A rare case of pulmonary sequestration with three aberrant artery branch supply

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ABSTRACT

Background: Pulmonary sequestration (PS) is a dysfunctional, congenital malformation with no apparent connection to the tracheobronchial tree, fed by an aberrant arterial connection from the systemic circulation. PS is a rare congenital malformation. The fact that very few cases in the literature are supplied by three aberrant arterial branches originating from the aorta makes our case even rarer.

Case Presentation: In our case, we will focus on a 24-year-old male patient who presented with complaints of cough and dyspnea, was investigated due to pneumonia-like symptoms resistant to antibiotic therapy and was diagnosed with intralobar PS (ILS). We performed left lower lobectomy on our patient with a hybrid approach combining video-assisted thoracoscopic surgery (VATS) and thoracotomy and achieved successful results.

Conclusion: ILS cases can be diagnosed late because they show pneumonic symptoms such as cough, chest pain, and dyspnea. PS should be considered in the differential diagnosis of recurrent pneumonia-like infections. The diagnosis should be confirmed by revealing aberrant arterial structures with imaging methods. In the patient's treatment decision, the surgical method should be carefully selected according to the patient's current condition. The hybrid approach allowed us to combine the advantages of commonly used VATS and thoracotomy while minimizing the disadvantages. This rare case, managed through an innovative surgical approach, has not only improved our clinical experience but also contributed meaningfully to the existing literature.

Keywords: Congenital pulmonary malformations, pulmonary sequestration, left lower lobectomy, hybrid thoracotomy, case report.

Type of Article: CASE REPORT **Speciality:** Thoracic Surgery

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Background

Pulmonary sequestration (PS) is a dysfunctional, congenital malformation with no apparent connection to the tracheobronchial tree, supplied by an aberrant arterial connection from the systemic circulation [1]. It provides its arterial supply mostly from the systemic circulation by the aorta and its primary branches, while its venous return is often drained into the pulmonary vein, and to a lesser extent into the azygos and hemizygous vein [2].

There are two subtypes: Extralobar PS (ELS) and intralobar PS (ILS) [3]. If the sequestrated lung tissue is surrounded by the common visceral pleura with the adjacent lung, it is called ILS; if it is surrounded by its separate visceral pleura, it is referred to as ELS [4]. ILS is most commonly seen in the left lung and is more frequently located in the lower lobes [3]. ELS, on the other hand, can be found in both supradiaphragmatic and infradiaphragmatic locations [5].

Arterial supply is mostly provided by a single abnormal artery branch, less often by two branches, rarely by three or more branches [3]. The definitive diagnosis of PS is made by imaging these abnormal arteries [1]. For this purpose, computed tomography angiography (CTA), magnetic resonance angiography (MRA), and digital subtraction angiography can be used as imaging methods [1].

Surgical treatment constitutes the curative treatment of PS [6]. Surgical treatments that can be performed in ILS are often wedge resection, segmentectomy, and lobectomy. Resection can be performed with robotic-assisted thoracic surgery, video-assisted thoracoscopic surgery (VATS), or thoracotomy methods [6,7].

In this study, we aimed to present a case of ILS located in the lower lobe of the left lung, supplied by three arterial branches originating from the thoracic aorta.

Case Presentation

A 24-year-old male patient was admitted to an external center with complaints of severe cough, shortness of breath, fever, fatigue, and muscle pain that had been ongoing for 1 month. Emphysematous areas were detected on the chest X-ray (Figure 1). Empiric treatment was initiated upon high white blood cell and C-reactive protein values in the patient's laboratory tests, and he used amoxicillin-clavulanate for 10 days. Despite the regression of the patient's symptoms, a thoracic computed tomography (CT) was performed due to the patient's continuing cough. A 32 × 21 mm patch-like consolidation area and focal emphysematous changes around it were reported on the thoracic CT examination in the medial basal segment of the left lung lower lobe (Figure 2). A pulmonology specialist evaluated the CT results and referred the patient to our clinic.

In the detailed history, the patient stated that there are periods, especially in the last 6 years, that begin with cough complaints, with increasing frequency, and then progress with pneumonic symptoms such as high fever, sputum production, and dyspnea, and that there are many hospital admissions for these reasons.

