Original Article

Departments of Respiratory Medicine and [1]Pathology, Peking Union Medical College Hospital, Peking Union Medical College and Chinese Academy of Medical Sciences, [2]Department of Rheumatology and Clinical Immunology, Peking Union Medical College Hospital, Peking Union Medical College and Chinese Academy of Medical Sciences, Key Laboratory of Rheumatology and Clinical Immunology, Ministry of Education, Beijing, China

Address for correspondence:
Prof. Jing Li, Department of Rheumatology and Clinical Immunology, Peking Union Medical College Hospital, Peking Union Medical College and Chinese Academy of Medical Sciences, Key Laboratory of Rheumatology and Clinical Immunology, Ministry of Education, Beijing 100032, China. E-mail: lijing6515@pumch.cn
Prof. Baiqiang Cai, Department of Respiratory Medicine, Peking Union Medical College Hospital, Peking Union Medical College and Chinese Academy of Medical Sciences, Beijing, 100730, China. E-mail: caibq2009@hotmail.com

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Anomalous systemic arterial supply of pulmonary sequestration in adult patients

Xiaomeng Hou, Ji Li[1], Jing Li[2], Baiqiang Cai

Abstract:

OBJECTIVES: This study described the characteristics of the systemic arterial supply of pulmonary sequestration (PS) in an attempt to better distinguish PS from other acquired lesions.

METHODS: We identified 25 patients hospitalized at the Peking Union Medical College Hospital during January 2013 to December 2015 with the assistance of medical catalogers. Twenty-three patients with a definite diagnosis of "pulmonary sequestration" clinically or pathologically were included in the study. The medical records, imaging information, and pathological data were reviewed retrospectively. The general characteristics of the patients and the features of the anomalous arteries were summarized.

RESULTS: Aberrant arterial supply of PS was found in all 23 (100%) cases. Among them, twenty patients received surgery, including 14 (70%) with aberrant arterial supply found before surgery, and the other 6 (30%) found during surgery. Nineteen (82.6%) patients had a single systematic arterial supply, with a median diameter of 8 mm. More than one arterial supplies were found in four (17.4%) cases. In 21 (91.3%) cases, the anomalous systemic artery originated from the descending thoracic aorta just adjacent to the sequestrated lung which it supplied, without the presence of accompanying bronchi. In twenty (87.0%) patients who received the surgical intervention, samples of 12 (85.7%) were proved to have elastic vessel walls, out of the 14 samples in which the anomalous systemic arteries were available for analysis.

CONCLUSIONS: There are no certain pathology diagnostic criteria for the diagnosis of PS. The detecting of the aberrant systematic artery and distinguishing it from the bronchial arteries corresponded to certain lung abnormalities are the keys to the accurate diagnosis of pulmonary sequestration in adult patients. We propose that the characteristic features of the anomalous arteries include: Originating from aorta and its main branches, adjacent to the sequestrated area, directly running into the sequestrated mass without accompanying bronchus branch, being large in diameter, and having elastic vessel wall.

Key words:

Aberrant systematic artery, congenital anomalous artery, pulmonary sequestration

Pulmonary sequestration (PS) is a rare congenital malformation which constitutes 0.15–6.4% of all congenital pulmonary malformations.[1] In the year 1946, Pryce coined the term sequestration to describe "disconnected bronchopulmonary mass or cyst with an anomalous systemic artery supply."[2] PS is usually divided into two types, the intralobar pulmonary sequestration (ILS), which shares the visceral pleura with an otherwise normal pulmonary lobe and the extralobar pulmonary sequestration (ELS), in which the extralobar sequestration is separated from normal lung tissue with an independent pleural covering.[3] To establish the diagnosis of classical PS, the presence of an aberrant systemic artery supply is essential. The assessment of the congenital features of these arteries, however, still lacks elucidation. In this retrospective study, we describe the characteristics of the systemic arterial supply in PS patients in an attempt to better distinguish PS from other acquired lesions, such as bronchiectasis.

