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■ Case report ■

A case of pulmonary vascular air embolism in a very-low-birth-weight infant with massive hydrops

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= Abstract =

Pulmonary vascular air embolism is a rare and, universally, almost a fatal complication of positive pressure ventilation in newborn infants. Here, we report a case of this unusual complication in a very-low-birth-weight infant who showed the clinical and radiological features of this complication along with pulmonary hypoplasia and massive hydrops. The possible pathogenesis has been discussed and a brief review of related literature has been presented. (Korean J Pediatr 2009;52: 1392-1395)

Key Words : Pulmonary embolism, Very-low-birth-weight, Hydrops fetalis

Introduction

Pulmonary vascular air embolism is a rare, and almost invariably fatal complication of positive pressure ventilation in newborn infants[1-5]. According to available statistics, there have only been 60 cases described in the world literature to date[6-8], and two cases of pulmonary vascular air embolism in the newborn were reported in Korea[6, 9]. Infants affected most notably have been very preterm neonates with respiratory distress syndrome who required mechanical support with high ventilatory pressures[10, 11]. The cause of death may not be obvious initially, unless this condition is suspected and appropriate radiologic examinations are performed[12]. It is documented that this is the first case of pulmonary vascular air embolism in a pulmonary hypoplasia associated with massive hydrops in Korea.

In this article, the pathophysiology and clinical characteristics of pulmonary vascular air embolism are discussed, and a brief review of related literature is also presented.

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Case report

A baby girl was born by vaginal delivery and weighed 1,140 g at 25^{+6} weeks' gestation to a 34-year-old woman (gravida 1, para 1). She had been diagnosed with hydrops fetalis at 24 weeks' gestation with negative TORCH and parvovirus titers. It was noted that the baby's Apgar Score was 0 and 2 at one and five minutes, respectively. As soon as she was intubated, the procedure of thoracenteses with intravenous catheters were performed on both sides on emergency in the delivery room, as there was whole body cyanosis, no heart rate and no respiratory effort. After pleural effusion was removed (40 mL on the right side, 30 mL on the left side) and intratracheal epinephrine was instilled, the heart rate was recovered at greater than 100/min. She was then immediately transferred to the neonatal intensive care unit (NICU).

In the NICU, her initial oxygen saturation was 82% and heart rate was 109/min with positive pressure ventilation. Apparently, she appeared to have a severely generalized edematous appearance. She had multiple bruises on the scalp, face and anterior chest wall as well. The umbilical cord vessels were exposed at the umbilical stump, being neatly cut. The insertion of an orogastric tube into the esophagus failed due to resistance. As a result, the aggravating severe respiratory insufficiency could only be treated with high frequency oscillatory ventilation, the insertion of three

chest tubes (two on the right side, and one on the left side), and nitric oxide. In addition, ventilatory requirements were further increased up to a mean airway pressure (MAP) of 19 cmH_2O, a stroke volume of 45 cmH_2O, a fractional inspired oxygen concentration of 100% and a nitric oxide of 20 ppm. Simultaneously, she was also commenced on dopamine, dobutamine, and continuous epinephrine.

By 16 hours of age, her clinical condition deteriorated, her intra-arterial blood pressure waveform dampened, and profound bradycardia developed with peripheral pallor of all the limbs. Blanching bright pink vessels against a generally cyanosed cutaneous background manifested on the right shoulder, neck and scalp (Fig. 1). At this stage, attempts to collect blood samples from an indwelling umbilical arterial catheter revealed the presence of air bubbles. Chest radiography subsequently showed air in the major vessels of the neck, heart chambers, and the abdomen (Fig. 2). Resuscitation was ultimately unsuccessful. As a consequence, she became progressively bradycardic and expired shortly thereafter.

