Case Report
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Anesthetic management of a patient with branchio-oto-renal syndrome

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Branchio-oto-renal syndrome (BOR) is a rare autosomal dominant disorder. The features include branchial cysts, hearing loss, ear malformation, preauricular pits, retrognathia, congenital heart disease, and renal abnormalities. However, anesthetic management of these patients has seldom been reported. We report a case in which general anesthesia was performed for dental treatment in a patient with BOR. Airway management, renal function, and hemodynamic changes can be of critical concern during anesthetic management. A 13-year-old girl diagnosed with BOR had severe right hearing loss, right external ear malformation, renal abnormalities, and postoperative patent ductus arteriosus (PDA). Dental extraction under general anesthesia was scheduled for a supernumerary tooth. The procedure was completed with sufficient urine volume, adequate airway management, and stable hemodynamics.

Keywords: Branchio-oto-renal Syndrome; Congenital Heart Disease; Difficult Airway; General Anesthesia; Renal Failure.

Branchio-oto-renal syndrome (BOR) is a rare autosomal dominant disorder, first described in 1976 [1]. The features include branchial cysts, fistulas, hearing loss, external ear malformation, retrognathia, congenital heart disease, and renal abnormalities [2-5]. However, anesthetic management of these patients has seldom been reported [1].

We report a case in which general anesthesia was performed for dental treatment in a patient with BOR. We have obtained consent for the case report from the patient.

CASE REPORT

A 13-year-old girl (height: 144.3 cm, weight: 41.4 kg) was diagnosed with BOR on the basis of a chromosomal study. Her clinical features included right facial nerve paralysis, hypoglossal nerve paralysis, severe right hearing loss, right external ear malformation, renal abnormalities, and postoperative patent ductus arteriosus (PDA). Dental extraction under general anesthesia was scheduled for a supernumerary tooth. Auscultation of the heart and lungs was within normal limits. She had no history of seizures. Preoperative echocardiography was unremarkable. A 12-lead electrocardiogram (ECG) showed normal sinus rhythm. No abnormalities were noted on renal function testing, chest X-ray, or laboratory data. In addition, clinical assessment of her airway was unremarkable, with Mallampati grade II and mouth opening 30 mm.

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On admission for dental extraction under general anesthesia, blood pressure (BP) was 112/52 mmHg, heart rate (HR) was 79 bpm, and oxygen saturation by pulse oximetry (SpO_2) was 100% on room air. On the day of surgery, no premedication was given, and she was transferred to the operating room. Anesthesia was induced with inhalation of sevoflurane 1-8% in 100% oxygen after the start of noninvasive monitoring for SpO_2 (100%), ECG (sinus rhythm), Bispectral Index (BIS value: 44-60, spectral edge frequency: 10-20 Hz, signal quality index: 95%), BP (105/45 mmHg), and HR (86 bpm). After loss of consciousness, mask ventilation with an oral airway was easily performed. Fentanyl 0.1 μg, atropine 0.2 μg, and rocuronium 30 mg were administered after peripheral intravenous access was obtained. Intubation was easily carried out with a 6.0-mm tracheal tube and a Macintosh laryngoscope blade (size 3). Cormack-Lehane classification was grade I, with a confirmed air leak (<25 mmHg). Anesthesia was maintained with isoflurane 0.8-1% in air and oxygen. BP was maintained at 80−108/40-48 mmHg, HR was 69-86 bpm, end-tidal carbon dioxide ($EtCO_2$) was 35-40 mmHg, and BIS value was maintained between 40-58 with SEF 10-15 Hz and SQI 95%. Dental treatment was completed in 204 min without any surgical and/or other anesthetic problems. There was minimal blood loss during the operation and she received a total of 639 mL lactated Ringer's solution with 1% glucose. Urine volume was 240 ml. At the end of surgery, we observed an air leak (<35 mmHg) and spontaneous respiration, then removed the tracheal tube. After extubation, her respiratory and hemodynamic status was stable. There were no significant postoperative changes and/or complications, and the patient was discharged 1 day later.

DISCUSSION

BOR is caused by a gene abnormality and has a prevalence of 1:40.000 births [2,4]. Major diagnostic criteria include branchial anomalies, deafness, preauricular pits, and renal anomalies. Minor criteria include middle ear anomalies, congenital heart disease, and facial and palatal abnormalities [4-6].

Risk factors for anesthetic management include decreased renal function, congenital heart disease, and a difficult airway.

Renal malformations have been reported in BOP [5,6]. The renal anomalies often remain asymptomatic. Patients with BOR may have congenital renal malformations that result in renal failure. Opioids and volatile anesthetics can reduce urine output. We carefully administered anesthetics with titration and avoidance of nephrotoxic muscle relaxants, nonsteroidal anti-inflammatory drugs, and antibiotics. Low doses of drugs were administered and careful fluid and urine management was performed.

Congenital heart disease is associated with BOR [1]. Therefore, hemodynamic monitoring is critical. Sevoflurane and isoflurane do not usually cause bradycardia in children undergoing general anesthesia, but an unpredictable incidence of bradycardia with volatile anesthetics has been reported [1]. In this patient with postoperative PDA, we carefully administered anesthetic agents with monitoring of hemodynamic changes and BIS, even though preoperative assessment of cardiac function, ECG, and chest X-ray revealed no abnormal findings.

A difficult airway was anticipated because of the presence of high palate, retrognathia, sleep apnea, and facial paralysis in BOP [1,5,6]. Preparation before anesthesia induction is essential in pediatric cases in which a difficult airway is predictable, although it was easy to control ventilation and intubation in this patient. The operating room was prepared, with different sized masks, tracheal tubes, nasal and/or oral airways, stylets, laryngeal masks, and a fiberscope and tracheostomy set. The control of urine volume, a difficult airway, and hemodynamic changes in terms of anesthesia management was the most important concern in this BOR patient. Therefore, it was necessary to assess blood urea nitrogen, creatinine, the airway, and cardiac status prior to anesthesia.

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