

# Patient-based Assessment of Treatment Options for Pulmonary Sequestration: Two Case Reports

## Pulmoner Sekestrasyon için Tedavi Seçeneklerinin Hasta Bazlı Değerlendirilmesi: İki Olgu Sunumu

Shukur Musayev<sup>1</sup>, Halil Bozkaya<sup>2</sup>, Kutsal Turhan<sup>3</sup>, Özgür Samancılar<sup>1</sup>

### Abstract

Pulmoner sekestrasyon (PS), nadir görülen bir konjenital pulmoner displazidir. PS, en önemli özelliği vaskülarizasyonu olan konjenital pulmoner displazinin nadir bir formudur. Genellikle torasik ve abdominal aortadan kaynaklanan atipik arterlerle vaskülarizasyon görülür. PS'nin interlobar (ILS) ve ekstralobar (ELS) olmak üzere iki formu vardır ve bunlar visseral plevra ile ilişkilerine göre ayırt edilebilir. Besleyici damarların cerrahi rezeksiyonu ve ligasyonu bu durum için standart tedavilerdir. Bu çalışmada, farklı sekestrasyon tiplerine sahip iki olgu, tedavi seçeneklerinin gözden geçirilmesiyle birlikte sunulmaktadır.

**Keywords:** Pulmoner sekestrasyon, embolizasyon, video yardımcı torakoskopik cerrahi.

### Öz

Pulmonary sequestration (PS) is a rare congenital pulmonary dysplasia. The most important feature is vascularisation with Pulmonary sequestration (PS) is a rare form of congenital pulmonary dysplasia, with the most significant feature being vascularization with atypical arteries, generally originating in the thoracic and abdominal aorta. There are two forms of PS, interlobar (ILS) and extralobar (ELS), which are distinguishable based on their relationship with the visceral pleura. Surgical resection and ligation of the feeding vessels are the standard treatments for the condition. In the present study, two cases with different sequestration types are presented along with a review of the treatment options.

**Anahtar Kelimeler:** Pulmonary sequestration, embolization, video-assisted thoracoscopic surgery.

<sup>1</sup>Department of Thoracic Surgery, Medica International Hospital, İzmir, Türkiye

<sup>2</sup>Department of Radiology, Medica International Hospital, İzmir, Türkiye

<sup>3</sup>Department of Thoracic Surgery, Ege University Hospital, İzmir, Türkiye

<sup>1</sup>Medica International İzmir Hastanesi, Göğüs Cerrahisi Bölümü, İzmir

<sup>2</sup>Medica International İzmir Hastanesi, Radyoloji Bölümü, İzmir

<sup>3</sup>Ege Üniversitesi Tıp Fakültesi Hastanesi, Göğüs Cerrahisi Anabilim Dalı, İzmir

Submitted (Başvuru tarihi): 20.05.2024 Accepted (Kabul tarihi): 02.08.2024

Correspondence (İletişim): Shukur Musayev, Department of Thoracic Surgery, Medica International Hospital, İzmir, Türkiye