Methods

We performed a systemic search with the assistance of medical catalogers, identifying 25 patients with PS hospitalized in the Peking Union Medical College Hospital, a top referral center in China, during January 2013 to December 2015. Only patients over 16 years old

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were eligible in this study. Medical records of the patients were reviewed retrospectively. Twenty-three patients with a definite diagnosis of "pulmonary sequestration" clinically or pathologically (ICD-10 code: Q33.201 or Q25.752) were included in the study. Two patients without a definite diagnosis of "pulmonary sequestration" were excluded from the study.

PS was defined as an area of lung, the parenchyma of which connected or disconnected with the bronchial system, is supplied by an aberrant artery arising from the aorta or one of its side branches.[2,4]

The clinical characteristics of all patients were collected from their medical records. Imaging information of the aberrant arterial supply of PS was recorded from the contrast-enhanced computed tomography (CT), which was extracted from an image database of our hospital. Twenty patients have received surgical operation for PS in our center. All pathological analysis results of the sequestrated tissue from a surgical procedure, both in gross anatomy and microscopic examination of the slides, were reviewed by a senior consultant pathologist. The diameters, origins, types of vessel wall, and accompanying components, i.e., bronchi and veins, of the anomalous arteries were summarized.

The study protocol was approved by the Institutional Review Board of Peking Union Medical College Hospital. Since this study was based on a review of medical records that had been obtained for the clinical purpose, the requirement of written informed consent was waived.

Numerical variables were described as mean values ± standard deviation or median (range), while categorical variables were described as number (percentage). The analysis was performed with the Microsoft Excel 2010 (Microsoft Corp., Redmond, WA, USA).

Results

A total of 23 patients with PS were qualified to be included in this study. The median age was 38.5 years (range: 21 years to 84 years) with a predominance of females (61%). All 23 patients were identified to have ILS, with a majority of 18 cases (78.3%) locating at the left lower lobe of the lung. The rest five cases (21.7%) showed an ILS location in the right lower lobe of the lung. The most common symptom of PS was a cough or expectoration (15/23, 65.2%), followed by hemoptysis (8/23, 34.8%), fever (6/23, 26.1%), and dyspnea (1/23, 4.3%). Furthermore, four cases (17.4%) were asymptomatic [Table 1].

Twenty patients (87.0%) received surgical treatment. The other three patients (13.0%) did not receive operation due to advanced age, asymptomatic clinical presentation, or declined consent to the surgery. Among the twenty patients who underwent operations, their surgical approaches included video-assisted thoracoscopic surgery in 13 cases (65.0%) and thoracotomy in seven cases (35.0%). Types of resection included segmentectomy in three cases (15.0%) and lobectomy in 16 cases (80.0%). One patient underwent aberrant systemic artery disconnection after reassuring the function of corresponding pulmonary artery and vein. Five patients (25%) developed postoperative complications, including four patients (20%) with hydrothorax and one patient (5%) with incision infection. The postoperative hospital stay of these patients ranged from 3 to 17 days, with a median stay of 6 days [Table 2].

Aberrant arterial blood supply profiles for the PS were identified in all 23 cases. Different technologies including contrast-enhanced CT in 13 cases (56.5%) preoperatively, CT angiography in four cases (17.4%) preoperatively [Figure 1a-c], and intraoperative observation in six cases (26.1%) were used in this study. Among the twenty patients who received surgery, the aberrant systemic artery was identified before surgery

Table 1: Demographic data and clinical manifestations of patients with pulmonary sequestration

Items studied	Number of case (%)
Number of cases diagnosed per year during 2013–2015	7.67
Male/female ratio	0.64:1
Age	
Median (range), years	38.5 (21–84)
Type (ILS/ELS)	23/0
Localization (LLL/RLL)	18/5
Clinical symptoms	
Cough and expectoration	15 (65.2)
Hemoptysis	8 (34.8)
Intermittent fever	6 (26.1)
Asymptomatic	4 (17.4)
Dyspnea	1 (4.3)

