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SCIENTIFIC ARTICLE - CASE REPORT

AWAKE INTUBATION FOR ANESTHESIA IN A PATIENT WITH MUCOPOLYSACCHARIDOSIS TYPE VI (MAROTEAUX-LAMY SYNDROME)

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ABSTRACT

Mucopolysaccharidoses (MPS) are a group of rare, progressive hereditary diseases caused by the deficiency of 11 lysosomal enzymes responsible for the degradation of glycosaminoglycans (GAGs). Enzymatic dysfunction leads to the progressive accumulation of unmetabolized GAGs in tissues and organs, including the airways, making management difficult and constituting a challenge for the anesthesiologist. This article reports the anesthetic management of a patient with Mucopolysaccharidosis type VI (Maroteaux-Lamy Syndrome) who underwent laparoscopic herniorrhaphy. After pre-anesthetic evaluation, general anesthesia with awake nasotracheal intubation was chosen, performed via fiberoptic bronchoscopy.

Keywords: Mucopolysaccharidosis VI, General anesthesia, Tracheal intubation, Bronchoscopy, Airway management.

INTRODUCTION

Mucopolysaccharidoses (MPS) represent a heterogeneous group of hereditary, progressive, and rare diseases. The etiology of these conditions lies in the deficiency of 11 distinct lysosomal enzymes, essential for the degradation of glycosaminoglycans (GAGs). The resulting enzymatic dysfunction leads to the progressive accumulation of unmetabolized GAGs in various tissues and organs, triggering cellular, tissue, and organ dysfunction. This cascade of events manifests clinically through cardiovascular, pulmonary, gastrointestinal, neurological, and musculoskeletal involvement. In addition, GAG deposition in the upper airways poses a significant challenge for anesthesiologists in airway management.[1,2]

The classification of MPS into seven main types (I to IX, although types V and VIII are no longer used) is based on the specific pattern of GAG deposition, which is determined by the absence or defect of one of the 11 lysosomal enzymes involved in the process. Most forms of MPS follow an autosomal

recessive inheritance pattern, except for type II, which has X-linked recessive inheritance. The estimated prevalence for the group of MPS forms is approximately one case per 25,000 live births. However, underdiagnosis may occur due to the existence of milder forms.[1,2]

Given the complex manifestations resulting from disease progression, patients with MPS often require surgical interventions under anesthesia. Airway management in this context may be substantially complicated by the accumulation of GAGs, which causes hypertrophy of the adenoids, tonsils, tongue, and laryngopharynx. Additionally, these patients may present with facial and airway anatomical abnormalities (except in type III), complicating both face mask ventilation and laryngoscopy or intubation procedures. The incidence of difficult intubation in this population varies considerably, ranging between 28% and 44%.[1,2]

In view of the potential challenges inherent to airway management in this specific group of patients, the present case report aims to emphasize the crucial importance of pre-anesthetic planning as an indispensable tool to ensure patient safety and the success of the anesthetic procedure.

CASE REPORT

A 45-year-old patient, 56 kg, 163 cm, with MPS type VI (Maroteaux-Lamy Syndrome), valvular heart disease (mitral and aortic bioprostheses), and glaucoma, was scheduled for laparoscopic herniorrhaphy. Current medications included Carvedilol 12.5 mg and Aspirin® 100 mg. No known allergies. Airway evaluation revealed Mallampati class III (Figure 1), thyromental distance less than 5 cm, mouth opening less than 3 cm (Figure 2), limited neck extension (Figure 3), and altered dentition. In view of the predictors of difficult airway, pre-anesthetic consultation included planning for awake fiberoptic intubation. The patient was informed about the procedure, its risks, and the need for cooperation.

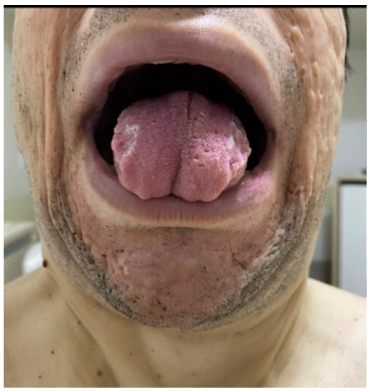

Figure 1: Mallampati III

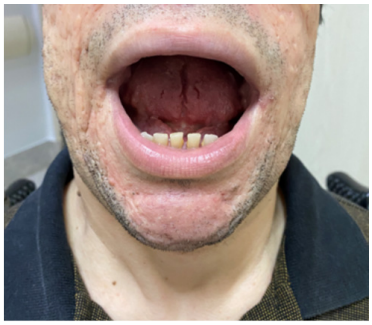

Figure 2: Mouth opening less than 3 cm.

