

STARD-compliant article: Comparison of pulmonary sequestrations with thoracic and abdominal aortic arterial supply

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Abstract

Pulmonary sequestrations (PS) are typically supplied by a vessel originating from thoracic aorta, or abdominal aorta. Differences in imaging features between these PS subtypes have not been described.

To analyze the imaging features of PS with arterial supply from the thoracic and abdominal aorta.

Retrospectively, 23 pathologically proven cases of pulmonary sequestration were analyzed and compared based on the site of feeding artery origin.

In 21 cases (21/23), the PS was soft tissue density. 1 (1/23) PS was purely cystic and another heterogeneous with both cystic and solid components (1/23). In 16/23 cases, the feeding vessel(s) arose from the thoracic aorta (male:female ratio 1:7) and in 7/23 cases from the abdominal aorta (male:female ratio 4:3). Feeding vessels from the thoracic aorta were duplicated in 7/16 cases. PS location ($P < .05$) and size ($P < .001$) differed based on the origin of the feeding vessel (thoracic aorta: 14/16 left lower lobe, mean volume 962.97 mL; abdominal aorta: 3/7 left lower lobe, mean volume 1120.89 mL). The feeding arteries themselves differed in size depending on their site of origin (thoracic aorta: mean diameter 7.0 mm \pm 2.7 mm, mean length 44.6 mm \pm 10.9 mm; abdominal aorta: mean diameter 3.3 mm \pm 0.6 mm, mean length 103.6 mm \pm 34.5 mm).

PS size and distribution differ depending upon the site of feeding vessel origin as does the size of the feeding vessel itself.

Abbreviations: CT = computed tomography, CTA = computed tomography angiography, PS = pulmonary sequestration.

Keywords: abdominal aorta, pulmonary sequestration, thoracic aorta

1. Introduction

Pulmonary sequestration (PS) is a rare congenital malformation consisting of a solitary non-functional mass of lung tissue

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The datasets used during the current study are available from the corresponding author on reasonable request.

The authors declare that there are no conflicts of interest.

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disconnected from the central respiratory tract with blood supply from the systemic rather than the pulmonary circulation.^[1] The etiology of PS is not clear, but the most widely accepted hypothesis is that an accessory lung bud forms caudal to the normal lung bud, migrating with it during development.^[2] PS is typically supplied by a vessel originating from the descending thoracic aorta, or less commonly the abdominal aorta. Differences in imaging features between these PS subtypes have not been described. In this retrospective study, 23 cases of pathologically proven PS were examined in an attempt to understand differences in imaging features between those with thoracic and abdominal aortic blood supply.

2. Methods

2.1. Participants

A total of 23 consecutive adults have received surgical operation for PS. All pathological results of the sequestered tissue from the surgical procedure were reviewed by a senior consultant pathologist. All of 23 cases (6 men and 17 women, age range 18–59 years, mean age of 43.8 years) were collected from patients hospitalized at the hospital from January 2012 to December 2015.

2.2. Test methods

In all cases, computed tomography (CT) and computed tomography angiography (CTA) examinations had been performed (Siemens, SOMATOM Definition AS+ 128 slice spiral

CT). The CT scanning parameters were as follows: tube current, 200 mA; tube voltage, 120KV; helical pitch, 0.984; matrix, 1.0 mm slice thickness. 60 mL contrast agent (300 mg iodine/mL, lopamidol injection) was intravenously injected through antecubital vein, followed by 40 mL saline solution at flow rates of 3.5 mL/s. 20 seconds following injection, an arterial phase scan was obtained. After another 15 seconds, a second phase of imaging was performed. All data was sent to a dedicated post-processing workstation where multiplanar reconstruction (MPR), maximum intensity projection (MIP), and 3-dimensional volume reconstruction (VR) images were obtained.

2.3. Image analysis

One radiologist, blinded to the diagnosis and patient characteristics, independently reviewed the CT imaging. The following data were obtained for analysis: PS location, volume, density of the PS, venous drainage of PS, and the dimensions (diameter and length) of supplying artery. If the feeding vessels from the aorta were duplicated (with R_1 & R_2 , L_1 & L_2), their diameter (R_0) and length (L_0) were calculated by the formulae: $R_0 = \sqrt{(R_1^2 + R_2^2)}$; $L_0 = (L_1 + L_2)/2$.