e-mail: shuko.musayev@gmail.com



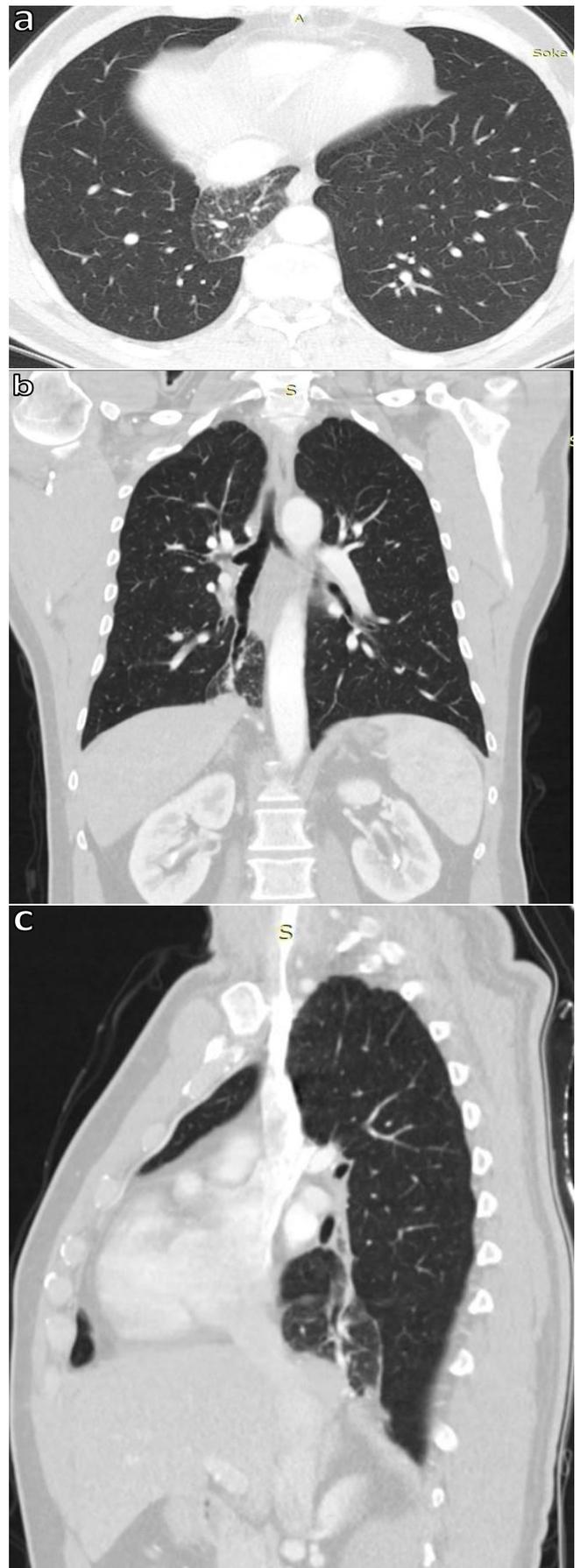
Pulmonary sequestration (PS) is characterized by a bronchial anomaly, parenchymal dysgenesis and vascularization of abnormal systemic artery. It accounts for less than 6.4% of all pulmonary congenital malformations (1). Such cystic structures are localized in the posterior basal segment of the right and left lung lower lobe in 80–90% of cases (2). The ILS form is more common (75–84% of the total), with 74% of feeding arteries identified as arising from branches of the thoracic aorta (3). The most common symptoms of PS include cough or expectoration, fever, hemoptysis and chest pain. Herein, we present a case of ILS with a systemic artery originating directly from the thoracic aorta, and an ELS sequestration with multiple branches of systemic arteries supplied from the abdominal aorta, both of which were safely resected in our clinic.

## CASE

**Case 1:** A 49-year-old non-smoking male was admitted with cough and hemoptysis. Laboratory results were normal, while a thorax computed tomography (CT) scan revealed limited lung parenchyma in the right lower lobe that was typical for ELS (Figure 1a, b and c). The dominant artery supplying the sequestered lung tissue was found to arise from the abdominal aorta. It was decided to perform an endovascular embolization as the first step and VATS resection on the following day.

Celiac trunk and phrenic angiographies were obtained selectively using a hydrophilic guide wire and an angiography catheter. A peripheral microcatheter (with microwire) was sent through the diagnostic catheter using the coaxial technique and a super selective catheterization was performed. A control angiogram revealed that the aberrant systemic artery was divided into three branches and supplied sequestration (Figure 2a). Each involved artery was super selectively embolized with mechanical peripheral fibrillar coils, and a post-embolization angiogram revealed the sequestered pulmonary tissue to be completely embolized (Figure 2b).

One day after the embolization, a sublobar resection was carried out with minimally invasive surgery. After double lumen intubation, the patient was placed in the left-lateral decubitus position. An extralobar sequestration connected to the lower right lobe was detected during the operation (Figure 3a). The adhesions of the lobe to the diaphragm were dissected and the branches coming from the abdominal aorta were exposed through three-port video-assisted thoracoscopic surgery (VATS) (Figure 3b). The atypical arteries were divided with a stapler and the sequestered segment was resected. The patient was discharged on the third postoperative day. A pathological examination revealed an extralobar sequestration. No hemoptysis recurred within the one-year follow-up period.



