



# A Pulmonary Sequestered Segment with an Aberrant Pulmonary Arterial Supply: A Case of Unique Anomaly

Minchul Kim, MD, Jeong Joo Woo, MD, PhD, Jin Kyung An, MD, PhD, Yoon Young Jung, MD, PhD, Yun Sun Choi, MD, PhD

All authors: Department of Radiology, Eulji Hospital, Eulji University, Seoul 01830, Korea

We presented a rare case of a 64-year-old man with a combined anomaly of the bronchus and pulmonary artery that was detected incidentally. Computed tomography showed a hyperlucent, aerated sequestered segment of the right lower lung with an independent ectopic bronchus, which had no connection to the other airway. The affected segment was supplied by its own aberrant pulmonary artery branch from the right pulmonary trunk. This anomaly cannot be classified with any of the previously reported anomalies.

**Index terms:** *Computed tomography; Airway anomaly; Aberrant pulmonary artery*

## INTRODUCTION

Pulmonary developmental anomalies are considered to span a continuum, ranging from an abnormal lung containing normal vessels to a normal lung containing abnormal vessels (1). Although many cases of pulmonary developmental anomalies have the classic features of a single anomaly, other cases have features of two or more anomalies. We encountered a unique case of a patient with segment of a hyperlucent lung with no normal bronchial connection and an aberrant pulmonary arterial supply, which does not match any other previously reported category. This case report was approved by our Institutional Review Board and the requirement for the patient's informed consent was waived.

Received November 21, 2015; accepted after revision December 28, 2015.

**Corresponding author:** Jeong Joo Woo, MD, PhD, Department of Radiology, Eulji Hospital, Eulji University, 68 Hangeulbiseok-ro, Nowon-gu, Seoul 01830, Korea.

• Tel: (822) 970-8290 • Fax: (822) 970-8346  
• E-mail: jjblue@eulji.ac.kr

This is an Open Access article distributed under the terms of the Creative Commons Attribution Non-Commercial License (<http://creativecommons.org/licenses/by-nc/3.0>) which permits unrestricted non-commercial use, distribution, and reproduction in any medium, provided the original work is properly cited.

## CASE REPORT

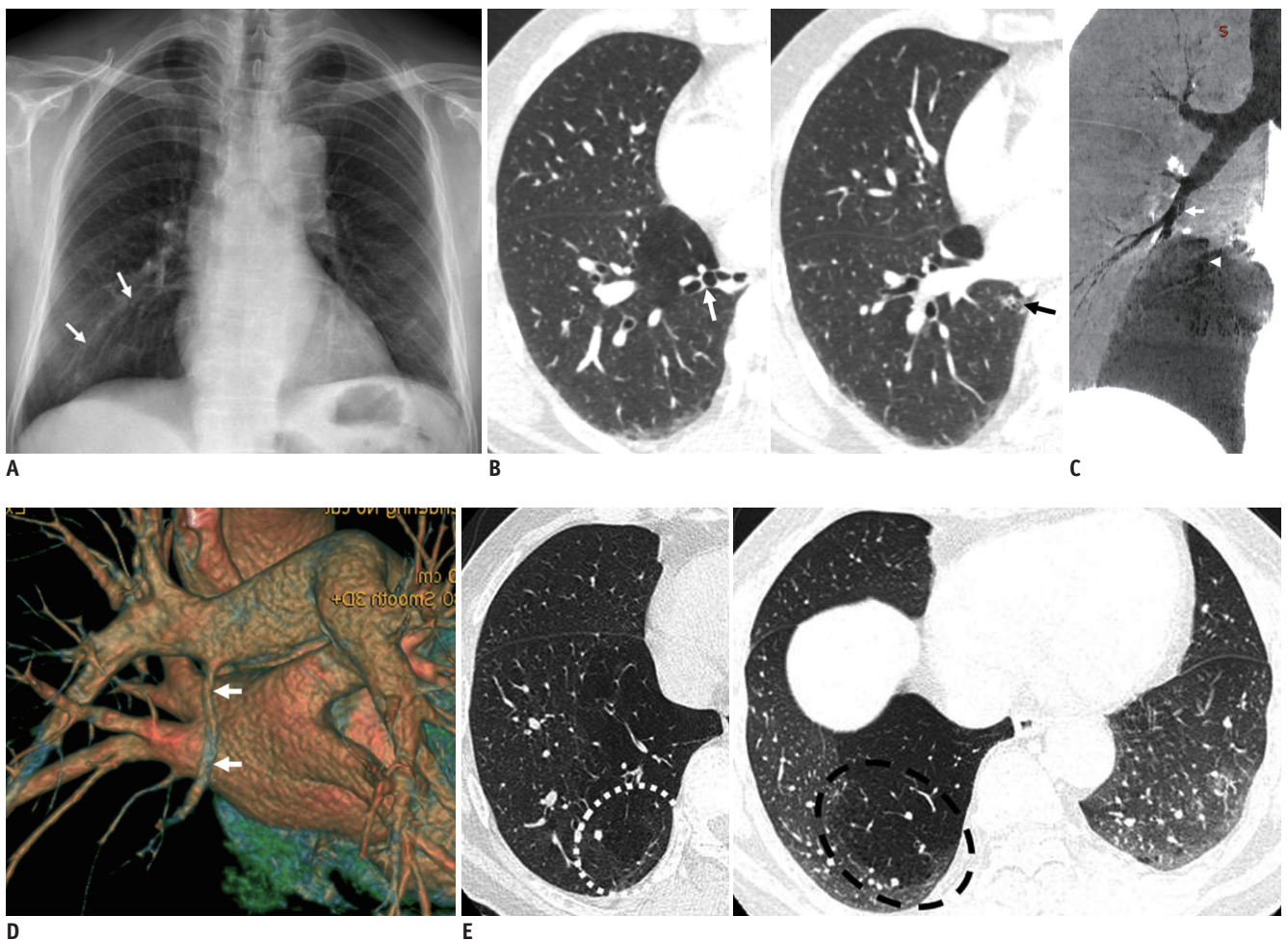
A 64-year-old man who was experiencing mild chest discomfort visited our hospital. His physical examination was unremarkable and there was no significant medical history. He underwent routine chest radiography (Fig. 1A) that showed an area of hyperlucency at the right lower lung zone, but no air-containing cysts or any other abnormal opacity.

