthesia Care (MAC) with sedation (e.g., dexmedetomidine) due to the complications mentioned above. For general anesthesia or procedural sedation, Total Intravenous Anesthesia (TIVA) with agents such as propofol and short-acting opioids is suitable. Use of supplemental oxygenation and an ICU backup for post procedure care and positive pressure ventilation should be sought. Intraoperatively, monitoring of SpO_2 blood or end-tidal carbon dioxide levels, if feasible.

In this case, after thorough counseling, the patient was shifted to the operating room. ASA standard monitors were attached and 22G IV cannula was secured. Positioning was done carefully ensuring patient comfort. The surgical procedure went uneventful with patient cooperation. LMA and endotracheal tube were kept as airway securing device with TIVA for sedation or maintenance of GA, rocuronium as muscle relaxant with sugammadex for rapid reversal of neuromuscular blockade, if required. ICU bed was available for postoperative mechanical ventilation and monitoring. Minimal literature is available on patients undergoing procedure under sedation or MAC.

In DMD patients, regional anesthesia is preferred when feasible, and minor surgeries such as eye procedures can be safely performed under topical anesthesia or MAC, with thorough perioperative counselling, vigilant monitoring, and a readily available backup plan for any complications.

1. Declaration of Patient Consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient consented to his images and other clinical information to be reported in the journal. The patient understands that his name and initials will not be published and due efforts will be made to conceal his identity, but anonymity cannot be guaranteed.

2. Conflicts of Interest

There are no conflicts of interest.

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