Anesthesia Management of a Patient with Kabuki Make-up and Wolff Parkinson White Syndrome

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INTRODUCTION

Kabuki Make-UP syndrome (KMS) is a rare genetic disorder associated with multiple organ abnormalities. It was first described by Niikawa et al., and Kuroki et al. simultaneously in a group of patients in 1981. (1,2). The facial features of these patients were like the make-up actors in Kabuki dance-drama, traditional Japanese theatre. KMS has been estimated to occur in 1/32000 in the Japanese population.

KMS is related with multiple anomalies such as facial features congenital heart disease, pulmonary dysfunctions, neurologic disorders (West syndromes), and skelotomuscle disease and possible airway difficulties. Facial features is described as long palpebral fissures, ptosis, arched eyebrows and prominent eyelashes and ears. Approximately one-third of patients have a cleft lip/palate, while two-third have a high arched plate. Patients with KMS may have some congenital heart defects, and the prevalence of these defects in this population has been estimated to be from 40-50%.

Cardiac abnormalities include atrial and ventricular septal defects, mitral and aortic valve disease. Joint laxity and dislocations are common in these patients. Hips, patellae and shoulders are especially affected, so caution must be taken while positioning the patient. In this case, Furlow operation was planned due to palatopharyngeal anomaly.

Figure 1: Kabuki Make Up Syndrome

CASE

A 8,5 years old female and 22 kg patient, was diagnosed with KMS and cleft palate. She was scheduled for Furlow palatoplasty. She had the typical facial features (Figure 1). The 2D Echo showed Mitral stenosis, Patent foramen ovale, left persistent vena cava and left atrial dilatation. Her 12 lead EKG was suggestive of Wolff Parkinson White (WPW). She was premedicated with 0,5mg/kg midazolam. Anesthesia was induced with 2mg/kg Propofol, 1µg/kg fentanyl and 0,5 mg/kg rocuronium. The laryngoscopy showed a Grade II Cormack and Lehane view of the larynx. The trachea was intubated easily with a size 5.0 cuff endotracheal tube. The patient was positioned carefully because of joint laxity. Anesthesia was maintained with sevoflurane 2% in a 50% air-oxygen mixture and fentanyl was added when needed. Vital signs remained stable during surgery. The patient was extubated after applying 2 mg/kg squammax. Tramadol 1mg/kg and 20 mg/kg paracetamol were given for postoperative analgesia. We don't show any complication during the operation.

Figure 2: Delta Wave in Wolff Parkinson White Syndrome

DISCUSSION

The anesthesiologist needs to evaluate the airway preoperatively and prepare for possible difficulty (2). The patients with KMS may have some congenital heart defects, and the prevalence of these defects in this population has been estimated to be from 40% to 50% (1). The goal during perioperative management of anesthesia was to avoid any factors that increases sympathetic activity such as pain, anxiety, fear, stress response of intubation/extubation, lighter plane of anesthesia, hypovolemia and we avoid also from bradycardia because of mitral stenosis. Therefore we prefer fentanyl despite remifentanyl. Patients with WPW syndrome are always at a risk of PSVP or atrial fibrillation during perioperative period. Anesthetic drugs that precipitate tachycardia should be avoided. Sevoflurane have no effect on AV node and Fentanyl provides adequate hemodynamic stability. Rocuronium is cardiac stable muscle relaxant. Squammax is also hemodynamic stable than anticholinergic agents.

CONCLUSION

Anesthesiologist should be aware of a possibly difficult tracheal intubation, cardiac lesions, arrhythmias, respiratory problems, neurological and musculoskeletal disorders when dealing with Kabuki Make-up syndrome.

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