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CASE REPORTS

A difficult airway approach in a merosin-deficient congenital muscular dystrophy patient: a case report

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Abstract Merosin-deficient muscular dystrophy is caused by an autosomal recessive mutation on laminin-α2 gene characterized by severe progressive muscle weakness associated with neuromuscular scoliosis and restrictive lung disease. In this case report, we describe an alternative airway approach performed in a child with anticipated difficult airway and merosin-deficient muscular dystrophy. Significant anesthetic implications may increase the perioperative risk, requiring accurate knowledge to anticipate an adequate management and provide patient-safety strategies.

Introduction

Congenital muscular dystrophies (CMDs) are autosomal recessive disorders with compromised synthesis and regeneration of the muscle contractile proteins. Besides all being rare and progressive, they differ in their clinical manifestations and genetic background.[1]

Merosin-deficient CMD (MD-CMD) has a prevalence of about 0.7/100,000 and is caused by a mutation on chromosome 6, in laminin-α2 gene (or merosin) present in skeletal muscle fibers.[2] It accounts for nearly 40% of all CMD cases, the most common and severe form. Regardless, few reports of its anesthetic management were published in the literature. This case report contributes to the current knowledge related to airway management in MD-CMD patients, sustaining a case-oriented approach to an anticipated difficult airway.

Case report

A nine-year-old boy, ASA III, diagnosed with MD-CMD during his first months of life, presented a severe dorsolumbar scoliosis and an inherent restrictive lung disease with diminished lung volumes and frequent respiratory infections. A previ-

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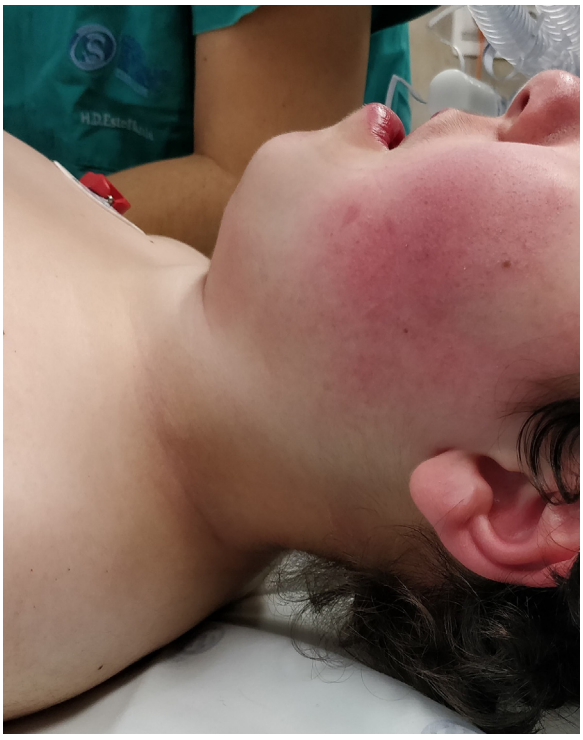

Figure 1 Preoperative evaluation of the airway.

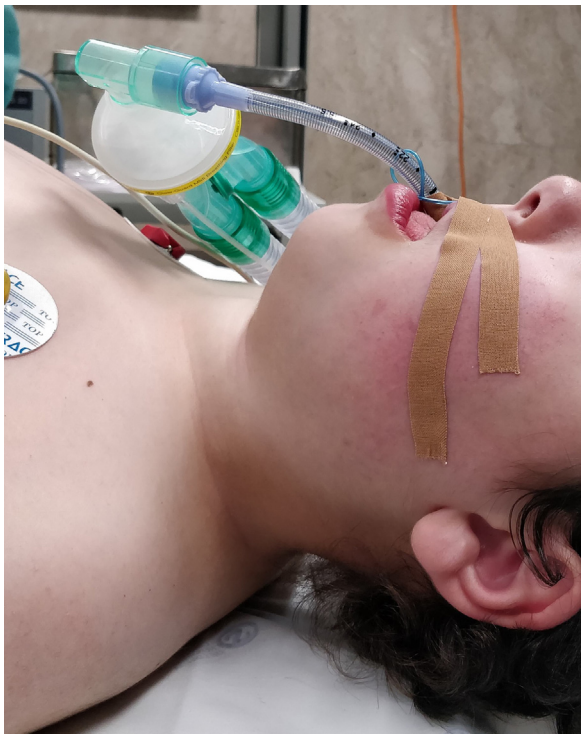

Figure 2 Status after airway management.

ous scoliosis correction surgery already showed that only videolaryngoscopy provided a successful airway approach due to Cormack-Lehane score IV on conventional laryngoscopy. Therefore, the child was now proposed for scoliosis definitive surgical treatment under intraoperative neurophysiology monitoring.

During preoperative assessment, the patient demonstrated reduced neck mobility, slight retrognathism and a persistent left flexion of the head, macroglossia and Mallampati III, predictors consistent with MD-CMD and a difficult airway approach as previously identified (Fig. 1).

