Case Report

Morquio Syndrome with Acute Cord Compression: An Anesthetic Challenge

Nischala Dixit, Bindu George, M Manjuladevi, Vineesh K Varghese, Archana Kalaichelvam

Department of Anesthesia and Critical Care, St John's Medical College and Hospital, Bengaluru, Karnataka, India

Abstract

Morquio syndrome with skeletal and extraskeletal manifestations is an anesthetic challenge. Airway management in children with Morquio syndrome can pose added risks of atlantoaxial instability, spinal cord compression, and distorted airway anatomy, leading to difficult mask ventilation and intubation. We report the anesthetic management of a 4-year-old child with Morquio syndrome who presented with acute cord compression and underwent emergency decompression of the stenotic spinal canal at the craniovertebral junction. Thorough preoperative evaluation, meticulous planning, and vigilant monitoring in the perioperative period are essential for safe anesthetic management.

Key words: Airway management, Morquio syndrome, spinal cord compression

Introduction

Morquio syndrome is a mucopolysaccharide storage disorder also known as mucopolysaccharidosis IV or lysosomal storage disorder. It is an inherited disorder of the metabolism with an autosomal recessive trait, existing in two forms: Morquio type A and type B. The enzymes deficient are N-acetylgalactosamine-6-sulfate sulfatase and β-galactosidase in type A and type B, respectively.[1] Morquio syndrome with skeletal and extraskeletal manifestations is an anesthetic challenge. Deposition of mucopolysaccharides in the soft tissue of the oropharynx adds to the risk in airway management, making mask ventilation and intubation difficult. Atlantoaxial instability results in the risk of subluxation and quadriparesis during airway manipulation.[2]

Case Report

A 4-year-old male child with Morquio syndrome presented with sudden weakness of all four limbs following a trivial trauma. Preanesthetic evaluation revealed short neck, large tongue, deformed pinna, low-set ears, barrel chest, knock-knee (genu valgum), flatfoot, and quadriparesis. Preoperative investigations included hemogram, urine examination, and renal and liver function tests, which were normal. Magnetic resonance imaging (MRI) spine revealed assimilation of the posterior arch of C_1, lying almost within the foramen magnum, causing narrowing of the spinal canal with significant compression of cord at the C_1-C_2 level [Figure 1], exaggerated lordosis of lumbar spine, and widened intervertebral disc spaces [Figure 2].

Sedative premedication was avoided in view of anticipated airway obstruction. Difficult airway cart was kept ready. Monitoring included electrocardiography (ECG), pulse oximetry, capnography, invasive blood pressure, and urine output. The child was induced with oxygen and graded doses of sevoflurane. After securing an intravenous (IV) access, glycopyrrolate 150 mcg and fentanyl 30 mcg were administered. Manual in-line stabilization of the cervical spine with the head in neutral position was maintained throughout airway manipulation. The airway was secured with 5 mm internal diameter (ID) uncuffed oral endotracheal tube with the patient on spontaneous ventilation. Anesthesia was maintained with titrated doses of atracurium and fentanyl, oxygen, air and isoflurane. IV fluid (250 mL) was administered during

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Address for correspondence: Dr. Bindu George, Department of Anesthesia and Critical Care, St John's Medical College and Hospital, Johnnagara, Bengaluru - 560 034, Karnataka, India. E-mail: bindu575@gmail.com

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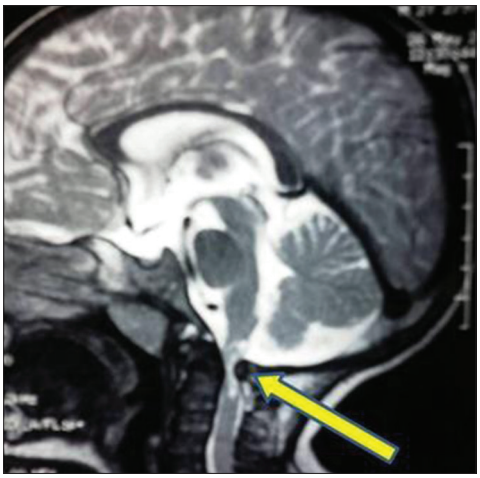

Figure 1: Arrow indicates spinal cord compression at the level of C_1 and C_2

Figure 2: Arrow indicates "fish mouth" appearance of vertebrae

surgery. The intraoperative period was uneventful. The patient was extubated when fully awake and shifted to the pediatric intensive care unit (PICU) for postoperative monitoring. The patient was discharged on the fifth postoperative day. Follow-up showed residual left hemiparesis after 1 month and complete recovery in 2 years.

