Anesthetic Management of a Child with Maroteau-lamy Syndrome

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Mucopolysaccharidoses (MPS) are diseases characterized by excessive storage and excretion of mucopolysaccharides. According to the sort of enzyme deficiencies, the MPS diseases are currently classified into seven types depending on mucopolysaccariduria patterns and the clinical features. Maroteaux-Lamy syndrome (MLS) is MPS type 6.

The etiology of MLS is an inherited deficiency of arylsulfatase-B, which causes an increased urinary excretion of mucopolysaccarides consisting predominantly of dermatan sulfate[1]. The clinical manifestations of MLS are a short statue, short neck, coarse facies, osseous changes, hepatosplenomegaly, umbilical and inguinal hernia, hearing loss and mental retardation[2].

Clinical interest in MLS has focussed on the problems caused by the accumulation of mucopolysaccharides in the cardiac tissues[3] and the frequent respirator infections which may increase postoperative morbidity and mortality. We present the anesthetic management of a child with MLS undergoing repair of an inguinal hernia.

Case Report

A 6 years old boy (weight 20 kg, height 102 cm) was admitted to our hospital for elective surgery to repair an inguinal hernia. He had previously been diagnosed as MLS by the clinical characteristics and a biochemical examination in our hospital when he was 3 years old.

His physical characteristics were a short neck, enlarged tongue, and thickened face (gargoylism). The physical examination revealed that he had mild hepatosplenomegaly, conductive deafness and a speech defect. In addition the chest roentogenogram showed cardiomegaly. There was systolic murmur, best heard near the apex. Furthermore, the echocardiograph revealed mitral regurgitation (grade II), but neither myocardial thickness nor poor ventricular contraction was observed. Preoperatively he always had copious secretions in his mouth and suffered from frequent respiratory infections. After his infection had been treated with antibiotics, he underwent operation.

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Table 1. Maroteaux-lamy syndrome in Japan: Summary of patients' abnormalities

Age (years)	Sex	Short neck	Enlarged tongue	Hearing loss	Mental retardation	Dysostosis multiplex	Hepatosplenomegaly	Recurrent pneumonia	Cardiac murmur	Valve disease	Inguinal or umbilical hernia
2	F	+	+	−	+	+	+	+	+	?	+
5	F	+	+	+	?	+	+	?	?	?	+
6	M	+	+	+	−	+	+	+	?	?	?
11	F	+	+	+	−	+	+	+	+	?	?
29	M	+	+	−	−	+	+	+	+	+	?
36	M	+	+	?	−	+	+	?	+	+	?
28	M	+	+	−	−	+	+	?	+	?	+
32	M	+	+	−	−	+	+	?	+	?	+
29	F	+	+	+	−	+	+	?	+	?	−
33	F	+	+	+	+	+	+	?	+	?	+
6	M	+	+	+	+	+	+	+	+	+	+
Incidence		11/11	11/11	6/10	3/10	11/11	11/11	5/5	9/9	3/3	6/7
(%)		(100)	(100)	(60)	(30)	(100)	(100)	(100)	(100)	(100)	(86)

(+: exist, −: absent, ?: unknown)

Preanesthetic medication consisted of simply 0.3 mg of intramuscular atropine. Special preparations for bronchoscopy and tracheotomy were made before induction, in view of possible difficulties in maintaining the airway during anesthesia. Anesthesia was induced via a mask with enflurane and nitrous oxide (N_2O) in oxygen. Muscular relaxation was obtained with succinylcholine 20 mg intravenously. His short and immobile neck and his enlarged tongue, however, made it difficult to manually ventilate. Tracheal intubation using the laryngoscope with a curved No. 2 blade was attempted several times. But, since his enlarged tongue and thickend oropharyngeal tissues prevented the visualization of the epiglottis, the orotracheal intubation was blindly performed with a single lumen tube with a 5.0 mm internal diameter. During the repeated attempts at intubation, the heart rate and the blood pressure remained within normal physiologic ranges; however, the oxygen saturation (Sp_{O_2}), measured by a pulse oximeter transiently decreaed to 92%.

