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Case Report

Anaesthesia considerations in a case of Morquio syndrome with bilateral mechanical vertebral artery occlusion: a case report

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ABSTRACT

Morquio syndrome (MS) or mucopolysaccharidosis (MPS) type IVA is a progressive lysosomal storage disorder with autosomal recessive inheritance. Deficiency of enzymes N-acetyl-galactosamine-6-sulphatase and beta-galactosidase which compromises the catabolism of the glycosaminoglycans (GAGs) leads to excessive accumulation of GAGs in soft tissue, cartilage and bone causing severe skeletal dysplasia and difficult airway. Other systems commonly involved systems are cardiovascular, respiratory, neurological, ophthalmological, otolaryngological and hepatosplenic. Most of the patients with MPS survive up to second or third decade only. We present a case of 7 years old male child, known case of Morquio syndrome who presented with acute cerebral arterial stroke due to bilateral mechanical vascular occlusion of vertebral artery, posted for occipito-cervical fusion. Understanding Morquio syndrome, its airway and the anesthetic implications and careful planning of the anesthetic technique enabled us to manage this patient successfully.

Keywords: Morquio syndrome, Difficult airway, Mechanical vascular occlusion of vertebral artery, Occipito-cervical fusion

INTRODUCTION

Morquio syndrome is a combination of MPS type IVA and B. Deficiency of enzymes N-acetyl-galactosamine-6-sulphatase and beta-galactosidase which compromises the catabolism of the GAGs leads to excessive accumulation of GAGs in soft tissue, cartilage and bone. GAGs (keratan sulphate - the predominant GAG) have a predisposition for the trachea and cornea, which accumulates in the hyaline cartilage of the anterior tracheal rings causing tracheal stenosis and tracheomalacia with position-depending obstruction leading to difficult airway.

CASE REPORT

A 7 years old male child weighing 19 kg, height-113 cm (<3rd percentile), (known case of MPS IV) diagnosed with Acute Posterior-cerebral arterial Stroke due to mechanical vascular occlusion of vertebral artery at the junction C2-C3, by suspected rotational subluxation of C2 into os-odontodium with thrombo-embolism was scheduled for Occipito-cervical fusion.

On pre-operative examination, positive findings were skeletal dysplasia, short stature: disproportionate rhizomelia, kyphoscoliosis, mild knock knees, pectus carinatum, odontoid hypoplasia, and skeletal dysostosis. It was noted that patient had hypotonia with power grade 5 in both upper limbs and grade 4 in both lower limbs. Developmental and intellectual assessment were found to be normal. Patient was on Tablet Aspirin 75 mgs half B.D and Syrup Levipil 200mg B.D.

On airway examination, mouth opening was 4-fingers, Mallampatti grade 2, adequate neck extension with

restricted flexion, Thyro-mental distance 5 cm. Examination of rest of the systems was normal.

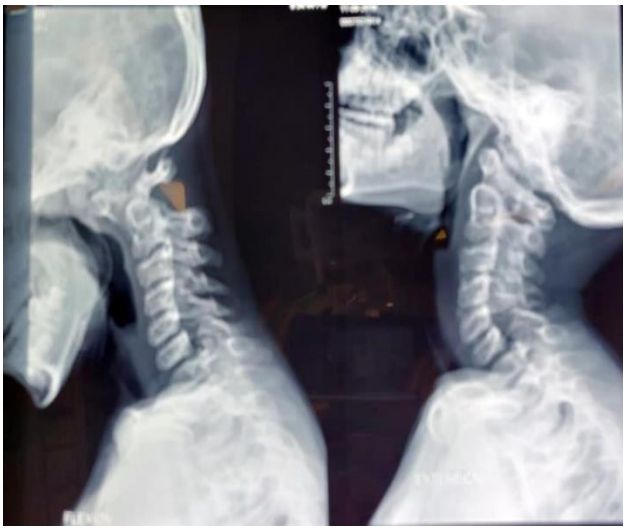

Figure 1: X-ray cervical spine lateral in flexion and in extension.