Discussion

Breakdown of glycosaminoglycans (GAGs), keratin sulfate, and chondroitin sulfate is partially dependent on the lysosomal enzyme N-acetylgalactosamine-6-sulfate sulfatase.[3] Their accumulation in connective tissue, ligaments, and bone leads to progressive skeletal dysplasia and multiorgan dysfunction.[3,4] These patients need multiple surgeries, which include adenoidectomy, tonsillectomy, cervical decompression and fusion, and corrective hip and knee surgeries, often done at a mean age of 7–10 years and requiring repeated anesthetic exposure.[3-6]

An experienced pediatric anesthetist familiar with mucopolysaccharidosis disorders is required to carefully manage such cases in tertiary hospitals.[4] Anesthetic considerations include airway, respiratory, craniofacial, cardiac, skeletal, ocular, and hepatic abnormalities [Table 1]. Airway abnormalities include large tongue, unstable short neck, submucosal and cartilaginous GAG deposits, and tortuous trachea and bronchi.[7] Otolaryngologists and pulmonary physicians are therefore needed to document airway abnormalities and evaluate respiratory function.[7] Atlantoaxial instability contributes to significant morbidity and mortality during procedures. Odontoid hypoplasia, ligamentous laxity, and extradural GAG deposition can result in atlantoaxial subluxation with cord compression, leading to cervical myelopathy.[3] Prophylactic cervical spine fusion at a mean age within the range 5.9–9.9 years is recommended before development of clinical symptoms of spinal cord compression and cervical myelopathy.[7] Any trauma with flexion or extension of the neck risks the integrity of spinal cord and can be life-threatening.[4] Early surgical intervention (within 2 weeks) is indicated in injuries that cannot be managed conservatively.[8]

Table 1: Multisystemic involvement in Morquio syndrome

System	Clinical manifestations
Ocular	Corneal clouding
ENT	Conductive deafness/mixed deafness
	Flattened nasal bridge
	Large tongue
Cardiovascular system	Valvular abnormalities
	Regurgitant lesions
	Cardiomyopathy
Respiratory system	Narrowed trachea
	Restrictive lung disease
	OSA
	Pulmonary hypertension
	Cor pulmonale
	Respiratory failure
Abdomen	Hepatosplenomegaly
	Inguinal/umbilical hernia
Musculoskeleton	Dysostosis multiplex
	Joint laxity
	Dysplastic hip
	Genu valgum
	Unstable knees
	Large elbow, wrist
	Flatfoot
Spine	Atlantoaxial dislocation
	Focal gibbus
	Kyphoscoliosis

ENT: Ear, nose, and throat, OSA: Obstructive sleep apnea

Regardless of choice of equipment, awareness that Morquio syndrome patients may pose difficulty in intubation and ventilation is important. Difficult airway equipment and skilled airway personnel should be made readily available.[9] Apart from manual in-line stabilization and conventional laryngoscope, various tools for difficult intubation have been used. Fiberoptic bronchoscope (FOB), lighted stylet, laryngeal mask airway as a conduit to introduce

FOB or endotracheal tube, videolarygoscope (angulated video intubation laryngoscope), and GlideScopes have been reported in the literature.[3,7,10] Manual displacement of the tongue anteriorly during intubation or using difficult airway tools (videolaryngoscope, GlideScope) that displace soft tissue easily are preferred.[3] The passage of a FOB may be difficult and traumatic due to mucopolysaccharide infiltration distorting or narrowing the airway anatomy. When significant difficulty is anticipated, awake intubation with local anesthetic in conjunction with the availability of tracheostomy has to be considered.[9] Short neck, inability to extend neck, pectus carinatum, and pathological tracheal rings and mucosa may render emergency tracheostomy a difficult task.[9]

Thorough clinical and radiological preoperative evaluation of the airway with respect to spinal cord function is thus paramount before any airway management and surgical intervention in the patient.[4] In view of the history of obstructive sleep apnea, difficulty in mask ventilation was anticipated in this case and sedative premedication was avoided. In the operation theater (OT), difficult airway equipment and skilled personnel including an otolaryngologist for emergency tracheostomy were readily available. Two-person mask ventilation was necessary, with one person facilitating manual in-line stabilization to keep the head and neck in neutral position. Correct positioning requires careful alignment of the mastoid process with the clavicle, minimizing flexion/extension of neck so as to ensure a neutral position of the neck.[3,9] Our patient was induced with a graded dose of sevoflurane, maintaining patency of the airway. Difficulty in direct laryngoscopy and intubation is noted with advancing age.[11] Fortunately, direct laryngoscopy and glottic visualization was easy, except that our patient required an endotracheal tube smaller than predicted, as described by Tomatsu *et al*.[3]

Positioning should be done with utmost care as these patients are prone to injury secondary to joint laxity and instability. While positioning from supine to prone, the associated pectus carinatum can create intrathoracic and abdominal compression, which may affect venous return.[4] Customized padding at pressure points helps in prevention of compression neuropathy. Monitoring of spinal cord function using somatosensory evoked potential or motor evoked potential helps to detect surgery- or anesthesia-related neurophysiological changes.[4] In our patient, surgical decompression of the stenotic spinal canal at the craniovertebral junction was done by removing the posterior part of the foramen magnum and assimilated posterior arch of C_1.

Adequate precautions are needed for safe extubation. Whether this happens immediately after surgery or later in the intensive care unit, monitoring for any upper airway obstruction is mandatory. Our patient was extubated in the OT and was shifted to the PICU for postoperative monitoring. Early decompression of the stenotic canal at the craniovertebral junction thus prevents permanent damage to the spinal cord. Continuous surveillance thereafter is essential.[4]

Conclusion

Paramount in the anesthetic care of a Morquio syndrome patient is a thorough preoperative evaluation of the airway and of cardiac, respiratory, and neurological function. Anesthetic management must focus on protection of the airway without compromising the integrity of the spinal cord. Early surgical intervention with decompression is recommended to prevent permanent damage to the spinal cord.

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Conflicts of interest

There are no conflicts of interest.

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