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Pulmonary sequestration: a retrospective analysis of 2625 cases in China

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Abstract

Objective: Pulmonary sequestration is a congenital lung malformation, which is often misdiagnosed as lung cancer, pulmonary cysts, mediastinal tumors, etc. Therefore, more research on the clinical characteristics of pulmonary sequestration should be carried out to improve the preoperative diagnosis rate. **Methods:** The study used was a retrospective analysis of 2625 cases of pulmonary sequestration well documented in the Chinese National Knowledge Infrastructure from 1998 to 2008. Analysis was performed on the patients' age, gender, symptom, chest computed tomography (CT) scan, chest radiograph, lesion localization, arterial supply, venous drainage, and incorrect preoperative diagnosis. **Results:** A total of 2625 cases of pulmonary sequestration was reported in the Chinese National Knowledge Infrastructure from 1998 to 2008, and the male:female ratio was 1.58:1. The symptoms of pulmonary sequestration were cough, sputum, fever, hemoptysis, and chest pain, with 13.36% of patients being asymptomatic. Chest CT scan showed mass lesions (49.01%), cystic lesions (28.57%), cavitary lesions (11.57%), and pneumonic lesions (7.96%). The sequestration was mainly located in the lower lobe, primarily in the left posterior basal segment (66.43%) and in the right posterior basal segment (20.16%). Pulmonary sequestrations were divided into two types, intralobar sequestration (83.95%) and extralobar sequestration (16.05%). Bilateral sequestrations were rare – only three cases had been reported. The arterial supply was mainly provided by branches of thoracic aorta (76.55%) and abdominal aorta (18.47%). The mean incorrect preoperative diagnosis rate was as high as 58.63%. A comparison between pediatric and adult patients indicated that the subtype ratio (intralobar/extralobar) was higher in the adult group than that in the pediatric group ($P < 0.001$). **Conclusions:** Clinical manifestations of pulmonary sequestration varied and preoperative diagnosis was often incorrect. To improve the preoperative diagnosis rate, we should take full advantage of symptoms, image performance, and localization characteristics. A certain early-onset age, recurrent pneumonias, mass or cyst lesion located in the lower lobe, and aberrant arterial supply are indicators for the diagnosis.

Keywords: Sequestration; Pulmonary; Congenital disorders; Neonatal abnormalities

1. Introduction

Pulmonary sequestration is a relatively rare congenital lung malformation. It is usually classified as either extralobar (ELS) or intralobar (ILS). ELS is separated from normal lung tissue by its own visceral pleura, whereas ILS is incorporated within normal lung tissue [1]. The numerous published reports dealt only with a small number of cases. Its clinical manifestations were varied, with a high, incorrect preoperative diagnosis rate. We researched on the clinical manifestations, diagnosis, and treatment characteristics of pulmonary sequestration through retrospective analysis of reported cases in the Chinese National Knowledge Infrastructure (CNKI) from 1998 to 2008, to subsequently improve the preoperative diagnosis rate.

2. Material and method

This study was a retrospective analysis of 2625 pulmonary sequestrations, which were published in the CNKI between 1998 and 2008. CNKI is the largest full-text databank of Chinese journals and the biggest medical digital library in China. The library collects material and provides information and research services to all areas of biomedicine and healthcare. Only reports on well-documented cases were considered. Separate statistic analysis was performed on patients' age, gender, symptom, chest computed tomography (CT) scan, chest radiograph, lesion localization, arterial supply, venous drainage, and incorrect preoperative diagnosis. The Fisher's exact test was used for comparing enumeration variables. $P < 0.05$ was considered statistically significant.

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3. Results

3.1. Sex distribution and age at diagnosis

A total of 2625 cases of pulmonary sequestration was reported in the CNKI from 1998 to 2008, including 1607 men and 1018 women. The male:female ratio was 1.58:1. The minimum and maximum ages at diagnosis were 1 month and 77 years, respectively. Age at diagnosis was different for ILS versus ELS sequestrations. ILS sequestrations were easily acquired pulmonary infections for having a connection to the tracheobronchial tree, thus leading to diagnosis at an early age (20 ± 8 years old). However, ELS sequestrations were generally asymptomatic or accidentally discovered on routine physical examination, resulting in diagnosis at a later age (38 ± 9 years old).

