LETTER

Careful pre-anesthetic evaluation leading to a successful anesthetic management of an undiagnosed Duchenne muscular dystrophy child

Dear Editor,

We report the anesthesiologic management of a 4.5 year-old-child with an undiagnosed Duchenne Muscular Dystrophy (DMD), who underwent an emergency operation after having swallowed a coin. The boy attended the emergency department with dysphagia and saliva drooling after having swallowed a coin, which was impacted in the cervical part of the esophagus. His parents reported complete vaccination history and bronchial asthma since infancy under treatment with inhalers. Up to two days before his admission, he had been treated for bronchitis with oral antibiotics. Although there was no family history of muscle disorder, his mother reported that he frequently complained of tight calves. On palpation both his calves were very tight while all other clinical examination was normal. Based on the history and the clinical examination, additional blood tests were performed: aspartate transaminase: 601 u/l, alanine transaminase: 335 u/l, lactate dehydrogenase: 2,336 u/l, creatine phosphokinase (CPK): 4,3461 u/l, alkaline phosphatase: 131 u/l. Electrolytes, blood sugar, urea, creatinine, serum Ca and PO4 and the hematological profile were normal. These biochemical results, in combination with the history and the clinical examination, raised the suspicion that the boy could be suffering from an undiagnosed form of muscular dystrophy. Before induction to anesthesia he received intravenous (IV) metoclopramide 0.15 mg/kg, dexamethasone 0.1 mg/kg, and ondansetron 0.05 mg/kg. Induction was obtained by IV propofol 3 mg/kg, and rocuronium 0.6 mg/kg. Rapid sequence intubation was applied. Anesthesia was maintained by propofol infusion 200 μg/kg/min, and bolus fentanyl 2 μg/kg. Intra-operatively blood pressure, cardiac rate, body temperature, O2 saturation and CO2 remained normal. Propofol was discontinued when TOF =90%. Reversion of muscle relaxation was obtained with IV sugammadex 2 mg/kg when TOF >90%. The child was extubated 50 minutes later and recovered promptly. After discharge, he was referred to the Medical Genetics Department, where the diagnosis was confirmed as DMD due to duplication of exon2 of the dystrophin gene.

In patients with DMD, certain problems can render general anesthesia extremely intricate, such as difficult intubation due to macroglossia, malignant hyperthermia after use of volatile anesthetic agents, and reduced laryngeal reflexes and prolonged time of gastric evacuation increasing the chances of aspiration. In addition, in DMD patients, no volatile gasses and no succinylcholine can be used for fear of rhabdomyolysis and hyperkalaemia[1]. Sugammadex, a drug known for its ability to reverse a rocuronium-induced neuromuscular block, was preferred in this boy since the use of succinylcholine was contraindicated[2]. Although we did not have a definitive diagnosis, we became suspicious based on the history of frequent cramps in the child's calf muscles, the tightness of his calf muscles on clinical examination and the extremely high value of CPK in biochemical tests. As the child was a chronic asthmatic, who had been treated with antibiotics for respiratory infection until two days before his admission, and also due to his dysphagia and saliva drooling from the foreign body impaction, we considered him as a good candidate for rapid sequence intubation for his anesthesia.

This case emphasizes the importance of a careful pre-anesthetic assessment that led to the clinical suspicion of DMD and finally to the successful anesthetic management of an undiagnosed child.

References

1. Segura LG, Lorenz JD, Weingarten TN, Scavonetto F, Bojanić K, Selcen D, et al. Anesthesia and Duchenne or Becker muscular dystrophy: review of 117 anesthetic exposures. Paediatr Anaesth. 2013; 23: 855-864.
2. de Boer HD, van Esmond J, Booij LH, Driessen JJ. Reversal of rocuronium-induced profound neuromuscular block by sugammadex in Duchenne muscular dystrophy. Paediatr Anaesth. 2009; 19: 1226-1228.

Conflict of interest

None.

Keywords: Duchenne muscular dystrophy, pre-anesthetic evaluation, foreign body, anesthesia

Vogiatzaki T, Chloropoulou P, Iatrou C

Department of Anaesthesiology, Democritus University of Thrace, University General Hospital of Alexandroupolis

Corresponding author: Dr Pelagia Chloropoulou MD, PhD, 19 Patriarchou Gregoriou str., Alexandroupolis, 68131, Greece, tel: +306947812132, fax: +302551031350, e-mail: peliachl@gmail.com