Case Report
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Anesthetic management of a patient with chromosome 6p duplication: a case report

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Chromosome 6p duplication is very rare and clinically characterized by short stature, mental retardation, and congenital heart diseases. Patients with mental retardation may present with poor oral health conditions. Dental treatment may need to be performed under general anesthesia in such patients. Our case report deals with induction of general anesthesia to a patient with chromosome 6p duplication, for dental treatment. The selection of a nasotracheal tube of an appropriate size, because of the patient's short stature, was especially important for airway management. In the present case, the patient with chromosome 6p duplication was intubated with a nasotracheal tube, which was not age-matched but adapted to the height and physique of the patient.

Keywords: Chromosome 6p; Dental Care; General Anesthesia; Tube Size.

Chromosome 6p duplication is a rare genetic syndrome, first described in 1977 [1]. The clinical features of this syndrome include features of delayed development such as short stature, mental retardation, and congenital heart diseases [1-5]. Some patients also present with deficits in cognitive skills and impaired cooperation, decreasing their quality of life [5,6].

Dental caries is a common oral problem in these patients. As opposed to growth in healthy children, growth in these children is impaired, necessitating careful induction of anesthesia with respect to its depth. Selection of a nasotracheal tube of appropriate size for a patient of short stature, bispectral index (BIS) monitoring, and maintaining stable hemodynamics are especially important during anesthetic management [7,8]. There are few reports in scientific literature dealing with anesthetic management of patients with chromosome 6p duplication [2]. Our report deals with the anesthetic management of a patient with chromosome 6p duplication undergoing dental surgery.

CASE REPORT

The patient was a 4-year-old girl, 86.2 cm in height and 10.2 kg in weight. She was diagnosed with chromosome 6p duplication based on a chromosomal study performed when she was an infant. The clinical manifestations were short stature, mental retardation, and congenital heart diseases including atrial septal defect (ASD) and mild pulmonary stenosis (PS). Her gait was clumsy with short steps and a swaying motion. Her speech was affected; she could articulate only few words. Her mother discovered caries in her teeth. Therefore,

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dental treatment was scheduled under general anesthesia. Preoperative echocardiography revealed a small ASD and mild PS. The 12-lead electrocardiogram (ECG) showed sinus rhythm without axis deviation and conduction disturbance. The tracheal diameter at the level of the 6th cervical vertebra (C6), in the chest X-ray, was 6.4 mm. Moreover, results of the biochemical examination were within normal limits.

At the time of admission, her heart rate (HR) was 100 bpm, blood pressure (BP) was 76/36 mmHg, and peripheral oxygen saturation (SpO_2) in room air was 98%. On the day of dental treatment, she was transferred to the operating room without being premedicated. Anesthesia was induced using 1.0-5.0% sevoflurane inhalation in oxygen, with noninvasive monitoring for SpO_2 (98%). After loss of consciousness, ECG (sinus rhythm), BIS [BIS value: 40-58, spectral edge frequency (SEF): 10-18 Hz, signal quality index (SQI): 95%], BP (92/42 mmHg), and HR (122 bpm) monitoring was started. Mask ventilation was performed with ease. Fentanyl 20 μg, atropine 100 μg, and rocuronium 10 mg were administered after peripheral intravenous access was obtained. At first attempt, the 5.0 mm nasotracheal tube, using fiberscope, did not advance through the vocal cord. A 4.5 mm tube did not advance through the narrow space in the vocal cord either. Finally, the patient was intubated with a 4.0 mm nasotracheal tube using a laryngoscope. The pressure of air leak around the tube was 20 cmH_2O. Betamethasone 2 mg was administered to prevent laryngeal edema. Anesthesia was maintained with 1.5-2.0% sevoflurane inhalation in air and oxygen (FiO_2: 0.4). BP (70-100/40-64 mmHg), HR (90-145 bpm) and BIS (40-58 with SEF 10-15 Hz and SQI 95%) were maintained at stable levels. SpO_2 and end tidal carbon dioxide ($EtCO_2$) levels were maintained at 98-100% and 35-40 mmHg. In total, 1.8 ml of 2% lidocaine containing 1:200000 adrenaline was administered for dental treatment. The dental treatment for caries was completed uneventfully. The surgery was completed in 197 minutes without any surgical and/or anesthetic complications. There was minimal blood loss during surgery; she received a total of 230 ml acetated Ringer's solution with 1% glucose; the urine output was 140 ml. She regained consciousness 13 minutes after stopping sevoflurane inhalation. She was extubated on confirmation of sufficient spontaneous respiration. The respiratory and hemodynamic conditions were stable on extubation.

DISCUSSION

Chromosome 6p duplication is an extremely rare disease. Chromosome 6p genetic defects are classified as insertions, inversions, or deletions. Patients manifest with clinical features such as delayed development, mental retardation, and congenital heart diseases [1-3].

Dental treatment is difficult even when it is not pre-schooled; moreover, it is complicated by presence of systemic diseases. Patients with developmental delay and intellectual disability tend to have complex dental problems and poor oral health [4]. Dental treatment under general anesthesia could be beneficial in these patients. Selection of adequate tube size, maintenance of BIS monitoring, and stable hemodynamics are important in the anesthetic management of such patients.

It is known that the required tracheal tube size might be unexpected in patients with short statue [6]. The appropriate tube sizes for children have been reported earlier [7-9]. There are simple formulae using age, height, and weight to predict the optimal tracheal tube size [7,8]. However, age-based formulae are known to be inaccurate in up to 60% of children [7].

In this case, nasotracheal intubation was needed for unrestricted surgical access to perform dental treatment. At first, a 5.0 mm nasotracheal tube was selected for intubation taking into account the internal diameter of her trachea at the level of C6 and her age. However, this tube or the tube with a diameter of 4.5 mm could not be inserted into the trachea using a fiberscope. Eventually, a 4.0 mm nasotracheal tube was inserted using a laryngoscope. Patients with short stature might have a narrow subglottic or tracheal airway when compared to

the airway in healthy patients. In addition, it is difficult to select the adequate tube size using fiberscope when compared to laryngoscope. We anticipated that it would be anatomically difficult to intubate her, and used the fiberscope to check the glottis and pharynx, before intubation. However, intubation was performed with a laryngoscope, to check the size of the tracheal tube and glottis. There are no reports on the adequate nasotracheal tube size. Therefore, we suggest that nasotracheal tubes a size smaller and a size larger than the predicted tube size should be readily available.

With several intubation attempts, there was potential postoperative edema that might have threatened the airway; hence, betamethasone was administered. At the end of the dental surgery, we checked for air leak and spontaneous respiration. Then, we removed tracheal tube in the preparation of difficult airway.

The depth of anesthesia was evaluated using BIS monitor. There was a recent report about the BIS monitoring of adequate depth of anesthesia using anesthetic agents [10]. However, in special needs patients such as those with mental retardation, a discrepancy between electroencephalogram and hemodynamics was reported; BIS value showed a sharp fall, although hemodynamics was stable during general anesthesia. In such cases, titration of anesthetics can be performed using information from the EEG, and BIS monitor maintained stable.

Congenital heart disease is frequently associated with chromosome 6p defects [2,3]. Based on the degree and type of heart disease, careful monitoring of hemodynamics is necessary. In this patient, the ASD and PS would be considered mild. Anesthetic agents and hemodynamic responses to them might cause significant circulatory disturbances. Hence, we carefully titrated the anesthetics during administration of anesthesia.

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