2.4. Statistical methods

The data were analyzed using SPSS v.17.0 for Windows (SPSS, Chicago, IL). Continuous variables were described as mean \pm standard deviations (SD). An independent sample Wilcoxon-test and chi-square test were used to analyze differences in imaging parameters between the PS with thoracic and abdominal aortic vascular supply. P values $<.05$ were considered statistically significant.

3. Results

3.1. Participants

The clinical presentation of these cases was variable. Cough and expectoration were the most common clinical symptoms, noted in 19 (82.6%) patients, followed by intermittent fever (4 cases, 17.4%), chest pain (3 cases, 13%), and hemoptysis (1 case, 4%). 4 cases (17.4%) were asymptomatic and PS were discovered incidentally. In 16/23 cases, the artery or arteries feeding the PS arose from the thoracic aorta (2 men and 14 women, age range 19–39 years, mean age of 27.6 years) and in 7/23 cases from the abdominal aorta (4 men and 3 women, age range 18–59 years, mean age of 46.7 years) (Table 1).

3.2. Test results

Out of 23 cases of PS, 16 were supplied by vessels originating from the thoracic aorta, and 7 by vessels arising from the abdominal aorta. 17 PS were located in the posterior basal segment of left lower lobe and 6 cases in the medial basal segment of right lower lobe (Tables 1 and 2). Statistically significant differences in location were found based on the site of aortic blood supply ($P <.05$).

Solid ($n=16$), mixed cystic-solid ($n=7$), and cystic ($n=1$) appearances of PS were identified on CT. The PS with thoracic aortic blood supply ranged from 1.1 cm \times 1.7 cm \times 3.3 cm to 5 cm \times 3.9 cm \times 4.8 cm (AP \times transverse \times cranial to caudal) in size versus a range from 4.6 cm \times 2.5 cm \times 2.8 cm to 7.3 cm \times 5 cm \times 7 cm for the PS with abdominal aortic supply blood.

In 14 cases, a single feeding artery was identified (Fig. 1A). In 7 cases, 2 separate feeding arteries arose from the thoracic aorta

Table 1
Clinical information and image findings of 23 cases.

No	demography		Clinical symptoms				Imaging findings		
	Sex	Age	Cough	Fever	Chest pain	Haemoptysis	Morphology	Feeding artery	Location
1	M	19	-	-	-	-	Solid	TA	RL
2	M	24	+	-	-	-	Solid	TA	LL
3	F	32	+	+	+	+	Mixed	TA	LL
4	F	27	+	-	-	-	Solid	TA	LL
5	F	33	+	-	-	-	Solid	TA	LL
6	F	36	+	-	-	-	Solid	TA	LL
7	F	21	-	-	-	-	Solid	TA	LL
8	F	28	+	-	-	-	Solid	TA	LL
9	F	20	+	-	-	-	Solid	TA	LL
10	F	25	+	+	-	-	Mixed	TA	LL
11	F	27	+	-	-	-	Solid	TA	LL
12	F	28	+	+	+	-	Mixed	TA	LL
13	F	19	+	-	-	-	Solid	TA	LL
14	F	35	+	-	-	-	Solid	TA	LL
15	F	29	-	-	-	-	Cystic	TA	RL
16	F	39	+	-	-	-	Solid	TA	LL
17	M	18	+	-	-	-	Solid	AA	LL
18	M	59	+	-	-	-	Solid	AA	RL
19	M	42	+	-	+	-	Solid	AA	RL
20	M	57	+	-	-	-	Mixed	AA	LL
21	F	49	+	+	-	-	Mixed	AA	RL
22	F	54	-	-	-	-	Solid	AA	RL
23	F	48	+	-	-	-	Mixed	AA	LL

M=Male; F=Female; TA=Thoracic aorta; AA=Abdominal aorta; LL=Left lung; RL=Right lung.

