Perioperative Management of a Patient with Kabuki Syndrome

Monica Cheriyan, MD; Arundathi Reddy, MD
Department of Anesthesiology, University of Kentucky, Lexington, KY.

Introduction

Kabuki Syndrome is a possible autosomal dominant syndrome characterized by facial features that include long palpebral fissures and eversion of the lower eyelids

Anesthesiologists should be familiar with other features that can provide challenges to administering anesthesia.

Case Report

A sixteen-year-old female with Kabuki Syndrome presented for full mouth dental restoration under general anesthesia because of her severe dental disease and situational anxiety making her unable to cooperate in a routine dental setting. She had a bicuspid aortic valve, gastroesophageal reflux disease, obstructive sleep apnea, epilepsy, tracheomalacia, and hypogammaglobulinemias. Additionally she had a history of prolonged sedation after general anesthesia and a possible difficult airway secondary to her tracheomalacia. Fortunately she had a smooth intraoperative course with a mask induction using sevoflurane, nitrous oxide, and oxygen. Post induction a 22 gauge peripheral IV was placed and a 5.5 nasal RAE was inserted using a Miller 2 blade, grade I view using the Cormack-Lehane classification system. Intraoperatively anesthesia was maintained with desflurane, she was kept relaxed with rocuronium and was successfully extubated at the end of the procedure.

In the PACU she was unable to maintain her oxygen saturation > 90% without supplemental oxygen and was admitted for post-operative hypoxia. She was weaned to room air within 24 hours and discharged home.

Discussion

➢ Kabuki syndrome includes several features that can make the administration of anesthesia complex
➢ Features include: abnormal dentition, cleft palate, pectus excavatum, cardiac defects, mental retardation, short stature, scoliosis, and obesity
➢ Anesthetic considerations include: taking note of dental anomalies, positioning with regards to hyperextensible joints, and alteration of metabolism of anesthetic drugs.

Conclusion

➢ This patient had a fairly unremarkable anesthetic course
➢ Proper planning and discussion with the patient and family are necessary beforehand to manage expectations

References