A Comparative Study of Intralobar Sequestration and Extralobar Pulmonary Sequestration

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Research article

Keywords: Thoracoscopic surgery, Pulmonary sequestration, Intralobar sequestration

Posted Date: October 18th, 2021

DOI: https://doi.org/10.21203/rs.3.rs-963651/v1

Abstract

Objective: This study aims to identify the difference between patients who have been diagnosed with either intralobar sequestration (ILS) or extralobar sequestration (ELS).

Methods: In this clinical study, 29 children with pulmonary sequestration (PS), diagnosed via physical examination and imaging at our hospital between January 2019 and January 2020, were enrolled. We compared whether statistical differences existed in the blood loss, operative time, and post-operative hospital stay between the two groups (ILS and ELS) after thoracoscopic pulmonary wedge resection.

Results: There were no significant differences in gender, operative age, preoperative weight, and isolated lung position between the ILS and ELS groups ($p > 0.05$). There was significantly more intra-operative bleeding in children with ILS than those with ELS ($p < 0.05$), and the operation time and postoperative hospitalization times were significantly longer for those with ILS($p < 0.05$). Upon microscopic evaluation after surgery, we found the appearance of ILS and ELS to be similar.

Conclusion: Different types of congenital PS have different influences onnewborns during and after operation. For children with ILS, surgery is more difficult and the postoperative recovery is slower than for children with ELS. For this reason, we suggest that more attention should be paid to the clinical treatment of children with ILS. Although ILS and ELS present with different manifestations, we found no evidence of a significant difference in the postoperative microscopy of the two conditions.

1. Background

Pulmonary sequestration (PS) is a rare congenital lung malformation with an etiology that has not yet been clearly defined[1–4]. Specialists in China suggest that PS consists of congenital pulmonary parenchyma with pulmonary vascular abnormalities. PS can be divided into either intralobar (ILS) or extralobar (ELS) based on the presence or absence of visceral pleura at the border of normal lung tissue. Most ILS patients develop symptoms in adolescence, typically presenting with recurrent pneumonia, hemoptysis, and dyspnea. ELS has a complete pleura and is usually not connected to the bronchi of normal lung tissue, thus rarely becomes infected[5–7].

As significant differences exist between ILS and ELS (clinical symptoms, pathological anatomy, venous reflux), it is important to differentiate between the two when a patient is diagnosed with PS. In this study, we retrospectively analyzed clinical data regarding treatment characteristics, surgical procedures, and outcomes of 29 pediatric patients diagnosed with PS, who were treated in our center from January 2019 to January 2020, with the aim of improving the awareness and treatment of PS.

2. Materials And Method

We extracted data from the medical records of all patients diagnosed with PS who underwent the Video-assisted thoracoscopic surgery (VATS) procedure at the Department of Pediatric Surgery in The Third

Affiliated Hospital of Guangzhou Medical University between January 2019 and January 2020. In total, 29 patients were included, of which 17 were diagnosed with ILS and 12 with ELS. Continuous variables were described as the mean ± standard deviation and categorical variables were described as rate or composition ratio. The continuous variables between ILS and ELS were compared via independent t-test, and the continuous variables were compared via chi-square test. $P < 0.05$ was considered statistically significant. No human subjects were involved in this study and all data used was de-identified, thus, ethical review and informed consent were waived by the institutional review board of The Third Affiliated Hospital of Guangzhou Medical University.

3. Results

3.1 Baseline characteristics

Out of a total of 29 children who presented with PS and were managed during the study period, 17 (58.6%) were diagnosed with ILS and 12 (41.4%) with ELS. Demographic data for these patients is shown in Table 1. There were 17 (58.6%) males and 12 (41.4%) females, and the age at follow up ranged from 55 to 313 days (median = 183.96 days). The most common PS location was the left lower lobe, where 23 (79.3%) of the lesions were identified, and the most common PS feeding artery was thoracic aorta, where 18 (62.1%) of the lesions were identified.