Table 2
Location and arterial supply of PS.

	Right lung	Left lung	Total
PS with thoracic aorta supply	2	14	16
PS with abdominal aorta supply	4	3	7
Total	6	17	23

(Fig. 1B). In 2 cases, a single artery supplying the PS arose from the thoracic aorta but then bifurcated upon entry into the sequestration (Fig. 1C, D). The mean diameter of feeding vessels with thoracic aortic origin was $7.0 \text{ mm} \pm 2.7 \text{ mm}$ (range 2.8–11 mm) versus $3.3 \text{ mm} \pm 0.6 \text{ mm}$ (range 1–4 mm) for those of abdominal aortic origin. The length of feeding vessels originating from the thoracic aorta was $44.6 \text{ mm} \pm 10.9 \text{ mm}$ (range 15–58 mm) versus $103.6 \text{ mm} \pm 34.5$ (range 59–160 mm) for those arising from the abdominal aorta.

3.3. Estimates

PS volume, feeding artery diameter, and feeding artery length all differed depending on whether supply was from the thoracic or abdominal aorta ($P < .05$) as determined by the rank sum test

Table 3
Volume and dimension (diameter and length) of supplying artery of PS.

	Feeding artery from thoracic aorta (mean)	Feeding artery from abdominal aorta (mean)	P
Volume of PS	962.97	1120.89	.000
diameter of feeding artery	1440.72	673.27	.000
length of feeding artery	505.50	1549.50	.000

(Table 3). In all cases, the draining vein was the inferior pulmonary vein ipsilateral to the PS. Associated findings included inflammatory pulmonary changes ($n=4$), emphysema ($n=3$), and bronchiectasis ($n=3$). The enhancement pattern was heterogeneous in the majority of cases ($n=19$) and homogeneous in 3 cases. All of the cases were intrapulmonary PS.

4. Discussion

PS is a rare congenital malformation consisting of nonfunctional pulmonary tissue without tracheobronchial communication but with anomalous systemic vascular supply. The incidence of PS