When we examined the results of the tests performed at the external center and the patient's history, we encountered some difficulties. In the tests performed, the return of the infection indicators to the normal range and the regression of the patient's symptoms showed that the empirical antibiotic treatment was successful. However, the cough complaints continued. We examined the common causes that could cause a chronic cough. When we performed spirometry (respiratory function test and reversibility test), we saw that the patient's results were within the normal range, these results allowed us to exclude the diagnosis of asthma. There was no known history of allergy, and the complaints of shortness of breath were accompanied by pneumonic symptoms such as fever and fatigue. The patient did not have symptoms and signs of gastroesophageal reflux disease. There was no complaint of postnasal drip. The patient was not exposed to irritant gases or air pollution in the environment where he lived and worked. He was not using any medications that could cause a cough, such as angiotensin-converting enzyme inhibitors. The patient had quit smoking 1 year ago and had a 5-pack/year smoking history. When the patient's thorax CT report and imaging were examined, multiple aberrant vascular structures originating from the thoracic aorta in the left hemithorax and coursing through the lung parenchyma were observed, but the vascular structures extended to a consolidated area, and there was an emphysematous area in the left lower lobe. The consolidated area suggested a pulmonary mass, while the aberrant vascular structures and emphysematous area strengthened the possibility of PS. The patient's absence of B symptoms, no family history of cancer, and young age were characteristics that ruled out malignancy.

We performed CTA imaging to evaluate aberrant arterial structures. CTA images reconstructed in maximum intensity projection and 3D allowed detailed evaluation of aberrant arterial structures and clear demonstration of the relationship between the arteries and the surrounding structures (Figures 3 and 4). The CTA report defined "intralobar tissue in the lower lobe of the left lung, not connected to the tracheobronchial tree and fed by three arterial branches originating from the thoracic aorta" and

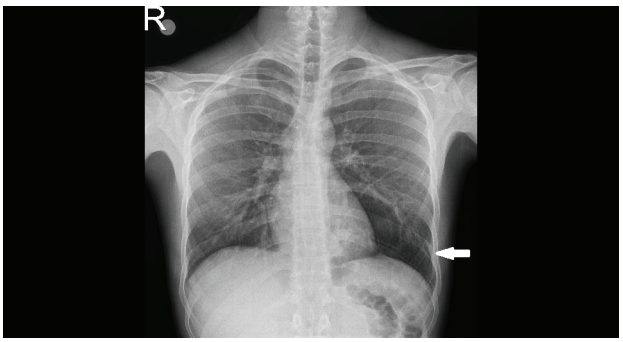

Figure 1. *Chest X-ray taken at an external center showed an emphysematous area in the left lower zone.*

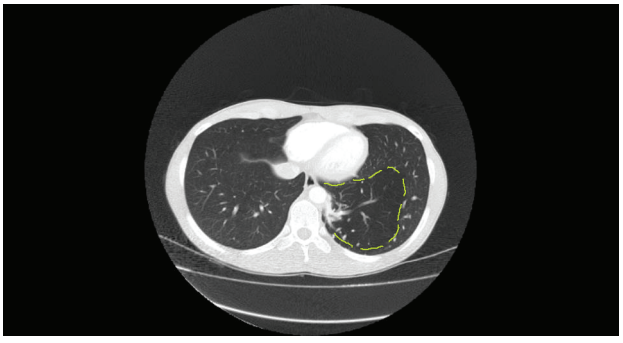

Figure 2. *The emphysematous area shown with green lines was observed on thorax CT taken at an external center.*