ILS=Intralobar pulmonary sequestration, ELS=Extralobar pulmonary sequestration, LLL=Left lower lobe, RLL=Right lower lobe

Table 2: Perioperative conditions of twenty pulmonary sequestration patients who received surgical operations

Perioperation conditions	Number of cases (%)
Preoperation diagnosis	
PS	14 (70)
Bronchiectasis	3 (15)
Lung mass	2 (10)
Pulmonary infection	1 (5)
Postoperation diagnosis	
PS	20 (100)
Surgical approach	
VATS	13 (65)
Thoracotomy	7 (35)
Types of resection	
Lobectomy	16 (80)
Segmentectomy	3 (15)
Vessel disconnection	1 (5)
Postoperative complications	
Hydrothorax	4 (20)
Incision infection	1 (5)
Postoperative stay (median [range], days)	6 (3–17)

PS=Pulmonary sequestration, VATS=Video-assisted thoracoscopic surgery

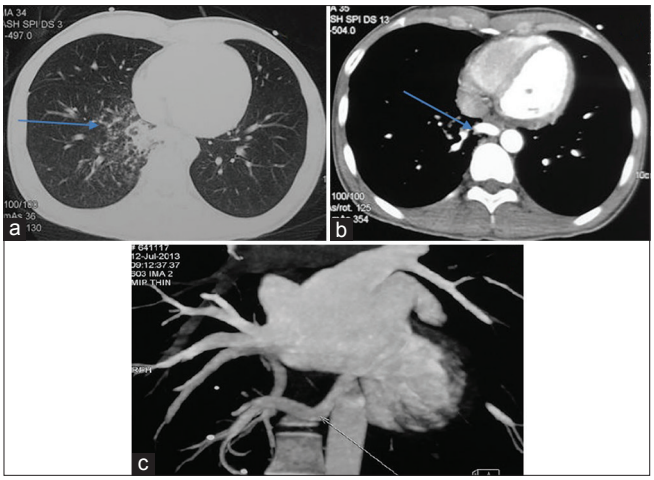

Figure 1: A 34-year-old male with recurrent infection in the right lower lung. Focal ground-glass density and patchy opacity at the right lower lung are evident on axial computed tomography (a) (fine arrow). An aberrant artery arising from the descending thoracic aorta, supplying the medial right lower lobe (b) (fine arrow). An aberrant artery arising from the descending thoracic aorta, running directly into the medial right lower lobe on computed tomography angiography and three-dimensional reconstruction of thoracic aorta (c) (fine arrow)

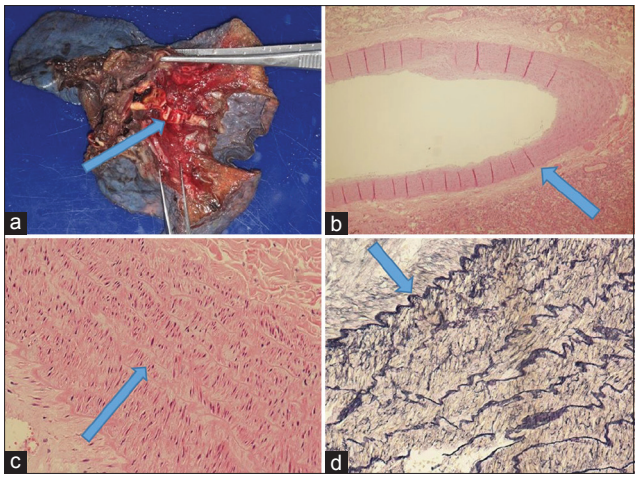

Figure 2: A 37-year-old male patient with pulmonary sequestration, samples taken from the left lung resection. (a) Opened visceral pleura of the lung, with one artery with a diameter of 10 mm visible (blue arrow). (b) A vessel visible in the lung tissue. Microscopic examination confirmed an artery (blue arrow) in its essence (H and E, ×30). (c) Multilayer elastic fiber (blue arrow) visible in the tunica media of the artery (H and E, ×150). (d) Multilayer elastic fiber (blue arrow) with positive staining visible in the tunica media of the artery (elastic fiber staining, ×150)

