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CASE REPORT
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Treatment of Pulmonary Lymphangiectasia with Chylothorax: Case Report

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ABSTRACT

Pulmonary lymphangiectasia is a rare congenital disease with unclear pathogenesis and controversial treatment. We report a case of an 8-year-old boy with pulmonary lymphangiectasia complicated by chylothorax, who was managed conservatively with thoracic drainage, nutritional support, and infection control over a three-month period. Although the pleural effusion did not significantly decrease, it gradually formed a stable encapsulated collection without progression. After 2 years of follow-up, the patient's encapsulated effusion gradually decreased. This experience suggests that in selected patients, a monitored conservative approach, emphasizing drainage maintenance and avoidance of invasive interventions, may allow for spontaneous stabilization, supported by serial ultrasound assessment.

Keywords: Pulmonary Lymphangiectasia • Pediatrics • Chylothorax • Pneumonia • Case Report

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INTRODUCTION

Pulmonary lymphangiectasia is the abnormal expansion of lymphatic vessels in the lung due to congenital abnormalities. It mainlyoccurs in the fetal period or neonatal period, manifested as pulmonary edema, pleural effusion, pericardial effusion, pulmonary infection, etc., with a high mortality rate. Some patients also present with systemic lymphangiectasia, especially intestinal lymphangiectasia, leading to poor prognosis. So far, there is no effective treatment for pulmonary lymphangiectasia or even for patients with chylothorax [1]. Recently, a child with pulmonary lymphangiectasia complicated with chylothorax was admitted to our hospital. The patient was cured and discharged after supportive treatments such as a fat-free diet, closed thoracic drainage, pleural irrigation, nutritional support, oral diuretic drugs, and plasma infusion.

CASE PRESENTATION

The patient was an 8-year-old boy who was admitted to the hospital due to recurrent pneumonia with bilateral pulmonary lymphangiectasia found for more than 4 years. The child developed fever with paroxysmal cough 4 years ago, and he was diagnosed with severe pneumonia, pulmonary lymphangiectasia, and constrictive pericarditis. After conservative treatment in the local hospital, such as anti-infection, ventilation support,

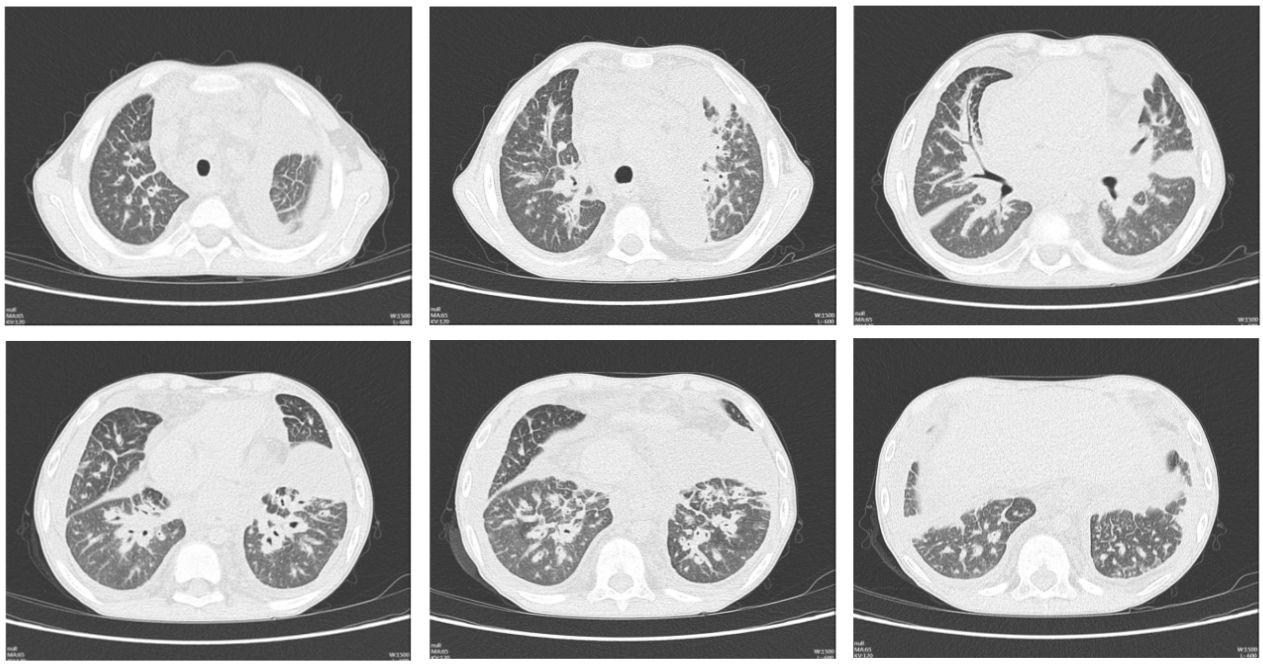

Figure 1: *Cross-sectional scan of the patient's chest CT.*

diuresis, and glucocorticoid anti-inflammatory therapy, he still had recurrent pneumonia and pleural effusion. Finally, the child came to our institution for further treatment.

