Ann Thorac Cardiovasc Surg 2012; 18: 560–563

doi: 10.5761/atcs.cr.11.01793

Case Report

Intralober Pulmonary Sequestration with Arterial Supply from Two Different Origins: A Case Report

Ersin Sukru Erden, MD,[1] Tulin Durgun Yetim, MD,[2] Ali Balci, MD,[3] Adnan Burak Akcay, MD,[4] Sibel Hakverdi, MD,[5] and Mesut Demirkose, MD[1]

Pulmonary sequestration is a rare anomaly, which does not have a connection with the bronchial system and gets its blood supply, generally, from the aorta or its branches. Anatomically, two different forms were described: intralobar and extralobar. Although 74% of intralobar pulmonary sequestrations get their blood supply from the descending thoracic aorta, they may get their blood supply from different arteries. Furthermore, there is more than one arterial anomaly in 14.8% of cases. We report an intralobar pulmonary sequestration, in which arterial blood supply is from two different origins (Arcus aorta and celiac trunk). To the best of our knowledge, this is the first case in the literature.

Keywords: congenital abnormalities, bronchopulmonary sequestration, pneumonectomy

Introduction

Pulmonary sequestration is a rare congenital malformation. Arterial blood is generally supplied by the aorta or its branches and does not have a connection with the bronchial system. It accounts for 0.15%–6.4% of all congenital pulmonary anomalies. Anatomically, two different types of pulmonary sequestration were identified; intralobar and extralobar forms. Intralobar pulmonary sequestration shares the same pleura with normal lung tissue, but the extralobar form is separated from normal lung tissue with its own pleura. Intralobar pulmonary sequestration is more common than the extralobar form, and it affects the posterior basal segments of lower lobes and involves the left lung inferior lobe more frequently. Venous drainage in pulmonary sequestration is provided by pulmonary veins, in 95% of the cases. It gets its arterial blood supply from the descending thoracic artery, in 74%, and from the abdominal aorta, in 19%, of cases. It may rarely receive its blood supply from intercostal, subclavian, innominate, internal thoracic, pericardiophrenic, celiac, splenic arteries. Furthermore in 14,8% of cases arterial anomaly is more than one.[1)]

Pulmonary sequestration cases, which receive arterial blood supply from more than one anatomical localization, have been reported in the literature.[2,3)] In this case report, we present an intralobar pulmonary sequestration in the inferior lobe of the right lung in which the arterial blood supply is from two different origins (arcus aorta and celiac trunk), to our knowledge this is the first case in the literature.

Case Report

A 27-year-old nonsmoking female patient was admitted

[1]*Department of Chest Diseases, Mustafa Kemal University Faculty of Medicine, Hatay, Turkey*
[2]*Department of Thoracic Surgery, Mustafa Kemal University Faculty of Medicine, Hatay, Turkey*
[3]*Department of Radiology, Mustafa Kemal University Faculty of Medicine, Hatay, Turkey*
[4]*Department of Cardiology, Mustafa Kemal University Faculty of Medicine, Hatay, Turkey*
[5]*Department of Pathology, Mustafa Kemal University Faculty of Medicine, Hatay, Turkey*

Received: September 15, 2011; Accepted: January 17, 2012
Corresponding author: Ersin Sukru Erden. Mustafa Kemal University, Faculty of Medicine, Department of Chest Diseases, 31000, Hatay/Turkey
Email: dreserden@gmail.com
*This case is presented as poster in Turkish Thoracic Society 14th Annual Congress, 2011.

to the Chest Disease outpatient clinic with complaints of coughing and sputum persisting from childhood. In her history, she defined that she used antibiotic treatment because of recurrent pneumonia attacks. Her body temperature was 36,7°C, pulse rate 82 per minute and blood pressure 80/60 mmHg. In her respiratory system examination, there was inspiratory rales and a decrease in respiratory sounds. There was no abnormality in other system examinations. Laboratory findings showed hemoglobin level 13.5 g/dL, hematocrit level 38.8%, white blood cell count 8500/μL and sedimentation 8 mm/h. Arterial blood gas analysis showed pH, 7.44; PCO_2, 33 mmHg; PO_2, 79 mmHg; HCO_3, 22.4 mEq/L; SaO_2, 96%. In the chest radiography, there was an elevation in the right hemidiaphragm and nonhomogeneous opacity in the medial part of the right lung inferior zone. In her thorax CT there was cystic bronchiectatic regions which shows air-fluid levels in posterobasal and mediobasal segments of right inferior lung lobe, and there was aberrant lung tissue which received arterial blood supply from the arcus aorta and celiac artery and venous drainage by pulmonary veins (**Fig. 1**). Pulmonary CT angiography demonstrated aberrant vascular structures which originates from the arcus aorta and celiac artery and supplies pulmonary sequestration (**Figs. 2** and **3**). The echocardiography was normal.

