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Anesthetic approach to a patient with Rett syndrome during tooth extraction

Ozkan Onal[1] and Aynur Kaplan[2]
[1]Anesthesiology and Reanimation Department, Yuksek Ihtisas Training and Educational Hospital, Turkey
[2]Anesthesiology and Reanimation Department, Ankara Tepebasi Dental Hospital, Turkey

Introduction: Rett Syndrome is a progressive developmental syndrome which occurs at the rate of 10000-23000 throughout the world and courses with autism, dementia, ataxia and loss of manual dexterity in girls. In this case report, a 26 year old female patient undergoing operation for tooth extraction is presented.

Case

26 year old female patient diagnosed with Rett syndrome was planned to undergo dental procedures under general anesthesia owing to many decayed teeth she has. She had mental and motor developmental retardation and was not able to walk. Atrophia was present in all extremities with limitation in manual skills, bruxism and shouting episodes. Laboratory tests were found to be normal. In Electrocardiogram, prolongation in QT was found. For anesthesia induction, propofol 3 mg kg-1, lidocain 1 mg kg-1, remifentanil 1 µg kg-1 were used at loading dose and tracheal intubation was performed with muscular relaxation by rocuronium 0.6 mg kg-1. In maintenance, propofol and remifentanil infusion was made. Overall 14 teeth underwent procedure without any complications. At the end of the procedure, propofol and remifentanil infusions were discontinued and patient extubated.

Discussion: In patients with Rett syndrome, irregular respiration, tachypnea and apnea attacks observed only when awake occur. Hypermetabolic states observed in patients with Rett syndrome indicate that anesthesia and anesthetic agents may increase the risk of malignant hyperthermia. Cardiac arrhythmias associated with prolonged QT interval occur frequently in patients with RS. In anesthesia, the use of thiopental and succinilcholin, should be avoided. As patients with Rett syndrome are extremely sensitive to sedation drugs, post operative recovery may be difficult. In addition, in patients with RS, due to the state of metabolic acidosis, the presence of low pain threshold, muscular degradation, and the tendency of potassium level to be triggered by anesthetic agents such as succinilcolin, the presence of abnormal EMG, loss of muscle, abnormal neural transmission and the risk of hyperthermia that can be triggered, prophylactic measures should be taken and operation should be carried out choosing the most suitable anesthesia method.

Coclusion: In patients with Rett syndrome, hemodynamic stabilization should be maintained by vigilant monitorisation in perioperative period due to the presence of malignant hyperthermia risk, apnea attacks that may occur during respiration and sleep, difficulty in intubation and extubation, prolongation of QT in ECG and abnormal T waves, which is important for safe anesthesia.

Biography

I. Ozkan Onal has completed his medical education from Ankara Gazi University Medical School and he has completed his anesthesia training from Ankara Hacettepe University Medical School and he is working in Ankara Yuksek Ihtisas training and educational hospital. He has published more than 15 papers about anesthesia.

drozkanonal@gmail.com