



Prolonged Compilation Due to Rett Syndrome: Case Report

Rett Sendromuna Bağlı Uzamış Komplikasyon: Olgu Sunumu

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Abstract

Rett Syndrome is a X-linked hereditary disease, accompanying neurological manifestations such as mental retardation, seizures, movement disorder, autistic behavior and abnormal respiratory pattern. It is an important clinical picture for anesthesiologists during perioperative period due to scoliosis and related respiratory problems, intubation difficulty due to micrognathia and abnormal respiratory pattern. In this case report we would like to present prolonged compilation due to Rett Syndrome and anesthetic approach in these cases.

Key words: RETT syndrome, extended compilation, general anesthesia.

Özet

Rett Sendromu, zeka geriliği, nöbetler, hareket bozukluğu, otistik davranış ve anormal solunum yolu örneği gibi nörolojik belirtilere eşlik eden X bağlantılı bir kalıtsal hastalıktır. Anesteziyologlar için perioperatif dönemde skolyoz mikrognati ve buna bağlı solunum problemleri nedeniyle entübasyon zorluğu ve anormal solunum yolu paternine bağlı önemli bir klinik tablo. Bu olgu sunumunda, Rett Sendromu ve anestezi yaklaşımından dolayı bu vakalarda uzun süreli derlemeyi sunmak istiyoruz.

Anahtar kelimeler: RETT sendromu, genişletilmiş derleme, genel anestezi.

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Introduction

The syndrome identified by Andreas Rett is a developmental disorder that occurs mostly in female children, with a mutation in the MECP2 gene. Children with Rett syndrome develop normally or almost usual up to 6-18 months of age. After this, the child enters the process of temporary stagnation or decline¹. The incidence was 1:10000-23000².

We would like to present a 23 year old patient with Rett syndrome who underwent prolonged complication after dental treatment under general anesthesia in the light of current literature.

Case

A 23-year-old patient with Rett syndrome weighing 30 kilograms was admitted for dental treatment under general anesthesia. The patient had a posterior constellation story under general anesthesia with diagnosis of scoliosis ten years ago. Neurological examination of the patient revealed stereotypic hand movements, autistic findings, and mental retardation. The patient preoperatively assessed was conscious, but not cooperatively and orally. The patient with motor developmental retardation was bed dependent. The respiratory examination revealed minimal rales. Atrophy was detected on the limbs. No convulsion history was reported. There was limitation of hand skills, and gingival creaking were observed. Cardiovascular examination was usual without any murmur. The patient's laboratory tests were normal. The electrocardiography (ECG) had QT extension. The patient was also consulted by Cardiology, Chest Diseases and Neurology and underwent elective surgery. The patient was taken to the operating room and the patient was operated on 20 gouge intra-vascular procedures on the left hand and 1/3 isodextrin infusion was started. ECG, pulse oxymetry, non-invasive artery monitorization and axillary heat monitoring were performed. Initial vital symptoms were as follows: Heart rate (HR): 80 beats / min, non-invasive blood pressure (NBCB): 120/75 mmHg, peripheral oxygen saturation (SpO2): 99%, axillary temperature: 36.5 °C. In anesthesia induction, propofol was used at a loading dose of 2 mg / kg, fentanyl 1 µg / kg, lidocaine 1 mg / kg, and tracheal intubation was performed with rocuronium 0.6 mg / kg provided muscle relaxation. 2% sevoflurane and 50% oxygen - 50% nitrous oxide were administered for anesthe-

sia. After intubation, the patient's heart rate was 90 beats / min, NBCB was 110/70 mmHg, SpO2 was 100%, and temperature was 36.6°C.

Dental examination and dental filling were performed by the patient's dentist. The process time was fifty minutes. Sevoflurane and nitrous oxide were closed after the procedure and the patient was inhaled for five minutes with 100% oxygen. A total of 4 mg/kg sugammadex was administered to the patients without any clinical collection. We could not measure Train Of Four Monitoring (TOF). The axillary temperature of the patient was 36.5°C and she also had pupillary myosis. The patient did not show clinical progress after Sugammadex and was treated with 50% oxygen-50% air. Patient started breathing and coughing at the end of the 45-minute collection period after Sugammadex administration. Sufficient breathing was obtained and the patient was awaked. The patient was admitted to postoperative intensive care unit with abnormal breathing pattern and stereotaxic movements. After one day of intensive care observation, the patient was dismissed.

Discussion

Rett syndrome is a genetic, neurological disorder that takes place mostly in female children, leading to death during the intrauterine period. Clinical features include; disorders of autistic behavior, mental retardation, respiratory disorders, loss of speech ability and handcrafts, unable to use for hands, abnormal respiratory pattern, motor dysfunctions, gastrointestinal motility disorders, scoliosis, autonomic dysfunction, somatic developmental disorders³. Therefore, preoperative evaluation of the respiratory pattern, arterial blood gas analysis, presence of autonomic dysfunction, reflux narrative, ECG, and seizure history should be carefully evaluated. The necessary laboratory analysis should be performed as these patients may have blood lactic acid increase⁴. Our patient had apnea and irregular respiratory pattern. We did not observe lactate increase, desaturation, or acid-base disorders in preoperative and intraoperative blood gas analyzes.

Careful ECG monitoring is required in the perioperative period as cardiac arrhythmias associated with extended QT interval are often seen in these patients⁵⁻⁶.

In our patient, there was no arrhythmia during perioperative follow-up with QT prolongation on preoperative evaluation. We also took care to avoid medications that could trigger cardiac arrhythmia and QT abnormalities during induction.

Patients with Rett Syndrome can show abnormal respiratory pattern, tachypnea and also apne seizures only during the awake period⁷. This is due to the impaired of behavioral control of the frontal lobe function⁸. Patients with rapidly progressive scoliosis can lead to serious respiratory insufficiency. Besides loss of muscle and impaired muscle tonus can cause difficulty in surgery positioning¹.

Administration of depolarizing muscle relaxant such as succinylcholine in patients with Rett Syndrome elevates serum potassium level. Because of this we used nondepolarizing agent as rocuronium.

These patients are very sensitive to anesthetic drugs. There are cases of prolonged sedation in the literature⁹. For this reason we administered optimal induction doses and avoided additional doses; but our patient had a prolonged compilation. After a total of 4 mg / kg sugammadex application at fifty-five minutes after induction, the patient started breathing and coughing at the end of the 45-minute compilation period.

Rett syndrome has many problems to be considered in anesthetic management. This is a syndrome that should be known in detailed manner by anesthesiologists in order to avoid perioperative complications.

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