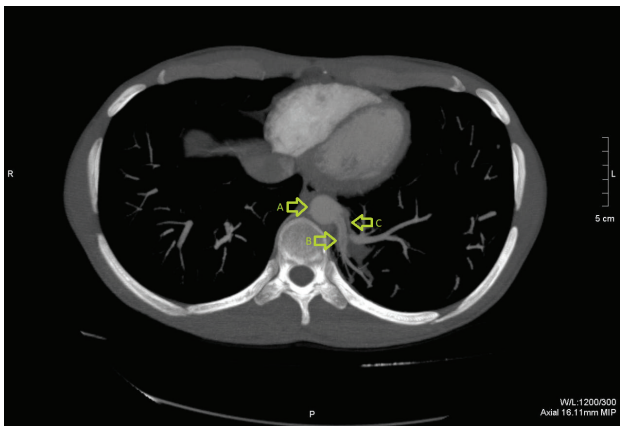

Figure 3. *CTA maximum intensity Projection (MIP) Aberrant arterial structures extending into the sequestered tissue on the left, originating from the descending aorta in the axial cross-section. A: descending aorta, B and C: aberrant arterials.*

the diameters of the arterial branches were stated as 5.4, 4.7, and 7.1, respectively, from proximal to distal.

The curative treatment of PS is surgical resection. For lung resection, we needed to examine the patient's pulmonary capacity preoperatively. Therefore, spirometry was repeated, and a 6-minute walk test was performed. The results were adequate for lung resection. Respiratory physiotherapy was started in the preoperative period. The commonly applied surgical methods for resection are VATS or thoracotomy, as seen in the literature. However, we preferred a hybrid approach in our patient. When we reviewed the literature, we found that the hybrid approach is an innovative method. The hybrid approach allowed us to combine the advantages of VATS and thoracotomy while minimizing their disadvantages.

The patient was informed about the operation. Written and verbal informed consent form was obtained. The patient was scheduled to undergo left lower lobectomy.

The patient was intubated under general anesthesia with a double-lumen endotracheal tube for selective single-lung ventilation. The patient was placed in the right lateral decubitus position, a 5 cm long hybrid thoracotomy incision was opened, and the pleural space was entered through the 5th intercostal space. A videothoracoscope was inserted through the port opened at the intersection of the 7th intercostal space and the midaxillary line. Intraparenchymal sequestration area was observed in the lower lobe during exploration via the videothoracoscope. In exploration, the presence of the superior pulmonary vein was confirmed, and it was observed that there was no separate venous branch for the sequestered tissue (Figure 5). It was understood that it would be safe and correct to perform a left lower lobectomy on the patient. By performing sharp and blunt dissection, the abnormal arterial structures were released, taped, and transected by a vascular stapler (Figure 6). Then, lower lobectomy was completed according to the standard procedure. The removed lower left lobe was separated for microscopic examination and sent to the pathology clinic. The patient's chest tube was removed on the 5th postoperative day. The patient was discharged on the 10th postoperative day. The pathology report was consistent with PS.

After our patient was discharged, for follow-up examinations were planned: first week, first, third, and sixth month. In these follow-up examinations, there was no problem in wound healing, and the patient's symptoms

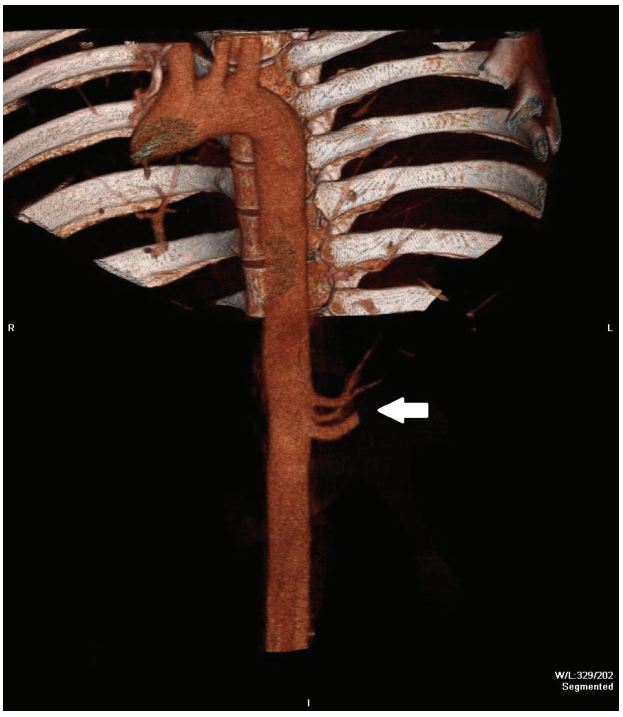

Figure 4. *Three aberrant vascular structure originating from the descending aorta in CTA 3D imaging.*

