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Case Report

Anesthetic Management of Prader Willi Syndrome pediatric patient undergoing Tonsillectomy Surgery: A Case Report

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ABSTRACT

Anesthetic Management of Prader Willi Syndrome pediatric patient undergoing Tonsillectomy Surgery: A Case Report.

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Prader-Willi syndrome (PWS) is a rare genetic disorder characterized by various abnormalities, such as hypotonia, high body mass index (BMI), craniofacial anomalies, and obstructive sleep apnea. Anesthetic management of these patients requires special perioperative planning and care due to multisystem involvement. In this case report we present the anesthetic considerations and perioperative management of a 9-year-old male patient with PWS, who underwent adenoidectomy-tonsillectomy surgery.

Keywords: Prader-Willi syndrome, anesthesia, tonsillectomy

INTRODUCTION

Prader-Willi syndrome (PWS) is an uncommon genetic disorder that affects both males and females[1]. This syndrome was first described by Prader et al in 1956 and is caused by abnormalities on chromosome 15 (region 15q11-q13)[2].The most characteristic manifestations of PWS are hypothalamic dysfunction, hypotonia, sucking and swallowing problems in infancy, hyperphagia in early childhood and also craniofacial anomalies, developmental delay, obstructive sleep apnea (OSA), mental retardation and behavioral problems or severe psychiatric problems[3]. Due to abnormalities of skeletal development and

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end-organ involvement, surgical procedures are often required in PWS patients. Scoliosis surgery, orchiopexy, adenotonsille-ctomy to treat OSA, ophthalmologic surgery in order to correct strabismus are surgical treatments that often undergo these patients[4].

Anesthesiologists may face problems like difficult intravenous (IV) access, cardio-pulmonary complications, thermoregulator management issues, or difficult airway management[5]. Here, we presented the anesthetic management of a of a pediatric male patient with PWS, who underwent adenoidectomy-tonsillectomy surgery in order to treat OSA.

CASE REPORT

We present a case of a 9 years old male patient, with PWS syndrome scheduled for adenoidectomy - tonsillectomy surgery, for OSA treatment. His weight was 30 kg and his height 124cm. Patient had been diagnosed with PWS syndrome 4 years prior.

His medical record showed that he had OSA (apnea-hypopnea index=10) and for the last two years the patient used bilevel positive airway pressure (BPAP), for treating OSA**.** His growth was slower than the normal pattern and he was receiving growth hormones. No other medication was administrated.

His preoperative physical examination was unremarkable concerning respiratory or cardiac symptoms. Cardiac and pulmonary evaluations were performed preoperatively with no signifycant findings. Physical exanination, also revealed a large tongue relative to mouth, malocclusion and a large mandibula. The Mallampati score was obtained and evaluated as score III. Preoperative laboratory tests were within normal values. He was cleared for surgery, assigned as ASA III physical status patient.

On arrival to the operating theater, standard monitoring was applied to the patient (heart rate, respiratory rate, non-invasive blood pressure, temperature, SpO_2) and a heated blanked was also applied under the patient to prevent hypothermia.

Anesthesia was induced initially with sevoflurane inhalation and an intravenous (i.v) line was inserted. Atropine 0.01 mg/kg, fentanyl 2μg/kg, lidocaine 1 mg/k propofol 1mg/kg titrated to loss of consciousness and loss of eyelash reflex. Endotracheal intubation was facilitated with rocuronium bromide 0.6 mg /Kg. A 4 mm endotracheal tube was inserted uneventfully with the use of video laryngoscopy due to suspected difficult airway. After intubation, the lungs were ventilated mechanically with FiO_2 0.4 and for anesthesia maintenance sevoflurane and remifentanil 0.05-0.1 mcg/kg/min IV infusion were used. Ventilation was adjusted to keep $P_{ET}CO_2$in the range 4.5-5.5 pKa. Intraoperatively patient's hemodynamic parameters were stable.

After ten minutes of surgical time, a drop of SpO_2 was noted (from 98% to 92%). Widespread wheezing was revealed on chest auscultation. Misplacement and occlusion of tracheal

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tube and breathing circuit were ruled out. With elevated airway pressure and up-slo-ping capnogram, bronchospasm was diagnosed. Instantaneously, FiO_2 and sevoflurane was increased, propofol 2 mg/ kg was added, salbu-tamol puffs sprayed down tracheal tube and methylprednisolone 1mg/ kg was given. Manual ventilation and recruitment maneuvers performed intraoperatively. Five minutes later, SpO_2 gradually improved with the appearance of normal capnogram. After that incidence, surgical procedure proceeded uneventfully. Duration of surgery was 80 minutes, without the appearance of any hemodynamic or other complications.

Neuromuscular blockade reversal was achieved with the use of TOF (Train of four) monitoring and with the administration of sugammadex 2 mg/kg. Patient was extubated in the operating room. He was alert, awake and able to follow commands. Postoperatively patient was transferred to the post-anesthesia care unit (PACU) with normal vital signs. Later, he was discharged from PACU (Aldrete score 9) to the ward, without any coplications.

DISCUSSION

Prader-Willi's syndrome (PWS) incidence is is about 1 in 15.000-25.000 births, and the male-to-female ratio is 3:2[6]. It has been described as having two nutritional phases: inability to gain weight due to sucking and swallowing problems, in infancy (phase 1), hyperphagia leading to obesity, in early childhood (phase 2)[3]. Major features are muscular hypotonia, skeletal deformities, mental alterations and obesity[3]. Preventing obesity is directly related to life expectancy. Patients with PWS have also craniofacial abnormalities like narrow bifrontal diameter, upslanting palpebral fissures, thin upper lip, and triangular mouth[7,8]. Endocrine disorders like hypogonadism and diabetes mellitus accompany obesity. Due to growth hormone (GH) deficiency, short stature and decreased bone mineral density may occur[9] Anesthetic management of these patients requires special perioperative planning and care due to multisystem involvement. Major concerns perioperatively include obesity and it's related complications: cardiovascular disorders, obstructive sleep apnea, or diabetes mellitus[10], and also muscular hypotonia, decreased motility of the gastrointestinal tract, lowered oesophageal sphincter tone, that may facilitate perioperative gastric aspiration in PWS patients.[7] Nasogastric aspiration and rapid sequence induction during endotracheal intubation can prevent gastric aspiration[11]. Hypothalamic dysfunction can cause temperature disregulation, so monitoring temperature is necessary to maintain normothermia. Perioperative positioning and accessing IV lines may be challenging due to obesity[12]. General or regional anesthesia could be applied in PMS patients. Both types of anesthesia are challenging. General anaesthesia may lead

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mainly to difficult airway management and regional anaesthesia's landmarks may be obscured due to morbid obesity. The appropriate equipment for difficult airway management should always be available in PWS patients. .

In our case, despite the fact that patient was not obese, broncospasm was diagnosed intraoperatively. We performed manual ventilation and recruitment maneuvers despite the possibility of atelectasis development[14]. Protective mechanical ventilation strategies were followed in order to protect the lungs from barotrauma[14]. Nafiu et al. showed that obese children were more likely to have complex ventilation problems and prolonged PACU stays, due to a high risk of postoperative airway obstruction [13]. İt must be noted that PWS patients are prone to pulmonary complications and restrictive lung diseases.

CONCLUSION

PWS is a genetic disorder that poses a significant challenge to anesthesiologists. Detailed physical examination and precision preparation are necessary in order to reduce morbidity and mortality. Early diagnosis of GH deficiency and its treatment has improved the quality of life of affected children. A multidisciplinary approach and comprehensive perioperative management may prevent complications.

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