3.2. Symptoms of sequestration

Out of the 2625 cases of pulmonary sequestration, there were 1923 cases which reported specific symptoms. The main symptoms of pulmonary sequestration were cough or expectoration, fever, hemoptysis, and chest pain. However, some were asymptomatic or accidentally discovered on routine physical examination. Because some patients had two or three types of symptoms at the same time, we needed to use the clinical manifestation rate (CMR) to show the probability of one specific symptom appearing in 1923 patients. CMR = (Cases of one specific symptom/1923) $\times$ 100%. The most common symptom of pulmonary sequestration was cough or expectoration (1303 cases, CMR was 67.76%), the second was fever (749 cases, CMR was 38.95%), the third was hemoptysis (532 cases, CMR was 27.67%), and the fourth was chest pain (214 cases, CMR was 11.13%). Furthermore, 257 cases were asymptomatic or accidentally discovered on routine physical examination, whose CMR was 13.36%. More details are shown in Table 1.

3.3. Imaging of sequestration

As many as 1106 cases of pulmonary sequestration were characterized by chest CT scan. They were mainly manifested into four categories: (1) mass lesion (542 cases, 49.01%); (2) cystic lesion (316 cases, 28.57%); (3) cavitary lesion (128 cases, 11.57%); and (4) pneumonic lesion (88 cases, 7.96%). More details are shown in Table 2.

Table 1. Symptoms of pulmonary sequestration (n = 1923).

Symptoms	No. of cases	CMR[a] (%)
Cough or expectoration	1303	67.76
Fever	749	38.95
Hemoptysis	532	27.67
Chest pain	214	11.13
Chest tightness or shortness of breath	92	4.78
Other symptoms[b]	14	0.73

[a] Clinical manifestation rate (CMR) meant the probability of one specific symptom appearing in 1923 patients. CMR = (Cases of one specific symptom/1923) $\times$ 100%.

[b] Other symptoms in 14 cases, including 11 cases of fatigue, two cases of weight loss, and one case of back pain.

Table 2. Chest CT appearances of pulmonary sequestration (n = 1106).

Chest CT appearances	No. of cases	Proportion (%)
Mass lesion	542	49.01
Cystic lesion	316	28.57
Cavitary lesion	128	11.57
Pneumonic lesion	88	7.96
Bronchiectasis	21	1.90
Encapsulated pleural effusion	6	
Other appearances[a]	5	

[a] Other appearances in five cases, including three cases of intrapulmonary cord-like shadow, and two cases of pulmonary atelectasis.

There were 1159 cases of pulmonary sequestration that had chest radiograph reports. They were mainly manifested into several categories: (1) mass lesion (537 cases, 46.33%); (2) cystic lesion (424 cases, 36.58%); (3) cavitary lesion (69 cases, 5.87%); (4) pneumonic lesion (118 cases, 10.18%); and (5) other appearances: nine cases of bronchiectasis, one case of thick lung markings, and one case of cord-like shadow.

3.4. Localization of pulmonary sequestration

There were 2037 cases of pulmonary sequestration that had lobe localization reports, including 1457 cases of left lower lobe (71.53%), 529 cases of right lower lobe (25.97%), 38 cases of left upper lobe (1.87%), five cases of left lingual lobe, six cases of right upper lobe, and two cases of right middle lobe.

There were 858 cases of pulmonary sequestration that had lung segment localization reports. They were mainly located in the posterior basal segment of the lower lobes. There were 570 cases of left posterior basal segment (66.43%) and 173 cases of right posterior basal segment (20.16%). More details are shown in Table 3.

3.5. Subtypes of pulmonary sequestration

Pulmonary sequestrations could be divided into two types: ILS and ELS sequestration. There were a total of 2231 cases of unilateral sequestration that had subtype reports, including 1873 cases of ILS sequestration (83.95%) and 358 cases of ELS sequestration (16.05%). Bilateral cases were rare. Only three bilateral cases had been reported, two were ILS bilaterally and one was mixed ILS/ELS bilaterally.

Table 3. Lung segment localization of pulmonary sequestration (n = 858).

Lung segment Localization	No. of cases	Proportion (%)
Posterior basal segment of left lower lobe	570	66.43
Posterior basal segment of right lower lobe	173	20.16
Medial basal segment of left lower lobe	45	5.24
Medial basal segment of right lower lobe	18	2.10
Apical segment of left lower lobe	16	1.86
Apical segment of right lower lobe	11	1.28
Lateral basal segment of right lower lobe	12	1.40
Lateral basal segment of left lower lobe	3	
Posterior segment of Left upper lobe	3	
Superior segment of Left lingual lobe	2	
Inferior segment of Left lingual lobe	1	
Apical segment of Left upper lobe	1	
Anterior basal segment of right lower lobe	1	
Anterior segment of right upper lobe	1	
Posterior segment of right upper lobe	1	

Table 4. Incorrect preoperative diagnosis of pulmonary sequestration (n = 713).

