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CLINICAL REPORT

Airway management in an infant with double aortic arch

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Abstract A 2-month old male was admitted due to repeated cyanotic attacks. He had suffered from stridor and retractive breathing since birth. Double aortic arch was diagnosed and the vascular ring formed by the double aortic arch was compressing the trachea. Multirow detector computed tomography showed that he had a right-dominant double aortic arch with left ductus arteriosus and an aberrant left subclavian artery, and that the narrowest part of the trachea, where the diameter was 2.0 mm, was located 9.0 mm above the carina. Airway management in patients with extreme narrowing of the trachea is challenging for anesthesiologists. He was scheduled for ligation and division of the left aortic arch and ductus arteriosus. In the operating theater, anesthesia was slowly induced with sevoflurane (0–4%) in oxygen. After mask ventilation was confirmed to be adequate, a 4.0 mm internal diameter endotracheal tube (ETT) was inserted and advanced smoothly beyond the tracheal stenosis. The tip of the ETT was placed just above the carina using a fiber optic bronchoscope (fiberscope) that was passed through the ETT. Since mechanical ventilation was adequate, vecuronium was administered. Surgery was conducted in the right lateral position and using a left thoracotomy approach. Anesthesia was maintained with sevoflurane (2–3%). After positioning, right one-lung ventilation was performed unexpectedly. However, anesthetic management was achieved without difficult ventilation during surgery. The tip of the ETT was pulled past the stenotic part before transfer to the intensive care unit (ICU). A patent trachea during spontaneous breathing under CPAP (5 and 2 cmH_2O) was confirmed with a bronchofiberscope in the ICU. After weaning from mechanical ventilation, he had the persistence of mild stridor despite improvement of respiratory symptoms.

Keywords Double aortic arch · Vascular ring · Murphy eye · Airway management · Infant

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Introduction

Vascular malformation of the double aortic arch may cause the formation of a vascular ring. The cause of a vascular ring includes a double aortic arch, Neuhauser's abnormality, which was defined as a right aortic arch with left ductus arteriosus and without a left bronchiocephalic arterial trunk, a right retro-esophageal (aberrant) subclavian artery, and a right aortic arch with a left bronchiocephalic arterial trunk and left ductus arteriosus (LBAT and LDA) [1]. Patients with a vascular ring present stridor, dyspnea and dysphasia due to tracheoesophageal compression [1], and symptoms of double aortic arch appear earlier than other varieties of vascular rings [2–4], but it is difficult to diagnose a vascular ring due to its unspecific symptoms [5]. As well as hemodynamic management, airway management of infants with double aortic arch can be a challenge for anesthesiologists, since there is only a small distance from the carina to the narrow part of the trachea (0–15 mm) and there is a risk of severe stenosis with respiratory failure [6]. Weaning from mechanical ventilation should be conducted carefully because previous studies have shown that respiratory symptoms can persist due to tracheomalacia and/or tracheal stenosis in pediatric patients postoperatively [7, 8]. Here, we report anesthetic management during surgery for double aortic arch in an infant.

Case report

A 2-month old male weighing 5.7 kg was admitted to our hospital with repeated cyanotic attacks. He had suffered from severe stridor and retractive breathing since birth. Double aortic arch was diagnosed by echocardiography and multirow detector computed tomography (MDCT) (Fig. 1). MDCT showed that he had a right-dominant double aortic arch with LBAT and LDA, that the vascular ring strongly compressed the trachea above the carina, that the narrowest part of the trachea was located 9.0 mm above the carina, and that the tracheal diameter of the narrowest part was 2.0 mm.