A right posterolateral thoracotomy was performed due to the diagnosis of pulmonary sequestration in the patient. During exploration, a thin vascular structure, which was entering to the lower lobe over the diaphragm, was observed, a second vascular structure was observed in anterior mediastinal face which was entering to the major fissure. The vessels were ligated and resected, and an inferior lobectomy was performed. In the histopathology, alveolar spaces were filled with hemorrhage and edema. Few thick-walled blood vessels and areas of bronchial dilatation were seen. There were lymphoid aggregates, which indicated chronic inflammation. Spaces were filled with macrophages and amorphous debris. Focal lymphoid aggregates surrounded the cysts. The patient, with normal postoperative follow-up, was cured and discharged.

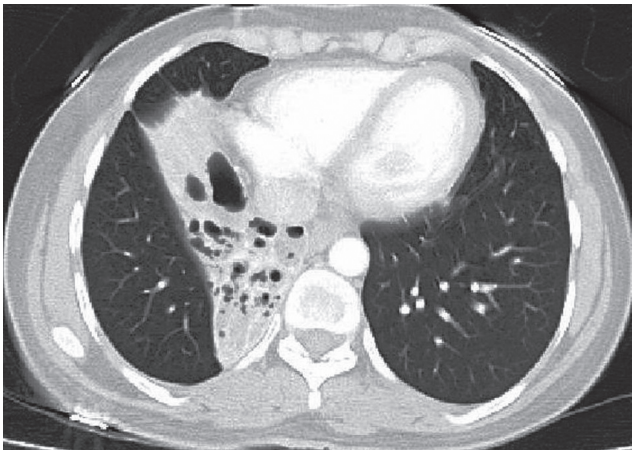

Fig. 1 Transverse reconstructive image of thoracic CT scan shows aberrant lung tissue.

Discussion

In the medical literature, bronchopulmonary sequestration was first described in 1946 as a bronchopulmonary mass or cyst without a bronchopulmonary tree connection, which gets its arterial blood supply from the aorta or one of its branches. Traditionally, pulmonary sequestrations are divided in two forms: intralobar and extralobar. Two thirds are seen in left lower lobe while the others are in right lower lobe.[4] Intralobar pulmonary sequestration accounts for 75% of all pulmonary sequestrations and its association with other congenital anomalies is lower than that for the extralobar form.[1] In our case, no other congenital anomaly was detected.

In the literature, there are theories to explain the pathogenesis of pulmonary sequestration, and Corbett and Humphrey summarized these theories.[5] The widely accepted hypothesis is the development of an accessory bud during development of the normal lung bud. The accessory bud, which develops independently from normal tracheobronchial tree in embryogenesis, receives its arterial blood supply, generally, from aorta.[5] However, in opposition to this theory, a current, more accepted theory suggests that obliterate bronchitis with lower lobe bronchial obstruction, caused by one, or more than one, necrotizing pneumonitis attack, results in pulmonary sequestration.[6] Moreover there were case reports in the literature which showed the congenital origin of the malformation. In the literature; bilateral intralobar sequestration, concomitant intralobar and extralobar sequestration, neonatal congestive heart failure as a result of intralobar sequestration, association of bronchogenic cyst with intralobar sequestration were reported, and it was clear that the systemic arterial blood supply was an embryologic developmental anomaly.[7] In our case, there was an intralobar pulmonary sequestration that received its arterial blood supply from two different and away from each other origin (to our knowledge this is the first case which gets its blood supply from the arcus aorta and celiac trunk), and in our opinion, this supports the hypothesis of the

