Pulmonary sequestration diagnosed at unusual age and location

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ABSTRACT

Background: Pulmonary sequestration (PS) is defined as nonfunctional tissue within lung, has separate arterial supply from lung itself and does not have any communication with the tracheobronchial tree. PS is divided into two types which are intralobar pulmonary sequestration (ILS) and extralobar pulmonary sequestration (ELS). PS is a rare congenital malformation of the lungs which is composed of 0.15% to 6.4% of all congenital lung malformation. PS is commonly located in the left lobes and its incidence, especially the incidence of ILS, in the pediatric population is higher than in the adult population. Diagnosis of intralobar pulmonary sequestration which is in the right lower lobe in the third decade of life is a rare situation. We present a case of 37-year-old man diagnosed with ILS located in the right lower lobe which has its own arterial supply directly from descending aorta. This rare case was treated with right lower lobectomy just after the coil embolization of its arterial supply with the help of interventional radiology.

Keywords: Pulmonary sequestration, coil embolisation of pulmonary sequestration.

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Background

Pulmonary sequestration (PS) is a disease in which a part of lung tissue lacks communication with normal tracheobronchial tree and have blood supply from systemic circulation. PS forms about 0.15% to 6.40% of all congenital pulmonary anomalies [1]. PS is considered a congenital anomaly of the lung in which separate tissue has not got any function within the lung parenchyma that does not have any proper bronchial and vascular connections. PS is divided into two varieties according to whether it has its own pleural covering or not. Extralobar pulmonary sequestration has its pleural covering, whereas intralobar pulmonary sequestration is implanted in its surrounding lung tissue [2]. ILS tends to be found in the posterolateral segment of the left lung, whereas extra lobar pulmonary sequestration (ELS) is generally found in lower lobes, mostly in the left costodiaphragmatic sinus [3]. Male:female ratio for PS is 1.58:1. Pulmonary sequestration is mainly located in left lobes. Although PS can be diagnosed at a wide variety of age range, the mean age for the diagnosis of ILS (20±8 years old) is less than ELS (38 ± 9 years old), since ELS is generally asymptomatic [4]. This paper is aimed to present 37-year-old man's ILS which is developed at the unusual location and treated with the remarkable method.

Case Presentation

37-year-old man who presented with recurrent, long-standing cough was referred to our clinic. He stated that he has dark, yellow and green-colored phlegm. He did not describe any chest pain, hemoptysis, dyspnea, or fever at the time of admission. At his previous hospital admission, he was diagnosed with bronchiectasis and sputum culture test was performed. The test result was compatible with pseudomonas aeruginosa so levofloxacin was chosen as a first-line therapy. His complaints were remnant after the treatment. As a result of ineffective antibiotherapy, he was referred to our clinic for further investigation. He does not have any chronic disease. He does not use any drugs on a regular basis. He does not have any malignancies or genetically inherited diseases in his family history. The patient has a history of pneumonia 10 years ago; however, he did not remember the exact treatment for the situation. In his physical examination in our clinic, there was not any outstanding pathological physical examination finding. His complete blood count, urea, creatinine, blood urea nitrogen, serum electrolytes which are sodium, chloride, potassium, bicarbonate; aspartate transaminase, alanine transaminase, direct and indirect bilirubin levels, alpha-1 antitrypsin level, pulmonary function test, blood glucose

level were in the normal range. In our clinic chest, radiography was performed and nodular opacity was noticed at his right lower lobe of the lung. Thorax computed tomography (CT) revealed a well-demarcated, air-filled multicystic mass predominantly in the mediobasal segment of the right lower lobe at the lung window of coronal reformation and axial images (Figure 1a and b). Systemic arterial branches arising from the right lateral wall of descending thoracic aorta (straight arrow) feeding the lesion can be seen in coronal maximum intensity projection (MIP) and volume-rendering reconstruction images (Figures 2 and 3). Right lower lobectomy is decided as definitive therapy. To reduce the risk of uncontrolled bleeding during surgery, detection of the exact location of vascularity was performed with coil embolization of the sequestrated segment's artery with the help of interventional radiology was performed. After that right lower lobectomy with mini thoracotomy. In this manner, a rare of its kind surgery technique, coil embolization backed mini thoracotomy, has succeeded without any complication. Resected lobe was 13 × 11.5 × 8 cm in diameter and consists of dilated, cystic bronchi filled with white purulent secretion. Histopathological investigation of the lesion was compatible with intralobar pulmonary sequestration. During his post-operative follow up, no complication was occurred, and the patient was discharged 6 days after his surgery. After right lower lobectomy, patient's symptoms were regressed. By the time this article was written, 3 months later of patient's surgery, there is not any sign of bronchiectasis, phlegm, or cough.

