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Case Report

The Use of Videolaryngoscope in a Difficult Intubation Patient with Beckwith-Wiedemann Syndrome

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Abstract: Beckwith-Wiedemann Syndrome is a congenital disease associated with macrosomia, macroglossia, abdomianl wall defects, hemihypertrophy, neonatal hypoglicemia, microcephaly and musculoskeletal system abnormalities. The aim of this case report is to present our anesthesia experience in a 10 year old patient with Beckwith-Wiedemann syndrome in whom could not be intubated with external laryngeal manipulation with conventional blades and lma could not be placed due to macroglossia, and was intubated at first attempt using videolaryngoscope.
Keywords: hemihypertrophy, neonatal hypoglicemia, microcephaly.

INTRODUCTION

Beckwith-Wiedemann syndrome is a congenital disease associated with macrosomia, macroglossia, abdominal wall defects, hemihypertrophy, and neonatal hypoglicemia. In addition, nevus flammeus, hepatomegaly, nephromegaly, exophtalmoembryonic tumors, psychomotor retardation, microcephalia, cardiac anomalies, musculo skeletal abnormalities and hearing loss may occur as well [1]. Difficult laryngoscopy occurs in 2-8% of all general anesthesia cases while difficult intubation is more seldom. If intubation is unsuccessful, it may be fatal albeit rarely. Awake fiberoptic intubation is the gold standard in difficult airways. However, it requires the cooperation of the patients and experienced practicioners. It is very difficult to use awake fiberoptic particularly in pediatric difficult intubation cases. Videolayngoscope is a portable airway tool which can be easily used and facilitates intubation by providing direct imaging of larynx. It is a good alternative in pediatric patients with difficult intubations since it is readily used and requires less experience than fiberoptic. The aim of this report is to present our anesthesia experience in pediatric patients with .Beckwith-Wiedemann Syndrome and difficulty of intubation.

CASE REPORT

A 1 year old boy at the wieght of 10 kg was diagnosed with undecended testis and operation was planned by pediatric surgery department. In preoperative pediatric examination, micrognathia, macroglossia, ridging in ear lobe, macrosomia and minimal atrial septal defect were detected. No pathology was found in routine biochemical and hematological investigations. Standard anesthesia monitorization was carried out vital finding prior to induction were as follows: SpO2 %93, heart rate 100 beat /min. blood pressure 90/47mmHg. Considering that airway obstruction caused by syndromic facial appearance and macroglossia may created difficulty in the protection of airway, face masks at different sizes, airways, laryngeal masks, blades at different sizes, and tracheal tubes and videolaryngoscope were kept ready. Following approximately 5 minutes of preoxygenation, anesthesia induction was made with 2 mg/kg propofol (Fresenius Kabi AB, Bad Homburg, Germany) without losing spontaneous respiration and the patient was ventilated with %50/50 air /oxygen. No problem was experienced in ventilation with face mask and intubation was attempted with external laryngeal manipulation via conventional blades. Following two unsuccessful attempts, lma was attempted to be placed. But, the attempt failed owing to macroglossia, then he was intubated at first attempt with videolaryngoscope (King Vision, King Systems, USA) guidance with size 4.0 mm ID cuffed tube. Anesthesia was maintained with propofol infusion (4mg/kg/h), %50 nitrous oxide and %50 oxygen. Plasma glucose level was measured with intervals in case hypoglicemia occurs and maintained with 5% dextrose fluid. After operation lasting about 90 minutes, neuromuscular block antagonization was performed with sugammadex. Patient was extubated without any complications and was followed for 30

minutes in recovery room before being transferred to clinic.

Fig-1: Photograph of patient (Beckwith-Wiedemann Syndrome)

DISCUSSION

Beckwith-Wiedemann Syndrome is a disorder of excessive growth marked by many anatomic and metabolic abnormalities, notably omphalocel, macroglossia and gigantism. In Beckwith-Wiedemann syndrome, abdominal anterior wall defects along with macroglossia are most common features [1]. Difficulty in laryngoscopy occurs in 2-8% of all general anesthesia cases. Diffiult intubation is less common. Failure in intubation is rarer, but may be fatal [2]. As it may lead to morbidity and mortality, difficult airway tools must always be kept ready. In intubation, gold standard is carrying out intubation with direct visualization of vocal cords [3]. Awake fiberoptic intubation is gold standard in difficult airway. Yet, it requires patients cooperation and experienced clinicians [4]. Especially in pediatric difficult intubation cases, awake fiberoptic application is very difficult. As to videolaryngoscope, it is aportable airway tool that facilitates intubation via direct visualization of larynx. Therefore, it is a very good option in pediatric patients with difficulty of intubation. Airway management, is particularly important in patients with Beckwith-Wiedemann syndrome. Probable anesthetic problems ascribed to physical characteristics of patients with Beckwith-Wiedemann syndrome are hypoglicemia and airway obstruction and difficulty in intubation caused by macroglossia. Upper airway obstruction and syndromic facial apperance makes it difficult to secure airway. Neurological sequels that may occur in association with hypoglicemia should also be borne in mind by the anethesist during operation.

CONCLUSION

Preoperative airway evaluation should be made carefully and cases with risk factors should be determined beforehand. Alternative and adjuvant airway tools should be kept ready for difficult intubation cases. It is our suggestion that videolaryngocoppe is an excellent alternative for airway management in pediatric patients in whom the use of fiberoptic is difficult.

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