

## Congenital hernia of the lung through the azygoesophageal recess

Young Seok Choi, M.D., Young Jun Son, M.D., Si Young Bae, M.D.  
 Kyung Sun Min, M.D., Young Kuk Cho, M.D., Woo Yeon Choi, M.D.  
 Young Youn Choi, M.D., Jae Sook Ma, M.D. and Tai Ju Hwang, M.D.

Department of Pediatrics, Chonnam National University Medical School, Gwangju, Korea

### = Abstract =

A lung hernia, defined as the protrusion of pulmonary tissue and pleural membranes through a defect in the thoracic wall, is a rare event. It can be congenital or acquired, and cervical, thoracic, or diaphragmatic in location. We report the rare occurrence of a congenital atraumatic lung herniation through the azygoesophageal recess. An 8-month-old male infant, who was born at 35 weeks gestation, had a chronic cough. Chest radiography showed haziness at the right lower lobe of the lung (RLL). Chest computed tomography (CT) revealed herniation of the RLL through the azygoesophageal recess. If persistent unilateral haziness is observed on chest radiography, the possibility of lung herniation should be considered. (*Korean J Pediatr* 2008;51:1123-1126)

**Key Words :** Lung hernia, Azygoesophageal recess, Children

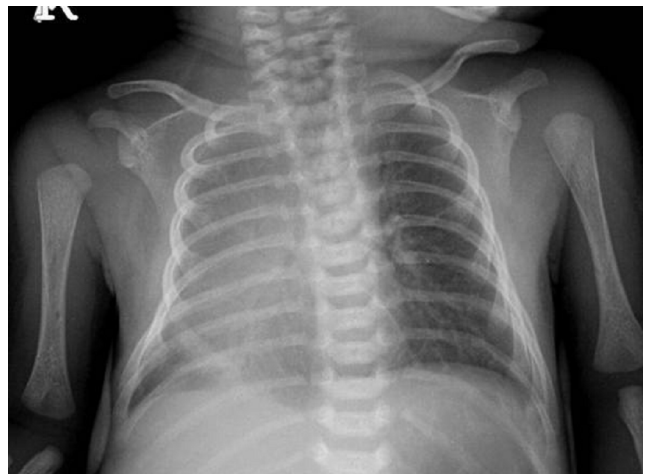
### Introduction

A lung hernia is a rare entity it is defined as the protrusion of the pulmonary tissue and pleural membranes through a defect in the thoracic wall<sup>1)</sup>. It can be congenital or acquired, and cervical, thoracic, or diaphragmatic in location. Lung hernias are most commonly identified following trauma they can also occur spontaneously after a bout of coughing<sup>2, 3)</sup>. We report the rare occurrence of a congenital atraumatic lung herniation through the azygoesophageal recess (AER) in an 8-month-old infant.

### Case report

An 8-month old boy was admitted to the Chonnam national university hospital (CNUH) due to a four-month history of cough. The birth history was significant for admission to CNUH for prematurity and low birth weight after an emergency Cesarean section birth due to an incompetent internal cervical os and preterm labor. The birth

weight was 2,030 g and the gestational age 35 weeks. The Apgar scores were 9 and 10 at 1 and 5 minutes. Tachypnea (60 breaths/min) and chest retractions were observed. The blood pressure and heart rate were in normal range. The arterial blood gas showed a pH of 7.35, a pCO<sub>2</sub> of 33 mmHg and a pO<sub>2</sub> of 89 mmHg. On the chest X-ray, the cardiac silhouette was found to be predominant in the right hemithorax (Fig. 1). The echocardiogram revealed dextro-position of the heart, a small muscular ventricular septal defect and mild left pulmonary artery stenosis. With the diagnosis of transient tachypnea of the newborn, the baby



**Fig. 1.** Chest radiograph shows right lung haziness and dextroposition of the heart at birth.

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Address for correspondence : Jae Sook Ma, M.D.

Department of Pediatrics, College of Medicine Dong-A University

Dong-gu Hak-dong 8, Gwangju 501-757, Korea

Tel : +82.62-220-6646, Fax : +82.62-222-6103

Email : cardiol@jnu.ac.kr

was treated conservatively and the symptoms improved. The infant was admitted to the neonatal intensive care unit for 21 days. When the weight increased to 2,300 g he was discharged. Four months later, he was admitted to CNUH due to acute bronchiolitis. After conservative treatment, he was discharged. Following the discharge, he had a chronic cough.

On physical examination, the blood pressure, heart rate and respiratory rate were within normal range; however, the baby had an increased temperature (38.8°C). The chest X-ray revealed haziness at the RLL (Fig. 2). The day after admission, a chest computed tomography (CT) scan revealed herniation of the RLL into the left chest through the azygoesophageal recess. This resulted in a decreased right thoracic volume and a minimal subsegmental atelectasis in the RLL (Fig. 3). There was no history of trauma to the thorax. After conservative treatment, the cough improved but the haziness in the RLL on the X-ray remained. The patient was discharged.

The patient was readmitted to CNUH because of pneumonia in both lower lung fields at 1 year 6 months of age. The chest CT scan showed pneumonia in both lower lung fields and subsegmental atelectasis in the RLL (Fig. 4). The lung hernia through azygoesophageal recess did not change. After conservative treatment, the patient was discharged. The follow up through 2 years and 9 months was without additional problems.