in 14 cases (70.0%) and seen during surgery in the other six cases (30.0%). Thirteen cases (13/15, 86.7%) were diagnosed before operation by contrast-enhanced CT, while four patients (4/4, 100%) were diagnosed by CT angiography. Four patients (4/4, 100%) were diagnosed during operation with CT noncontrast-enhanced scan only before the intervention. No digital subtraction angiography was performed to identify the anomalous artery in this study.

All anomalous systemic arteries in this study originated from the descending thoracic aorta. A majority of the PS (19 cases, 82.6%) have a single systematic arterial supply, the diameter of which ranging from 3 to 14 mm (median diameter: 8 mm). Two systematic arteries were found in one case (4.3%), while multiple systematic arterial supplies were found in three cases (13.0%). Most patients (21 cases, 91.3%) had their anomalous systemic arteries originating from the thoracic descending aorta just adjacent to the sequestrated lung which it supplied, without entering the hilum or the presence of accompanying bronchi [Figure 1b and c]. In the other two cases (8.7%), the systemic artery originated from the hilum level of the aorta, running a winding course to the sequestrated area with a bronchus. In the twenty patients who received the surgical intervention, fourteen samples taken during the procedures were considered to contain anomalous systemic arteries by the primary pathologists, among which 12 (85.7%) were proved to have elastic vessel walls when reviewed by the consultant pathologist [Figure 2a-d] [Table 3].

Table 3: Characteristics of anomalous arteries in pulmonary sequestration patients of the present study

Characteristics	Number of cases (%)
Total	23
Number of AA	
Single	19 (82.6)
Double	1 (4.3)
Multiple	3 (13.0)
Origin of AA	
DTA	23 (100)
Level of the sequestrated area	21 (91.3)
Level of hilum	2 (8.7)
Diameter of AA (median [range]), (mm)	8 (3–14)*
AA with elastic vessel wall	12 (85.7)#
AA accompanied with or without bronchus	
Accompanied without bronchus	21 (91.3)
Accompanied with bronchus	2 (8.7)

*Figures from the 19 cases with single AA, #Figures from 14 samples in which the anomalous systemic arteries being available for analysis. AA=Anomalous arteries, DTA=Descending thoracic aorta

Discussion

In the year 1946, Pryce coined the term sequestration.[2] After that many theories of the pathogenesis of PS have been proposed, while the most widely accepted hypothesis suggested that PS was the consequence of an insult to the tip of developing lung buds. In early developing stages, the tips of the dividing bronchial buds are supplied by systemic capillary plexus derived from the primitive aorta. This plexus regresses with further lung maturation as the developing pulmonary artery takes over.[5-7] The nature of the insult, probably more importantly the timing and severity of the insult, determines the morphology of the final lesions.[4,8]

PS is categorized into ILS and ELS. ILS is the most common form, constituting 92.5% of series from our hospital from 1990 to 2013 by Sun and Xiao[9] With the development of diagnostic technology in China, increasing cases of PS, especially ELS, have been diagnosed correctly and given proper treatment in other local hospitals. Therefore, there were no ELS patients included in our series. It also suggests that ILS is more difficult to be distinguished. PS are manifested as a cough, purulent sputum, fever, etc.[10] They all have symptoms

Table 4: Characteristics of anomalous arteries in pulmonary sequestration patients reported from other case series

