Management of anesthesia in biotinidase deficiency

Sir,

We present the anesthetic management in a 2-year-old boy with biotinidase deficiency (BD). BD is an autosomal recessive inherited metabolic disorder, which was first described in 1985 by Wolf *et al*.[1] The incidence of BD is approximately 1:60.000 newborns in the world.[1] 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). It is also associated with respiratory problems (apnea, dyspnea, tachypnea) and immune deficiency findings.[1-4] Treatment with biotin results in a rapid clinical and biochemical improvement. However, if untreated, the disease can lead to coma and death. A 2-year-old boy weighing 12 kg was scheduled for a probing procedure due to lacrimal duct obstruction. BD was diagnosed and treated with biotin when the patient was 4-month-old. Patient has no episode of seizure over the last year and has used biotin 20 mg/day without antiepileptic drug. Physical examination was normal except the crusted lesions on skin and scalp of recent chicken pox infections. Laboratory findings were normal except anemia due to iron deficiency (hemoglobin: 10.2 g/dL, serum iron: 26.8 μg/dL, ferritin: 10.9 ng/mL, serum iron binding capacity: 406 μg/dL). Intravenous (IV) access had been established before instituting anesthesia in the preoperative holding area. Patient was pre-medicated with IV midazolam 0.05 mg/kg. Monitoring in the form of electrocardiography, non-invasive blood pressure and pulse oximetery were instituted. Patient's heart rate was 150/min, blood pressure was 125/75 mmHg and SpO_2 was 99% before induction of anesthesia. Induction was facilitated with propofol 2 mg/kg and fentanyl 1 μg/kg IV and then, size two classic laryngeal mask airway (LMA) was inserted. Pediatric semi-closed circle breathing system was used. While preserving the patient's spontaneous ventilation, anesthesia was maintained with 1 MAC (minimum alveolar concentration) of sevoflurane in 60/40 NO_2-O_2. End-tidal carbon dioxide was maintained between 30 and 40 mmHg. Hemodynamic stability was maintained during the surgery. At the end of the surgery, which lasted 25 min, the patient was awake from anesthesia uneventfully. After then, the patient was taken to the post-operative care unit. While the patient had normal vital signs, we transported him to the ward.

In our patient who had not diagnosed any neuromuscular pathology, we used a safe approach of low dose propofol and fentanyl and inserted LMA without neuromuscular blocking agents because hypotonia is a common pathology in these children with BD. Thus, anesthetic management should be planned carefully to avoid hypoxia, acidosis and hemodynamic instability in all such cases.

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