On the day of surgery all essential difficult airway equipment's and techniques like tube exchanger, fibreoptic bronchoscope, LMA, ENT support for tracheostomy were available inside the OT before induction. After attaching the monitors - NIBP, SpO2, ECG, oesophageal temperature probe, precordial stethoscope and adequate preoxygenation and premedication (Injection Midazolam 0.2 MG and Injection Glycopyrolate 0.08 MG and Injection Fentanyl 20 MCG I.V.)

Patient was induced in supine position with Injection Propofol 30 MG I.V. and bag and mask ventilation with oxygen and 2% sevoflurane. Intubation was done using 5.5 mm Cuffed flexometallic endotracheal tube using bougie after satisfactory check laryngoscopy. Straps were applied to patient in prone position, all pressure points and soft tissues, eyes were padded. Minimum Alveolar Concentration of Sevoflurane was maintained between 0.8 and 1 during surgery with Oxygen: Nitrous oxide: 1:1 and Sevoflurane. Injection dexamethasone 4MG and Injection hydrocortisone 100MG IV were given intraoperatively.

Surgical and anaesthetic courses were uneventful intra-operatively. After the surgery leak test was performed which was negative.

All essential difficult airway equipments and techniques like tube exchanger, fibreoptic bronchoscope, LMA, ENT support for tracheostomy were also made available before extubation. Total anaesthetic time was recorded to be 200 minutes.

Table 1: Systemic involvement in patient with Morquio syndrome.

System	Manifestation
Cardiac	Valvular regurgitant/stenotic lesion
	Reduced myocardial compliance
	Hypertension
	Coronary disease
	Arrythmia
	Cardiac failure
Respiratory	Macroglossia
	Short neck
	Hanging epiglottis
	Sub-glottic narrowing
	Oro-pharyngeal deposits
	Large adenoids/tonsils
	Restricted opening of temporo-mandibular joint
	Restrictive/obstructive patterns
Vertebral	Cranio-vertebral anomalies
	Odontoid hypoplasia
	Cervical cord compression
Skeletal	Short stature
	Curved metaphysis
	Short diaphysis
	Poorly developed epiphysis
	Wide acetabuli
	Hypoplastic femoral heads
	Pigeon chest
	Genu valgum
	Equino-valgum
Others	Prognathism
	Hepatomegaly
	Corneal clouding
	Glaucoma
	Degenerative retinal lesions
	Hearing loss

DISCUSSION

Morquio syndrome was described independently in 1929 by Morquio in Uruguay and Brailsford in the UK. Surgery in MPS patients is associated with a high mortality rate. A study comprising 932 patients enrolled in the MPS I registry that underwent a total of 4,762 procedures showed 30-day risk of death/procedure and death/patient rates of 0.7 % and 4.2%.[5]

A recent study including 17 MPS patients reported difficult mask ventilation in 20/141 anaesthetics (14.2 %), difficult intubation in 25 % and failed intubation in 1.6 %.[4] Older patients had more difficult intubations, confirming findings from previous studies.[3] Sometimes, failure to intubate requires an emergency tracheostomy.

When planning surgery in MPS patients, it is important to weigh the benefits against the risks of the procedure. Anaesthetic risk factors should be carefully evaluated preoperatively MPS type, age, previous anaesthetics and disease-targeting therapies (ERT, HSCT).

The main goal of the additional preoperative evaluations is to delineate the upper and lower airway anatomical and functional abnormality with ENT examination, sleep study and Pulmonary function tests. Coronal and axial MRI or CT of the airways is recommended in order to assess tracheal stenoses which can be found in a very proximal position. During intubation optical head and neck position should be given.