Table 1 The clinical date of patients with pulmonary sequestration (n = 29)

Characteristic	
Type of sequestration(n)	
Intralobar	17
Extralobar	12
Abdominal	3
Location(n)	
Left lower lobe	23
Right lower lobe	6
Sex(n)	
Male	17
Female	12
Weight(kg),mean	7.74±1.94
Age(days),mean	183.96±128.67
Feeding artery(n)	
Thoracic Aorta	18
Abdominal Aorta	3
Not determined	8

3.2 Localization and arterial supply between ILS and ELS

In both ILS and ELS patients, the most common feeding artery was the thoracic aorta (ILS: 54%), and the lesions were most likely to occur in the left lower lobe (ILS: 54%). There was no significant difference in gender, location, and supplying artery between the two patient groups (Table 2).

Table 2 The comparison between ILS and ELS patients

Characteristic	ILS	ELS	*p*
Newborn Sex(n)			*p*=0.703
Male	9	8	
Female	8	4	
Location(n)			*p*=0.354
Left lower lobe	12	8	
Right lower lobe	5	1	
Abdominal	0	3	
Feeding artery(n)			
Thoracic Aorta	12	6	
Abdominal Aorta	2	1	
Not determined	3	5	
Weight(kg),mean	8.07±1.75	8.39±2.78	*p*=0.276

3.3 Surgical procedures between ILS and ELS patients

Thoracoscopic surgical resection was performed on all 29 patients. Of these 29 patients, none of the cases required conversion to thoracotomy. A comparison between the ILS and ELS surgical procedures is shown in Table 3. It can be seen that ILS patients had longer operating room times (46.79±15.86 vs. 40.39±17.10), more intraoperative bleeding (2.70±2.15 vs. 2.12±2.42), and longer postoperative hospital stays (5.21±0.94 vs. 4.94±0.24) compared to ELS patients.

Table 3 Comparison of surgical procedures between ILS and ELS

Surgical procedures	ILS	ELS	*p*
Operative time(min),mean	46.79±15.86	40.39±17.10	*p*=0.005
Blood loss(ml),mean	2.70±2.15	2.12±2.42	*p*=0.048
Post-operative hospital stay(day),mean	5.21±0.94	4.94±0.24	*p*=0.020

3.4 The postoperative pathology between ILS and ELS

Macroscopically, it could be seen that lesions from ELS patients were isolated, in that they were clearly demarcated from normal lung tissue, and the surface boundaries of isolated lung was clear. On the other hand, lesions from ILS patientsshowed no own pleural covering, meaning the boundaries between the lesion surface and normal lung tissue were not clear. Postoperative pathological specimens of all 29 children were analyzed after sectioning and HE staining. Microscopy revealed no significant differences between tissue from ILS and ELS patients. The diseased lung tissue in both cases was composed of cystic dilated bronchi, bronchioles, and alveoli. Additionally, cystic interstitial fibrous hyperplasia with chronic inflammatory cell infiltration and lymphatic follicular formation were observed. Submembranous vascular dilatation and vascular proliferation were prominent, with vascular wall thickening and different lumen sizes also visible.

4. Discussion

Congenital PS is a lesion of the lung tissue which occurs during embryonic development. The lesion is without respiratory function and shows an abnormal arterial supply. Although PS is commonly divided into two types (ILS and ELS), the clinical significance of this classification for patients with congenital PS is still controversial. In this study, we retrospectively analyzed the clinical data of 29 cases of congenital PS, including 17 cases of ILS and 12 cases of ELS. We found that ILS and ELS tend to occur in the lower lobe of the left lung. PS in other locations have also been recorded in the literature (the upper lobe, lobar isolating lung in the neck, mediastinum and abdomen), however, as this occurs rarely, they are often misdiagnosed as tumors. Therefore, for masses of unknown nature at these sites, the differential diagnosis of PS should be considered[8].

