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Case Report

Airway Management in Pediatric MPS: A Case Report

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ABSTRACT

Mucopolysaccharidoses (MPS) refer to a group of disorders classified as congenital metabolic defects, characterized by airway management challenges because of facial deformities, macroglossia, short neck, hypertrophic tonsils and adenoids, kyphoscoliosis, an immobile jaw, narrowed nasal passages, and atlantoaxial instability. In this report, we present a case of successful management of a difficult airway in a pediatric patient with MPS.

Introduction

Mucopolysaccharidoses (MPS) refer to a group of disorders classified as congenital metabolic defects, characterized by the absence of enzymes required for the metabolism of complex carbohydrates known as glycosaminoglycans (GAG) [1]. The prevalence of congenital metabolic disorders linked to complex carbohydrates ranges from 1 in 1,500 to 1 in 7,000 live births [2-3].

The general clinical manifestations of MPS include bone malformations, accumulation of intermediate metabolites of heparan sulfate or chondroitin sulfate type glycosaminoglycans in soft tissues, neurological alterations, decreased height, cardiovascular changes, and restrictive pulmonary alterations secondary to distortions in the rib cage, which will cause coarse facial features, ear, nose, and throat (ENT) issues, organomegaly, cervical spine instability accompanied by spinal cord compression, along with diminished vision and hearing, joint contractures, and various hernias [1,4]. The MPS group is classified into seven subtypes based on the affected genes and altered enzymes; these pathologies are categorized as monogenic disorders of autosomal recessive inheritance, except for MPS type II cases, which are characterized as diseases with an X-linked inheritance pattern with survival depending on the severity of the cases [5]. Most patients with MPS succumb to infectious and neurological complications, such as intracranial hypertension, as well as cardiovascular complications associated with heart failure syndrome and valvular failures [6-7]. Airway management poses challenges in these patients because of facial deformities and the aggregation of mucopolysaccharides in the nasopharynx [8].

Case Report

A 7-year-old known case of MPS presented to Mofid Pediatrics Hospital in Tehran, Iran, with a progressive decline in fine motor skills, accompanied by craniocervical junction stenosis and severe cord compression observed in a magnetic resonance imaging (MRI) study (Figure 1), without dislocation noted in a dynamic computed tomographic (CT) scan. The patient

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weighed 17 kg and exhibited a broad face, a relatively short stature, and a flattened midface. She had prominent, rounded cheeks, orbital hypertelorism, and a broad nose, along with a short neck (Figure 2). The patient's parents did not report any history of cardiovascular disease, and the past medical history was not significant. There was a surgical procedure in the patient's past history due to hydrocephalus leading to shunt insertion.

The patient presented to our operating room for cervical spine decompression. The patient had a 22-gauge intravenous (IV) line in the dorsal aspect of the right hand. She received 1 mg of IV midazolam for premedication and was transferred onto the operating bed. She received 10 mcg of IV fentanyl and inhaled sevoflurane, with careful increments in mean alveolar concentration (MAC) to achieve complete unresponsiveness. With spontaneous breathing and assurance of proper mask ventilation, 50 mg of IV propofol was administered. After complete apnea without using neuromuscular blocking agents, we attempted intubation three times with an armored cuffed endotracheal tube (ETT) size 5.0 using videolaryngoscopy, but it was unsuccessful. We switched to a standard ETT and performed intubation using its firm properties. The tube was then exchanged for an armored one using a tube exchanger. After intubation, the patient received 4 mg of IV cisatracurium and was placed in a prone position, secured with a neurosurgical Mayfield and tapes (Figure 3). A three-hour surgical procedure was performed, resulting in cord decompression and removal of the first cervical spine vertebra and foramen magnum. Anesthesia was maintained using sevoflurane and remifentanil IV infusion.

The surgical procedure was uneventful, and the patient’s vital signs remained stable throughout the operation. The amount of blood loss was minimal. At the end of the procedure, the patient was moved to the intensive care unit (ICU) intubated, and after five days, she was extubated.