Anesthesia was maintained with 1% enflurane and 60% N_2O in oxygen. Muscular relaxation was sustained by intermittent administrations of vecuronium. The Pa_{O_2}, Pa_{CO_2}, and Sa_{O_2} values and potassium and sodium ion concentrations in his arterial blood were all maintained within normal ranges. No severe changes in blood pressure and heart rate were observed throughout the surgical procedure. After the operation, the muscular relaxation was countered by an intravenous administration of 0.5 mg atoropine and 1.5 mg neostigmine. Copious secretions in his respiratory tract were aspirated and after complete recovery of spontaneous respiration, his trachea was extubated. Following extubation, no obstruction in the respiratory tract was observed. The postoperative course was uneventful and the patient was discharged 5 days after the operation.

Discussion

The incidence of MLS is thought to be very rare. In a nation-wide study on MPS in Japan, eleven cases of MLS have been identified in 634 cases of MPS. In table 1, we have reviewed the clinical manifestations and abnormalities of these cases. The three possible anesthetic problems caused by these physical characteristics are as follows: (a) perioperative management of airway, (b) treatment of frequent respiratory infection, and (c) evaluation of cardiac function. These may be associated with decreased postoperative mortality and morbidity.

(a) *The perioperative management of the airway*

Preoperatively, the alleviation of airway problems must be considered to be the highest priority. The difficulty in manual ventilation and tracheal intubation is attributed to the thickend soft tissues that narrow the upper airway, the inability of cervical extention, and the copious secretions. In addition, sedating the patient with barbiturates, tranquilizers or opiates may depress perioperative ventilatory drive[4]. Whereas, premedication with belladonna drugs to minimize secretions is well advocated. Some doctors, however, do not administer belladonna drugs to patients with MPS because these drugs increase the thickness of secretions[4]. During induction, the excessive extension of the neck and the small abnormal cervical vertebrae may cause spinal injury[5]. The administration of high concentrations of both inhalation anesthetics and muscle relaxant to patients in the supine position, may cause the tongue to fall into the retrolingual space which can lead to severe airway obstruction. Although an anesthesiologist may often

encounter difficulties trying the manually ventilate via a mask during general anesthesia, intubation either under light anesthesia or without muscle relaxation may cause a high incidence of laryngospasms. Furthermore, since the large adenoid and thickened tissues prevent the visualization of the vocal cord, repeatedly and traumatically attempted endotracheal intubation may cause dramatic hemorrhage.

The preoperative evaluation of the relationship between the larynx and other structures by the lateral cervical roentogenogram and the respiratory tract by computed tomography may be useful[6]. Various skillful techniques of intubation in MPS have been described: blind intubation, fiberoptic intubation, endotracheal intubation by a retrograded guidewire, and transtracheal intubation[7–9]. These techniques can also be applied to patients with MLS. The preparation for an emergency tracheotomy during the perioperative period is neccessary, however, the patient's vulnerable and narrow trachea may be intolerant of the long-term intubation. In addition, during anesthesia, the Sp_{O_2} and endtidal CO_2 pressrue should be monitored. Since the recovery period is often accompanied with respiratory distress, close observation of the spontaneous respiration and careful monitoring of Sp_{O_2} are recommended[10].

(b) *The treatment of frequent upper air tract infections*

Because the patients with MLS have frequent respiratory infections (recurrent pneumonia)[6], preoperative treatment with antibiotics is required.

(c) *The evaluation of the cardiac function*

The cardiac characteristics of MLS, such as cardiomyopathy, valvular disease and coronary artery stenosis, have been previously described[3,11–13]. Valve lesions are observed with higher incidence in MLS than in other types of MPS[11]. Semenza et al. reported that the echocardiograph confirmed valvular thickining in all their MLS cases[6]. The preoperative evaluation of the cardiac function, espesially by echocardiograph, is therefore necessary and recommended.

MLS patients can potentially live long and in their long life time they repeatedly undergo operations for their symptoms which can be treated with surgical therapy[14]. Thus, as documented in literature[7,14,15], anesthesiologists encounter patients undergoing frequent surgical procedures such as adenoidectomy, tonsilectomy, hernia, repair, shunting for hydrocephalus, spinal fusion for atlantoaxial subluxation, and cardiac valve and hip replacements.

In conclusion, we reported on a case of a child with MLS and discussed the anesthetic problems associated with treating the disease. We believe that anesthesia for MLS should be, in principle, the same as for other types of MPS. Since the natural course of the syndrome toward an advanced stage aggravates and complicates the pathophysiological status of patients, a detailed preanesthetic evaluation for each surgical procedure is necessary. Furthermore, as upper airway obstruction during induction and difficulties associated with intubaiton are critical problems for an anesthesiologist, various skillful techniques should be employed to overcome airway problems associated with MLS.

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