

Anesthetic Management of Leigh Syndrome

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ABSTRACT

Leigh syndrome is an extremely rare disorder in infants and children. It is characterized by a progressive neurodegenerative course with subacute necrotizing encephalomyelopathy, and it presents with developmental delay, seizures, dysarthria, ataxia, and myopathy. We present a patient with Leigh syndrome who required general anesthesia for percutaneous endoscopic gastrostomy. The respiratory, cardiac, and renal system functions should be very carefully evaluated in the perioperative period. The use of long-acting opioids and neuromuscular agents increases postoperative respiratory complications related to neuromuscular myopathy. Considering that lactic acidosis can develop in the perioperative period, it is recommended to give pre-perioperative fluids with dextrose.

Keywords: Child, anesthesia, Leigh syndrome

Introduction

Leigh syndrome is a rare infant and childhood disease. In this disease, there are mitochondrial and nuclear genomic mutations. The disease progresses with neurodegeneration and subacute necrotizing encephalomyelopathy and clinically manifests as growth retardation, convulsion, dysarthria, ataxia, and myopathy. Patients with this disease may require general anesthesia induction for endoscopic interventions, dental problems, and the like (1, 2).

In this case, anesthesia induction in a child with Leigh syndrome in whom surgery was planned due to percutaneous endoscopic gastrostomy is reported.

Case Report

Motor and growth retardation was found in a girl who was born mature with a weight of 3600 g, in whom neuromotor development was normal until 1 year of age, and who faced difficulties in eating and walking later. Changes secondary to pyruvate deficiency were observed on performing radiological examinations: mitochondrial complex IV deficiency was found in the laboratory and high levels of lactate and pyruvate were found in the CSF and urine. Surgery was planned due to percutaneous endoscopic gastrostomy for the patient who was 6 years and who weighed 10 kg and who was followed up with a diagnosis of Leigh syndrome.

In the physical examination, it was observed that the patient was conscious but could only provide limited cooperation; she had abdominal type of respiration and hyperventilation, and her heart sounds were normal. The patient was taken to an operating room without sedation before the surgery. Following standard anesthesia monitoring, anesthesia induction was performed with 0.1 mg/kg of midazolam and 2.5 mg/kg of ketamine; it was then observed that her heart rate decreased from 140-160 beats/min to 65 beats/min. Her heart rate increased after atropine administration. The patient was intubated without using neuromuscular blockers, and anesthesia maintenance was provided with 0.1-0.5 mcg/kg/min of remifentanyl and 2% sevoflurane. The mean arterial pressure was 35-40 mmHg and heart rate was 135-145 beats/

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min when 2% dextrose-isolate mixture was infused during the surgery. The surgery was completed in 45 min without any complications. The patient was extubated after providing adequate spontaneous respiration and consciousness and was sent to a ward after being observed in the post-anesthesia care unit for 2 h.

Discussion

Leigh syndrome is a neurodegenerative disease in which multiple organ exposure is seen along with abnormalities in mitochondrial energy production during infancy and childhood. Inheritance has been reported as X chromosome-based autosomal recessive or autosomal dominant. The most important feature is that the lactate level in the CSF, which is found in the laboratory examination, is higher than the level in blood. Clinical features depend on the defects of the pyruvate dehydrogenase complex, pyruvate carboxylase, cytochrome C oxidase (complex IV), and NADH-CoQ reductase (complex I). Complex IV deficiency is the most common biochemical enzyme deficiency; lack of these enzymes reportedly leads to an increase in lactate and pyruvate levels in serum, the CSF, and urine (1-3).

During radiological examinations, focal, bilateral, symmetric, spongiform lesions, particularly those localized to the brainstem and thalamus, can be detected (1). Clinical findings of Leigh syndrome can be observed in different clinical pictures according to the enzyme deficiency. Hypotonia, ataxia, nystagmus, choreoathetosis, and hyperventilation are the most important clinical findings. In addition to these, symptoms such as peripheral neuropathy and apneic seizures may occur, and the cause of death has often been reported as central respiratory failure (1, 2).

Neurological, respiratory, cardiac, and renal functions should be evaluated in detail before anesthesia induction. Increased airway reactivity during the preoperative period may lead to an increase in airway problems. Patients should be in the best possible condition in terms of respiratory function before anesthesia induction. Existing hypotonia and spasticity require a cautious approach to be followed during the use of neuromuscular agents and opioids. The use of long-acting opioid and neuromuscular blockers reportedly increases postoperative respiratory complications and may prolong the recovery

time. Due to these drawbacks of general anesthesia induction, Gozal et al performed percutaneous endoscopic gastrostomy in five children under deep sedation (1). Patients with Leigh syndrome who have respiratory insufficiency before anesthesia induction can also experience severe problems with respiratory failure after anesthesia induction and can even die (4). Considering that lactic acidosis may develop during the perioperative period and considering degenerative mitochondrial dysfunction, it has been suggested to administer dextrose liquids and avoid lactated Ringer's solutions during the pre- and perioperative periods (1, 3).

Conclusion

Patients with Leigh syndrome are at risk during anesthesia induction; different systems can be affected, and metabolic and respiratory problems can be seen. Multisystem evaluation should be performed before surgery, and possible problems related to post-surgery should be considered.

Informed Consent: Informed consent was obtained from patients who participated in this study.

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