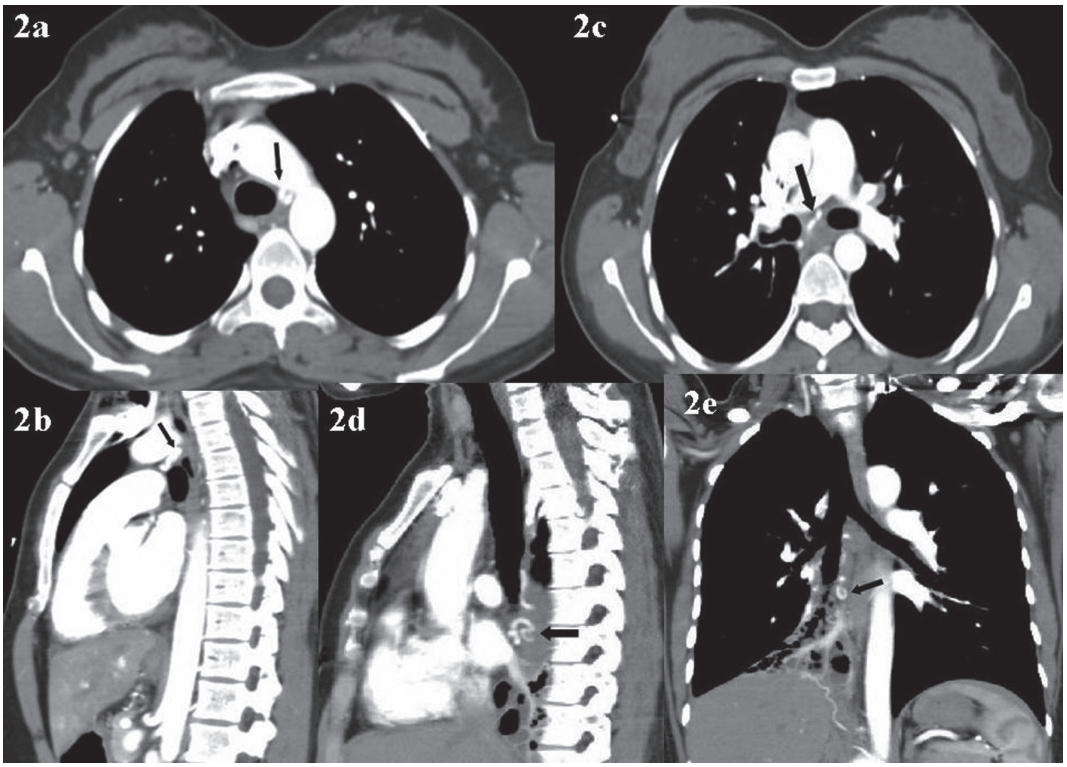

Fig. 2 (**a**) Transverse reconstructive image of thoracic CT scan shows the aberrant vascular structure originating from aortic arc (arrow). (**b**) Sagittal reconstructive image of thoracic CT scan shows the aberrant vascular structure originating from aortic arc (arrow). (**c**) In transverse reconstruction, this vessel extends to the pulmonary sequestration (arrow). (**d**) In sagittal reconstruction, this vessel extends to the pulmonary sequestration (arrow). (**e**) In coronal reconstruction, this vessel extends to the pulmonary sequestration (arrow).

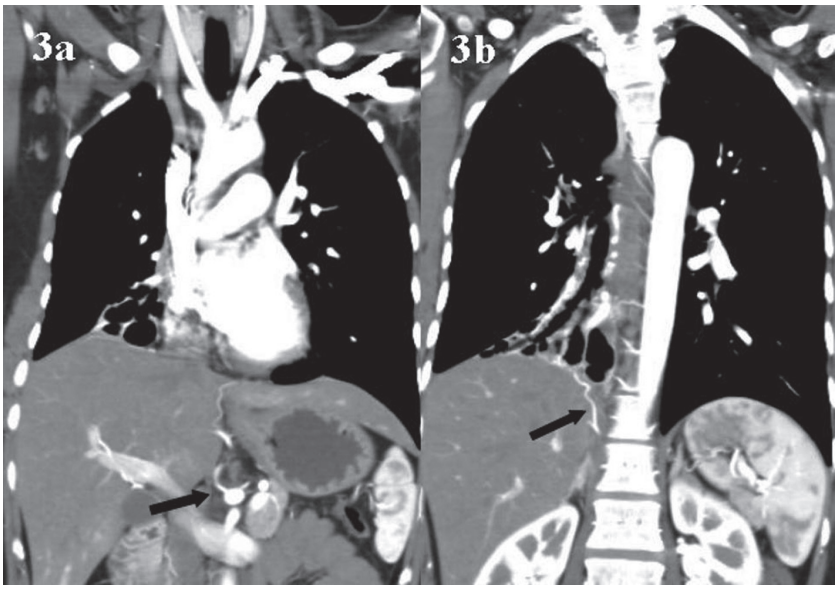

Fig. 3 (**a**) Coronal reconstructive image of thoracic CT scan shows the aberrant vascular structure originating from celiac trunks (arrow). (**b**) In coronal reconstruction, this vessel extends to the pulmonary sequestration (arrow).

congenital origin of the disease.