**Fig. 2.** Chest radiograph shows right lung haziness and dextroposition of the heart at 8 months.

## Discussion

A lung hernia is a rare clinical finding. Roland, first described herniation of pulmonary tissue in 1499<sup>4)</sup>. Maurer and Bladess<sup>5)</sup> established the definition of lung herniation in 1946 as "a protrusion of the pleural-covered lung beyond its normal boundaries through an abnormal opening in the thoracic enclosure"<sup>5)</sup>. Morel-Lavalée<sup>6)</sup> categorized hernias of the lung according to their site and etiology<sup>6)</sup>. To date, hundreds of cases of lung hernias have been reported, 82% have been reported as acquired and 18% congenital<sup>2)</sup>.



**Fig. 3.** Chest CT scan at 8 months shows herniation of the right lower lung into the left through the azygoesophageal recess (arrows), resulting in decreased right thoracic volume and minimal subsegmental atelectasis in the right lower lung.



**Fig. 4.** Chest CT scan at 1 year, 6 months shows persistent herniation of the right lower lung into the left through the azygoesophageal recess and subsegmental atelectasis in the right lower lung and pneumonia in both lower lobes.

Acquired hernias are classified as spontaneous in 30% and traumatic in 52% according to underlying disease. The location of 65-83% of all lung hernias is thoracic and the remaining 17-35% cervical<sup>6,7)</sup>.

Spontaneous hernias occur in both the thoracic and cervical regions and are associated with an abnormal increase in intrathoracic pressure associated with straining, coughing or heavy lifting in addition to a weakness in the thoracic wall. A spontaneous pulmonary hernia has recently been reported in Recklinghausen disease (NF-1), associated with osseous intercostal abnormalities<sup>8)</sup>. Spontaneous thoracic wall hernias are more common in patients with chronic obstructive airway disease, presumably because of chronic coughing and hyperinflation, perhaps combined with long-term steroid administration<sup>9)</sup>. It is likely that a proportion of cases goes unnoticed or gives rise to only minimal symptoms that do not lead to medical consultation, and therefore the true incidence of the condition is difficult to estimate. This could explain why most pulmonary hernias reported in the literature are not spontaneous but are related to various traumatic conditions.

A lung hernia usually presents with few symptoms. A bulging, spongy, crepitant mass can be palpated if it protrudes from the chest wall. The chest radiograph may or may not demonstrate a lung hernia. However, a CT scan is diagnostic. As this condition is rare, Jacka et al.<sup>10)</sup> and Reardon et al.<sup>11)</sup> did not perform CT scan as part of their evaluation<sup>10,11)</sup>. CT scan of the chest not only provides the dimensions of the hernia, information about the thoracic cage and pleural space, but also helps rule out neoplastic or inflammatory causes<sup>12)</sup>. CT scan may also be helpful in differentiating other chest wall conditions, such as traumatic subcutaneous emphysema. In general, however, lung hernias are asymptomatic. Although spontaneous regression is uncommon, conservative management is the treatment of choice. Surgical repair is indicated when incarceration, pain, recurrent infection, and airway obstruction occur<sup>13)</sup>.

The AER is described in anatomic texts as the right retroesophageal pouch or the "space of Holzkecht"<sup>14)</sup>. The AER is the space within the mediastinum that extends from the azygos arch caudally to the aortic hiatus and right hemidiaphragm. This space extends across the spine a variable distance it is bordered anteriorly and medially by the esophagus, the azygos vein and left atrium. Heitzman reported that the AER represents the intrusion of the medial part of the right lower lobe of the lung, the crista pulmonis,

into the mediastinum<sup>14)</sup>. The configuration of this recess can be used as an indicator of disease in the mediastinum<sup>15,16)</sup>.

Recognizing a lung herniation is important for the pediatrician, who is often the first medical contact in patients with this abnormality. Identifying a lung herniation may help patients avoid invasive treatment, an unnecessary workup and therapy that is not beneficial and possibly harmful. There is no prior case reported of lung herniation through the azygoesophageal recess. In our patient, the chest radiograph did not clearly show the lung tissue herniation through the azygoesophageal recess. CT scan was performed to evaluate the lung and confirmed the diagnosis. This case illustrates that a persistent unilateral haziness detected on a chest X-ray, should be followed up by a chest CT scan to rule out the possibility of lung herniation.

**한 글 요약**

**Azygoesophageal recess를 통한 선천 폐탈장**

전남대학교 의과대학 소아과학교실

최영석 · 손영준 · 배시영 · 민경선 · 조영국  
최우연 · 최영륜 · 마재숙 · 황태주

폐조직과 흉막의 흉곽의 결손 부위를 통한 돌출로 정의되는 폐탈장은 선천성 또는 후천적일수도 있으며 부위별로 경부, 흉부, 횡경막 탈장이 있다. 8개월된 남아가 4개월간의 지속적인 기침과 흉부 방사선 촬영에서 폐우하엽의 지속적인 음영 증가소견 있어 시행한 흉부 전산화 단층촬영에서 azygoesophageal recess를 통한 폐탈장이 관찰되었다. 아직까지 보고된 적이 없는 azygoesophageal recess를 통한 폐탈장이 저자들에 관찰하게 되어 보고하는 바이다.

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