The vital signs of the child were stable upon admission. Chest examination showed barrel-shaped chest changes, weak breath sounds in both lower lungs, and scattered wet rales in both lungs. Chest CT (Figure 1-2) showed bilateral pleural effusions, partially encysted, especially on the left side. Multiple patchy, fuzzy, dense shadows were seen in both lungs, and the bronchial walls of both lungs were thickened, especially in the lower lobes of both lungs. The interlobular septa of both lungs were thickened, and some showed reticular changes. Patchy, slightly hyperdense shadows were seen in the anterior mediastinum. The trachea and main bronchus in the thorax were normal. Multiple slightly larger lymph nodes were seen in the bilateral supraclavicular fossa and mediastinum. Abdominal CT (Figure 3) showed increased cardiophrenicangle, retroperitoneum, and mesenteric fat density. Multiple small lymph nodes were seen in the mesentery and retroperitoneum.

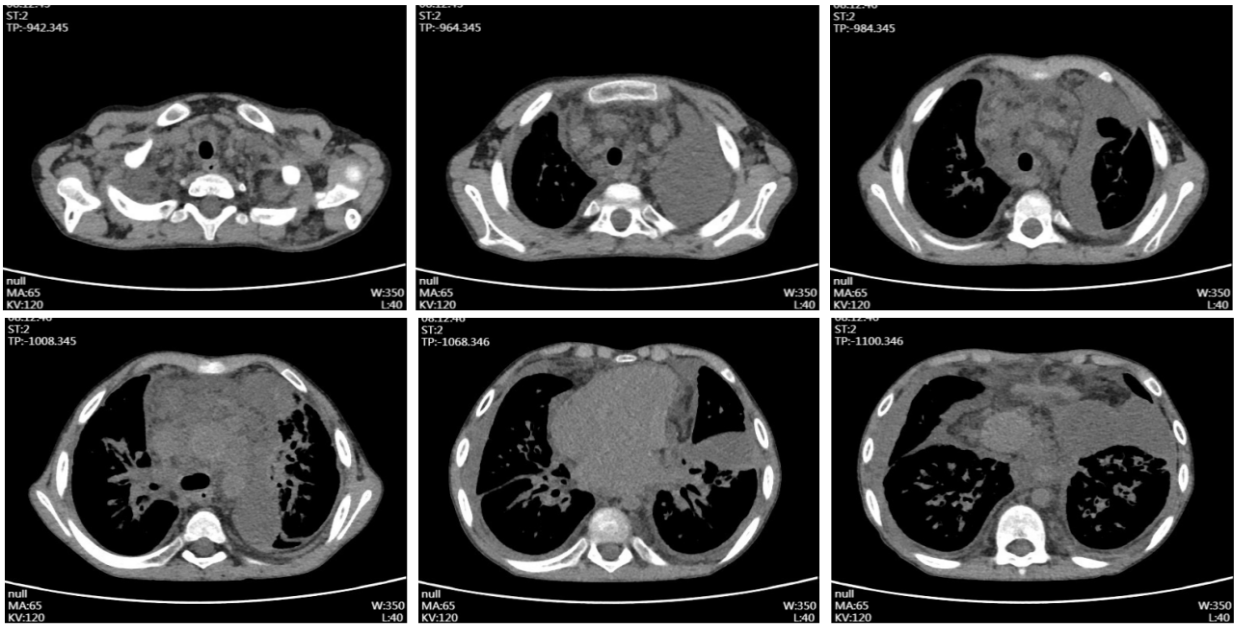

Figure 2: *Cross-sectional mediastinum window scan of the patient's chest CT.*

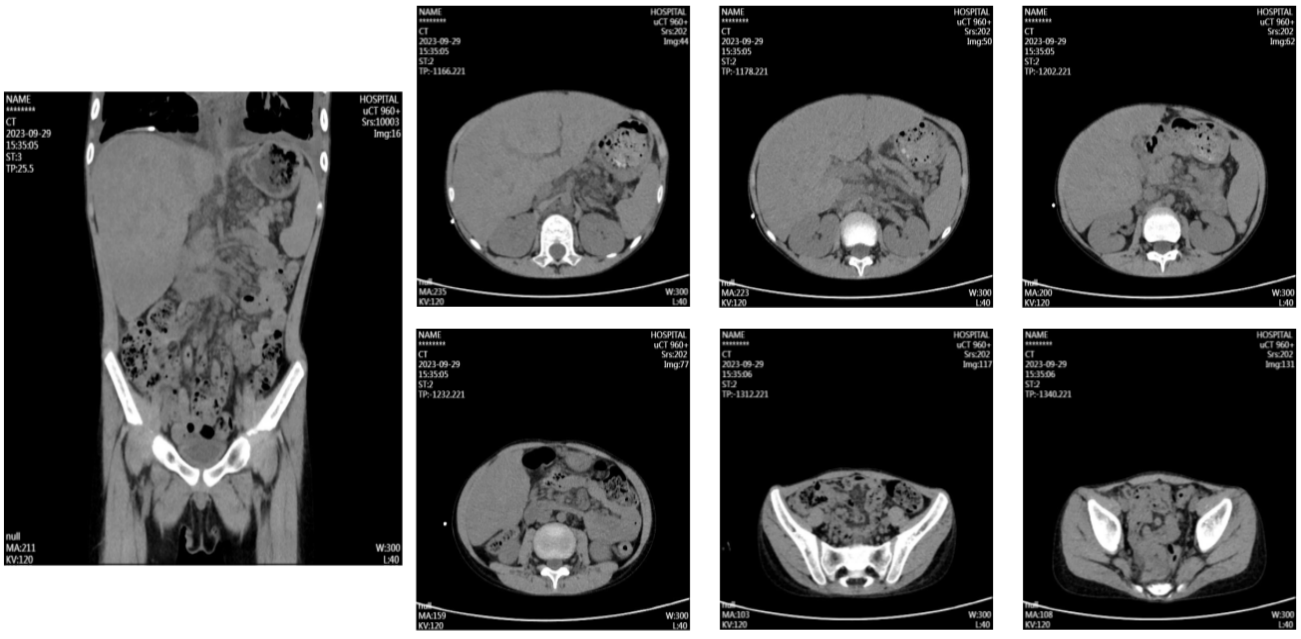

Figure 3: *Cross-sectional and coronalscans of the patient's abdominal CT.*

When the patient was admitted to our institution, we placed bilateral thoracic drainage tubes. The drainage fluid was light red, and the daily drainage volume was about 100 ml to 200 ml. Laboratory tests suggested drainage fluid as a chylous pleural effusion. Due to the long-term low-fat diet and repeated catheter drainage of bloody pleural effusion, the