Anesthetic approach to Niemann–Pick Type C patient for dental treatment

Sir,

Niemann–Pick disease is an autosomal recessive lipid storage disease, characterized with differentiating levels of hepatosplenomegaly and progressive psychomotor retardation (this disease exhibiting heterogenic symptoms has four subtypes; Type A and B, in which sphingomyelinase values are normal, and Type C and D, in which sphingomyelinase values are close to normal values). Disease emerges with early childhood ataxia and progressive dementia, and the most evident features are early childhood hepatosplenomegaly, vertical supranuclear ophthalmoplegia, ataxia, dysarthria, mental-motor retardation, and seizures.[1,3] In this report, we present our anesthetic approach to an uncooperative 3-year-old female patient with hepatosplenomegaly, mental-motor retardation, developmental retardation, deglutition, speech deficiency, ataxia, and seizures who admitted to our clinic in collaboration with our pedodontics clinic for preventive dentistry practice.

Preventive measures such as topical fluoride application to teeth and incision of the excess mucosa over the left upper and lower primary second molars to obtain eruption was planned, and the patient was consulted to our department for the surgical procedure and the need of sufficient mouth opening under anesthesia for the uncooperative patient. Mallampati was Grade 3, sternomental and thyromental distances were short. After 6 h of fasting, sedoanalgesia with monitored anesthesia care was planned because of the presence of difficult intubation criteria. Patient was taken to the operation room without premedication, was monitorized and oxygenized 2 L/min. Five minutes after EMLA® application to the left-hand dorsal side, peripheral intravenous cannulation was obtained. Metoclopramide, to minimize vomiting possibility by increasing lower esophagus sphincter pressure, and ranitidine, to neutralize stomach acid was administered. Afterward, 0.5 mg midazolam and 20 mg ketamine were administered intravenously. After termination of the 20 min operation, patient was observed for 10 min and sent to service while hemodynamically stable.

Our choice of anesthetic in addition to local infiltration anesthesia (Ultracain®) was ketamine, which has analgesic, hypnotic, and amnestic effects but does not repress pharyngeal and laryngeal reflexes and does not evoke cardiovascular and respiratory depression.[3] The potential effect of ketamine of elevating convulsion threshold was prevented with midazolam. We are in the opinion that this method, which did not require additional anesthetic methods, was free of side effects and safe for the 3-year-old child with Niemann–Pick Type C disease.

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Conflicts of interest

There are no conflicts of interest.

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