

A Case of Congenital Bronchial Defect Resulting in Massive Posterior Pneumomediastinum : First Case Report

Ji Eun Jeong, M.D.¹, Chi Hoon Bae, M.D.², and Woo Taek Kim, M.D.¹

*Department of pediatrics¹, Department of Thoracic and Cardiovascular Surgery²,
Catholic university of Daegu School of Medicine, Daegu, Korea*

Bronchial defects in neonates are known to occur very rarely as a complication of mechanical ventilation or intubation. This causes persistent air leakage that may form massive pneumomediastinum or pneumothorax, leading to cardiac tamponade or cardiorespiratory deterioration. Early diagnosis and treatment of bronchial defects are essential, as they can be accompanied by underlying severe lung parenchymal diseases, especially in preterm infants. We encountered an extremely low birth weight infant with an air cyst cavity in the posterior mediastinum that displaced the heart anteriorly, thereby causing cardiopulmonary deterioration. During exploratory-thoracotomy, after division of the air cyst wall (mediastinal pleura), we found a small bronchial defect in the posterior side of the right main bronchus. The patient had shown respiratory distress syndrome at birth, and she was managed by constant low positive pressure ventilation using a T-piece resuscitator after gentle intubation. As the peak inspiratory pressure was maintained low throughout and because intubation was successful at the first attempt without any difficulty, we think that the cause of the defect was not barotrauma or airway injury during intubation. The fact that the margin of the defect was very clear also suggested a congenital origin. To our knowledge, this is the first case of congenital bronchial defect in English literature.

Key Words: Bronchial defect, Posterior pneumomediastinum, Congenital air leak syndrome, Extremely low birth weight infants

A bronchial defect that causes a massive air leak is a surgical emergency. Some cases of bronchial defects present in neonates, mainly due to injury from intubation, tracheal suction, or mechanical ventilation.¹⁻⁴ Rarely, bronchial defects may also occur secondary to congenital pulmonary airway malfor-

mation or parenchymal lung disease, but congenital bronchial defect has never been reported before. Air leakage from the bronchial defect can form a retrocardiac pneumomediastinum or paramediastinal air cysts.⁵ Most cases of pneumomediastinum resolve spontaneously, but with bronchial defects, continuous air leakage may lead to hemodynamic compromise, especially cardiac tamponade.^{1-3, 5} If a bronchial defect is suspected, early diagnosis and aggressive intervention is required.

Received: 11 September 2015

Revised: 21 September 2015

Accepted: 22 September 2015

Correspondence to: Woo Taek Kim, M.D.

42472, 33, Duryugongwon-ro 17-gil, Nam-gu, Daegu, Korea
Division of Neonatology, Department of Pediatrics, School of Medicine, Catholic University of Daegu, Daegu 42472, Korea
Tel: +82-53-650-4250, **Fax:** +82-53-622-4240

E-mail: woodykim@hanmail.net

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The Korean Journal of Perinatology · pISSN 1229-2605 eISSN 2289-0432 · e-kjp.org

Case report

A female baby was born at 26 weeks 5 days gestation via cesarean section for preterm labor, and her

birth weight was 980 g. The pregnancy was a result of in vitro fertilization, and the mother was hospitalized for vaginal spotting with amniotic membrane bulging and cervical dilatation from 23 weeks gestation until delivery. The mother was 38 years old who had no significant medical or family history. After birth, the baby showed weak respiration with cyanosis and her heart beat was less than 100 beats per minute (bpm). She received positive pressure ventilation with a T-piece resuscitator, which was set at 15 cmH₂O of peak inspiratory pressure (PIP) and 5 cmH₂O of positive end expiratory pressure. She was successfully intubated at the first attempt without any difficulty. The diameter of endotracheal tube (E-tube) was 2.5 Fr, and which distance from upper lip was 6.5 cm. After surfactant administration, she was mechanically ventilated at 17 cmH₂O of PIP lowered to 13 cmH₂O.



Fig. 1. Cylindrical air shadow is seen in the mediastinum after the birth.

