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Cataract Surgery Anesthesia Management of a Patient with Biotinidase Deficiency

Biyotinidaz Eksikliği olan Hastanın Katarakt Cerrahisi Anestezi Yönetimi

Anesteziyoloji ve Reanimasyon

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Özet

Biyotinidaz enzim eksikliği, multisistemi içeren otozomal resesif nadir bir doğumsal hastalıktır. Hastalığın biotin ile tedavisi, hızlı bir biyokimyasal ve klinik iyileşme ile sonuçlanır. Bu enzim eksikliği, enfeksiyonlara karşı duyarlılık artışı, nörolojik, nöromüsküler, solunum, dermatolojik ve immünolojik problemleri içerir. Bu hastalarda anestezi yönetimi; öncelikle komorbidite ve konjenital anomalilere bağlı sorunları belirlemek ve peroperatif hastanın güvenliğini sağlamak için kapsamlı preoperatif stratejiler uygulamaktır. Bizde çalışmamızda, BD olan 10 aylık bir erkek çocuğun katarakt cerrahinde başarılı anestezi yönetimini sunuyoruz.

Anahtar kelimeler: *Biyotinidaz eksikliği*, *Katarakt cerrahisi*, *Anestezi*

Abstract

Biotinidase deficiency is an autosomal recessive rare congenital disease involving multisystem. Treatment of the disease with biotin results in rapid biochemical and clinical improvement. This enzyme deficiency includes increased susceptibility to infections, neurological, neuromuscular, respiratory, dermatological and immunological problems. Anesthesia management in these patients; The aim of this study is to determine the problems related to comorbidity and congenital anomalies and to apply comprehensive preoperative strategies to ensure the safety of the patient peroperatively. In this study, we present the successful anesthesia management of a 10-month-old boy with BD in cataract surgery.

Keywords: *Biotinidase Deficienc*, *Cataract Surgery*, *Anesthesia*

Introduction

We present the anesthetic management of a 10-month-old boy with biotinidase deficiency (BD) for cataract surgery. BD is a rare autosomal recessive metabolic disease with estimated incidence of approximately 1:60,000. BD most of the time presents at the age of 2-3 years [1]. Absence or deficiency of biotinidase impairs the cleavage of biotin from biocytin causing a deficiency of free biotin, slowing the functioning of biotin dependent carboxylases[2]. The clinical spectrum of BD depends on the severity of the defect. This impairment causes abnormalities in fatty acid synthesis, amino acid catabolism, and gluconeogenesis which may manifest as various clinical findings such as vomiting, lethargy and hypotonia, epilepsy, skin rash, conjunctivitis, alopecia, ataxia and increased susceptibility to infections. It is also associated with respiratory problems (apnea, dyspnea, tachypnea) and immune deficiency findings [3]. Anesthetic management in patients with BD presents various difficulties. New epileptic activity, acidosis, prolonged neuromuscular blockade (NMB) and increased sensitivity to malignant hyperthermia are anesthesia-specific problems. In addition, increased risk of aspiration due to gastroesophageal reflux, pharyngeal muscle weakness and airway complications due to oral secretions make anesthesia difficult in these patients. Therefore, anesthesia management is very important in this rare disease [3,4].

Case Report

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A 10-month-old boy weighing 6 kg was scheduled for a probing procedure due to cataract. BD was diagnosed ın the outer center and treated with biotin 5 mg/day. The patient had frequent respiratory tract infections. A severe respiratory tract infection occurred during the past two months. Physical examination revealed growth and mental retardation, dermatitis, alopecia, reduced skin turgor-tonus, dry mucous membrane, lethargy and hypotonia.. Neurological symptoms (mental retardation, hypotonia) were reviewed by paediatric neurology. In preoperative evaluation, laboratory findings were normal except anemia due to iron deficiency and AST: 167 U/l, ALT: 157 U/I. The patient was taken to the operating room without premedication. Following the routine monitoring of the patient, 8% sevoflurane (Sevorane®, Abbott, Ilionis, USA) and FiO2 1.0 oxygen was inhaled and induction was started. The vascular route was opened with 24 G branules. Patient's heart rate was 145/min, blood pressure was 115/65 mmHg and SpO2 was 90% before induction of anesthesia. Induction was facilitated with lidocaine 5 mg, propofol 2 mg/kg and remifentanyl 0.25 µg/kg ıv and then, size one classic laryngeal mask airway (LMA) was inserted. Following successful LMA insertion, anaesthesia was maintained with 1 minimum alveolar concentration (1 MAC) of sevoflurane in N2O+O2. Ventilation was controlled manually inorder to maintain end tidal carbondioxide concentration (EtCO2) between 35-45 mmHg. Hemodynamic stability was maintained during the surgery. A 10 mg kg-1 paracetamol (Parol 1 g flakon Atabay Kimya, İstanbul, Türkiye) suppository was applied rectally to provide post-surgical analgesia. The surgery lasted 1 hour and the patient recovered from anesthesia uneventfully. When the hemodynamic and respiratory parameters were within normal limits, he was transferred to the service.

Case Discussion

There are several considerations in anesthetic management for BD patients. Clinical manifestations of BD include neurological (seizures, ataxia, hypotonia, developmental delay, hearing and vision loss), neuromuscular (muscle weakness, spinal cord diseases), dermatological (seborrheic dermatitis, alopecia, skin rash), and metabolic abnormalities (chronic lactic acidosis, organic aciduria) [2]. Therefore, pre-anesthesia assessment and all preparations before anesthesia induction should be well done.

Premedication with metoclopramide and histamine-2 receptor antagonists or proton pump inhibitors per intravenously at 30 minutes before surgical procedure for the risk of gastric aspiration is recommended. Benzodiazepines can be used for sedation and preventive effect of the new epileptogenic activity. However, preoperative deep sedation should be avoided[2]. We did not apply preoperative sedation in our patient. These children tend to have upper respiratory tract infection of viral agents, especially because of immunodeficiency. Our patient was evaluated by a pediatric specialist in the preoperative period. In the minor surgical procedures, LMA anesthesia is recommended by using low-dose propofol and opioids such as alfentanil, remifentanil and fentanil without NMB agents. In major surgical procedures, total intravenous anesthesia with propofol and remifentanil must be preferred as it is a safe technique for children with hypotonia or seizures. High doses of opioids are not recommended in hypotonic children. At induction of anaesthesia, as thiopental has a property of inhibiting epileptic activity, it must be used especially in patients with seizures. Etomidate should be avoided because of its myoclonic movements. The use of ketamine is controversial in patients with seizures [5].

Because hypotonia is a common pathology in these children, NMB agents should be avoided, if possible. If needed, rocuronium must be preferred initially because this agent may be reversed by sugammadex. Because of risk of hyperkalaemia, malignant hyperthermia and epileptiform activity, succinylcholine should be avoided[6] . In our case as the patient had muscle weakness, we used low dose propofol and remifentanyl as a safe approach and inserted LMA without NMB agents because of hypotonia. In patients with BD, anesthetic management should be planned carefully to avoid epileptic activity, hypothermia, acidosis, hemodynamic instability, respiratory problems and malignant hyperthermia.

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Information Presantation

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