**Figure 1a:** Views of right lower lobe ELS on thoracic CT image; axial (a), (b) and sagittal (c)



**Figure 2:** Angiography of the aberrant artery during embolization; Feeding artery on control angiogram (a), Artery after embolization with a coil (b)

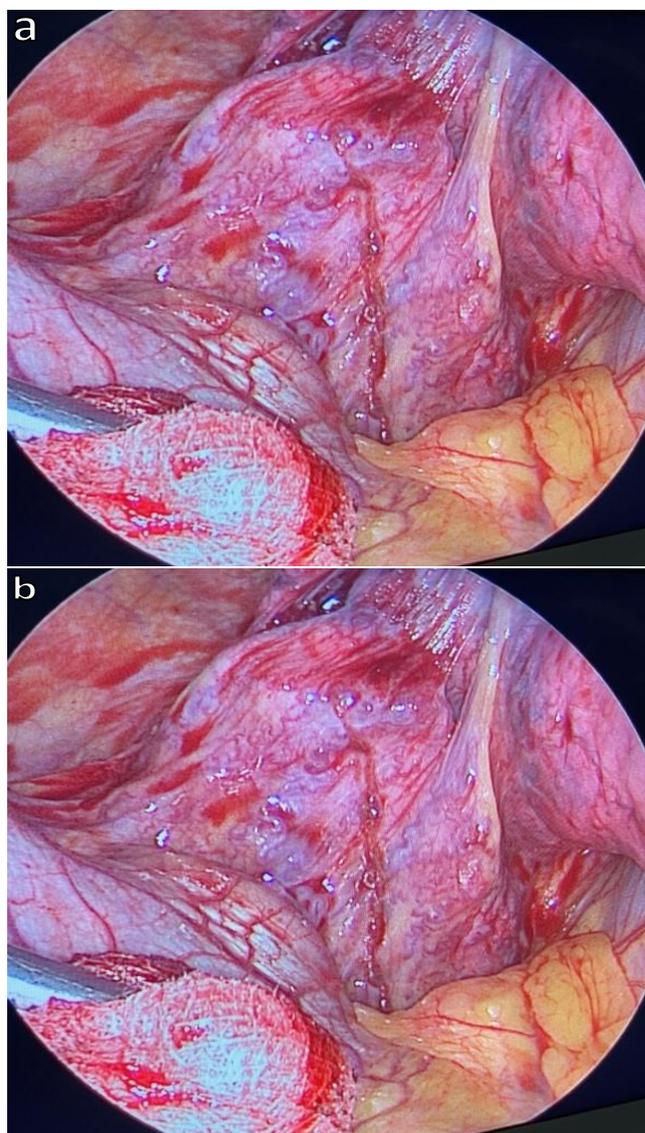
**Case 2:** A 35-year-old non-smoking female patient was admitted to our clinic with a history of repeated left-sided pneumonia. The cause of the pneumonia was investigated with a CT scan that revealed diffuse and prominent segmental cystic bronchiectasis in the lower lobe superior and anteromedio-basal segments of the left lung, and partially in the posterior basal segment in the paravertebral areas and perihilar regions. The lesion was seen to be supplied by a 5.9 mm diameter systemic artery arising directly from the thoracic aorta, and was identified as ILS (Figure 4a, b and c). The patient was planned to undergo left lower lobectomy by VATS.

After double-lumen intubation, one utility and two port incisions were made on the left side. Upon exploration, the left lower lobe was identified as destroyed and ineffective, and an abnormal aberrant branch directly originating from the thoracic aorta was noted to be supplying the lobe. The feeder arterial branch was first dissected and devised with a vascular stapler (Figure 5a and b), after which, the left lower lobe was resected with a left

lower lobectomy. The patient was discharged on the fifth postoperative day. A pathological examination revealed an intralobar sequestration in the left lower lobe. The patient was noted to be in good health throughout the 6-month follow-up.