An infantogram taken 20 minutes after birth showed a ground glass appearance of the lung field with abnormal mediastinal air (Fig. 1). The air formed a septated cyst that enlarged progressively, but the patient remained hemodynamically stable (Fig. 2). Two days after birth, her condition deteriorated with tachypnea and chest wall retraction. Sinus tachycardia developed more than 200 bpm, and respiratory acidosis occurred. More oxygen and ventilation with high frequency oscillation were required to maintain proper oxygenation. Chest computed tomography (CT) showed a large air-cystic cavity with internal septation in the posterior mediastinum. The tracheo-bronchial tree and the esophagus were displaced anterior to the cyst (Fig. 3). The heart was also displaced anteriorly by the cyst, but there were no signs of cardiac tamponade or pneumopericardium on transthoracic echocardiography. Although appropriate supportive treatment was administered, her medical condition deteriorated. Exploratory-thora-



Fig. 2. Leaked air formed large cyst in retrocardiac area on 2nd day of life.

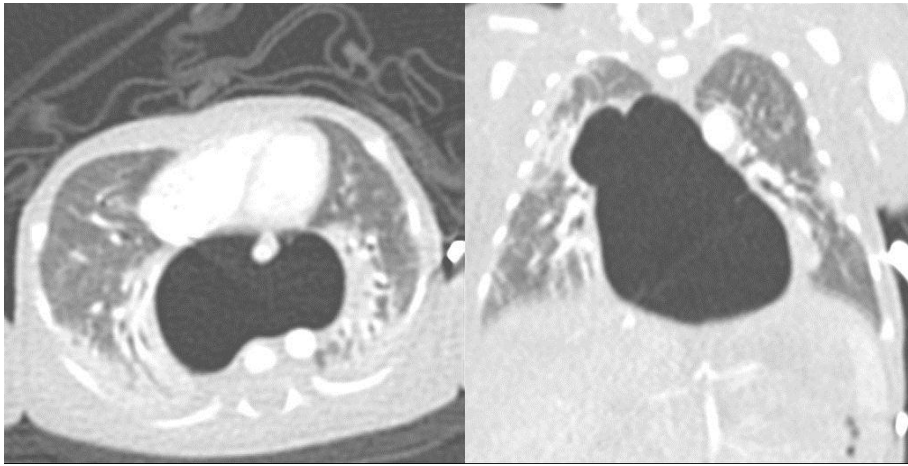


Fig. 3. The septated air cyst compresses the heart and the esophagus is displaced.

cotomy was performed. She was placed in the lateral decubitus position, a posterolateral thoracotomy incision was made, and the thorax was entered through the 5th intercostal space. A thin-walled clear cystic lesion was observed (Fig. 4). The cystic wall was found to be extended mediastinal pleura, that was pseudo-cyst. On dividing the pleura, continuous air leakage was found from a posterior side of the right main bronchus. There was a focal defect which diameter was more than 1 mm. The defect was oval shape and the margin was very smooth and clear. It was primarily closed with a 5-0 PDS suture, and covered with a fibrinogen-based collagen fleece (TachoComb). The thorax was then closed layer by layer with a chest tube in place. Immediately after surgery, the air cyst was shown to have disappeared on chest radiography, and the baby's clinical condition was stable. The chest tube was removed the day after the surgery, and extubation was performed on the 8th postoperative day.

Discussion

Air leak syndrome is defined as the phenomenon in which air escapes from the tracheo-bronchial tree

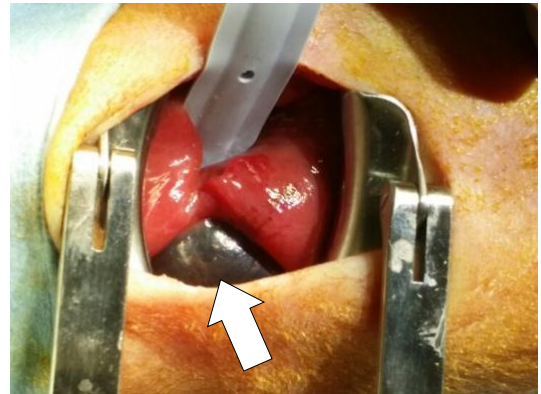


Fig. 4. The arrow indicates clear cystic wall, which is composed by mediastinal pleura.

and collects in various body spaces where it is not normally present.⁶ It occurs more frequently in the newborn period owing to the immature lungs. The incidence of air leaks in newborns is inversely related to birth weight and gestational age.⁷ Immature lungs require higher airway pressure for proper ventilation, which causes barotrauma. Extremely low birth weight infants are especially more exposed to processes such as intubation or the use of a suction catheter, which increase the risk of air leaks occurring.