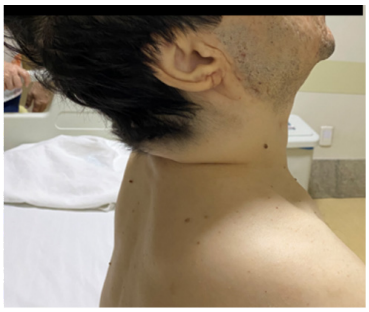

Figure 3: Limited neck extension.

Multiparametric monitoring was performed with pulse oximeter, cardiac monitor, pneumatic cuff, and capnography after intubation. The patient was admitted with blood pressure of 129 × 85 mmHg, heart rate of 84 bpm, and peripheral oxygen saturation (SpO_2) of 100%. Venous access was obtained in the right upper limb with a 20G intravenous cannula. Pre-oxygenation was initiated with face mask oxygen at 10 L/min, and sedation was performed with Dexmedetomidine 25 mcg and Midazolam 1 mg. Atropine 0.5 mg was administered due to its antisialagogue action. Superior laryngeal nerve block was performed with 3 ml of 2% lidocaine without vasoconstrictor (Figure 4), and glossopharyngeal nerve block with lidocaine spray.

Airway evaluation was performed by videolaryngoscopy (Figure 5), visualization being hindered by bleeding, with a Cormack-Lehane grade 4 view. Fiberoptic bronchoscopy was chosen (Figure 6), via the nasal route, with difficulty in tube advancement, requiring two attempts. After visualization of the vocal cords and access to the tracheal carina, a size 6 tracheal tube was passed slowly, requiring 3 minutes and 30 seconds for tube passage and cuff inflation (Figure 7). Intravenous anesthetic induction was then performed with Sufentanil 20 mcg + Propofol 120 mg + Cisatracurium 6 mg, and mechanical ventilation was initiated. Inhalational anesthesia was maintained with Sevoflurane 2%.

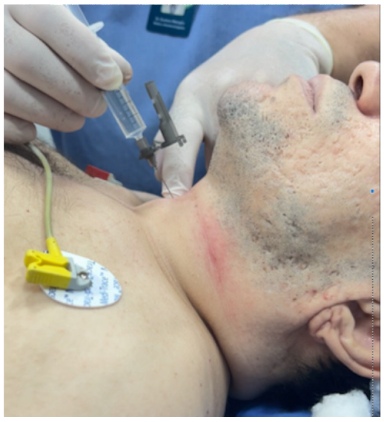

Figure 4: Superior laryngeal nerve block.

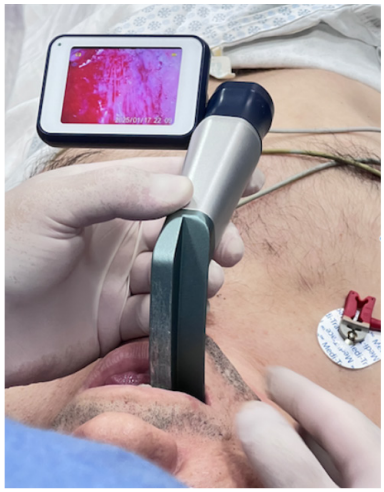

Figure 5: Airway evaluation by videolaryngoscopy.

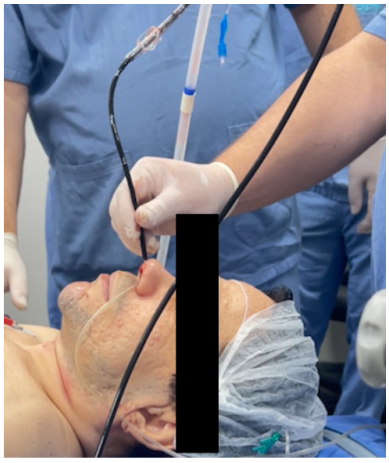

Figure 6: Nasotracheal tube placement via bronchoscopy.

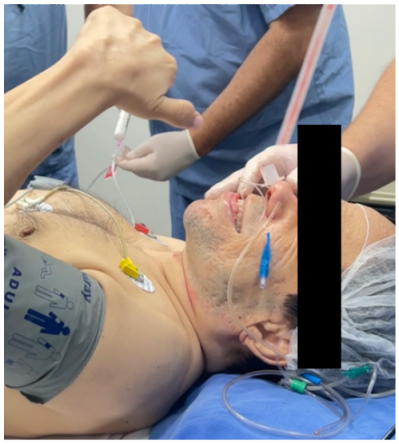

Figure 7: Anesthetic induction after nasotracheal tube placement.

The procedure was uneventful, and extubation was performed at the end of surgery after reversal with Atropine 1 mg and Neostigmine 2 mg. The patient remained in the post-anesthesia care unit for one hour and was then discharged to the ward.