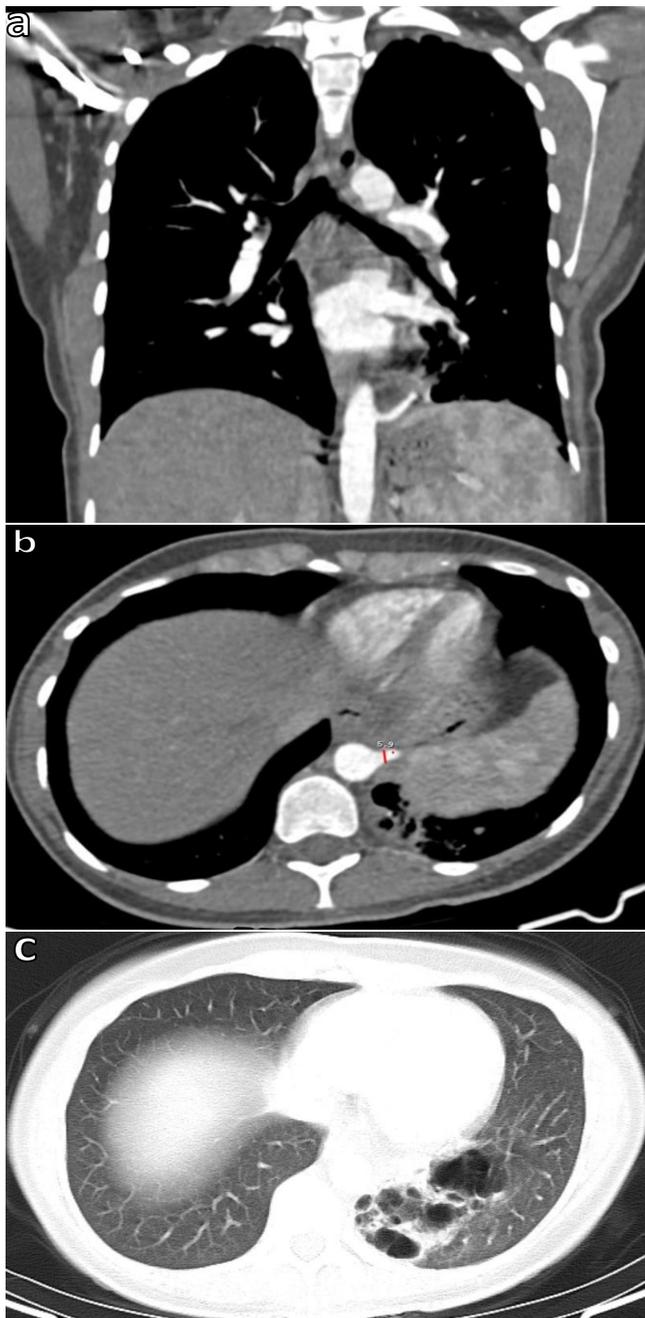
## DISCUSSION

PSs are rarely encountered malformations that are supplied by atypical arteries originating from the thoracic aorta, and more rarely from the abdominal aorta. The location of arterial supply and venous drainage should be determined preoperatively, and so preoperative imaging studies are of vital importance. In our first case, three arteries originating from the abdominal aorta were found to supply the ELS, while in the other case, a 5.9 mm diameter artery originating from the thoracic aorta was found to supply the ILS.