Discussion

Clinical features of PS include cough, chest pain, having a history of recurrent pneumonia with fever, and hemoptysis. Symptoms of PS can mimic of those pneumonia [4,5]. Pediatric population acquired symptoms more frequently than adult patients. However, some of the cases with pulmonary sequestration diagnosed without any symptoms at routine physical examination or incidentally [4]. Radiologically, nodular opacity can be seen in chest X-ray [6]. Spiral CT and magnetic resonance angiography (MRA) are the imaging method of choice for identifying the aberrant artery and the venous return[7]. Computed tomography is considered enough to see PS as a pulmonary mass, an abnormal structure in the lung with its vascular drainage [10]. In our case, coronal and axial reformation, lung window of CT scan shows a well-demarcated, air-filled multicystic mass predominantly in the medio basal segment of the right lower lobe. Note the arterial supply directly from descending thoracic aorta (Figure 1). Histologic features of PS include normal lung tissue containing proliferation of bronchial structures which remind of congenital adenomatoid malformation. Different amounts of fibrosis and inflammation related cells can be seen [5].

During differential diagnosis procedure, other congenital lung malformations (CLMs) and bronchiectasis should be in consideration. CLMs encompass congenital cystic adenomatoid malformation, bronchial atresia, congenital lobar emphysema, and bronchogenic cyst. Bronchogenic cysts are mucus filled lesions that generally arise from bronchial tree in mediastinal location. Cystic adenomatoid malformations are lesions where atypical development of the distal bronchi-alveolar structures results in a multicystic lung mass. Congenital lobar emphysema exists as overdistension of the pulmonary lobes and is thought to be a result of an abnormality of the bronchial tree resulting in focal bronchomalacia. Airway collapse during expiration results in distal air trapping, focal overdistension, and emphysematous change [8]. Although some of the CLMs have emphysematous changes within the lesion just as

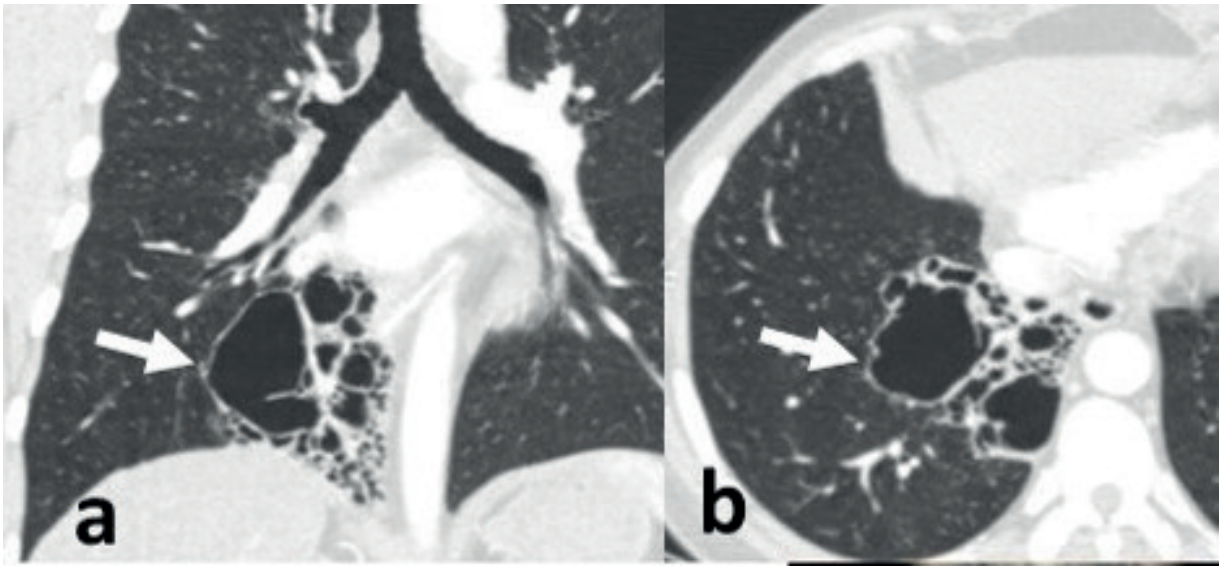

Figure 1. *(a) Coronal reformation and (b) axial lung window of CT scan shows a lobulated area that contains cystic, bronchiectasic fields with decent borders (arrow).*

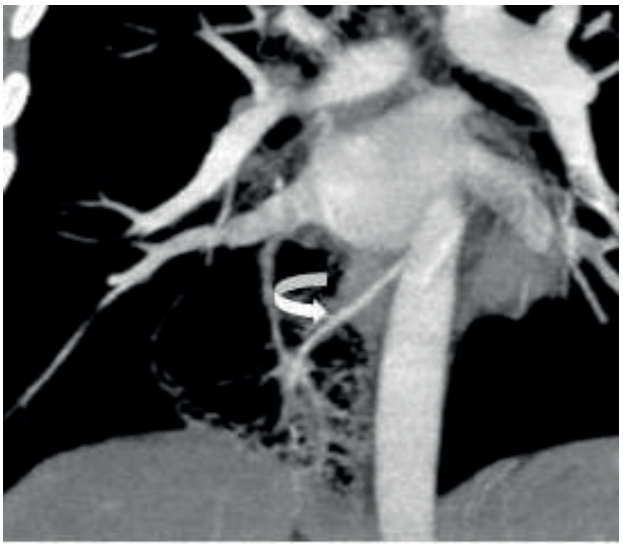

Figure 2. *Coronal MIP image. Note the systemic arterial branches feeding the pulmonary sequestration (straight arrow).*