Intraoperatively, an inhaled induction with sevoflurane ensuring spontaneous ventilation, without opioid adjuvants, was maintained until intubation, allowing an adequate level of unconsciousness and also a safe and dynamic airway assessment. Initially, McCoy laryngoscopy evidenced a Cormack-Lehane IV, confirming the difficult airway assessment. Secondly, McGrath videolaryngoscopy showed an anterior glottis and intubation could not be productively accomplished even with a tube introducer. To improve the airway approach success, McGrath videolaryngoscopy was combined with an optic fiberscope as a guide for intubation, which was successfully achieved with a 6.0-cuffed tube (Fig. 2). The entire intubation process lasted 20 minutes.

Maintenance with remifentanil and propofol total intravenous anesthesia without neuromuscular blockade was performed avoiding any interference with evoked potential acquisition. The surgical procedure was uneventful and the patient remained intubated in the Pediatric Intensive Care Unit for 24 hours. Extubation was planned after weaning of sedation and mechanical ventilation under Anesthesiology team supervision. No further complications occurred during postoperative period.

Discussion

It is widely recognized that MD-CMD patients typically require orthopedic procedures early in life, namely scoliosis correction; however, anesthetic management is particularly challenging and burdened with perioperative complications.[3] Before the procedure, an experienced multidisciplinary team should be involved to implement patient-safety strategies and provide an integrated care. This should include a comprehensive evaluation of the patient's global functional condition and anticipation of difficult airway predictors regarding their poor head control, marked neck and trunk hypotonia, and limited mouth opening,[1] as well as the increased risk of rhabdomyolysis.[3]

In what difficult airway concerns, fiberoptic intubation is the gold-standard, although it does not always provide a successful approach and no single airway device or technique will adapt in every clinical situation.[4] The option of a videolaryngoscopy approach after a Cormack-Lehane IV identification was an initial attempt to optimize glottis visualization as part of the process of difficult airway algorithm. Although the absence attempt of a flexible fibroscopy alone was an eventual limitation, we believe that a simultaneous approach of these two airway devices provided a better visualization through the videolaryngoscope screen and an easier control of the tube tip with the fiberscope in order to secure the airway. As stated by other authors,[5] this combined technique is advantageous to decrease tissue trauma particularly in patients with high Cormack-Lehane scores. Therefore, maintenance of spontaneous ventilation combined with a videolaryngoscopy-assisted fiberoptic intubation should be considered as an alternative to difficult but non-emergent airway scenarios in Pediatric Anesthesi-

ology, specially when there is an association of a higher Cormack-Lehane score with other clinical difficult airway predictors.

Concerning timing of tracheal extubation, most authors suggest performing it awake after a complete reversal of neuromuscular blockade.[3] Non-invasive ventilation should be considered as a resource for immediate postoperative period, bearing in mind the possibility of respiratory failure and need of Intensive Care Unit admission.

Standard ASA monitoring must always be used and neuromuscular blockade stimulation is recommended mostly in cases of severe hypotonia. Albeit MD-CMD patients are not prone to malignant hyperthermia, its theoretical risk still exists as a muscular dystrophy, along with anesthesia-induced rhabdomyolysis. Both may induce arrhythmias and respiratory failure exacerbation. Hence, general recommendations include a trigger-free anesthetic technique and prefer an inhaled induction as a less traumatic experience. Multimodal analgesia seems especially important to reduce the inherent risk of most complications.

Learning points

Muscular dystrophies share significant perioperative risk, namely a difficult airway approach or MH. A combined airway approach involving a McGrath videolaryngoscopy and a flexible fiberscope provides a valid alternative approach of an expected difficult airway in a child with MD-CMD. A multidisciplinary strategy guarantees the most effective perioperative care, minimizing any issues related to the disease and anesthetic management.

Conflicts of interest

The authors declare no conflict of interest.

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References

1. Jimenez N, Song K, Lynn AM. Hemodynamic instability during prone spine surgery in a patient with merosin-deficient congenital muscular dystrophy. Paediatr Anaesth. 2013;23:294–6.
2. Pregardien C, Pirotte T, Veyckemans F. Pressure-support ventilation in a child with merosin-deficient congenital muscular dystrophy under sevoflurane anesthesia. Acta Anaesthesiol Belg. 2016;67:139–41.
3. Echeverry-Marín PC, Bustamante-Vega ÁM. Anesthetic implications of muscular dystrophies. Rev Colomb Anestesiol. 2018;46:228–39.
4. Matioc AA. Use of the Airtraq® with a fibreoptic bronchoscope in a difficult intubation outside the operating room. Can J Anesth. 2008;55:561–2.
5. Sgalambro F, Sanfilippo F, Santonocito C, Caltavuturo C, Grillo C. Virtual laryngoscopy and combined laryngoscopic-bronchoscopic approach for safe management of obstructive upper airways lesions. Br J Anaesth. 2014;113:304–6.