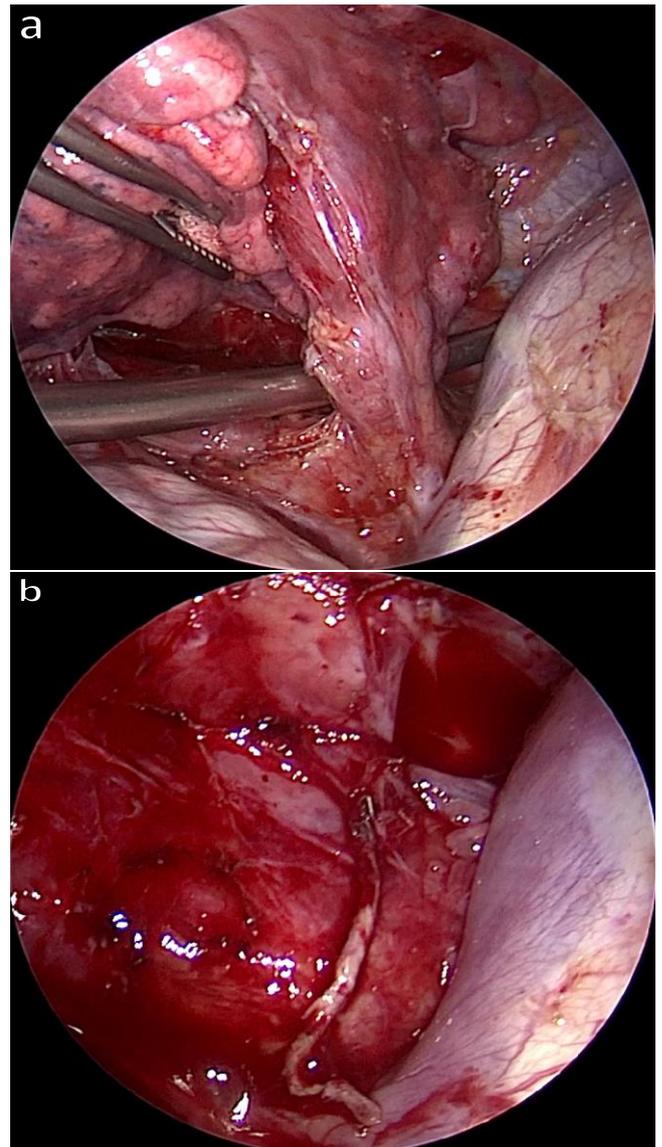


**Figure 3:** Intraoperative thoracoscopic findings; Intraoperative image of ELS (a), feeding artery arising from the abdominal aorta (b)

Currently, the most widely accepted treatment for PS is surgical resection, which can prevent possible infection. In the treatment of PS, both conventional thoracotomy and VATS can be used for the sublobar resection or lobectomy. In a study by Zhang et al. (4), 15 patients underwent thoracotomy and six underwent minimally invasive surgery within 2 years. The authors reported VATS to be a better approach than thoracotomy for PS resection with the continuous development of thoracoscopic techniques. The outcomes of the minimally invasive surgery detailed in the present study support the findings reported Zhang et al., related to the easy pain palliation, shorter hospitalizations and aesthetic advantages.



**Figure 4:** Coronal and axial view of the feeding artery on a thoracic CT image (a, b and c) diffuse and prominent segmental cystic bronchiectasis on a thoracic CT image



**Figure 5:** Intraoperative thoracoscopic findings; Feeder artery arising from the aorta after dissection (a), Image after the division of the feeder artery arising from the aorta (b)

Recurrent infection rates increase due to the cystic structure of PS, as a result, the vessels may be more fragile and thinner walled with inflammation. All of the above reasons can contribute to serious intraoperative bleeding, especially in patients with multiple arteries arising from the abdominal aorta, as in our first case, due primarily to the deep foci of bleeding in the diaphragmatic hiatus, making hemostasis difficult. Preoperative embolization has been reported to be especially effective in the prevention of intraoperative bleeding (5). In our second case, the surgical intervention was performed without embolization due to the large diameter of the single artery supplying the sequestration, arising directly from the thoracic aorta. The treatment of cases with such sequestrations should be decided upon on a patient-based basis.

