Case Report

Dental procedure under opioid-free balanced anaesthesia in a child with Rett syndrome who convulsed on every attempt to feed: Case report

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ABSTRACT

Rett syndrome is a genetic neurodevelopmental disorder which occurs in females and includes autism, spasticity, hypotonia, microcephaly, scoliosis, stereotyping, abnormal respiratory control and seizures. They are at an increased predisposition for QT interval prolongation and cardiac arrhythmias. An 8-year-old severely intellectually impaired girl with Rett syndrome was referred to us for anaesthesia for multiple dental abscess drainage and rehabilitation. Her frequency of convulsions had increased and she convulsed every time an attempt was made to feed her. The pain of chewing exacerbated the convulsions. The cornerstone of our management was to provide adequate pain relief, anaesthesia without muscle relaxant and opioids, and judicious use of local anaesthetics. We discuss the anaesthetic management and its advantages and limitations in this case report.

Key words: Abscess drainage, loco-regional, opioid-free, Rett syndrome

INTRODUCTION

Rett syndrome is a rare genetically inherited neurodevelopmental disorder that is characterized by progressive deterioration of the central nervous system.[1,2] It primarily affects young females, becoming apparent at 6–24 months of age, and its most characteristic feature is the deceleration of head growth.[3] Other clinical features of neurodevelopmental arrest such as loss of acquired fine motor skills, stereotypical hand movements, severe expressive and receptive language and gait apraxia follow.[4] Rett syndrome is also associated with multi-organ complications such as behavioural abnormalities, mental retardation, seizures, cardiorespiratory dysfunction, gastrointestinal problems and musculoskeletal disorders.[5-9]

CASE

An 8-year-old girl weighing 28 kg was scheduled for elective multiple dental abscess drainage and rehabilitation. She was diagnosed with Rett syndrome when she was 9 months old and currently suffered from refractory epilepsy and global developmental delay. Because of communication impairment associated with her developmental disability, it was imperative to involve her mother during the entire perioperative course.

In her preoperative examination, she appeared drowsy and spasticity was noted in her limbs. Mallampati score evaluation was not possible.

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She only responded to her parents and exhibited discomfort from the pain she was experiencing from her intraoral abscess. She was maintained on several antiepileptic medications: topiramate 25 mg, clobazam 5 mg, levetiracetam 500 mg, lacosamide 50 mg and oxcarbazepine 300, each of the above given twice a day. Her epilepsy had worsened with increased frequency and was thought to be aggravated by the pain that her intraoral abscess was causing. She exhibited epilepsy during chewing due to pain it elicited. Her laboratory tests including coagulation function and electrolytes were normal.

After obtaining informed consent from her parents, the patient was kept nil by mouth for 6 hours after solid food before surgery and no premedication was administered. The antiepileptic medications were given at 6 am in the morning with sips of water and she was scheduled for the surgery at 8 am. We planned to perform an opioid- and muscle relaxant free-balanced general anaesthesia with nasotracheal intubation.

In the operating room, the standard American Society of Anesthesiologists monitors were connected to the patient. She was pre oxygenated and anaesthesia was induced with sevoflurane in air and oxygen via face mask. Intravenous access was secured after achieving inhalational induction, followed by administration of 700 mg of magnesium sulphate as an IV infusion over 30 min (1.5 mL of 50% $MgSO_4$ added in 15 mL of 5% dextrose). Gentle nasotracheal intubation was performed under propofol 85 mg and lidocaine 25 mg 1.5 mL 2% lignocaine diluted to 10 mL in normal saline) IV without the use of any muscle relaxant. The vocal cords were anaesthetised using 0.5 mL of 2% lidocaine-adrenaline spray during direct laryngoscopy. A 5 mm ID cuffed endotracheal tube was used followed by throat pack insertion. anaesthesia was then maintained on 50% oxygen in the air and 2% sevoflurane. This was supplemented by buccal infiltration of 5 mL of 2% lidocaine with 1:200,000 adrenaline by the surgeon. She was also administered 500 mg of acetaminophen IV for analgesia.

The procedure took approximately 120 min. Vital signs were stable throughout the procedure. The estimated blood loss was less than 10 mL. After the return of spontaneous ventilation, the throat pack was removed, the trachea was extubated and the patient was transferred to the paediatric intensive care unit (PICU) for postoperative care. For postoperative analgesia, the patient was administered 60 mg of diluted IV diclofenac twice a day and 500 mg of IV acetaminophen four times a day.

DISCUSSION

The anaesthetic concerns in our patient included refractory epilepsy, the potential for delayed recovery from anaesthesia, drug interactions between antiepileptic drugs and anaesthetic drugs and risk of triggering arrhythmia or QT prolongation. There are no reports about performing an opioid- and muscle relaxant-free balanced general anaesthesia in patients with Rett syndrome, which we adopted to address the aforementioned anaesthetic concerns.

We secured IV access under volatile anaesthesia to reduce the stress and pain that can potentially trigger her epilepsy. We administered magnesium sulphate for its anticonvulsant, analgesic and anti-arrhythmic properties. Opioid-free analgesia with the use of local anaesthetic infiltration and acetaminophen was our mode of analgesia and we avoided the use of muscle relaxants and reversal agents that could potentially induce arrhythmias or QT prolongation. Although opioids are not contraindicated in children with Rett syndrome, we avoided them because they are more prone to respiratory depression due to opioids.[9]

Kako *et al*.[2] described an anaesthetic approach using opioids and rocuronium for a case of a patient with Rett syndrome scheduled for spine surgery. They advocated the use of sugammadex to reverse rocuronium-induced neuromuscular blockade as opposed to neostigmine which risks precipitating cardiac arrhythmias. Unlike these authors, we planned an anaesthetic technique devoid of muscle relaxants and opioids thus minimising the possibility of arrhythmias and delayed recovery.

Önal and Kaplan[10] reported a case of an adult patient with Rett syndrome who presented for tooth extraction. They described the use of remifentanil and propofol infusions, as well as rocuronium for muscle relaxation. They further stated that the operation should be carried out choosing the most suitable anaesthesia method and do not advocate one specific technique of anaesthesia. We avoided opioids and muscle relaxants so as to avoid their adverse effects, and used locoregional anaesthesia. Judicious use of local anaesthesia along with multimodal analgesic technique helped achieve excellent pain relief and an uneventful procedure.

Pérez-Moreno *et al*.[11] described the case of a 26-year old woman with Rett syndrome who underwent humerus fracture fixation. They concluded that regional anaesthesia is the technique of choice in this group of patients and suggested avoiding drugs like benzodiazepines which have excessive sedative effects. The mainstay of analgesia in our case, though being a dental procedure, was locoregional along with the selected drugs for multimodal analgesia.

We managed this case successfully by combining the local infiltration and tailor-made general anaesthesia.

CONCLUSION

A simple concept of locoregional anaesthesia, with avoidance of opioid and muscle relaxant, is feasible for dental rehabilitation surgeries in cases of Rett syndrome. The mainstay is to maintain adequate depth of anaesthesia and sufficient analgesia to allow rapid recovery.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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