Preoperative misdiagnosis	No. of cases	Proportion (%)
Pulmonary cyst	258	36.19
Lung cancer	150	21.04
Bronchiectasis	110	15.43
Pulmonary abscess	55	7.71
Pneumonia	37	5.19
Inflammatory pseudotumor	34	4.77
Mediastinal tumor	26	3.65
Pulmonary tuberculosis	22	3.09
Pulmonary bulla	11	1.54
Empyema	3	
Diaphragmatocele	3	
Hemothorax	2	
Hiatal hernia	1	
Arteriovenous fistula	1	

3.6. Arterial supply and venous drainage of pulmonary sequestration

A total of 1808 cases of pulmonary sequestration reported arterial supply. They originated from thoracic aorta (1384 cases, 76.55%), abdominal aorta (334 cases, 18.47%), intercostal artery (36 cases, 1.99%), diaphragmatic artery (28 cases, 1.55%), aortic arch (eight cases, 0.44%), subclavian artery (six cases), pulmonary artery (five cases), left gastric artery (four cases), coronary artery (two cases), and celiac trunk (one case).

A total of 813 cases of pulmonary sequestration reported the number of arterial supply, including 643 cases of single supplying artery (79.09%), 130 cases of two supplying arteries (15.99%), and 40 cases of over two supplying arteries (4.92%).

As many as 476 cases of pulmonary sequestration reported venous drainage, of which 433 cases reflowed to pulmonary vein (90.97%), 20 cases to azygos vein (4.20%), 18 cases to semi-azygos vein (3.78%), four cases to inferior vena cava (0.84%), and one case to diaphragmatic vein.

3.7. Incorrect preoperative diagnosis

A total of 713 cases of pulmonary sequestration reported incorrect preoperative diagnosis, 36.19% of which were misdiagnosed by pulmonary cyst and 21.04% were misdiagnosed by lung cancer. More details are shown in Table 4. There were 103 papers that reported incorrect preoperative diagnosis rate. The mean incorrect preoperative diagnosis rate was 58.63%.

3.8. A comparison between pediatric and adult patients

A total of 282 cases was diagnosed as adult or pediatric pulmonary sequestration expressly, with 132 adult and 150 pediatric cases. Subtype ratio (ILS/ELS) was higher in the adult group than that in the pediatric group ($P < 0.001$). Pediatric patients acquired symptoms more frequently than the adult patients ($P = 0.046$). There was no significant difference in sex or arterial supply between the two groups. More details are shown in Table 5.

4. Discussion

Pulmonary sequestration was first reported by Huber in 1877, which was named as 'sequestration' by Pryce in 1946 [2]. The pathogenesis of pulmonary sequestration was that a part of lung tissue developed into a separate lesion in the embryonic period, which had no respiratory function but had aberrant arterial supply [3].

Pulmonary sequestration was mainly located in the left lower lobe (71.53%) and the right lower lobe (25.97%). According to whether pulmonary sequestration and normal lung tissue had common visceral pleura, it could be divided into two types: ILS sequestration and ELS sequestration. The 83.95% of pulmonary sequestration was ILS sequestration, which shared common visceral pleura with the adjacent lung tissue. ELS sequestration accounted for 16.05%, which was wrapped by independent visceral pleura. Bilateral sequestrations were rare. Only three bilateral cases had been reported, two were ILS bilaterally and one was mixed ILS/ELS bilaterally. Subtype ratio (ILS/ELS) was higher in the adult group than that in the pediatric group; this was different from Raemdonck's review of 28 cases, which indicated no significant difference between the two groups [4].

Symptoms of pulmonary sequestration were varied. They were all symptoms reminiscent of pneumonia. ILS sequestrations were easily acquired pulmonary infections, manifested as cough, purulent sputum, fever, etc. However, ELS sequestration was generally asymptomatic or accidentally discovered on routine physical examination, which was often misdiagnosed as lung cancer [5], pulmonary cysts, or mediastinal tumors. The most useful diagnostic methods for pulmonary sequestration are CT angiography (CTA) (Fig. 1), magnetic resonance angiography (MRA), and digital subtraction angiography (DSA), which could show the aberrant arterial supply [6,7]. Furthermore, color Doppler also could show the supplying artery of pulmonary sequestrations that were close to diaphragm or liver. Ultrasonography is noninvasive and safe, making its use ideal in prenatal and postnatal settings. However, ultrasonographic findings are nonspecific in the antenatal and neonatal periods, and the differential diagnosis is wide in these findings. It also has limited value in adults with both ILS and ELS [8,9]. Selective DSA angiography could accurately show the aberrant arterial supply. Earlier, it was a gold diagnostic criterion for pulmonary sequestration. However, because of the high specificity and sensitivity of current CTA, MRA, and other noninvasive examination, invasive angiography

Table 5. Comparison between pediatric and adult patients.