Previous studies in literature recommend the treatment of small (<3 cm) interlobar sequestrations with endovascu-

lar embolization alone. Zhang et al. (4) achieved complete response in two patients and partial (<60%) response in five patients after the embolization of sequestrations of <3 cm. Recanalization after embolization is not uncommon, and is related to the time interval following the procedure (6). Based on this, we opted to operate on the patient the day after the embolization to avoid recanalization. In contrast, Cho et al. (7) recommended surgical resection in all patients, including asymptomatic patients, to exclude the risk of infection, post-embolization complications and other pathologies. In our first case, the patient was presented with hemoptysis. Since PS is directly related to the tracheobronchial system, we opted for curative treatment surgery after embolization. Anatomical resections are the most preferred procedure, and have produced excellent long-term results, especially in symptomatic patients who present with hemoptysis. There is a lack of consensus on the optimum time interval between embolization and surgery in hybrid surgical treatments of pulmonary sequestration. VATS resections have been performed immediately, 4 days or 4 weeks after the embolization procedure in different studies (8,9). We opted to carry out a surgical resection 24 hours after the endovascular embolization in the absence of any problematic intraoperative events (9).

## CONCLUSION

We believe that the treatment options in PS should be evaluated on a patient-by-patient basis. We recommend preoperative endovascular embolization as the safer surgery option, especially for sequestrations with feeder artery(ies) originating from the abdominal aorta. In cases of sequestration supplied by a single large-diameter artery arising directly from the thoracic aorta, surgical treatment may be considered without preoperative embolization. In symptomatic PS cases in which there is a direct relationship with the tracheobronchial system, embolization alone is not sufficient, and surgical resection may be appropriate as a curative treatment.

## CONFLICTS OF INTEREST

None declared.

## AUTHOR CONTRIBUTIONS

Concept - S.M., H.B., K.T., Ö.S.; Planning and Design - S.M., H.B., K.T., Ö.S.; Supervision - Ö.S., S.M., H.B.,

K.T.; Funding - S.M.; Materials - S.M.; Data Collection and/or Processing - S.M.; Analysis and/or Interpretation - S.M.; Literature Review - S.M.; Writing - S.M.; Critical Review - Ö.S.

## REFERENCES

1. Savic B, Birtel FJ, Tholen W, Funke HD, Knoche R. Lung sequestration: report of seven cases and review of 540 published cases. *Thorax* 1979; 34:96-101. [\[CrossRef\]](#)
2. Wei Y, Li F. Pulmonary sequestration: a retrospective analysis of 2625 cases in China. *Eur J Cardiothorac Surg* 2011; 40:e39-42. [\[CrossRef\]](#)
3. Fabbri N, Tamburini N, Galeotti R, Quarantotto F, Maniscalco P, Rinaldi R, et al. A rare case of intralobar pulmonary sequestration: combined endovascular and video-assisted thoracoscopic approach. *AME Case Rep* 2018; 2:19. [\[CrossRef\]](#)
4. Zhang SX, Wang HD, Yang K, Cheng W, Wu W. Retrospective review of the diagnosis and treatment of pulmonary sequestration in 28 patients: surgery or endovascular techniques? *J Thorac Dis* 2017; 9:5153-60. [\[CrossRef\]](#)
5. Pauels L, De Waele M, Medart L, Debruche M. Two unusual variants of pulmonary intra-lobar sequestration. *Interact Cardiovasc Thorac Surg* 2022; 35:ivac189. [\[CrossRef\]](#)
6. Garcia-Olivé I, Sanz-Santos J, Centeno C, Radua J, Andreo F, Sampere J, et al. Predictors of recanalization in patients with life-threatening hemoptysis requiring artery embolization. *Arch Bronconeumol* 2014; 50:51-6. [\[CrossRef\]](#)
7. Cho MJ, Kim DY, Kim SC, Kim KS, Kim EA, Lee BS. Embolization versus surgical resection of pulmonary sequestration: clinical experiences with a thoracoscopic approach. *J Pediatr Surg* 2012; 47:2228-33. [\[CrossRef\]](#)
8. Zhou H, Tang S, Fu Q, Yu L, Liu L. Hybrid surgery in treatment of pulmonary sequestration with abdominal aorta feeding vessel: a case report. *J Cardiothorac Surg* 2018; 18:13:44. [\[CrossRef\]](#)
9. He B, Sun MS, Niu Y, Zhang JB, Nie QQ, Zheng X, et al. Hybrid and endovascular treatment of pulmonary sequestration: two case reports and literature review. *Ann Vasc Surg* 2020; 69:447.e1-447.e8. [\[CrossRef\]](#)