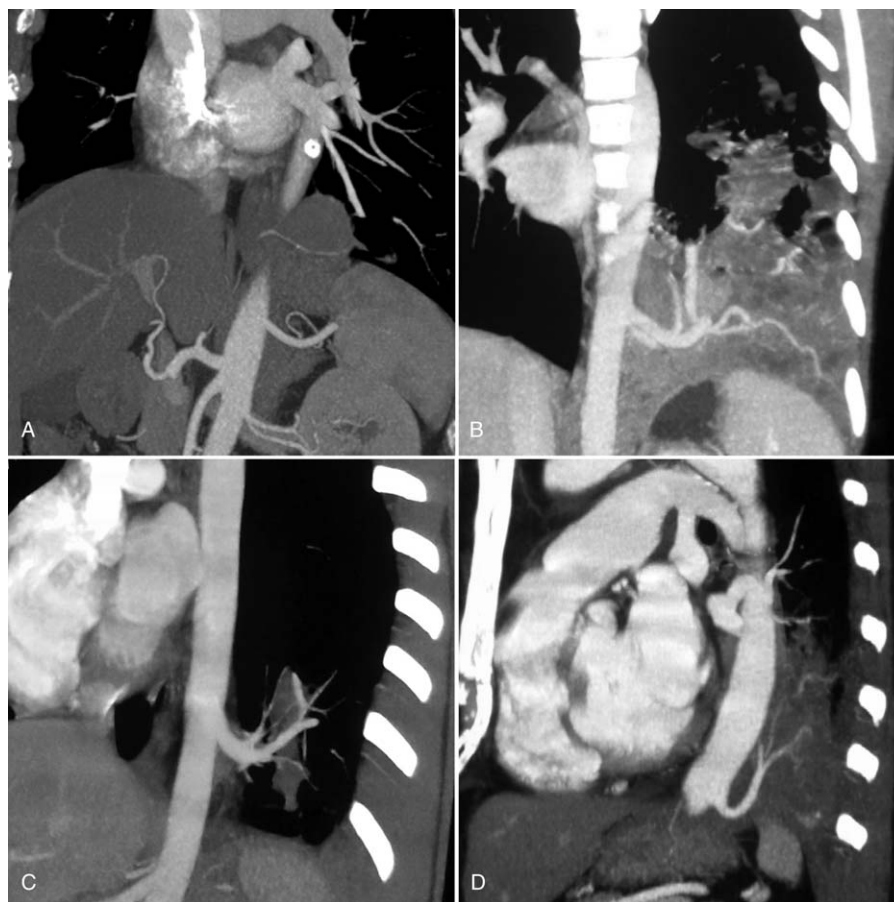


Figure 1. MIP images demonstrating variations in pulmonary sequestration vascular supply in 4 different patients. In (A), an aberrant artery arising from the left gastric artery feeds a left lower lobe pulmonary sequestration. In (B), 2 arteries (B) arising from the thoracic aorta feed the left lower lobe sequestration. In (C and D), the single feeding vessel originating from the descending thoracic aorta bifurcates following entry into the PS. MIP=maximum intensity projection, PS=pulmonary sequestration.

ranges between 0.15% and 1.8%.^[3,4] Identification of abnormal systemic blood supply is critical for the diagnosis. The entity was named by Pryce in 1946, who hypothesized an etiology related to vascular traction whereby a persistent abnormal aortic branch pulled with it embryonic lung tissue during development.^[5]

PS typically occurs in the left lower lobe and right lower lobe, rarely in the right middle lobe. The feeding artery can originate from either the thoracic or abdominal aorta.^[6,7] Bilateral sequestrations are rare.^[8,9] A few case reports have reported PS blood supply from an aberrant fissural artery, the internal mammary artery, and even the right coronary artery.^[10–13]

In the present study, 17 cases of PS were located in the posterior basal segment of the left lower lobe and 6 in the medial basal segment of right lower lobe (left/right: 16/7). The sequestrations supplied by the thoracic aorta were preferentially found in the left lower lobe, whereas those supplied by the abdominal aorta were evenly distributed between the left and right lower lobes.

PS is divided into 2 types depending on the relationship of the sequestered lung with the pleura. Intralobar sequestrations do not have pleura separate from the adjacent normal lung and are predisposed to infection. Extralobar PS are less common, separated from surrounding normal lung by their own pleura, commonly asymptomatic, and often found incidentally on imaging or routine physical examination.^[7] In this study, all PS were intralobar, and as expected, the majority were associated with symptoms of recurrent cough and fever; although, 4 cases were incidentally discovered during a routine medical examination. On imaging, the key to accurate diagnosis of PS is to identify the aberrant systemic arterial supply. In the past, DSA was the gold standard for diagnosis of PS, but given the high specificity and sensitivity of non-invasive modalities such as CTA and MRA, DSA has been supplanted and is rarely used in this application.^[14–18]

Timely surgical treatment should be considered for most patients with PS, especially for patients with recurrent pulmonary infections or in whom there is concern for malignancy.^[19] The morphological features of PS (volume, feeding artery length and diameter) are important for pre-surgical planning. In the present study, the mean PS volume was smaller in those with a blood supply from the thoracic aorta. Feeding arteries from the thoracic aorta were greater in diameter but shorter than those arising from the abdominal aorta. The feeding artery is most commonly solitary but was duplicated in a minority of cases. In each case, both feeding arteries arose from the thoracic aorta. In 2 additional cases, a single artery arose from the thoracic aorta bifurcating upon entry into the PS. Failure to identify and report these variations could complicate surgical treatment. Catheter embolization is a minimally invasive treatment alternative for PS in neonates,^[20] and pre-embolization planning and identification of all feeding arteries is critical for this technique as well.

There were some limitations in this study. First, Small sample in this study prevented us to generalize the results. Second, Patients were exposed to ionizing radiation in CTA scanning. Last, this study is a retrospective cohort.

In conclusion, PS with blood supply from the thoracic aorta are preferentially located in the left lower lobe, tend to be larger in size, and are fed by larger caliber arteries than PS with blood supply from the abdominal aorta.

