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CLINICAL REPORT

A novel association of alveolar capillary dysplasia, atypical duodenal atresia, and subglottic stenosis

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Abstract Alveolar capillary dysplasia (ACD), which is a rare and lethal congenital pulmonary anomaly found in newborns, begins its onset or causes deterioration of the infant's condition some time after birth. Various congenital anomalies in combination with ACD have been reported, except for subglottic stenosis. Therefore, we aim to report a novel association in a case of ACD with the combination of atypical duodenal atresia and subglottic stenosis. The male infant was scheduled for duodeno-duodenostomy because a double-bubble sign was observed on a chest radiograph. He arrived at the operating theater without any symptoms. After induction of general anesthesia, although mask ventilation was performed without difficulties throughout the entire procedure, oxygen saturation values of the upper and lower extremities dissociated after several attempts of intubation. Surgery was canceled because of instability of the respiratory condition. Respiratory insufficiency worsened progressively, and the infant died at 5 days of age. An autopsy confirmed ACD and revealed cartilaginous subglottic stenosis, which had made intubation difficult. This report highlights the hazards of the onset and worsening of ACD, and the importance of thorough echocardiography before surgery when atypical duodenal atresia is suspected. Anesthesiologists should also be prepared for the difficulty of intubation.

Keywords Alveolar capillary dysplasia · Atypical duodenal atresia · Subglottic stenosis

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Introduction

In the pediatric field, several diseases begin their onset or cause deterioration of the infant's condition some time after birth. Alveolar capillary dysplasia (ACD), which is a rare but lethal congenital pulmonary anomaly found in newborns, is also one of these diseases. ACD is clinically associated with persistent pulmonary hypertension of the neonate (PPHN) [1]. Histopathologically, dysplasia of the alveolar walls and capillaries with/without misalignment of pulmonary vessels is observed [2]. Ever since its first report by Janney et al. [2] in 1981, only about 100 cases have been reported in the literature together with other abnormalities [3–6]. However, subglottic stenosis has not been reported. To date, in the anesthesia literature, ACD has not been reported and is poorly understood despite the fact that ACD is a life-threatening disease and important for anesthesiologists.

In this paper, the authors describe the anesthetic precautions for a patient with atypical duodenal atresia and discuss the potential difficulties in intubation due to airway abnormalities associated with ACD.

Case report

The patient was diagnosed prenatally with duodenal atresia by fetal ultrasonography at 31 weeks of gestation because of polyhydramnios. Two thousand milliliters of amniotic fluid was drained at 32 weeks of gestation. Genetic testing of the fetus by FISH was normal. The male infant was delivered vaginally at 37 weeks of gestation with a body weight of 2,528 g and height of 49 cm. At 1 and 5 min, his APGAR scores were 9 and 9, respectively. Since duodenal atresia was suspected after birth, the patient was transferred

to our hospital. A chest radiograph showed slight hyperlucency with normal cardiac size. No abnormal cardiac physical findings, including hypoxia, stridor, tachypnea, tachycardia, and cardiac murmurs, were observed. Because echocardiography performed at the previous hospital reported no abnormalities except patent ductus arteriosus, additional preoperative echocardiography was not performed. Abdominal radiograph showed large gastric dilatation. Although the typical double-bubble sign was not observed, duodenal atresia was confirmed following upper gastrointestinal barium study. Therefore, duodeno-duodenostomy was planned. There were no dysmorphic external features.

The patient arrived at the operating theater at 5.5 h of age. He showed normopnea with normal ausculatory findings, while oxygen saturation was 97% on room air. An orogastric tube had already been placed. Atropine 0.025 mg was given IV. General anesthesia was induced with thiamylal. Laryngoscopy was facilitated with vecuronium. Twenty minutes after induction and following four attempts at intubation, oxygen saturation gradually decreased to 85 in 100% oxygen despite the fact that the lungs were easy to ventilate. A capnogram was used the entire time and showed endotidal CO_2 during the mask ventilation. Since oxygen saturation in the lower extremities (80%) was less than in the upper extremities (91%), a patent ductus arteriosus (PDA) and possible pulmonary hypertension were suspected. A 2.5-mm ID (3.5-mm OD) tracheal tube was attempted at first, but could not pass the cricoid ring. A serially smaller diameter 2.0-mm ID (3.0-mm OD) tracheal tube (Portex blue line, Kent, UK) was passed through the cricoid ring, but only with effort. Even with this size, moderate resistance at intubation was noted. There was no air leakage. Since subglottic stenosis in the full-term infant is defined as a subglottic airway diameter of less than 4 mm at the level of the cricoid cartilage [7], a clinical diagnosis of subglottic stenosis was established in this patient. Surgery was canceled because positioning for surgery or slight compression to the abdomen caused depression of oxygen saturation. The patient left the operating theater when oxygen saturation in the upper extremities could be maintained at 98 in 48% oxygen. Further investigation of causative factors of PPHN was planned.