Typically, there is a history of coughing and mucopurulent sputum in intrapulmonary sequestration cases in early adult life, and there are recurrent pneumonia attacks, resulting from pyogenic bacterias.[7] Savic et al. reported that more than 50% of the cases were symptomatic after the second decade and 15% cases were asymptomatic.[1] Minor hemoptysis is a frequent complaint; however, massive hemoptysis, which threatens life, was also reported. Moreover, heart failure, as a result of left to left shunt in intralobar sequestration, was reported. In the physical examination, signs are usually nonspecific, and there may be findings consistent with infection.[8]

Traditionally, angiography is the gold standard because it demonstrates excellent arterial and venous blood supplies; however, angiography is an invasive procedure, and there is exposure to radiation and it requires anesthesia, in children. After the development of noninvasive techniques for multidetector CT and MR imaging, these noninvasive techniques are being used instead of conventional angiography or subtraction angiography.[8] CT angiography provides the opportunity to evaluate arterial blood supply, venous drainage and changes in lung parenchyma in a single test, and differential diagnosis and preoperative evaluation can also be done.[9] We also evaluated our patient with CT angiography and demonstrated intralobar pulmonary sequestration with arterial blood supply from two different origins and venous drainage by pulmonary veins, and we made the differential diagnosis before the operation. We think that with recent developments in imaging techniques, multislice pulmonary CT angiography is sufficient for the diagnosis of intralobar pulmonary sequestration, imaging of the arterial-venous blood circulation, and differential diagnosis and preoperative evaluation. The choice of treatment in intralobar pulmonary sequestration is surgery.[10] Surgical treatment is not only required for symptomatic cases but is also required for asymptomatic cases because complications such as infection, pulmonary hemorrhage, pulmonary hypertension, congestive heart failure may occur.[11] The standard surgical technique for sequestration is wedge resection or lobectomy by thoracotomy. With advances in technology, video-assisted thoracoscopic surgery was used with success in the surgical management of sequestration with minimal invasiveness.[12,13] In our case, because of recurrent episodes of pneumonia, surgical treatment was considered as the treatment of choice. Lobectomy with thoracotomy and arterial ligation was performed because the arterial blood supply was from two different and away from each other origins.

In conclusion with this case, we report the first intralobar pulmonary sequestration in the literature, which received its arterial blood supply from both the aorta and celiac trunk, which was diagnosed with CT angiography and treated surgically.

References

1) Savic B, Birtel FJ, Tholen W, et al. Lung sequestration: report of seven cases and review of 540 published cases. Thorax 1979; **34**: 96-101.
2) Kanazawa S, Miyake T, Ishida A, et al. Intralobar pulmonary sequestration supplied by multiple anomalous arteries: report of a case. Surg Today 2001; **31**: 701-4.
3) Asai K, Urabe N, Yajima K. Anomalous systemic arterial supply to the basal segments of the left lung with two aberrant arteries. Jpn J Thorac Cardiovasc Surg 2005; **53**: 573-6.
4) Silverman ME, White CS, Ziskind AA. Pulmonary sequestration receiving arterial supply from the left circumflex coronary artery. Chest 1994; **106**: 948-9.
5) Corbett HJ, Humphrey GM. Pulmonary sequestration. Paediatr Respir Rev 2004; **5**: 59-68.
6) Prasad R, Garg R, Verma SK. Intralobar sequestration of lung. Lung India. 2009; **26**: 159-61.
7) Frazier AA, Rosado de Christenson ML, Stocker JT, et al. Intralobar sequestration: radiologic-pathologic correlation. Radiographics 1997; **17**: 725-45.
8) Abbey P, Das CJ, Pangtey GS, et al. Imaging in bronchopulmonary sequestration. J Med Imaging Radiat Oncol 2009; **53**: 22-31.
9) Kang M, Khandelwal N, Ojili V, et al. Multidetector CT angiography in pulmonary sequestration. J Comput Assist Tomogr 2006; **30**: 926-32.
10) Hirai S, Hamanaka Y, Mitsui N, et al. Surgical treatment of infected intralobar pulmonary sequestration: a collective review of patients older than 50 years reported in the literature. Ann Thorac Cardiovasc Surg 2007; **13**: 331-4.
11) Tsunezuka Y, Sato H. Intralobar pulmonary sequestration with three aberrant arteries in a 75-year-old patient. Chest 1998; **114**: 936-8.
12) Avgerinos D, Reyes A, Plantilla E, et al. Video-assisted thoracoscopic surgery for intralobar pulmonary sequestration. Cases J 2008; **1**: 269.
13) Gonzalez D, Garcia J, Fieira E, et al. Video-assisted thoracoscopic lobectomy in the treatment of intralobar pulmonary sequestration. Interact Cardiovasc Thorac Surg 2011; **12**: 77-9.