Case Report

Cardiac Arrest in a Case of Mucopolysaccharidosis after Tracheostomy

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Abstract

Airway management during induction of anesthesia is difficult in some metabolic disorders like mucopolysaccharidosis (MPS). In this article we report an 11 years old child with Hurler-Scheie syndrome how was admitted to operating room for tracheostomy under general anesthesia. Mask ventilation was difficult and endotracheal intubation was impossible and anesthesiologist ventilated the patients lung by inserting laryngeal mask airway. The purpose of this article is to increase awareness among anesthetists and surgeons about the anatomical and pathophysiological changes in these syndrome which may lead to severe airway problems and complications in perioperative period.

Keywords: Mucopolysaccharidosis  
Hurler-Scheie syndrome  
Difficult Intubation

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Introduction

Mucopolysaccharidosis (MPS) are a group of inherited disorders of connective tissue metabolism in which lysosomal enzyme deficiency leads to deposition of mucopolysaccharides or glycosaminoglycans in the airway, cornea, brain, heart, liver, spleen, bones,… producing different symptoms. The perioperative management of a case of MPS is considered as a challenge to the anesthetist. It represents one of the worst airway problems in pediatric anesthesia.

Case

An 11 years old male (weight 19 kg) child of Hurler-Scheie Syndrome was exposed to general anesthesia for tracheostomy. He was diagnosed as MPS at the age one. The features of Mucopolysaccharidosis gradually progressed over the years. He developed stiffness of joints of upper and lower limbs. He was short in stature (85cm) and had an apathetic face with saddle deformity of nose, antimonogolian eyes, small lower jaw, glossoptosis and a normal mental function. He had a history of sleep apnea, respiratory distress, inspiratory and expiratory stridor from the age of two. He had surgery for cleft palate repair, adenotonsilectomy and ear ventilation tube (VT) replacement and inguinal hernia repair when was 4, 6, and 7 years old respectively in our center without any event (difficult intubation). His age was not correlated with his appearance. He had multiple hospitalizations because of respiratory problems. He had severe respiratory distress in supine position. His tongue was very large with limited mouth opening and malampatti grade 4 views. He had a short neck with limited extension. ECG, NIBP and SpO2 monitors were attached. He was preoxygenated with 100% oxygen in sitting position. Anesthesia was induced with fentanyl 10 µg, midazolam 1mg, propofol 20 mg, lidocaine 10mg. He had spontaneous ventilation. Mask ventilation was difficult and endotracheal intubation was impossible with direct laryngoscopy. It wasn't possible to pass fibroptic bronchoscope transducer through mouth and nose because of huge tongue. The patient ventilated through larynged mask airway (LMA) No 2 and was stable during tracheostomy. Maintenance of anesthesia was with halothane 1-1.5%. The LMA was removed when he was awake. In recovery room the general condition of patient was deteriorated and Spo2 declined. We couldn't ventilate the patient through tracheostomy tube and because of airway obstruction the patient had cardiac arrest that did not respond to complete sequence of CPR with early defibrillation.

Discussion

Hurler syndrome or MPS 1 H is the prototype of MPS and is the most severe form of it. It has been described, as posing worst airway problem in pediatric anesthesia. MPS are rare conditions, incidence varying from 1 in 24,000-500,000 population. Knowledge of the anesthetic implications of the disease is essential to prevent any catastrophe. The anesthesiologist may face diverse problems while anesthetizing these patients. The most important and life threatening problem is difficulty in maintaining airway while anaesthetizing such patients because of the anatomical changes in upper airway due to deposition of mucopolysaccharides in tongue, tonsils, adenoid, epiglottis, glottis and trachea. They also have excessive tracheobronchial secretions with frequent upper respiratory infection. Chest deformity along with deposits in lower respiratory tract and lung interstitium may cause obstructive lung disease and diffusion defects leading to hypercapnia, hypoxia and elevated airway pressures. In preanesthesia survey the type of MPS syndrome should be confirmed as it has a great implication for the degree of difficulty anticipated during airway management. Child’s intelligence and behavior are important. In Children with MPS, behavior may vary from uncooperative belligerent to placid, cooperative and lovable. Child’s favorable sleeping position should be inquired, since this may be the position in which airway is held open. History of snoring and sleep apnea should be asked. Thorough examination of cardiovascular system and respiratory system is required. Cardiovascular defects and its implications in anesthesia should be taken care. Lung function should be optimized preoperatively by energetic physiotherapy and
antibiotic treatment of preexisting pulmonary infection. In all cases of MPS excessive deposition of mucopolysaccharides continues throughout the life with a special predilection for tracheal cartilages, therefore clinical features worsen in all of them as age advances. In children with MPS, maintaining airway with facemask is difficult as they have macroglossia, narrow upper airway and limited temporomandibular joint mobility. Airway management often becomes more difficult by insertion of an oropharyngeal airway as it pushes their long high anterior epiglottis over the laryngeal inlet or may buckle the posterior end of large tongue causing occlusion of the airway. Nasopharyngeal airway may sometime improve the airway but there is often difficulty in inserting it due to deposits in the nasopharynx. Kemphome et al. used tongue suture in a patient to pull the enlarged tongue forward to relieve obstruction. Laryngoscopy may be unduly difficult in patients with MPS as they have oral and laryngeal deformity, rigidity of the area and copious viscous oral secretion. A high laryngeal inlet (C2) and an anteriorly inclined larynx will usually make intubations difficult. Awake fiberoptic intubation is the method of choice in these children. But because of subnormal mental function, it will not be easy to perform awake fiberoptic intubation in these children. In our case it was impossible because of huge tongue. Blind nasal intubation, was also tried. LMA has been used successfully in these patients specially when intubation failed (as our case). It was also considered as an excellent aid to fiberoptic intubation in more difficult cases of MPS. Tracheostomy is difficult in these children as they have short neck, mucopolysaccharide deposits anterior to trachea and a relatively narrow trachea. In a case report it was found to be impossible even at postmortem. Cricothyrotomy is not recommended in MPS patients as their cricothyroid membrane, cricoid cartilage and thyroid cartilage are often thickened and deformed by mucopolysaccharides deposits making rapid dissection difficult and vocal cord damage likely. Distal tracheal obstruction can not be bypassed by endotracheal intubation or tracheostomy and may prove fatal. Thus severe airway difficulty can occur while anaesthetizing such children. In all cases inhalation induction with maintenance of spontaneous ventilation is preferred but in mentally retarded and uncooperative patient intravenous induction is more satisfactory. In all these patients spontaneous ventilation should be maintained until adequate airway control is achieved. Therefore as a rule of thumb spontaneous ventilation should be maintained until the airway is secured. Recovery after GA in patients with MPS is often slow and accompanied by periods of breath holding, apnea, bronchospasm, cyanosis and respiratory arrest. Therefore anesthetic sequences which ensure early return of consciousness and airway reflexes were strongly recommended. Regional anesthesia offer a valuable and a safe alternative for children with MPS undergoing lower abdominal, perineal, upper and lower extremity. In conclusion, in children with MPS, anesthesiologist and surgeons should be aware of the expected complications. The benefit of the surgical procedure should be balanced against the risk of exposing the child to general anesthesia. The parents of the patient should properly be informed of the risk involved. Anesthesia should ideally be given by the anesthesiologists who are experts in handling pediatric airway problems and resuscitation, in a center in which pediatric intensive care facilities are available.

References