Postmortem examination revealed a generalized edema and high arched palate, as well as hyaline membrane disease at the early stage with diffuse alveolar hemorrhage. At the same time, both lungs had collapsed, and diffuse hypoxic damage was observed with multifocal hemorrhage of

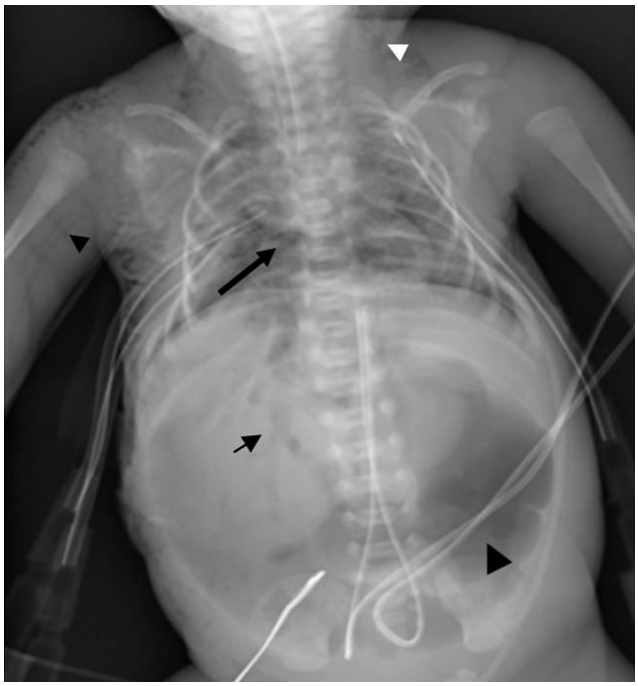

Fig. 2. Chest X-ray reveals a decreased lung volume. Endotracheal tube, umbilical catheters, and chest tubes are in place. Air is present within the cardiac chambers (large arrow), and branching lucencies within the liver indicate hepatic venous air (small arrow); pneumoperitonium is also observed (large arrowhead). Air in the neck is located within both carotid arteries and jugular veins (small white arrowhead). Linear lucencies in the axillae represent intravascular air in the axillary artery and vein (small black arrowhead).

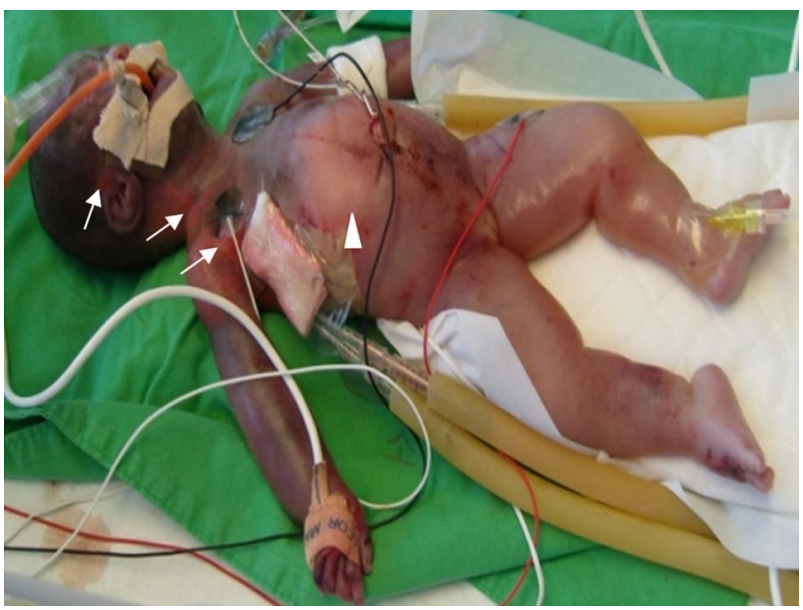

Fig. 1. The patient presents a massive generalized edema. Blanching bright pink vessels against a generally cyanosed cutaneous background are observed on the neck, right shoulder, and scalp (arrow). Subcutaneous emphysema is observed on the abdomen (arrowhead).

the thymus, kidney, spleen, esophagus, and the hemoperitoneum. However, postmortem examination was unable to disclose the cause of this massive fatal air embolism.