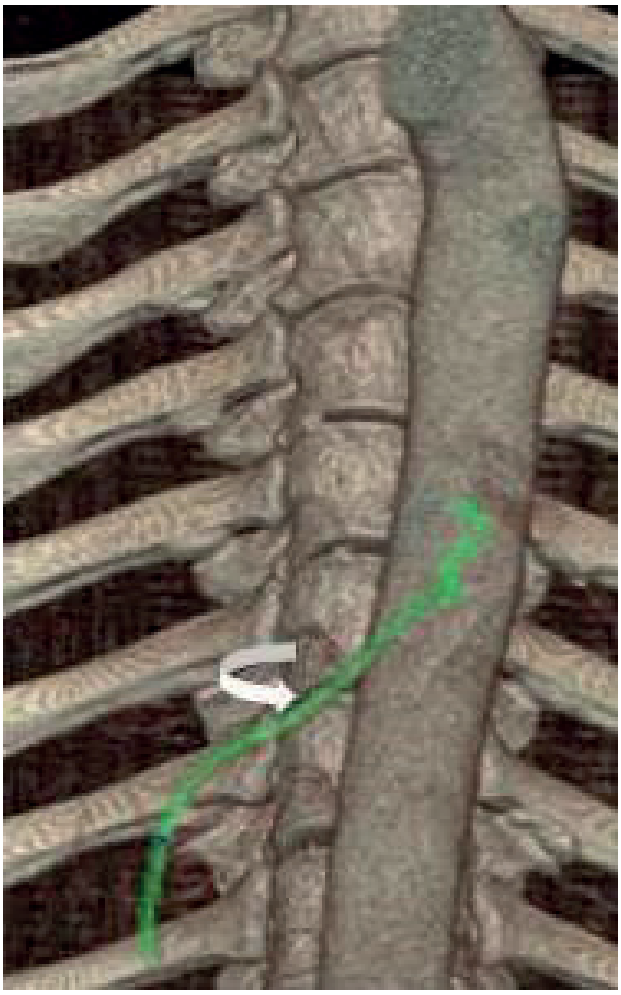

Figure 3. *Volume-rendering (VR) image show the systemic arterial branch arising from the right lateral wall of descending thoracic aorta (straight arrow).*

in pulmonary sequestration, neither bronchiectasis nor CLMs have their own arterial supply arising from systemic circulation separated from lungs.

Treatment options for symptomatic PS include surgical resection of the anomaly. Surgery is recommended as a conclusive treatment for pulmonary malformations since postponed surgery of misdiagnosed condition prompt to recurrent infections and its complications, prolonged hospitalization, and inappropriate treatment. [9].

For clinicians, while assessing patients who are diagnosed and has been followed with bronchiectasis considering pulmonary sequestration as one of the possible preliminary diagnosis leads to early and proper surgical treatment for patients. Our patient, 37-year-old man diagnosed with intralobar pulmonary sequestration at his right lower lobe is a rare case both for age and location if compare the mean age for the diagnosis of ILS (20 ± 8 years old) and ILS tends to be found in posterolateral segment of left lung, whereas ELS is generally found in lower lobes [3,4]. Not only its unusual location and age but also being rare sample of its kind by applying coil embolization to arterial supply of pulmonary sequestration in order to increase surgical safety, accuracy, and ease hemostasis. Our surgical treatment is combining interventional radiology with classical surgery to minimize risk of uncontrolled bleeding during surgery. Moreover, we believe our successful surgical approach to right lower lobe resection with coil embolization to arterial supply of sequestrated segment is about to have unique contribution to current literature and guide to clinicians for the cases resemble with in our clinic.

Conclusion

For followed up young and middle-aged patients with bronchiectasis, sequestration should come into mind at the differential diagnosis stage, that's why contrast-enhanced CT has a significant role for detecting anomalies at the vascular structures. To conclude, with the help of careful history taking and radiological evaluation sequestration could be accurately diagnosed and surgical complications could be prevented by embolization of the vessel which is originating from the aorta.

What is new?

We believe our successful surgical approach to right lower lobe resection with coil embolization to arterial supply of sequestrated segments is about to have a unique contribution to current literature and guide clinicians for the cases that resemble within our clinic.

Conflict of interest

The authors declare that there is no conflict of interest regarding the publication of this article.

Funding

None.

Consent for publication

Written and informed consent was taken from patient to publish this case report.

Ethical approval

Ethical approval is not required at our institution to publish an anonymous case report.

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Summary of the case

1	**Patient details**	Man, 37 years old
2	**Symptoms**	Long-standing cough, Phlegm
3	**Final diagnosis**	Intralobar Pulmonary Sequestration
4	**Medication**	Right Lower Lobectomy after Coil Embolization
5	**Clinical procedures**	Surgery
6	**Clinical specialty**	Thoracic Surgery and Interventional Radiology