	Publication year	Number of cases	Number of AA	Origin of AA	Diameter of AA (mm)	AA with elastic vessel wall	AA accompanied with interstitial pulmonary components
Savic *et al*.[1]	1979	547	318 with single, 55 with more than one	318 from DTA 99 from AbA 16 from intercostal arteries	6.3 (160 from TA) 6.6 (46 from AbA)	Most/60 descriptions of the histological structure	#
Tsolakis and Kollias[11]	1997	8	#	5 from DTA 2 from AbA 1 from intercostal arteries	#	2/2 ELS #/6 ILS	#
Yamanaka *et al*.[12]	1999	4	4 with single	4 from DTA	7, 8, 15, 5 respectively	#	#
Wei and Li[10]*	2011	2625	643 with single, 130 with two, 40 with multiple	1384 from TA 334 from AbA 36 from intercostal artery 28 from diaphragmatic artery 8 from aortic arch 6 from subclavian artery 4 from left gastric artery 2 from coronary artery 1 from celiac trunk	#	#	#
Mori *et al*.[13]	2013	4	4 with single	4 from DTA	10, 16, 15, 15 respectively	#	#
Sun and Xiao[9]	2015	72	#	62 from TA, 5 from AbA, 4 from phrenic artery, 1 from intercostal artery	#	#	#

*This is a meta-analysis article, #No data. TA=Thoracic aorta, DTA=Descending thoracic aorta, AbA=Abdominal aorta, AA=Anomalous arteries

similar to that of pneumonia. PS patients were susceptible to pulmonary infections. Without the presentation of pulmonary infection, PS is asymptomatic and only diagnosed incidentally as with 17.4% of patients in our study.[1]

A number of case series on PS were reviewed along with this study.[1,9-13] The characteristics of anomalous arteries described in this literature are summarized in Table 4. Few had done a pathological study for constituents of the vessel wall. The accompanying component of the anomalous systemic arteries in PS had not been studied in the previous publications [Table 4]. The systemic arteries of PS are potentially dangerous and should be identified before surgery. In this study, the aberrant systemic arteries were detected before surgery in 14 cases (70.0%) by contrast-enhanced CT (13/15, 86.7%) or CT angiography (4/4, 100%) and seen during surgery in the other six (30%), including four patients had taken CT noncontrast-enhanced before operation, and two patients (2/15, 13.3%) had taken contrast-enhanced CT. It is suggestive that a contrast-enhanced CT or CT angiography before the operation is essential for detecting potential abnormal arterial vessels. CT angiography is more useful to disclose PS. For patients with a pulmonary mass of unknown etiology, the possibility of PS should be considered preoperatively. A targeted observation of the systemic blood supply by enhanced CT or CTA is very important to reduce the risk of fatal intraoperative bleeding.

In PS, the aberrant arteries with elastic vessel walls represent a congenital anomaly, which cannot be reproduced by any form of interference to the pulmonary artery circulation. In dog models, in response to the ligation of the pulmonary artery, a large number of bronchial arteries with vessel walls of muscular nature were formed.[14,15] Several investigators also indicated that the aberrant arteries in PS are distinct from bronchial arteries.[1,4] A preponderance of elastic vessel walls in the aberrant arteries was identified in our study, consistent with the findings reported by Savic *et al*.[1] This evidence could prove crucial in recognizing the congenital features of anomalous arterial supplies, as well as distinguishing it from bronchial arteries.

The aberrant systemic arteries of ILS are usually large in diameter. In Savic's case series, the mean diameter values of the aberrant arteries were 6.3 mm (originating from the thoracic aorta) and 6.6 mm (originating from the abdominal aorta).[1] The mean diameter values of the anomalous arteries in Yamanaka's literature review were 9 mm at the origin, further dilating aneurysmatically (mean 20 mm in diameter) when entering the sequestrated lobe.[12] In our study, the median diameter of the aberrant arteries was 8 mm, far exceeding the normal value of bronchial arteries.[16] A relatively large diameter is also consistent with a congenital origin of the aberrant arterial supply.

