Management of an anticipated difficult airway in Hurler’s syndrome

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Abstract

Management of an anticipated difficult airway in Hurler syndrome. Hurler syndrome is a subtype of Mucopolysaccharidosis (MPS) type 1. Mucopolysaccharidosis (lysosomal storage diseases) are a group of inherited disorders caused by deficiency of specific lysosomal enzyme required for normal degradation of glycosaminoglycans (GAGs). Administration of general anaesthesia in patients who have congenital syndromes such as Hurler’s is often a challenge because of progressive airway, craniofacial and skeletal abnormalities that may make both the ventilation and intubation difficult. We encountered difficult mask ventilation and endotracheal intubation was not possible and finally ventilated with laryngeal mask airway in a known case of Hurler syndrome posted for umbilical hernia repair.

Key words: Anesthesia, difficult airway, Hurler syndrome, mucopolysaccharidoses

Introduction

Hurler syndrome is a subtype of mucopolysaccharidosis (MPS Type I) named after a pediatrician Gerturd Hurler in 1919.[1] The MPS are a group of inherited metabolic disorders caused by a deficiency of specific lysosomal enzymes.[2,4] Administration of general anesthesia in patients who have congenital syndromes such as Hurler’s is often a challenge because of progressive airway, craniofacial and skeletal abnormalities that may make both the ventilation and intubation difficult. Anesthesiologists can expect to see more of these patients in future as new therapies such as bone marrow transplant and enzyme replacement therapy extend life expectancy.[3,6] We present our experience of managing anticipated difficult airway in a known case of Hurler syndrome.

Case Report

An 18-year-old man diagnosed as a case of Hurler syndrome (MPS Type I) was scheduled for repair of umbilical hernia. His medical history was significant for frequent upper respiratory tract infection, chronic otitis media and obstructive sleep apnea. He had undergone bilateral inguinal hernia repair at the age of 4 years without anesthetic complications.

Pre-operative physical examination revealed a dysmorphic individual with short stature weighing 20 Kg, height 111 cm and the age did not correlate with his appearance. Patient was co-operative, but slow in responding to commands. Head and neck findings included short neck with limited movement, macrocephaly, coarse facial features, depressed nasal bridge [Figure 1] with copious oral secretions and flexion deformity seen at elbows, wrists and knee joints. The airway assessment was Mallampati class 4. Systemic examination was noted for pectus excavatum, protuberant belly with an umbilical hernia and enlarged liver extending 7 cm below right costal margin. Laboratory investigations such as hemogram, urine routine and liver function tests were within the normal limit. Chest X-ray showed broadened anterior portion of ribs (oar shaped/paddle ribs) with widened medullary cavity [Figure 2]. Ultrasound of the abdomen revealed hepatomegaly with fatty infiltration and umbilical hernia with bowel loops and omentum as contents. Echocardiogram showed mitral valve prolapse with moderate mitral regurgitation. Biochemical test on
urine showed the presence of MPS by toluidine blue spot test. Specific enzyme assay revealed deficient α-L-iduronidase enzyme activity.

Management

We planned for general anesthesia either with endotracheal intubation or laryngeal mask airway (LMA). From the physical examination, difficult intubation was anticipated. Infective endocarditis prophylaxis was given. In the operation theater, difficult airway cart was kept ready except for fiberscope. On arrival to the operating room, an 18 gauge intravenous (IV) line was established and was premedicated with Ranitidine 50 mg IV and atropine 0.4 mg IV 15 min prior to induction. Basic monitors were connected. After preoxygenation with 100% oxygen anesthesia was induced with fentanyl 20 mcg and propofol 40 mg IV, titrated to maintain spontaneous respiration. To confirm mask ventilation oral Guedel airway size 2 was inserted and the ventilation was not possible. Immediately, we removed the airway and were able to accomplish adequate mask ventilation. Check laryngoscopy was attempted in deeper plane with the patient breathing spontaneously. The laryngoscopic view was limited to the tip of the epiglottis even with optimal external laryngeal manipulation and copious amount of secretions were present. An attempt was made to guide 5 mm tracheal tube with stylet under epiglottis and was not successful. Meanwhile, patient developed bronchospasm with bilateral rhonchi and the oxygen saturation dropped to 80%. Patient responded to intermittent positive pressure ventilation with 100% oxygen by mask, deriphylline 100 mg and hydrocortisone 100 mg IV. Spontaneous respiration regained and the saturation picked up to 100%. Again anesthesia was deepened with injection propofol 20 mg with halothane 1% and classic LMA size 2 was inserted. We were able to ventilate adequately through LMA. Then anesthesia was maintained with oxygen, nitrous oxide and halothane 0.2%. A bolus of atracurium 10 mg IV was given to achieve muscle paralysis and the patient was connected to volume controlled ventilation. Surgery lasted for ½ h and at the end patient was reversed with atropine 0.4 mg and neostigmine 1 mg IV. LMA was removed after ensuring adequate respiration and patient being fully awake. The patient remained hemodynamically stable throughout the procedure. Supplemental, humidified oxygen (6 L/min) by mask, dexamethasone 4 mg and post-operative analgesia with diclofenac 20 mg IV was provided in post-anesthesia care unit. Post-operative period was uneventful and he was discharged on the 2nd post-operative day.