The multidetector computed tomography scan was performed on a 64-slice CT system (Discovery CT 750HD; GE Healthcare, Milwaukee, WI, USA). Precontrast and postcontrast CT scans were performed with iodinated non-ionic contrast, 90 cc/3 cc/s, injected using a pressure injector. Then, a series of inspiratory and expiratory high-resolution CT (HRCT) scans were obtained at 0.6-mm collimation and 1-cm intervals. CT showed an area of increased lucency at the right lower medial lung, and a tortuous, ectopic bronchus with bronchiectasis was identified within the hyperlucent area (Fig. 1B); furthermore, communication between the ectopic bronchus and the right bronchial tree was absent, and the right medial basal segmental bronchus was obliterated (Fig. 1C). The ectopic bronchus communicated with no other structures such as the esophagus or mediastinum, but

showed communication with a peripheral bronchiolectasia that ran upward and had a bubbly appearance (Fig. 1B). The mediastinal setting in the CT scan revealed an anomalous pulmonary artery arising from the posterior inferior aspect of the right main pulmonary artery and supplying the affected region. A three-dimensional volume-rendering image was reconstructed to properly demonstrate the abnormal pulmonary artery (Fig. 1D). The segmental vein drained into the right pulmonary vein.

Expiratory HRCT scans were obtained in an attempt to show air trapping within the abnormally lucent lung. These images showed numerous, moderately thin-walled,

air-containing cysts, 0.5 to 1.0 cm in diameter that were mainly located in the peripheral zone of the hyperlucent segment (Fig. 1E). Several areas within the lucent segment that contained small cysts showed an increase in attenuation density during exhalation, but this increase had a delayed rate and a lesser degree, than the density increase seen in the normal contralateral lung (Fig. 1E). The patient's symptoms improved and he refused to undergo further evaluation.



**Fig. 1. Pulmonary sequestered segment with aberrant pulmonary arterial supply in 64-year-old man.**

**A.** 64-year-old man with hyperlucent right lower lobe (arrows). **B.** Lung window CT at full inspiration shows hyperlucent abnormal lung parenchyma located in lower medial zone of right lower lobe. White arrow (image on left) indicates tortuous bronchiectatic bronchus and black arrow (image on right) indicates peripheral bronchiolectasia with bubbly appearance, connected to ectopic bronchus. **C.** Oblique coronal minimal intensity projection reconstruction shows proximal obliteration of right lower lobe segmental bronchus (arrow) and no visible communication between bronchial tree and ectopic bronchus (arrowhead) in hyperlucent lung. **D.** Three-dimensional volume rendering image shows anomalous segmental pulmonary artery (arrows) arising from posteroinferior aspect of right pulmonary trunk. **E.** HRCT taken in inspiratory (image on left) and expiratory (image on right) phases. There are multiple small cyst-like or emphysematous lesions (in dotted line) in peripheral zone of affected hyperlucent segment. In expiratory phase HRCT, airtrapping in affected area is prominent, in comparison with normal contralateral lung. HRCT = high-resolution CT

## DISCUSSION

The two major components of this case were 1) hyperlucent aeration of a sequestered segment with an independent ectopic bronchus that had no connection with the other bronchus or organs such as the esophagus and 2) an aberrant pulmonary artery from the right pulmonary trunk supplying the involved lung segment. These findings indicated a continuum of maldevelopment involving the pulmonary parenchyma or the pulmonary vessels, or a combination of both; this spectrum has been called the "sequestration spectrum" (1). Thus, this case represents a spectrum of congenital bronchopulmonary malformations that has not been reported previously.