According to Cagle et al,⁷ pneumomediastinum is the third most common air leak syndrome in newborn after pneumothorax and pulmonary interstitial em-

physema. That is often associated with other air leak syndromes, but sometimes can occur spontaneously.⁸⁻¹¹ Leaked air gathers at the hilum and is deposited usually between the thymus and heart, but may also enter the retrocardiac area, causing posterior pneumomediastinum. In the past, this has also been termed infra-azygos pneumomediastinum, paramediastinal cyst, pneumatocele, and pulmonary ligament air, among other terms, based on chest radiography findings. However, as the diagnosis can now be made accurately on CT scans, these terms are disused.¹² The diagnosis of posterior pneumomediastinum is more difficult than that of anterior pneumomediastinum, which has specific features such as the 'spinnaker sign'.¹³ Furthermore, leaked air can compress the heart and spread more to other sites such as the pericardium, peritoneum, or subcutaneous tissue of the chest and neck. Therefore, posterior pneumomediastinum presents more complications and a higher mortality rate.^{12, 13} Lateral radiography and CT are helpful for diagnosis and echocardiography is needed for the assessment of cardiac function. Posterior pneumomediastinum is more commonly linked to iatrogenic bronchial defects or lacerations than pulmonary parenchymal disease since the introduction of surfactants.^{2, 12} All bronchial defects of neonates reported to date were due to barotraumas or difficult intubations.¹⁻⁴ Unlike previously reported cases, it is considered to be our patient did not received barotrauma or intubation-associated injury. Although she had respiratory distress syndrome, we avoided over-distension or unintentional high pressures using the T-piece resuscitator, rather than a bag valve mask. If the bronchus was torn by the E-tube or stylet, the defect was formed in the anterior or inferior site. It is difficult to tear the posterior bronchus during intubation, because we

use a slightly curved E- tube upward. We decided that exploratory-thoracotomy was needed because serial chest radiography showed an increased air cyst or pneumomediastinum, and further anterior displacement of the heart. Hemodynamics and respiration were slowly deteriorated. We considered that placing a drainage catheter in the cyst cavity could have caused air-hunger sign and inadequate ventilation, because the defect was large and the air leakage was massive.

Said et al.¹ reported a similar case to ours. They presented a baby with an air cyst on chest radiography immediately after birth, who showed deterioration of cardiovascular function including tachycardia. They found a small bronchial defect in the bronchus intermedius on thoracotomy. The baby was stabilized and no further air leakage was found after the closure of the defect. They thought that this defect was due to bronchial injury. However, considering that the baby was full term, did not have other lung diseases, and that there were no problems during intubation, congenital bronchial defect may be considered in that case. Purohit et al.³ reported another case, in which they found pneumomediastinum and pneumothorax on chest x-ray radiography four hours after birth, in a premature baby. They also found a bronchial defect near the bronchus intermedius on tracheogram. They did not perform a bronchoscopy or surgery, and therefore the shape and margins of the defect could not be described. Unlike our case, the baby had many other problems such as Rh incompatibility, high PIPs of up to 25 cmH₂O, and tension pneumothorax. They thought that the cause of the bronchial injury was multifactorial, including congenital weakness of the bronchial wall at the tearing site. They suggested that even though intubation was easy, they may have torn the right bronchus inadvertently. It is notable that the

baby improved after chest tube insertion only instead of surgery. To verify the shape or the margins of the defect is a significant factor in finding the cause.

Newborns, especially preterm infants, who are on ventilator in the incubator for suspected pneumomediastinum should be treated medically at first. Most of them respond well to conservative treatment.⁷ However, as preterm infants often have multiple underlying diseases and complications, they could have unstable hemodynamics anyway in the early days of life. It is difficult to use CT or other diagnostic tools such as bronchoscopy or tracheogram in them. Nonetheless, accurate diagnosis and early intervention is mandatory for saving the lives of these infants.

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