child had poor nutrition, hypoalbuminemia, and moderate anemia. Conservative supportive treatments such as albumin supplementation, plasma infusion, correction of water and electrolyte disturbances, and parenteral nutrition were given after an indwelling deep venous catheter. Appropriate diuretics were also administered. At the same time, because the patient had pneumonia on admission, antibiotics, expectorants, and aerosol inhalation of airway spasmolytic drugs were administered. The patient not only had pulmonary lymphangiectasia but also had abdominal lymphangiectasia. Therefore, the patient often had abdominal pain, which was relieved by treatment with nonsteroidal anti-inflammatory drugs.

Table 1: Changes in pleural effusion from ultrasound examination during treatment.

Date	Effusion on the left side (cm * cm)	Effusion on the right side (cm * cm)
10-Oct-2023	3.09 * 0.94	1.58 * 0.87
13-Oct-2023	0 * 0	4.74 * 2.57
16-Oct-2023	8.04 * 3.63	2.28 * 2.21
20-Oct-2023	0 * 0	4.59 * 2.28
25-Oct-2023	3.50 * 1.86	4.93 * 2.89
30-Oct-2023	3.26 * 1.84	NA
3-Nov-2023	3.89 * 2.82	2.4

After repeated pleural irrigation to keep the drainage tube unobstructed, regular review of chest ultrasound showed that the child's pleural effusion gradually formed an encyst. Under the condition of maintaining the original diet structure unchanged, the drainage tube was removed, and the patient had no discomfort symptoms after 2 weeks of observation. Reexamination of the ultrasound showed that the encapsulated effusion was gradually absorbed and reduced.