C1-C2 reduction and fusion in patients with MS is a procedure of significant perioperative concern. Careful attention needs to be given to the alignment of the head and neck and the ‘‘head should be positioned posterior to the body and looking up’’ This head and neck alignment is favoured by patients themselves because it optimizes airway patency and facilitates unobstructed breathing, giving the patients a somewhat peculiar posture that is easily discernible on examination. The airways of patients with MS easily obstruct when the neck is flexed, demonstrated by flow–volume loop, tracheal tomography, and fiberoptic tracheography reported by Pritzker et al. They noted anterior buckling of the posterior tracheal wall during flexion of the head, which caused a slit-like narrowing of the tracheal lumen. The need for a posture of optimal head and neck extension cannot be overemphasized in children with MS. While the causative mechanism is unknown, it might be attributed to a loss of tensile integrity of the tracheal walls due to a combination of abnormal hyaline cartilage composition and GAG deposits in the sub mucosal tissue. Other structural and morphological airway abnormalities result from sub-mucosal GAG deposits in the upper airways (tongue, floor of mouth, epiglottis, ary-epiglottis folds, and tracheal wall), all of which combined impart a rigid anatomy. Although rare, vocal cord paralysis has also been described in children with MS.

At least 2 anesthesiologist should be available during mask ventilation where one person manages the mask with anterior displacement of tongue while second person applies positive pressure breaths. Use of LMA in failed intubation maybe possible but is not guranted to provide adequate ventilation. It is possible to intubate via LMA which is well placed. Awake FOB maybe considered when all other methods have failed. When nasal FOB is attempted, bleeding from nostrils can attenuate the view. Application of vascular constrictore can be considered (adrenaline-soaked gauzes/otrivin nasal drops) ENT specialist should be available in case emergency tracheostomy is required.

Extubation represents another major anaesthetic risk factor in MPS patients, who can develop postobstruction pulmonary oedema or be unable to maintain an airway after late extubation, requiring urgent reintubation or tracheostomy

Extubation should not be performed before the patient is fully awake, coughing vigorously, breathing adequately and moving deliberately. Patients are best extubated early after surgery. This allows early assessment of neurological status and reduces airway swelling from intubation. If postoperative intubation is required for several days, fibreoptic bronchoscopy can be used to assess the extent of any swelling of the laryngeal area or obstruction from blood clots or other debris. As before, an adequate respiratory effort, a leak around the endotracheal tube and other measures necessary to ensure safe extubation should be followed.

All essential reintubation techniques like tube exchanger, fibreoptic bronchoscope, LMA, ENT support for tracheostomy should be made available before extubation.

Neurophysiological monitoring may also be considered in patients undergoing procedures other than spinal surgery, particularly patients at increased risk of spinal cord compression and for long procedures or procedures requiring head movement.

Most serious anaesthetic complications occurring during surgery in MPS patients are associated with airway obstruction, with accompanying difficulty in ventilation and oxygenation, resulting in significant cardiovascular compromise.

Following is a list of serious anaesthetic complications that may occur during anaesthesia in patients with MPS: inability to ventilate or intubate. Temporary airway obstruction: can cause negative pressure (potentially obstructive) pulmonary oedema. Complete airway obstruction (mostly during induction or at extubation): can cause profound hypoxaemia and cardiac arrest. Post-intubation problems: stridor, lower airway collapse/infection and need for reintubation or tracheostomy.

CONCLUSION

The MPS patient poses a major challenge to the anaesthetist. The anaesthetic risk can be reduced considerably if the anaesthetist anticipates potential problems that may arise in these patients during and after the procedure, including difficult intubation and ventilation, and cardiac and cervical spine issues. This requires a thorough preoperative evaluation and knowledge of the pathophysiology underlying the respiratory and cardiac manifestations, as well as cervical and tracheolaryngeal anatomy in these patients. Therefore, these difficult decisions should ideally be made by a multidisciplinary team in a tertiary referral centre experienced in the perioperative management of MPS patients. Anaesthesia in patients with an unstable

spine or for spine surgery is particularly difficult and requires additional care and thought in the choice of anaesthetic, monitoring and postoperative care.

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