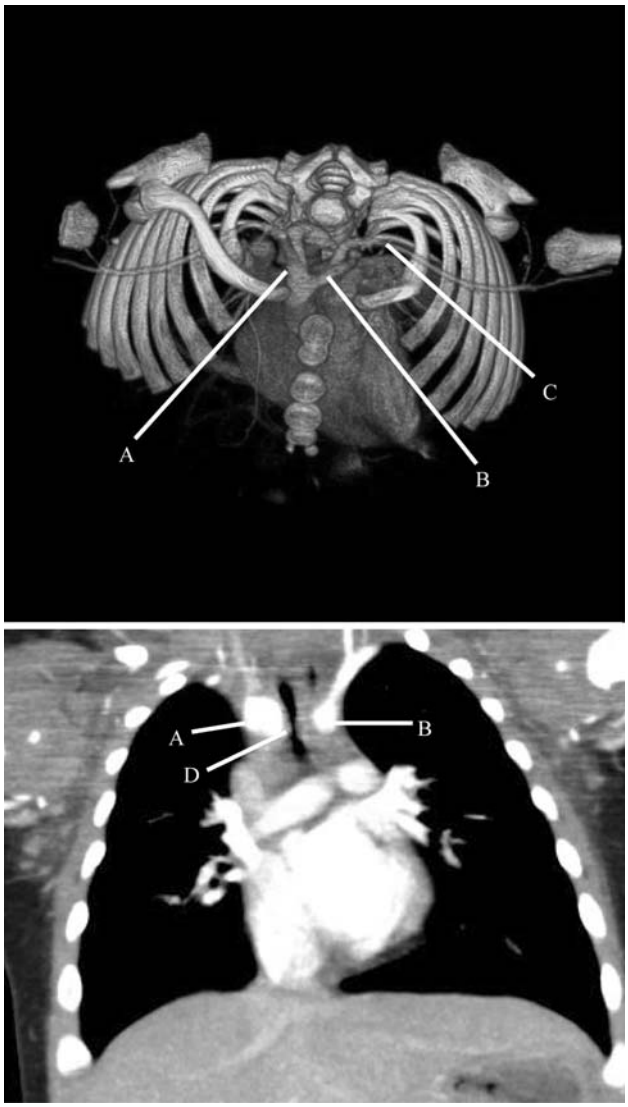

Fig. 1 MDCT showing the double aortic arch and CT showing tracheal compression. The trachea runs into the vascular ring formed by the right aortic arch and left aortic arch. *A* right aortic arch, *B* left aortic arch, *C* left subclavian artery, *D* tracheal stenosis

He was scheduled for ligation and division of the left aortic arch and ductus arteriosus. He was premedicated with chloral hydrate 30 mg kg^{-1} administered into the rectum. In the operating theater, slow induction of general anesthesia with sevoflurane (0–4%) was performed. After it had been confirmed that the mask ventilation was adequate, a 4.0 mm ID (5.5 mm outside diameter) endotracheal tube (ETT) with a Murphy eye was inserted and advanced smoothly beyond the tracheal stenosis, which was visualized with a 2.2 mm external diameter fiberscope through the ETT. The tip of the ETT was placed just above the carina using the fiberscope. Since mechanical ventilation was adequate for bilateral lungs, vecuronium 0.5 mg was administered intravenously. Surgery was conducted in the right lateral position. Although right one-lung ventilation was performed unexpectedly after positioning for surgery, the ETT was kept in place since ventilation was not difficult. Anesthesia was maintained with sevoflurane (2–3%) and an O_2/air mixture (fractional inspired oxygen [F_IO_2] = 0.6). His arterial blood gas analysis values showed that the partial pressure of carbon dioxide [$PaCO_2$] = 41 mmHg, partial pressure of oxygen [PaO_2] = 204 mmHg, and base excess [BE] = 1.8 mmol/l. Anesthetic management was performed without difficult ventilation during ligation and division of the left aortic arch and ductus arteriosus under one-lung ventilation. The tip of the ETT was withdrawn above the tracheal stenosis, and an improvement in the tracheal narrowing was confirmed with a fiberscope before transportation to the intensive care unit (ICU). In the ICU, a 2.2 mm external diameter fiberscope was used to observe the tracheal stenosis during spontaneous breathing with different levels of CPAP ranging from 5 to 2 cm H_2O, confirming oxygen saturation by pulse oximetry. Restenosis related to edema did not occur, and airway management proceeded without difficulty after surgery. Successful weaning from mechanical ventilation was achieved on the third postoperative day, and the patient had only mild stridor 1 month later.