DISCUSSION

The anatomical and physiological particularities of patients with MPS impose significant challenges for anesthetic management, especially regarding the airway. Findings such as high epiglottis, limited cervical mobility, micrognathia, restricted mouth opening secondary to temporomandibular joint dysplasia, pectus carinatum, tortuous and narrow trachea, tracheomalacia, skeletal dysplasia, scoliosis, and spinal cord compression, notably in the craniocervical and thoracolumbar regions, are frequently observed.[1,2]

In addition, the occurrence of odontoid hypoplasia, which predisposes to atlantoaxial instability, requires cervical stabilization in situations that demand movement, aiming to maintain the neck in a neutral position. In procedures requiring cervical manipulation, such as laryngoscopy, the risk of subluxation and spinal cord injury is considerable. In prolonged surgeries involving cervical and cranial manipulation, intraoperative somatosensory evoked potential monitoring is strongly recommended.[2]

The accumulation of GAGs in the pulmonary parenchyma impairs ventilation, making patients more susceptible to pulmonary infections, obstructive or restrictive lung diseases, upper airway resistance syndrome, and obstructive sleep apnea. Bronchospasm crises and oxyhemoglobin desaturation are also common clinical manifestations.[1,4]

Given the complexity of the airway in these patients and the need for specialized care, the Salford MPS Airway Score was developed. This comprehensive score evaluates the upper and lower airways through fifteen parameters, each scored from zero (normal) to three (severe abnormality). The sum of the fifteen parameters quantifies the severity of airway involvement. Clinical evaluation (parameters 1–6), nasoendoscopy (parameters 7–10), cross-sectional imaging studies such as computed tomography or magnetic resonance imaging (parameters 11–13), and pulmonary function tests (parameters 14–15) are used to determine the score.[3]

To ensure patient safety and design the ideal anesthetic strategy, a complete preoperative assessment is required, since MPS is a multisystemic disease. Comprehensive airway inspection is fundamental to evaluating the feasibility of general anesthesia, which strategies may be used, and which alternatives may apply. Older age is associated with a higher risk of difficult intubation, which, if unforeseen, may be fatal during management. Despite the challenges inherent in airway management in patients with MPS—from face mask ventilation to mechanical ventilation—general anesthesia remains the technique of choice and routine for many anesthesiologists. However, in patients with a predictably difficult airway, procedures requiring general anesthesia should be avoided whenever possible.[4,5]

As an alternative, neuraxial anesthesia should be considered. Musculoskeletal involvement, such as scoliosis, multiple dysostosis, and atlantoaxial instability, and restriction of joint mobility make it difficult to achieve the ideal positioning of the patient for regional block techniques. In addition, poor cooperation, often associated with intellectual disability and behavioral disorders, also limits the use of regional or neuraxial anesthesia in this population.[4]

The application of epidural anesthesia is difficult due to the need for patient cooperation, ideal positioning, and the time required for the technique. In addition, there are reports of epidural block failure, attributed to the deposition of mucopolysaccharides in the epidural space, preventing the distribution of the local anesthetic. It is also believed that mucopolysaccharide deposition may occur around the nerve fiber sheaths, making regional anesthesia more difficult. Failure of the technique due to mucopolysaccharide deposition has not been reported in spinal anesthesia.[4,5]

In the reported case, the choice of general anesthesia with awake intubation for laparoscopic herniorrhaphy was made after the pre-anesthetic consultation with the patient. The patient's airway was evaluated, possible predictors and complications were established, and alternative anesthetic plans were defined. In addition, the patient was questioned about past surgical history, which revealed no complications or adverse events. The patient's cognitive level, degree of understanding, and ability to cooperate with the procedure were also assessed. In the absence of a history of complications with previous general anesthesia, general anesthesia was chosen as the anesthetic technique. Given the predictors of difficult airway access, awake intubation was planned, with videolaryngoscopy evaluation and bronchoscopy if necessary. The choice of anesthetic technique was only possible due to the patient's understanding of the procedure and his collaboration.

CONCLUSION

MPS are a group of rare, progressive hereditary diseases caused by lysosomal enzyme deficiencies. The accumulation of GAGs in tissues and organs, including the airways, poses a significant challenge for the anesthesiologist. This case report describes the anesthetic management of a patient with Mucopolysaccharidosis type VI (Maroteaux-Lamy Syndrome) who underwent laparoscopic herniorrhaphy. Due to the predictors of difficult airway observed during pre-anesthetic evaluation, general anesthesia with awake nasotracheal intubation performed via fiberoptic bronchoscopy was chosen, an approach that proved effective in ensuring patient safety and the success of the surgical procedure.

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