

Successful spinal anesthesia in a patient with mucopolysaccharidosis type I under femoral fracture reduction and external fixation

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ABSTRACT

Introduction: Mucopolysaccharidosis (MPS) is an inherited lysosomal storage disorders with glycosaminoglycans accumulation in tissues. MPS patients undergoing intratracheal intubation anesthesia show high mortality, with serious anesthetic complications associated with airway thickness and narrow. The regional anesthesia is a useful alternative to general anesthesia, however performing spinal anesthesia in MPS patients are rarely documented.

Case presentation: We report a case of a boy with MPS type I undergoing femoral reduction and external fixation under spinal anesthesia in combination with sevoflurane inhalational induction, getting rid of difficulties associated with intubation.

Conclusion: Sevoflurane inhalational induction with spinal anesthesia without tracheal intubation is a safe choice for MPS I patient.

KEYWORDS

Mucopolysaccharidosis, Children, Spinal anesthesia, Sevoflurane, Airway management

INTRODUCTION

Mucopolysaccharidosis type I (MPS I) is the most common of the mucopolysaccharidosis (MPS) disorders and is caused by a deficiency of the lysosomal enzyme α -L-iduronidase. It is a genetic disease with autosomal recessive inheritance.^{1,2} Common symptoms include growth retardation, short stature, intellectual disability, multiple dysostosis, joint stiffness, corneal clouding, cardiomyopathy, valvulopathy, respiratory insufficiency, recurrent respiratory infections, and hepatosplenomegaly.^{2,3} Patients with MPS I have multiple comorbidities requiring frequent surgical procedures with anesthesia, and they have facial and airway characteristics that may create a challenge for anesthetic airway management. Therefore, airway-related anesthetic complications in patients with MPS I are also a part of pediatric anesthesia. Clark et al⁴ found that the overall incidence of difficult tracheal

intubation in a case series of various types of MPS ranged from 28% to 44%. Regional anesthesia seems to be a logical choice for these patients. We herein report the anesthetic management of a 15-year-old boy with MPS I and femoral fractures. In view of probable femoral fractures with pneumonia, he was considered for surgery.

CASE REPORT

A 15-year-old boy weighing 26.5 kg presented for correction of femoral fractures. He had typical intellectual disability, a protruding tongue, and stiff limbs. He had a normally sized head on a small body (118 cm), malnutrition, and a short neck. Moist rales were auscultated, and oral secretions were observed. He had a history of recurrent respiratory infections and required intermittent auxiliary sputum suction. He also required noninvasive ventilator-assisted ventilation during sleep. He

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had undergone four operations under general anesthesia: left and right hernia sac high ligation, correction of an umbilical hernia, and blepharoplasty. Laboratory tests showed an activated partial thromboplastin time of 49.5 s, but other parameters were within the reference range. A chest radiograph showed pneumonia. A two-dimensional echocardiogram showed aortic and mitral valve regurgitation with an ejection fraction of 67%. However, an electrocardiogram was normal. The patient had difficulty communicating because of consciousness disturbance. He had not received enzyme replacement therapy, resulting in worse symptoms.

The patient's parents were informed that the child faced grave risks during anesthesia, including possible death. We explained to them that we were planning to perform the surgery under spinal anesthesia with the minimal amount of initial general anesthesia that was necessary. We also explained that in the event of failure of spinal anesthesia, we might need to intubate the trachea with the possibility of difficult tracheal intubation.

Monitors for electrocardiography, oximetry, blood pressure measurement, and capnography were attached to the patient. Anesthesia was induced smoothly by inhalation of sevoflurane 8% in oxygen. Once the patient was asleep, he was turned to his right side. Satisfactory gas exchange was maintained, although his breathing was noisy. He was prepared for subarachnoid puncture. A 22-gauge lumbar spinal needle was introduced in the L3–4 interspace after local infiltration of 1% lidocaine. Clear cerebrospinal fluid flowed out of the needle, and 3 mL of 0.5% ropivacaine was injected. During the surgery, the patient was given oxygen via a mask, and his vital signs were stable. After an uneventful 1-hour procedure involving femoral fracture reduction and external fixation, he was transported to the general ward. No postoperative complications associated with anesthesia occurred.

DISCUSSION

In this case, the patient had complex findings suggesting that sevoflurane inhalational induction combined with spinal anesthesia without tracheal intubation would be the best management option. Respiratory management was difficult, which was consistent with some recently reported case series of patients with MPS.^{4,5} Older age is associated with an increased risk of difficult intubation.⁶ Difficulty with intubation may prove fatal. When possible, procedures requiring general anesthesia should be avoided to minimize risk. Therefore, we depended on the regional block to be the mainstay of our anesthesia in the present case.

α -L-Iduronidase deficiency results in the progressive accumulation of glycosaminoglycan within lysosomes with subsequent multiorgan dysfunction and damage, including restricted joint movement, skeletal deformity,

abnormal cardiac valves, or frequent respiratory infections. In patients with severe MPS I, spinal deformities and neurological problems may be evident. Epidural anesthesia in a patient with MPS I is difficult. Many children with MPS I have intellectual disability. Odontoid hypoplasia may place them at risk for atlantoaxial subluxation. Additionally, masks may not fit properly because of their abnormal facies, and intubation for general anesthesia may be difficult. Tracheal extubation is also difficult. Most of these children require multiple surgical procedures during their life. Organ dysfunction can increase the risk of perioperative complications. Therefore, regional anesthesia, such as spinal anesthesia, can be used in such patients, especially those with intellectual disability.

Vas and Naregal⁷ described a 9-year-old boy with Hurler disease (the most severe variant of MPS I). They speculated that failure of epidural anesthesia in such patients might be because of deposition of mucopolysaccharides in either the general epidural space or in the sheath of the nerve fibers, preventing direct access of the local anesthetic to the nerve.⁷ Their supposition was confirmed by postmortem studies of patients with MPS. No such problems are encountered during spinal anesthesia because the drugs are deposited in the subarachnoid space. However, some patients may not remain calm and cooperate during induction of spinal anesthesia.

In this report, anesthesia was induced by inhalation of sevoflurane before spinal anesthesia. Compared with intravenous anesthetics, sevoflurane has an effect on analgesia and sedation and partial skeletal muscular relaxation. The patient in the present case had pneumonia. Sevoflurane does not usually stimulate the respiratory tract and increase respiratory secretions. Additionally, it is readily accepted because it is not pungent and its induction period is short. Sevoflurane is appropriate for a short operation with application by mask to avoid difficult tracheal intubation and extubation. We chose sevoflurane for these reasons in the present case. Sevoflurane can also be exhaled quickly after delivering the spinal anesthesia. The intellectual disability and potential behavioral problems in this patient did not worsen after this anesthetic protocol. In addition, preoperative evaluation of the vertebral column is the foundation of spinal anesthesia. Spinal anesthesia requires positioning the patient in spinal flexion. Regional anesthesia reduces the complications of general anesthesia while also saving hospitalization expenses and hospitalization time.

In conclusion, spinal anesthesia avoided potentially serious airway problems in a patient with MPS I. Compared with the uncertainty of epidural anesthesia, spinal anesthesia might provide safe and effective anesthesia.

CONFLICT OF INTEREST

The authors have no conflict of interest relevant to this article.

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