Discussion

Patients with a vascular ring present with respiratory and gastrointestinal symptoms of differing severities because of tracheoesophageal compression [1]. The double aortic arch manifests earlier symptoms than those associated with other varieties of vascular rings [2–4], and life-threatening episodes can occur [5, 6, 9]. Neonates and infants with a double aortic arch suffer severe symptoms such as cyanosis (12%), apnea (9%) and respiratory arrest (7%) [10], and they should be taken care of without delay. Therefore, proper airway management is important for saving their lives.

Multirow detector computed tomography allows a detailed understanding of the anatomical forms of the cardiovascular and respiratory systems [11, 12]. We were able to determine the anatomy of our patient's respiratory and cardiovascular systems preoperatively. This enabled us to decide on suitable airway management and the surgical procedure, including positioning and approach.

In infants, there is only a small distance from the carina to the narrow part of the trachea [6]. Therefore, it is difficult to decide on the appropriate location for the ETT tip. Difficulty with ventilation when the tip was located at or above the stenosis has been reported [13]. In the present case, placing the ETT in the trachea above the stenosis was expected to result in difficult ventilation because of the presence of extreme narrowing, so the tip of the ETT was placed just above the carina and below the tracheal narrowing. Right one-lung ventilation occurred in the right lateral position, and this could not provide adequate ventilation compared to bilateral lung ventilation. One-lung ventilation was not necessarily required to perform this surgery. We noted that one-lung ventilation for patients with double aortic arch is not necessarily safe management, since Alsenaidi et al. [10] showed that 17% of patients with double aortic arch had cardiac anomalies such as a ventricular septal defect, an arterial septal defect, a patent ductus arteriosus, and tetralogy of Fallot. However, we didn't reposition the ETT because ventilation was not difficult during surgery and the arterial blood gas analysis values were permissible. As a result, the positioning of the tip of the ETT above the stenotic part was avoided.

The end of the ETT, which had a beveled structure that opened on the left side, could be obstructed by the right broncheal wall. When the ETT was advanced into the right bronchus, the Murphy eye of the ETT prevented the ETT from being obstructed completely [14], while it was unclear whether the Murphy eye contributed to adequate ventilation, since the end of the ETT was not obstructed in this case.

Double aortic arches in infants and children may cause significant and persistent respiratory symptoms postoperatively due to residual tracheomalacia and/or tracheal stenosis for more than 6 months [5, 8, 10]. Tuo et al. [15] reported that a neonate with severe congenital tracheal stenosis with double aortic arch required tracheoplasty. The severity of persistent respiratory symptoms should be evaluated before weaning from mechanical ventilation [6]. Mok et al. [16] demonstrated that bronchography with varying CPAP was a better method for evaluating tracheomalacia than dynamic CT. Soong [17] reported that ultrathin flexible bronchoscopy was useful for assessing tracheomalacia. In the present case, a 2.2 mm external diameter fiberscope was used to observe his trachea during spontaneous breathing with different levels of CPAP ranging from 5 to 2 cm H_2O, since bronchography was not performed in our institution as it is a stressful intervention for infants. This patient was successfully weaned from mechanical ventilation after confirming that the airway had been opened, and he exhibited postoperative persistence of stridor in spite of an improvement in respiratory symptoms.

In conclusion, we experienced a case of double aortic arch in an infant that required anesthetic management during surgery. The main anesthetic problem is to prevent life-threatening events caused by the difficult management of the airway. Safe airway management was achieved by positioning the ETT below the tracheal narrowing and assessing the airway tract carefully while weaning from mechanical ventilation.

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