Research Paper

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PERIOPERATIVE ANESTHETIC MANAGEMENT OF A PATIENT OF RUBINSTEIN TAYBI SYNDROME FOR SQUINT SURGERY

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ABSTRACT

We report a successful anesthetic management of a case of Rubinstein Taybi (RT) Syndrome with seizure disorder posted for squint surgery. Anesthetic management becomes challenging as this is a multisystem disease involving airway, cardiac, musculoskeletal, respiratory and urogenital system

KEYWORD

Rubinstein Taybi syndrome, difficult intubation, squint surgery.

ARTICLE HISTORY

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INTRODUCTION

Rubinstein Taybi syndrome was first described by Rubeinstein and Taybi in 1963. The incidence is estimated to be approximately 1 in 3,00,000 in general population. It occurs due to mutation of chr 16p13.3 an EP300 gene coding for CREBB protein[1]. It is an autosomal dominant disorder chiefly characterized by short stature, moderate to severe mental retardation, broad thumb and first toe and distinctive craniofacial features[2]. It is also associated congenital heart diseases, feeding difficulties, gastroesophageal reflux and other congenital malformations.

Major concern in anesthetic management of squint surgery includes occurrence of oculocardiac reflex and more incidences of postoperative nausea and vomiting(PONV)[3]. Some literature draws attention to the risk of succinylcholine triggering malignant hyperthermia

CASE REPORT

We report a successful anesthetic management of a patient with RT syndrome with seizure disorder for squint surgery.
A 12 year old female patient weighing 35 kg, known case of RT syndrome was posted for squint surgery. She had history of mental retardation, delayed development and speech difficulty. She was suffering from seizure disorder since age of three and was on tab. Valproate 200 mg TDS. Last seizure episode was reported four months back.

On examination, she had slanting eyes, beaked nose, small mandible and high arched palate suggestive of difficult laryngoscopy and intubation. Airway examination showed modified Mallampatti grade II. There were no signs of congenital heart disease and respiratory illness.

Routine haematological and biochemical investigations were within normal limits. ECG was normal and EEG showed signs of multifocal seizures.

The patient was posted first on list. She received morning dose of valproate with sips of water. Preoperative sedation was avoided. Aspiration prophylaxis in the form of tablet Rantac 75 mg was given the night before surgery. Written informed consent was obtained from the parents.

In operation theatre, all equipments necessary to manage a difficult airway were arranged like different size and types of mask, alternative size of blades, bougie, intubating LMA, PLMA. After shifting the patient to operation theatre, standard monitors like pulse oximeter, non invasive blood pressure, ECG and temperature probe were attached.

Patient was induced with O_2 and sevoflurane. After induction, IV line was secured. 70 mcg fentanyl was administered. After confirmation of mask ventilation, rocuronium 20 mg was given. Direct laryngoscopy was performed with Macintosh blade no. 2 which revealed CL grade II. A 6.5 mm ID cuffed endotracheal tube was placed in first attempt. Anesthesia was maintained with isoflurane in O_2 and N_2O and rocuronium 5 mg as and when required. Heart rate varied from 70/ min to 90/ min with a normal sinus rhythm. One episode of bradycardia was noted during pulling of medial rectus muscle owing to oculocardiac reflex. Bradycardia was relieved on removing the surgical stimulus.

Intraoperative fluid included ringer lactate 500 ml. Ondansetron 3 mg was administered at the end of the surgery to avoid PONV. Paracetamol 500 mg was administered to take care of postoperative pain relief. Surgery lasted for about 90 minutes. At the end of surgery, residual neuromuscular block was reversed with neostigmine(1.7 mg) and glycopyrrolate(0.35 mg) and trachea was extubated. Patient was shifted to recovery room for observation. The patient was pain free and was shifted to the ward after 2 hours. Postoperative course was uneventful. She was discharged on 3rd postoperative day.

DISCUSSION

Rubinstein Taybi syndrome was first reported in 1963[1]. Multisystem involvement like airway, cardiac, musculoskeletal, respiratory and urogenital system poses challenge to anaesthesiologist for managing such cases[4].

Premedication in the form of sedatives should better be avoided considering the risk of respiratory depression and upper airway problems [5]. So, we did not give any sedative agent preroperatively.

The airway management is challenging in such patients in

view of craniofacial abnormalities, laryngotracheal abnormalities, obstructive sleep apnoea, gastroesophageal reflux disease, increased risk of aspiration and mental retardation (noncooperation). The abnormalities of head and face include microcephaly, antimongoloid faces, high arched palate, widely spaced eyes, broad nasal bridge and an unusually small, hypoplastic lower jaw with small mouth opening and bucked upper incisors. They have choanal atrsia, deviated nasal septum, upper and lower airway narrowing, postcricoid web and layngomalacia[5,6]. These all features poses intubation as well as ventilation difficulties. In our patient, the airway difficulty was anticipated. All possible measures to manage difficult airway were kept ready.

Other major airway complication is gastric content aspiration due to increased incidence of gastro-esophageal reflux[7]. This risk increases mainly during induction and emergence. Recurrent aspiration often leads to acute and chronic pneumonia. Though the use of supraglottic airway device may be beneficial to avoid stimuli in view of ocular surgery, but it may be limited due to possibility of airway collapse and inaccessibility to airway under surgical drapes[4]. So we secured the airway with an endotracheal tube.

Cardiac abnormalities like patent ductus arteriosus, atrial septal defect, aortic coarctation, and bicuspid aortic valve are present in 33% of these patients[4]. Our patient was not diagnosed with any of the cardiac abnormalities. These cardiac abnormalities also predispose to arrhythmias with arrythmogenic drugs like succinylcholine, atropine and neostigmine. Succinylcholine should be avoided in these patients because it is reported to be responsible for supraventricular tachycardia and early atrial and ventricular contractions[8].

Occurrence of OCR is common in squint surgery. Pressure on eyeball or traction of extraocular muscles can produce various cardiac manifestations which includes sinus bradycardia, ventricular ectopic beats and even asystole by stimulation of vagus nerve fibres in sinus node[9,10]. In our case, one episode of bradycardia was noted during pulling of medial rectus muscle which terminated by itself on removing the stimulus by surgeon.

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