In our study, a majority of the aberrant systemic arteries originated from the descending aorta. Moreover, the origins of these aberrant arteries were close to the level of the sequestrated mass in the lung, leading them directly to the sequestrated region as a supply. This finding suggests that the aberrant

artery and the sequestrated lung mass are closely related in position during embryonic development.[4,17]

In general, pulmonary arteries and bronchial arteries go through the hilum in accompany with bronchi. Most of the aberrant systemic arteries examined in our cases penetrated directly the adjacent pleura to the sequestrated lung, without going through the hilum nor accompanied by any bronchial branch[16,17] indicating that the aberrant arteries may originate from the systemic capillary plexus like the hypothesis,[5-7] presenting a different way of evolution.

Conclusions

PS in many patients is underdiagnosed until their adulthood. There are no certain pathology diagnostic criteria for the diagnosis of PS. Detection of special systemic arteries plays a decisive role in the PS diagnosis. Some bronchial arteries corresponding to certain lung abnormalities could be confused with the anomalous systemic arteries of PS. In some cases extremely difficult to distinguish. In the light of this study, the representative features of the anomalous arteries in PS are summarized, including originating from aorta adjacent to the sequestrated area, directly supplying the sequestrated mass without accompanying bronchi, being large in diameter, and having elastic vessel walls histologically. Moreover, these features are not presented in bronchial arteries.

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Conflicts of interest
There are no conflicts of interest.

References

1. Savic B, Birtel FJ, Tholen W, Funke HD, Knoche R. Lung sequestration: Report of seven cases and review of 540 published cases. Thora×1979;34:96-101.
2. Pryce DM. Lower accessory pulmonary artery with intralobar sequestration of lung; a report of seven cases. J Pathol Bacteriol 1946;58:457-67.
3. Abbey P, Das CJ, Pangtey GS, Seith A, Dutta R, Kumar A. Imaging in bronchopulmonary sequestration. J Med Imaging Radiat Oncol 2009;53:22-31.
4. Clements BS, Warner JO. Pulmonary sequestration and related congenital bronchopulmonary-vascular malformations: Nomenclature and classification based on anatomical and embryological considerations. Thora×1987;42:401-8.
5. Boyden EA. Bronchogenic cysts and the theory of intralobar sequestration: New embryologic data. J Thorac Surg 1958;35:604-16.
6. Reid LM. Lung growth in health and disease. Br J Dis Chest 1984;78:113-34.
7. Gerbeaux J. Paediatric Respiratory Disease. 3rd ed. London: John Wiley and Sons; 1985.
8. Stovin PG. Early lung development. Thorax 1985;40:401-4.
9. Sun X, Xiao Y. Pulmonary sequestration in adult patients: A retrospective study. Eur J Cardiothorac Surg 2015;48:279-82.
10. Wei Y, Li F. Pulmonary sequestration: A retrospective analysis of 2625 cases in China. Eur J Cardiothorac Surg 2011;40:e39-42.
11. Tsolakis CC, Kollias VD, Panayotopoulos PP. Pulmonary sequestration. Experience with eight consecutive cases. Scand Cardiovasc J 1997;31:229-32.
12. Yamanaka A, Hirai T, Fujimoto T, Hase M, Noguchi M, Konishi F. Anomalous systemic arterial supply to normal basal segments of the left lower lobe. Ann Thorac Surg 1999;68:332-8.
13. Mori S, Odaka M, Asano H, Marushima H, Yamashita M, Kamiya N, *et al*. Anomalous systemic arterial supply to the Basal segments of the lung: Feasible thoracoscopic surgery. Ann Thorac Surg 2013;96:990-4.
14. Liebow AA, Hales MR, Bloomer WE, Harrison W, Lindskog GE. Studies on the lung after ligation of the pulmonary artery; anatomical changes. Am J Pathol 1950;26:177-95.
15. Smith RA. A theory of the origin of intralobar sequestration of lung. Thora×1956;11:10-24.
16. Osiro S, Wear C, Hudson R, Ma XX, Zurada A, Michalak M, *et al*. A friend to the airways: A review of the emerging clinical importance of the bronchial arterial circulation. Surg Radiol Anat 2012;34:791-8.
17. Hislop AA. Airway and blood vessel interaction during lung development. J Anat 2002;201:325-34.