In the present case, thin section CT revealed proximal obliteration of the medial basal segment of the right lung, which is a feature of bronchial atresia. However, there was no evidence of mucocele or bronchocele at the affected segment, as is frequently seen in bronchial atresia; thus, this case was distinguished from bronchial atresia. A tortuous, bronchiectatic ectopic segmental airway was identified within the affected segment. Similar findings were reported in a pulmonary sequestration report (2).

On lung window images, CT clearly showed that the medial basal segment of lung lacked normal communication with the tracheobronchial tree or any adjacent organ or mediastinum. However, the manifested segment appeared as an area of hyperlucency that remained at the end of forced expiration, confirming air trapping. The mechanisms by which the segment was aerated are unclear. Stern et al. (3) suggested that aerated pulmonary sequestration is caused by collateral pathways between the medial basal segment and the right lung through pores of Kohn that are probably located at the level of the lung parenchyma, as might also be applicable to the current case. In a review on the CT appearances of cases with aerated bronchopulmonary sequestration (4), the presence of lucent or low-attenuation areas of "emphysema" is emphasized. The emphysema in patients with sequestration is suggested to result from collateral ventilation and air trapping (5). In our case, the emphysematous small cystic feature within the involved segment is clearly seen on HRCT, which can be a potential collateral aeration channel. The location of emphysema or aircysts, which were found mainly at the peripheral zone and adjacent to the normal lung, also supports this idea. Furthermore, the ectopic bronchus is possibly aerated by a small airway connected to peripheral bronchiolectasia,

as in our case. On CT, the ectopic bronchus, ran upward and tapered into bronchiolectasia with an unusual bubbly appearance. This unexpected finding could be a clue to collateral aeration channel in addition to the peripheral collateral ventilation through pores of Kohn.

This patient also showed an aberrant pulmonary artery from the right pulmonary trunk supplying the involved lung segment. This is a discrepant feature, since most vascular anomalies in the "sequestration spectrum" are supplied by aberrant systemic arteries. Moreover, in reviewing 524 lungs or lobes of resection performed on 426 patients, Cory and Valentine (6) described no variation of the pulmonary artery in the primary lung hilum. However, our case showed early branching of a segmental pulmonary artery before the right main pulmonary trunk entered the lung hilum. A similar vascular abnormality has not been reported previously, to the best of our knowledge. If overlooked, anatomical variations of the lung vessels may be important obstacles during lung surgery. This case suggested pulmonary artery abnormality at the hilar level. Moreover, in early fetal development, the airways appear to act as a template for pulmonary artery development and central pulmonary artery sprouting or angiogenesis for up to approximately seven generations (counting the artery to each lung as the first generation) (7, 8). The findings in our case were suggestive of a unique combination of congenital airway and vascular anomalies, rather than an acquired lesion.

In summary, we presented a case of combined anomalies of a hyperlucent lung segment supplied by ectopic bronchus and an aberrant pulmonary artery. Our patient is currently receiving regular expectant follow-up, with particular attention to infection or impaired pulmonary function.

### Acknowledgments

The authors would like to express their appreciation to Bong-eun Cha, radiology technician at Eulji hospital, who contributed to CT image reconstruction.

### REFERENCES

1. Panicek DM, Heitzman ER, Randall PA, Groskin SA, Chew FS, Lane EJ Jr, et al. The continuum of pulmonary developmental anomalies. *Radiographics* 1987;7:747-772
2. Ferretti GR, Jouvan FB, Coulomb M. MDCT demonstration of intralobar pulmonary sequestration of the right upper lobe in an adult. *AJR Am J Roentgenol* 2005;185:1663-1664
3. Stern EJ, Webb WR, Warnock ML, Salmon CJ. Bronchopulmonary sequestration: dynamic, ultrafast, high-resolution CT evidence of air trapping. *AJR Am J Roentgenol*

- 1991;157:947-949
4. Ikezoe J, Murayama S, Godwin JD, Done SL, Verschakelen JA. Bronchopulmonary sequestration: CT assessment. *Radiology* 1990;176:375-379
  5. Culiner MM, Wall CA. Collateral ventilation in "Intralobar pummonary sequestration." Report of a case. *Dis Chest* 1965;47:118-122
  6. Cory RA, Valentine EJ. Varying patterns of the lobar branches of the pulmonary artery. A study of 524 lungs and lobes seen at operation of 426 patients. *Thorax* 1959;14:267-280
  7. deMello DE, Sawyer D, Galvin N, Reid LM. Early fetal development of lung vasculature. *Am J Respir Cell Mol Biol* 1997;16:568-581
  8. Hall SM, Hislop AA, Pierce CM, Haworth SG. Prenatal origins of human intrapulmonary arteries: formation and smooth muscle maturation. *Am J Respir Cell Mol Biol* 2000;23:194-203