Discussion

MPS are a group of rare genetic lysosomal storage disorders characterized by the deficiency in or complete lack of necessary lysosomal enzymes required for the stepwise breakdown of glycosaminoglycans (GAGs, also known as MPS). Consequently, fragments of GAGs accumulate intracellularly in the lysosome resulting in cellular enlargement causing disruption/dysfunction of structure and function of tissues. Of the total 11 MPS disorders, there are seven major types.[6,7] Most commonly the disorders are divided into mild and severe variants. In that MPS Type I, the severe variant is known by its eponym as Hurler’s syndrome (Type IH) and the mild variant is Schie’s syndrome (Type IS). Anything apparently not confirms to this division is called Hurler/Schie syndrome (Type I HS) by most pediatricians.[8] All the subtypes of MPS I (Type IH, HS and S) share an identical enzyme deficiency, but demonstrate distinctly different clinical features.[9]

Hurler syndrome, the most common MPS, occur 1 in 100,000 live births. There is a mutation of the α-L-iduronidase enzyme,
which leads to excessive accumulation of partially degraded GAGs dermatan sulfate and heparan sulfate within cells.\textsuperscript{[4]} For the Anesthesiologist, the establishment and maintenance of an adequate airway represent the most commonly encountered problem in MPS I.\textsuperscript{[9]} These include a short neck with limited mobility, relatively high epiglottis, a deep cranial fossa that narrows nasopharynx, hypoplastic mandible, ankylosis of temporomandibular joint, infiltration of pharyngeal tissues and tracheal cartilages, intraluminal narrowing of the conductive airways and excessive secretions, macroGLOSSia, friable oral mucosa and tonsillar hypertrophy.\textsuperscript{[22]}

The incidence of difficult and failed intubation in patients with Hurler syndrome are 54% and 23% respectively.\textsuperscript{[10]} Investigations include urine spot tests, which are readily available to screen for MPS, but are associated with false-positive and false-negative results. Heparan, keratan and dermatan sulfate can be distinguished by electrophoresis techniques to differentiate between the MPS. The diagnosis is confirmed by direct enzyme assay of leucocytes or fibroblast.\textsuperscript{[5]}

In our case, in spite of antisialagogue we observed a copious amount of secretions in the oral cavity. Diaz and Belaine\textsuperscript{[4]} have stated that sedate premedications should be avoided for fear of upper airway obstruction, use of antisialagogue may help reduce secretions and antibiotic prophylaxis is recommended for all patients with MPS.\textsuperscript{[11]}

Local or regional anesthetic technique are unsuitable as the sole form of anesthesia in young and uncooperative children and it may fail because of deposition of MPS in the nervous system.\textsuperscript{[12]} In spite of anticipated difficulty in securing the airway, we chose general anesthesia in this case. Intraperitoneal procedures, including umbilical herniorrhapsy with its potential for peritoneal cavity and bowel manipulation will require general endotracheal anesthesia to provide a secure airway for controlled ventilation during neuromuscular paralysis and to protect lungs from aspiration of gastric contents.\textsuperscript{[13]} We chose to induce with propofol titrated to maintain spontaneous ventilation. Airway manipulation is much easier to perform in deeply sedated spontaneously ventilating patients.\textsuperscript{[6]} Many authors preferred inhalational induction with halothane or isoflurane in older patients and with severe somatic involvement in MPS. IV induction with titrated doses of thiopentone or ketamine proved more satisfactory in severely retarded and uncooperative patients as long as spontaneous ventilation can be maintained until adequate airway control is assured.\textsuperscript{[4,9,14]} We used halothane for the maintenance of general anesthesia. Diaz and Belani reported rarely does functional hepatic impairment contraindicate halothane inhalation.\textsuperscript{[4]} Baines and Keneally reported preponderance of inhalation induction, primarily with halothane. Although, the incidence of coronary artery and cardiac valvular involvement is reportedly high in these patients, isoflurane-induced intracoronary steal pose less risk to MPS patients.\textsuperscript{[4,9]}

We experienced difficult mask ventilation after oropharyngeal airway insertion. Many authors have mentioned the similar problem and stated that the oropharyngeal airway pushes the epiglottis back and down so that it occludes the laryngeal inlet or buckle the posterior third of the long tongue causing airway obstruction.\textsuperscript{[3,4,14]} A flexible nasopharyngeal airway softened by pre-soaking in warmed fluids may offer a better artificial airway in this type of cases.\textsuperscript{[4]} We managed to secure the airway with LMA. If LMA insertion had become difficult or unsuccessful, we planned retrograde intubation or as the last resort tracheostomy as an alternate approach to secure the airway in this case. LMA has been used successfully and found to be very useful in these patients by many authors specially when the intubation attempt has failed.\textsuperscript{[13-15]} Proseal LMA is a better option for upper gastrointestinal surgeries. Even though, there was adequate mask ventilation, we avoided muscle relaxant because of difficulty in securing the airway. As always, the ability to maintain a patent airway during either spontaneous or assisted ventilation does not necessarily mean that such control will continue after the administration of muscle relaxants.\textsuperscript{[4]} Regarding alternate approaches to airway management, tracheostomy carries significant risks because of short neck and MPS deposits anterior to trachea.\textsuperscript{[3,6,14]} Cricothyroidotomy is not recommended for MPS patients whose cricothyroid membrane, cricoid cartilage and thyroid cartilage are often thickened and deformed by MPS deposits making rapid dissection difficult and vocal damage likely.\textsuperscript{[14,15]} In case of difficult intubation, multiple attempts at tracheal intubation will predispose a patient to symptomatic glottic and subglottic edema.\textsuperscript{[4]} To prevent that we administered humidified oxygen and dexamethasone post-operatively. We realized from this case that flexible fibreoptic intubation could have been attempted at the onset maintaining spontaneous respiration under sedation. Alternatively, intubation could have been performed through fibreoptic technique or an intubating LMA under general anesthesia. Unfortunately, fiber optic bronchoscope was not available with us.

To conclude, paramount in the anesthetic care of such patients is a thorough knowledge of MPS, careful pre-operative and perioperative management plan and preferably they should be anesthetised by an experienced anesthesiologist if disaster is to be averted.
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