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Author contributions

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References

- [1] Landing BH, Dixon LG. Congenital malformations and genetic disorders of the respiratory tract (larynx, trachea, bronchi, and lungs). *Am Rev Respir Dis* 1979;120:151–85.
- [2] Clements BS, Warner JO. Pulmonary sequestration and related congenital bronchopulmonary-vascular malformations: nomenclature and classification based on anatomical and embryological considerations. *Thorax* 1987;42:401–8.
- [3] Gezer S, Tastepe I, Sirmali M, et al. Pulmonary sequestration: a single-institutional series composed of 27 cases. *J Thorac Cardiovasc Surg* 2007;133:955–9.
- [4] Cooke CR. Bronchopulmonary sequestration. *Respir Care* 2006;51:661–4.
- [5] Pryce DM. Lower accessory pulmonary artery with intralobar sequestration of lung; a report of seven cases. *J Pathol Bacteriol* 1946;58:457–67.
- [6] Van Raemdonck D, De Boeck K, Devlieger H, et al. Pulmonary sequestration: a comparison between pediatric and adult patients. *Eur J Cardiothorac Surg Off J Eur Assoc Cardiothorac Surg* 2001;19:388–95.
- [7] Wei Y, Li F. Pulmonary sequestration: a retrospective analysis of 2625 cases in China. *Eur J Cardiothorac Surg Off J Eur Assoc Cardiothorac Surg* 2011;40:e39–42.
- [8] Oliveri Aruete F, Candelario Caceres A, Alonso Mallo E. Bilateral intralobar pulmonary sequestration in a young adult. a case report. *Arch Bronconeumol* 2017;53:281–2.
- [9] Wu L, Lin L, Jiang L, et al. Bilateral intralobar pulmonary sequestration with a bridging isthmus in a young adult. *Ann Thorac Surg* 2016;102:e257.
- [10] Hamaji M, Burt BM, Ali SO, et al. An incidental and uncommon pulmonary sequestration with an uncommon feeding artery. *Int J Surg Case Rep* 2013;4:861–2.
- [11] Xie D, Xie H, You X, et al. Pulmonary sequestration with aberrant arteries arising from the renal artery and the internal thoracic artery. *Ann Thorac Surg* 2013;96:e131.
- [12] Laborda A, Ruiz JR, de Gregorio MA. Congenital pulmonary sequestration supplied by the right coronary artery. *Arch Bronconeumol* 2016;52:389.
- [13] Van Langenhove G, Convens C, Seynaeve P, et al. Intralobar pulmonary sequestration supplied by the right coronary artery. *Catheter Cardiovasc Interv Off J Soc Cardiac Angiograph Interv* 1999;47:218–20.
- [14] Buyukoglan H, Mavili E, Tutar N, et al. Evaluation of diagnostic accuracy of computed tomography to assess the angioarchitecture of pulmonary sequestration. *Tuberk Toraks* 2011;59:242–7.
- [15] Franco J, Aliaga R, Domingo ML, et al. Diagnosis of pulmonary sequestration by spiral CT angiography. *Thorax* 1998;53:1089–92.
- [16] Au VW, Chan JK, Chan FL. Pulmonary sequestration diagnosed by contrast enhanced three-dimensional MR angiography. *Br J Radiol* 1999;72:709–11.
- [17] Xu H, Jiang D, Kong X, et al. Pulmonary sequestration: three dimensional dynamic contrast-enhanced MR angiography and MRI. *J Tongji Med Univ Tong Ji Yi Ke Da Xue Xue Bao* 2001;21:345–8.
- [18] Zhang M, Zhu J, Wang Q, et al. Contrast enhanced MR angiography in pulmonary sequestration. *Chin Med J* 2001;114:1326–8.
- [19] Bratu I, Flageole H, Chen MF, et al. The multiple facets of pulmonary sequestration. *J Pediatr Surg* 2001;36:784–90.
- [20] Lee KH, Sung KB, Yoon HK, et al. Transcatheter arterial embolization of pulmonary sequestration in neonates: long-term follow-up results. *J Vasc Interv Radiol J Vasc Interv Radiol* 2003;14:363–7.