OLGU SUNUMU

Anesthetic Management of a Patient with Beckwith-Wiedemann Syndrome

Beckwith-Wiedemann Sendromlu Hastada Anestezi Yönetimi

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ABSTRACT
Beckwith-Widemann Syndrome (BWS) occurs in the neonatal period at a frequency of approximately one in 13 700 live births. This syndrome consists of abnormalities, such as; macroglossia, omphalocele, visceromegaly, gigantism and neonatal hypoglycemia. It is expected that airway management would be complicated by macroglossia which might cause difficult ventilation and endotracheal intubation. Therefore preparations for airway difficulty should be considered before anesthesia induction.

Another problem in BWS is postoperative analgesia. Because opioids can depress breathing and non-steroid antiinflammatory drugs can increase bleeding, postoperative analgesia should be planned carefully.

We present anesthetic management of an eight years old patient with BWS having big tongue undergoing tonsil-adenoid resection.

Keywords: Beckwith-Widemann syndrome; airway management; postoperative analgesia.

ÖZET

BWS’da diğer bir problemde postoperatif analjezidir. Çünkü opioid kullanımı solunumu deprese edebilirken, non-steroid antiinflammatuar ilaclarla kanamaya yol açabileceğiinden postoperatif analjezi yönetimi dikkatlice planlanmalıdır.

Biz büyük bir dile sahip, adenotonsillektomi ameliyatı yapılacak 8 yaşında Beckwith-Widemann sendromlu hastada anestezi yönetimini sunduk.

Anahtar Kelimeler: Beckwith-Widemann sendromu; havayolu yönetimi; postoperatif analjezi.

INTRODUCTION
Beckwith-Widemann Syndrome (BWS) was first described by Beckwith (1) in 1963 and by Wiedemann (2) in 1964. This syndrome is characterized by macroglossia, omphalocele, visceromegaly, gigantism and hypoglycemia. Although perioperatif anesthetic management may be complicated by anatomical airway abnormalities and hypoglycemia, there have been few reports concerning these problems.

Adenotonsillectomy is a common surgical procedure in children. Postoperative analgesic choice is very important for patient with BWS. Lots of agents such as non-steroid antiinflammatory drugs (NSAIDs), opioids are used for postoperative pain. NSAIDs have lower risk of postoperative nausea and vomiting but increase bleeding due to their antiplatelet effects (3,4). Opioids provide satisfactory analgesia but can
depress breathing and delay discharge from hospital for its prolonged sedation (5). Paracetamol is non-opioid analgesic that is devoid of these risks and also has little antiplatelet activity and does not effect bleeding time like NSAIDs (6). An intravenous (iv) formulation of paracetamol recently has become available, and it achieves target plasma concentration more rapidly with reduced variability compared with the rectal and oral formulations (7).

We present and discuss anesthetic management of eight years old patient with BWS undergoing tonsil and adenoid resection.

CASE REPORT

8 years old, 25kg, 128cm patient was admitted to otorhinolaringology clinic with a complaint of snoring and apne periods during sleeping. Grade 4 tonsil hypertrophy was diagnosed and operation was planned. Then he was admitted to our anesthesia policlinic for pre-examination there we learned that he had been monitored as BWS for five years. In physical examination; macroglossia, earlobe fissures, hepatomegaly, asymmetry in facial and extremity were seen. Also he is dumb and has mental and motor regression. Other findings of physical examination were unremarkable. Routine laboratory data were within normal ranges for ages, an electrocardiogram (ECG) and chest roentgenogram, echocardiography were normal. Hepatomegaly, renomegaly and splenomegaly were seen in his ultrasonography. In oropharyngeal examination his mallampati score was IV.

The patient was premedicated with 250 micrograms atropine intravenously and ten minutes later he was brought to the operating room. Monitors consisted of ECG, non invasiv blood pressure and pulseoximeter. Nasopharyngeal, oropharyngeal airways, laryngeal mask, ILMA-fastrach were also prepared for difficult airway. We administered 20mg lidocaine, 1mg midazolam and 25mcg Fentanyl intravenously and topical anesthetic for awake laryngoscopy. After an assessment was made that there would be no difficulty in intubation by laryngoscopy, anesthesia was induced with 50mg propofol. When mask ventilation was seen easy 12,5 mg Rocuronium was administered intravenously. Laryngoscopy and orotracheal intubation were performed easier then expected. Anesthesia was maintained with nitrous oxide and oxygen and sevoflorane (1-2%) under volume controlled ventilation. The intraoperative course was uneventful. No remarkable hemodynamic changes were observed throughout the surgical procedures. During bleeding control, we started 15mg/kg paracetamol infusion for analgesia. After he awakened perfectly, we extubated him at the end of surgery. But we did not take away the oropharyngeal airway for probable airway obstruction. He was discharged from postoperative care unit, when he was hemodynamically stable and awake.

DISCUSSION

BWS presents with variable anomalies which include omphalocele, macroglossia, visceromegaly and severe cardiac defects (8,9). Macroglossia is the most common feature. It occurs in approximately 95% of patients (10). Upper airway obstruction and difficulties in endotracheal intubation due to macroglossia are critical problems for anesthetic management. Because of macroglossia and sleep apne patients with BWS often requires either awake vocal cord inspection or awake tracheal intubation. This can be facilitated using topical anesthetics. Kim et all (11), saw the glottis by laryngoscopy following diazepam administration and then they started their anesthesia with administering of thiopental sodium. We administered midazolam, fentanyl intravenously and topical anesthetic to the mouth for making laryngoscopy easier in awake position. Y. Kimura et all (12), recommended using cuffed tracheal tube in patients with BWS because of the appropriate tracheal size can not be predicted. We also used cuffed tracheal tube for our patient.

In the late 1970s a joint retrospective study found 13 cases of BWS. Twelve of these were found to have cardiovascular abnormalities (13). During anestesia pre-examination our patient was consulted by pediatric cardiology. Echocardiography was seen normal and cardiomegaly was not found.

Postoperative pain is very important for children. Alhashemia and Daghistani found earlier recovery for postoperative care unit discharge in pediatric patients undergoing dental restoration with iv paracetamol when compared intramuscular meperidine (14).
Uysal et al found that the postoperative analgesia and early readiness for discharge from post operative care unit provided by iv paracetamol 15mg/kg was similar to the outcome with iv tramadol 1mg/kg in children after adenotonsillectomy (15). We used 15mg/kg iv paracetamol for postoperative pain and comfortable awake extubation in our patient.

As a conclusion, in patients with BWS a comprehensive preoperative examination should include assessment of the airway, cardiovascular status, endocrin status and electrolytes. Preparations for difficult airway management should be made. We recommended iv paracetamol for postoperative analgesia and awake extubation in patients with BWS.

REFERENCES