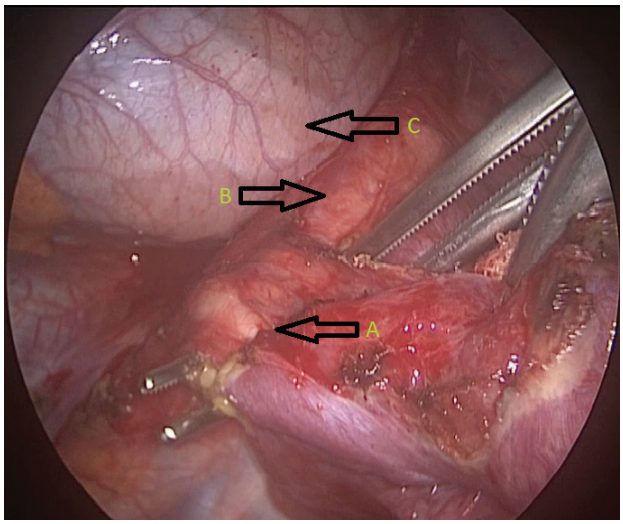

Figure 5. *Exploration of aberrant arterial structures intraoperatively. A: Aberrant arterial structures, B: Thoracic aorta, and C: Pericardium.*

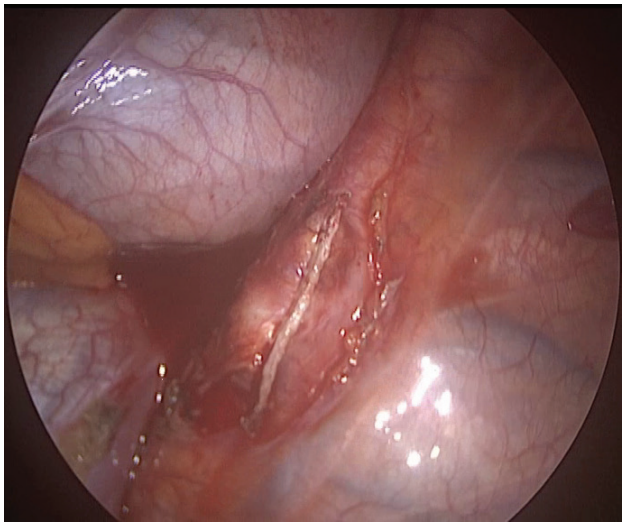

Figure 6. *Image of aberrant arterial structures originating from the thoracic aorta after being transected by a vascular stapler.*

did not recur. Blood tests and radiological imaging were normal.

Discussion

PS was first described in 1861 by Kektozic and Rokitansky as an accessory pulmonary lobe [1]. PS are rare among congenital pulmonary malformations and their frequency is less than 6.4% [3,8]. It is mostly seen in the left lung, and its occurrence in the lower lobes is high [3]. There are 2 subtypes, ILS and ELS, and ILS is the most common [3,9]. Our patient had ILS tissue, including the posterior, basal, and lateral segments of the lower lobe of the left lung.

ELS is separated from the adjacent lung parenchyma by its pleural membrane. Although it is often asymptomatic, it can be diagnosed starting from the antenatal period due to its frequent association with other congenital anomalies [10].

The ILS is covered by the pleura of the lung parenchyma to which it is adjacent, and causes frequent pneumonia attacks in childhood. ILS is usually diagnosed in the first 1-2 decades of life [11]. In line with the literature, our patient had frequent pneumonia attacks for the last few years and was diagnosed in the 2nd decade.

The arterial supply of PS is mostly provided from the systemic circulation through the abnormal artery originating from the aorta and its branches [12]. In the largest retrospective study to date, conducted by Wei and Li [3] which examined 2,625 cases of PS, 130 cases were reported in which 2 abnormal arterial branches provided the arterial supply of sequestration tissue, and 40 cases were reported in which three or more arterial branches provided [3]. This study shows that the case of PS with three or more aberrant arterial branches is extremely rare. Our case report describes a case whose arterial supply was provided by three aberrant arterial branches.