Discussion

Pulmonary vascular air embolism represents a very rare, but potentially life-threatening event with a high mortality rate that occurs as a consequence of the entrainment of air into the vasculature[1-3, 13, 14]. The exact mechanism of the air leaking into the vascular space is less likely to be clinically explained. In essence: a direct communication between a source of air and the vasculature must exist, and a pressure gradient favoring the passage of air into the circulation[1-3, 15]. Rupture of alveoli into pulmonary capillaries due to barotraumas through alveolar-capillary or bronchovenous fistulae is thought to be the main cause of massive air embolism in infants[1, 12, 18, 19]. In some cases, there was evidence of trauma related to the introduction of chest tubes. In general, lung perforation is noted to occur in 25-30% of infants with respiratory distress syndrome who have chest tubes inserted for drainage of pneumothoraces [20]. Furthermore, laceration of the lung tissue is reported to favor reversal of the intra-bronchial pressure-pulmonary venous pressure gradient, thereby increasing the risk of pulmonary vascular air embolism[17]. In the present case, the infant had three chest tubes, nonetheless, evidence of trauma to the lung during the postpartum examination was not observed.

In our patient, we postulate that such a condition of pulmonary vascular air embolism may have been to do with a marked pulmonary hypoplasia, which was complicated with a hydrops fetalis in the phase directly after birth, especially in a very low birth weight infant. However, there is limited literature available of air embolism associated with these conditions. A review of the cases published since then identifies that the affected infants are usually very preterm neonates with respiratory distress syndrome. Only one paper refers to air embolism in an infant following pulmonary hypoplasia with hydrops fetalis[8].

Clinical signs were observed to include a sudden collapse with pallor, poor perfusion, mottling, narrowed or flat pulse pressure, apnea, cyanosis, hypotension, and bizarre electrocardiogram irregularities, varying from tachycardia to bradycardia, with the latter being more common. More specifically, a millwheel murmur was heard in several cases, and the heart sounds were also noted to be distant and diminished. In addition, blanching and migrating areas of cutaneous pallor were observed in several cases, and as in our case[3, 10]. Diagnosis is supported by the withdrawal of gas from an indwelling umbilical arterial catheter and radiographic examination before death, knowing that free air can be present in both the arterial and venous systems, as well as in the heart[14]. At the present time, there is no effective treatment for pulmonary vascular air embolism[6, 9]. Establishing clear guidelines for prevention is less likely to be possible due to the fact that the avoidance of high airway pressures, which is rarely applied if there is an alternative, would be advantageous. Hence, high frequency ventilation may eventually offer an alternative, despite the fact that the use of a surfactant applied down the airways may reduce the need for extremely high ventilation pressures. In infants with a pneumothorax, it may be possible to reduce the incidence of pulmonary vascular air embolism using soft rubber catheters, instead of stiff plastic chest tubes for drainage, as these may be less traumatic to the surface of the lung[6, 12, 16]. Considering the possibility that pulmonary vascular air embolism occurs despite the use of high frequency ventilation and soft chest tubes, in this case, pulmonary vascular air embolism may be closely associated with immaturity and an underlying hypoplastic lung condition associated with massive hydrops fetalis.

As stated earlier, it is possible to identify the effects of rare complications of pulmonary vascular air embolism with pulmonary hypoplasia in massive hydrops fetalis on a ventilated premature newborn. Most of the time, pulmonary vascular air embolism is ultimately lethal. The presence of pallor, sudden unexplained deterioration and discrepancy between the baby's clinical condition and high readings of a continuous oxygen monitor should alert clinicians to this complication. For now, the best approach is prophylaxis, that is, to minimize barotrauma in the treatment of preterm lung disease.

한 글 요 약

초극소 미숙아에서 발생한 중증 태아 수종을 동반한 폐혈관 공기 색전증 1례

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폐혈관 공기 색전증은 인공 환기 요법으로 치료 받는 신생아에서 드물게 발생하지만 매우 치명적인 합병증이다. 저자들은 특징적인 임상소견, 방사선소견을 보이며, 중증의 태아 수종과 폐형성저하증을 동반한 초극소 미숙아에서 발생한 폐혈관 공기 색전증 1례를 경험하였기에 문헌고찰과 함께 보고하는 바이다.

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