	Total	Sex		Subtype		With symptoms		Arterial supply	
		M	F	IL	EL	Yes	No	TAA	Others
Adult group	132	66	66	116	16	92	40	126	6
Pediatric group	150	92	58	106	44	120	30	144	6
P value		0.056		<0.001		0.046		0.821	

M: male; F: female; IL: intralobar; EL: extralobar; and TAA: thoracic or abdominal aorta.

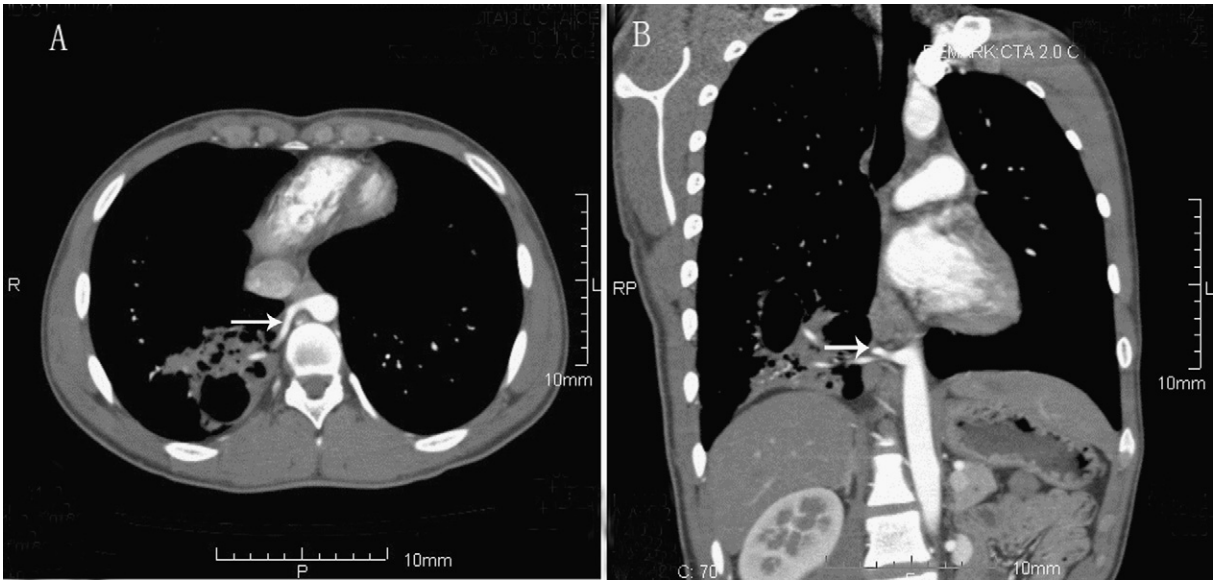

Fig. 1. Contrast-enhanced computer tomography imaging of pulmonary sequestration. (A) and (B) clearly showed the arterial supply originated from the descending aorta (arrow) and extended into the mass.

was even replaced by CTA or MRA [10,11]. Pulmonary sequestration was often misdiagnosed. The average incorrect preoperative diagnosis rate was 58.63%. Arterial supply of pulmonary sequestration mainly originated from thoracic aorta (76.55%) and abdominal aorta (18.47%). Venous drainage of pulmonary sequestration mainly reflowed to pulmonary veins (90.97%).

Timely surgical treatment should be considered for most patients, especially for patients with recurrent pulmonary infection or cancer that cannot be excluded [12]. ELS sequestration only needed tumor resection, but ILS pulmonary sequestration required lobectomy. Most of the pulmonary sequestration had single supplying artery, but 20.91% of them still had over one supplying arteries. Therefore, we should pay particular attention to the unusual supplying arteries. We should tightly suture the tissue which abnormal supplying arteries could not be excluded from, so as to prevent extensive bleeding. Video-assisted thoracic surgery (VATS) lobectomy for pulmonary sequestration was first reported by Wan et al. [13] in 2002. The need for thorough preoperative investigation should be emphasized in order to delineate the vascular anatomy, especially when a minimally invasive approach is to be adopted. In addition, the development of interventional treatment of pulmonary sequestration has opened up new methods. Lee et al. [14] succeeded in transcatheter arterial embolization of pulmonary sequestration in neonates. Compared with surgical treatment, interventional therapy and VATS were more patient friendly and less invasive.

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