Imaging methods have an important place in the diagnosis of PS and in determining the surgical method. The decision to embolize aberrant arterial structures and the surgical method to be chosen may be possible with the effective and accurate use of imaging methods in the preoperative period. Color Doppler ultrasonography and magnetic resonance imaging in the antenatal period, and Thoracic CT and CTA/MRA in childhood and adulthood are the most commonly used examinations [2,6]. For a definitive diagnosis, aberrant arteries feeding the PS tissue should be demonstrated with aortography, but this is an invasive method. Today, CTA/MRA is used instead of aortography [1]. We also demonstrated the aberrant arteries feeding our patient's PS with CTA (Figure 4).

Curative treatment for PS is surgical resection. Thoracotomy or VATS are widely used surgical methods that should be evaluated and decided on according to the patient. The combination of surgical resection with embolization to be applied to aberrant arterial structures provides advantages by reducing the risk of bleeding. However, the necessity of performing the procedure in two separate sessions and the complications specific to embolization constitute disadvantages [13,14]. The VATS method has advantages such as cosmetic gain with smaller incisions, reduced postoperative pain, and shortened hospital stay [3,11]. However, there are some difficulties in managing possible complications. Thoracotomy has advantages such as wide and three-dimensional exploration, greater maneuverability, and effective and rapid management of complications. In addition, it has disadvantages such as infection risk and cosmetic loss due to wide incision, postoperative pain, and prolonged hospitalization. In a previous case of ELS originating from the abdominal aorta in our clinic, we performed embolization by placing a coil into the vessel, and since the coil was under the diaphragm, we cut the aberrant artery on the diaphragm with a stapler, and there was no risk of bleeding [15]. However, in this case, we did not consider embolization because all three aberrant arteries originated from the aorta at a level above the diaphragm. Because the coil placed into the vessel with embolization could have remained between the stapler and could have posed a bleeding risk. We decided to perform a hybrid thoracotomy in our patient to benefit from the advantages of VATS and thoracotomy as a surgical method. Our choice of surgical treatment was influenced by the hybrid approach's ability to provide more extensive and detailed exploration, more space and maneuverability for safe division and cutting of the three anomalous arteries, its superiority in managing possible complications such as bleeding, less postoperative pain, and cosmetic gain with a small thoracotomy incision [16].

In most of the cases reported in the literature, arterial supply has not been clearly reported [8,9]. With this study, we believe that performing a successful surgical treatment by performing thoracotomy left lower lobectomy on a rare case of ILS fed by three aberrant arteries directly originating from the thoracic aorta will make a very important contribution to the literature and take our clinic's experience in this field to the next level.

Conclusion

PS is among the rare congenital lung malformations. Although ELS can be asymptomatic, it may be diagnosed at earlier ages due to associated congenital anomalies and advanced fetal diagnostic methods available today. However, ILS cases can be diagnosed late because they show pneumonic symptoms such as cough, chest pain, and dyspnea. Especially in the case of recurrent pneumonia attacks that do not respond completely to treatment in childhood and young adulthood, intralobar sequestration should be considered, and surgical treatment of the patient

should be planned without wasting time. To plan the most appropriate surgery and prevent unexpected intraoperative complications, the aberrant arterial supply of the ILS must be identified during the preoperative period using appropriate imaging techniques.

What is new?

PS is a rare congenital malformation. The fact that there are very few cases in the literature that are supplied by three aberrant arterial branches originating from the aorta makes our case even rarer. The hybrid approach allowed us to combine the advantages of commonly used VATS and thoracotomy while minimizing the disadvantages. We believe that we have made positive contributions to the literature while improving our clinical experience with this rare case and innovative surgical method.

List of Abbreviations

CT	computed tomography
CTA	computed tomography angiography
ELS	extralobar pulmonary sequestration
ILS	intralobar pulmonary sequestration
MIP	maximum intensity projection
MRA	magnetic resonance angiography
PS	pulmonary sequestration
VATS	video-assisted thoracoscopic surgery

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 informed consent was obtained from the patient.

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 (gender, age)	24 years, male
2	Final diagnosis	Pulmonary sequestration
3	Symptoms	Fever, cough, sputum
4	Medications	Symptomatic treatment given
5	Clinical procedure	Surgery
6	Specialty	Thoracic surgery