DISCUSSION

Pulmonary lymphangiectasia is a rare disease due to abnormal development of lymphatic vessels in the lungs. Noonan et al. initially classified pulmonary lymphangiectasia into three types:

1. Systemic lymphangiectasia that primarily accumulates in the gastrointestinal tract and less affects the lungs.
2. Secondary pulmonary lymphangiectasia due to pulmonary venous obstruction
3. Primary pulmonary lymphangiectasia.

Pulmonary lymphangiectasia was later modified into two categories: primary and secondary pulmonary lymphangiectasia due to improvement in clinical symptoms and advances in neonatal intensive care [1].

The cause of primary pulmonary lymphangiectasia is

unknown. Because this disease is a congenital anomaly of lymphatic development, we hypothesized that lymphangiectasis may be associated with mutations in receptors or signaling pathways related to lymphatic vessel development and lymphatic valve formation, such as vascular endothelial growth factor receptor (VEGFR), RAS/MAPK, PI3K/AKT, TNFRSF13B, GDF2, and NF-κB signaling pathways [2-4]. In addition, some patients have Noonan syndrome. Whether there is a link between the two diseases is not known [5].

Lung biopsy has long been considered the gold standard for the diagnosis of pulmonary lymphangiectasia. It can also be distinguished from lymphangioma, interstitial emphysema, congenital cystic adenomatoid malformation, and other diseases [6]. However, according to previous studies, lung biopsies were performed mostly in children who died of pulmonary lymphangiectasis. When pulmonary lymphangiectasiscan be diagnosed on typical CT images and clinical symptoms [7,8], does lung biopsy in living children, especially those combined with severe chylothorax, aggravate the condition? Is a lung biopsy necessary?

In our management of this patient, we did not use chemical pleurodesis or surgical procedures such as thoracic duct ligation, because we believed that such procedures would necessarily increase the pressure in the pulmonary circulation and that pulmonary edema would worsen when there was no place for the chylous fluid to go. We also did not use octreotide or other somatostatin regimens recommended in guidelines or reviews. However, studies have shown that it can reduce the pressure of lymph reflux and promote fistula healing. However, its pathology is lymphangiectasia. For patients with abdominal lymphangiectasia, the curative effect is not exact. Moreover, it is only temporary symptomatic treatment, and the side effects brought by it should be the focus of consideration [9].

What we do is to keep the chest tube unobstructed, and in the case of adequate drainage, maintain the stability of the internal environment and avoid infection. According to the results in Table 1, the volume of fluid collection in the patient did not decrease significantly, but gradually encapsulatedfrom the ultrasound images. In our experience, as long as the child is developing, the lymphatic fistula will gradually heal itself. In addition, we also recommend ultrasound as a follow-up examination for patients with pulmonary lymphangiectasia because of its convenience and radiation-free advantages [10,11].

At present, the diagnosis and treatment of patients with pulmonary lymphangiectasia is still controversial [1,11]. Because this disease is a congenital abnormality of lymphatic development, it may be cured by regulating lymphatic regeneration or structural remodeling. For now, the existing treatment is simply supportive care, not treatment of the cause. This case also provides a reference for the diagnosis and treatment of pulmonary lymphangiectasia. It is believed that with the deepening of research, the disease will eventually be cured.

CONCLUSION

This case demonstrates that for selected pediatric patients with pulmonary lymphangiectasia complicated by chylothorax, a conservative management approach centered on thoracic drainage, nutritional support, and infection control may promote spontaneous encapsulation and stabilization of effusion during follow-up. By avoiding invasive interventions and employing serial ultrasound monitoring, clinicians can safely manage such patients, supporting a time- and growth-based self-limiting treatment pathway.

FUNDING

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CONFLICT OF INTEREST

The authors declare no conflicts of interest.

ETHICS APPROVAL AND CONSENT TO PARTICIPATE

Data from our present study were collected from patients' progress of treatment. Informed consent was obtained from the patient's guardian and was approved by the Ethics Committee of The Affiliated Guangdong Second Provincial General Hospital of Jinan University.

AUTHOR CONTRIBUTIONS

Shaoyi Zheng contributed to data curation and writing the original draft. Weiguang Long was involved in the conceptualization of the report and the patient's treatment. Supervision was provided by Wenlin Wang. Bin Cai and Yang Liu participated in writing the review and editing the manuscript. Fen Lei was responsible for patient management.

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