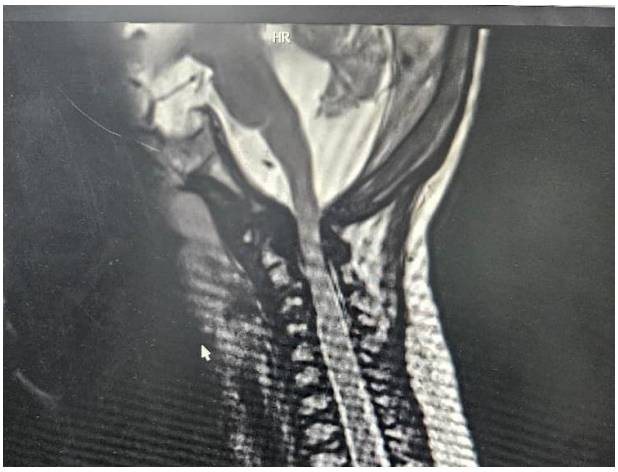

Figure 1- Sagittal view of spinal cord with severe cord compression of cervical spine

Figure 2- Patient’s facial features

Figure 3- Patient positioning

Discussion

Individuals with mucopolysaccharidosis (MPS) often undergo surgical interventions because of organ involvement and skeletal deformities in the extremities. According to registry data, at least one operation was required in 75% of cases, with patients typically undergoing a median of three to four procedures [9]. Airway management in these patients is often challenging, and the risk of difficult intubation should always be considered due to features such as macroglossia, short neck, hypertrophied tonsils and adenoids, kyphoscoliosis, restricted jaw mobility, narrowed nasal airways, and atlantoaxial instability [10]. According to Gonuldas et al. (2014), MPS patients present significant intubation challenges due to limited mouth opening, Mallampati class 3 or 4 airway scores, and recurrent adenoid hypertrophy [11]. Studies have shown that patients with mucopolysaccharidosis have a higher prevalence of dental caries, periodontal disease, malocclusion, and delayed tooth eruption. Additionally, cystic lesions or enlargement of the dental follicle are common. These oral manifestations—including gingival hyperplasia, diastemas, macroglossia, high-arched palate, and condylar defects—can pose significant challenges for anesthesiologists during airway management [12-13].

In this report, we present a case of successful management of a difficult airway in a pediatric patient with MPS. We identified the features of difficult intubation, and considering the nature of MPS, we utilized videolaryngoscopy instead of direct laryngoscopy, refraining from the use of neuromuscular blocking agents until successful intubation.

The incidence of difficult endotracheal intubation ranges between 28%-44% [14]. As shown by Arn et al. in 2012, surgery in MPS patients is linked to a high mortality rate due to airway obstruction, leading to difficulties in ventilation and oxygenation, which can cause significant cardiovascular compromise [15]. In 1994, Walker et al. reported a high rate of difficult intubation (11/29) and failed intubation (3/29) in a retrospective study of 110 surgical procedures in MPS patients [16]. Another study by Frawley et al. in 2012 demonstrated challenging mask ventilation in 14.2%, difficult intubation in 25%, and failed intubation in 1.6% of 17 MPS patients across 141 anesthetics [17].

Kurdi et al. in 2008 reported a case of difficulty/inability of ventilating with a face mask and intubation (can't intubate, can't ventilate (CICV)) of a 10-year-old boy with MPS [18]. Madoff et al. (2019) demonstrated a higher rate of difficult intubation in older patients compared to pediatric patients under 12 years of age. However, the difficulty of using laryngeal mask airways (LMAs) and mask ventilation did not show significant differences [19]. Scaravilli et al. reported that 29% of intubations were difficult for MPS patients, with older age being associated with a higher risk of difficult intubation [20]. Similar to our report, in their 2023 report, Tumer et al. examined 31 surgical procedures on MPS patients, revealing that video-laryngoscopy was the most frequently used alternative intubation method [21].

Extubation poses another significant anesthetic risk for MPS patients, who may experience post-obstruction pulmonary edema or struggle to maintain an airway after delayed extubation, necessitating urgent reintubation or tracheostomy [22].

Conclusion

This case reinforces the importance of proactive airway assessment and planning for increased risks of difficult endotracheal intubation, mask ventilation, and potential post-extubation respiratory complications unique to MPS. A multidisciplinary approach, including close airway monitoring and readiness for rapid reintubation or tracheostomy, is essential to minimize anesthesia-related morbidity and mortality in this population.

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