Lung biopsy was not feasible due to the critical condition. However, the clinical features and the results of echocardiography—the narrowing of the pulmonary artery—suggested ACD. Echocardiography during mask ventilation and just after intubation revealed that the estimated pulmonary artery pressure was 38 mmHg, whereas the systemic systolic blood pressure was 40–50 mmHg. Several drug therapies were started in the NICU. However, inhaled nitric oxide (NO) therapy was effective only transiently, and other medications including prostacyclin (prostaglandin I_2, PGI_2) and isoproterenol were ineffective. Extracorporeal membrane oxygenation (ECMO) was not performed because it has been known that patients with suspected disease cannot be weaned from ECMO and this would just prolong the course [7]. Respiratory insufficiency worsened progressively, and the infant died at 5 days of age. An autopsy revealed dysplasia of the alveolar walls and capillaries with misalignment of the pulmonary vessels. Paradoxical dilatation of the duodenum and cartilaginous subglottic stenosis were also observed.

Discussion

Although the combination of duodenal atresia and subglottic stenosis has been reported [8], the addition of ACD leading to oxygenating difficulties precluded continuing the surgery. In the presence of PPHN and a PDA, the former refractory to medical management, the risk of desaturation and frank hypoxia together with a potentially escalating acidosis presented insurmountable challenges to manage during surgery. In order to stabilize the neonate's condition, surgery was canceled, and the child's medical condition was assessed further for treatment and prognosis by the neonatologists. Ever since its first report [2], various kinds of congenital anomalies in combination with ACD have been reported, such as cardiovascular anomalies, genitourinary tract malformations, and gastrointestinal abnormalities [6]. To the best of our knowledge, however, subglottic stenosis has never been associated with ACD. At the same time, this is the first report of a patient with ACD in the anesthetic literature.

ACD can be differentiated from idiopathic PPHN by the following characteristics in the former: (1) no predisposing factors, including asphyxia, prematurity and infection; and (2) there is a latent period from birth to the onset of the disease with a mean duration of 48 h, namely, a 'honeymoon period' in the former [2, 9]. Michalsky et al. [10] reported that 75% of ACD patients had normal APGAR scores but deteriorated 1.5 h to 30 days after birth. If it was before onset time, patients would have seemed normal by the time they arrived at the operating theater. This time lag can be a pitfall to anesthesiologists without experience of this disease. Anesthesiologists may not be aware that the patient's condition is so severe that it can be fatal. Furthermore, its rarity makes it more difficult to diagnose or suspect ACD during induction of anesthesia. Since we applied oxygen saturation monitors on both the upper and lower extremities and the neonatologist who had previously diagnosed ACD attended at the operating theater, it did not take us long to suspect ACD. Although combination therapy of inhaled NO and intravenous PGI_2 has been

reported effective to reduce persistent pulmonary hypertension in ACD [11], its effectiveness was only transient, and the patient ultimately died. None of the therapies, including NO, PGI_2, and even invasive ECMO, have been effective so far to save patients with ACD [2–6]. It seems that lung transplantation is the only effective therapy [12]. However, it would be very difficult to diagnose correctly and find a donor under such an urgent circumstance.

Before the development of PPHN, there are no known data highly indicative of ACD. We speculate the atypical dilatation of the distal blind end in duodenal atresia might be a clue in the diagnosis of ACD. Two reasons can be proposed. First, Usui et al. [5] previously reported three cases of ACD with the same findings. While Haraida et al. [13] speculated that intestinal dysmotility caused the dilatation, Usui et al. suggested blood flow disturbance as a relevant cause of duodenal atresia and ACD since there was no intestinal dysfunction or histologic abnormality in their cases. In our case, although functional abnormality was not confirmed because the correction of duodenal atresia could not be done, a histologic abnormality was not found. Second, embryologically, the duodenum, laryngeal cartilage, and lung commence to develop around the 4th–5th weeks from the endoderm, the 4th and 6th branchial arches, and the lung bud (6th branchial arch), respectively [14]. Abnormal vasculogenesis in the organogenesis period might have disturbed blood flow, resulting in peristalsis dysfunction of the distal blind end of the fetal duodenum accompanied by a paradoxical dilatation and misalignment of pulmonary vessels, and subglottic stenosis.

Jacobs et al. [15] reported that as much as 67% of infants with pulmonary artery hypertension have airway obstruction including subglottic stenosis, tracheobronchomalacia, and laryngomalacia. Therefore, difficulty in mask ventilation or intubation would be encountered frequently in these patients. Subsequent use of a venous catheter through the oral cavity or cricoid membrane [16], or emergency tracheostomy may be required.

Anesthesia in emergency pediatric cases is often jeopardized by undiagnosed abnormalities. Although there may be an argument for performing a preoperative echocardiography on neonates without abnormal cardiac physical findings, we strongly recommend it if duodenal atresia with an atypical bubble sign is suspected. At the same time, airway assessment by ultrasonography may be of help in case of subglottic stenosis. Special caution should be exercised in cases of atypical duodenal atresia.

Conflict of interest This case was supported solely by departmental resources.

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