The Anesthetic Management of a Patient with Cohen Syndrome

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Cohen syndrome is a rare genetic disorder caused by autosomal recessive inheritance and is characterized by the following features: mental retardation, infantile hypotonia, micrognathia, narrow and high-arched palate, microcephaly, prominent upper central incisors, poor dentition, short stature, and truncal obesity. Some patients have strabismus, myopia, optic atrophy, and total blindness. A small number of cases present with heart defects or mitral valve prolapse. Only approximately 100 cases have been reported in the world literature. The administration of general anesthesia in patients with Cohen syndrome can be a challenge because most of these patients are mentally retarded and uncooperative and have facial malformations that may make intubation difficult. We present our experience with the anesthetic management of a patient with Cohen syndrome.

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Case Report

A 28-yr-old woman with Cohen syndrome (1) was scheduled to undergo magnetic resonance imaging (MRI) of her right knee. The patient had been diagnosed with Cohen syndrome as an infant because of mental retardation, micrognathia, prominent upper central incisors, and obesity (2). The patient had moderate mental retardation and retinitis resulting in total blindness (3) but no heart defects or mitral valve prolapse (4). On physical examination, the patient followed some commands from her mother. She weighed 68 kg and was 4 ft 11 in. tall. She had micrognathia, truncal obesity, a short neck, a small mouth with macroglossia, prominent upper central incisors, and poor dentition. She was not able to open her mouth widely. Airway assessment was Mallampati Class 3. The remainder of the physical examination was unremarkable.

We invited the mother to come into the MRI procedure room with the patient. The standard noninvasive monitors were placed. Multiple laryngoscope blades, a fiberoptic bronchoscope, a laryngeal mask airway, and a jet ventilator for possible transtracheal jet ventilation were available. After a peripheral IV line was placed with the help of the mother, midazolam 2 mg was given IV. The patient became cooperative, and the mother left the room. After the patient breathed pure oxygen, anesthesia was induced with fentanyl 50 μg, lidocaine100 mg, and propofol 150 mg IV. The lungs were easily ventilated. Rocuronium 50 mg IV was given. Direct laryngoscopy was performed with a Macintosh No. 3 blade. The cords could not be visualized. A fiberoptic bronchoscope was introduced orally, and it revealed that the patient had a small U-shaped epiglottis that partially covered the glottic opening. A 7.0-mm endotracheal tube was successfully placed into the trachea over the fiberoptic bronchoscope. General anesthesia was maintained with sevoflurane and oxygen. The case proceeded uneventfully. After reversal of neuromuscular blockade, the patient was fully awake from the anesthesia and was alert with stable vital signs, and the endotracheal tube was removed. She did well after this procedure and was discharged home the same day.

Discussion

There are few reports in the medical literature regarding Cohen syndrome (5–7), primarily because the disease is rare and very few patients reach adulthood. One critical concern is the possibility of a difficult airway caused by craniofacial deformities in an uncooperative patient with mental retardation. An airway management plan should be established after an airway history is obtained and the airway is examined, in accordance with the ASA difficult airway algorithm. Micrognathia, a narrow and high-arched palate, microcephaly, and prominent upper central incisors are important features to consider in the airway management of a patient with Cohen syndrome.

Because this patient was difficult to tracheally intubate and was highly unlikely to cooperate with us, we did not attempt an awake fiberoptic intubation. General anesthesia was induced and ventilation could be

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performed. The patient was neuromuscularly blocked, and intubation was achieved via a fiberoptic technique. An intubating laryngeal mask airway was our backup method for intubation. We also had the means to immediately ventilate with a subglottic technique via transtracheal jet ventilation. An otolaryngologist was available to provide a surgical airway. Although we could have intubated the patient's trachea in the operating room, we believed that we could create the same environment and resources in the MRI suite, so we performed tracheal intubation there.

In conclusion, Cohen syndrome is a rare congenital syndrome with craniofacial deformities associated with the possibility of difficult airway management and cardiac abnormalities (8). In such patients, we suggest that preparations for alternative techniques of intubation be made and that equipment for managing the difficult airway be readily available.

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