Prenatal fetal ultrasound is the standard for the diagnosis of congenital PS at present, however, compared to other imaging modalities, the ultrasound examination has higher requirements on the instrument and the operator. Chest enhanced CT can clearly display the supplying artery and abnormal vein, something that is not possible via ultrasound[9, 10]. Magnetic resonance imaging (MRI) is able to detect the relationship between lesions and abnormal supplying arteries in the systemic circulation without the use of contrast agents, however, MRI requires higher coordination in children and is not as useful as enhanced CT in revealing the features of PS[11, 12].

Currently the largest study of PS-related data, Wei et al.[13] retrospectively analyzed 2,625 cases of PS in the Chinese database, including 132 adult patients. The results of this study showed that the arterial blood supply of PS-affected lung tissues was predominantly provided by the thoracic (76.55%) and abdominal (18.47%) aortic branches. Wani et al.[14] found that the abnormal supply of systemic arteries to originate from the descending thoracic aorta (72%), the abdominal aorta, the abdominal axis or the splenic artery (21%). These results are consistent with the findings of our study, which suggest that the arterial blood supply for PS isolating lung tissue is mainly from thoracic aortic branches[15, 16].

A difference in venous drainage between ILS and ELS has also been reported in the literature, with Zhang et al.[17] finding that: in patients with ILS, the venous drainage was mainly via the pulmonary veins, and;

in patients with ELS, the venous drainage was via the azygos vein or hemiazygos vein. The study also presented a difference in infection rate, with 71.17% and 31.37% for ILS and ELS, respectively. The existence of these differences between ILS and ELS in regard to symptoms, and pathological anatomy mean that it is of great clinical significance to classify PS into these two groups. However, the significance of ILS and ELS classification on surgical difficulty, treatment and prognosis of PS in children has not yet been further investigated.

In this study, a retrospective analysis of 29 children with PS was performed, resulting in the finding that the classification of these patients into ILS and ELS groups was significant for predicting their surgical outcomes. The duration of operation and postoperative hospitalization were longer in most children with ILS ($p < 0.05$), and intraoperative bleeding was more frequent in children with ILS ($p < 0.05$). More children with ILS were prone to infection, due to the lack of complete boundary between the pleura and normal lung tissue, and lack of connection to the normal trachea and bronchial tree. For the same reason, ILS lesions were also more likely to affect the surrounding normal lung tissue. The normal lung tissue after infection is brittle, more likely to bleed when touched, and more difficult to suture after bleeding. It is also more likely to cause thoracic adhesion, making intraoperative separation difficult. Due to the higher probability of lung tissue infection in children with ILS, more care should be taken, including adequate preoperative preparation, in comparison to children with ELS.

In our study, the histologic appearance of ILS and ELS showed no significant difference, with both PS types being composed of cystic dilated bronchi, bronchioles and a little immature alveolar tissue. This is consistent with the report of Richard et al. [18], who found that the histologic appearance of ELS consisted of lung tissue with acini displaying uniformly dilated bronchioles, alveolar saccules, alveolar ducts, and alveoli. As ILS and ELS appear histologically similar, the differentiation between the two is based mainly on imaging and intraoperative findings.

In summary, congenital PS is more likely to occur in the left lower lung regardless ofILS or ELS type, and most of the blood supply to the isolating lung tissue originates from the thoracic aorta. The type of congenital PS has an influence on neonates during and after operation, with ILS patients being more difficult to operate on and taking a longer time to recover. Upon histological examination, ILS and ELS showed no significant difference, thus, the differentiation between ILS and ELS needs to be made based on imaging results and intraoperative findings.

Declarations

Acknowledgments

Not applicable.

Funding

None declared.

Availability of data and materials

Please contact author for data requests.

Authors' contributions

WZH and YG conceived the report. The authors XDQ and WGF contributed equally to this work. All authors approved the final manuscript.

Ethics approval and consent to participate

Not applicable.

Consent for publication

Consent for publication of this study in its entirety was obtained from the patient.

Competing interests

The authors declare that they have no competing interests.

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