Residência RP Pediátrica
Publicação Oficial da Sociedade Brasileira de Pediatria
ISSN-Online: 2236-6814

Submitted on: 11/05/2020
Approved on: 03/25/2021

CASE REPORT

Unilateral congenital pulmonary lymphangiectasia: case report

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Keywords:
Pulmonary lymphangietasia, Newborn, Lymphatic vessels, Child.

Abstract

Pulmonary lymphangiectasia is a rare developmental disorder in which lymphatic vessels proliferate and dilate. Based on pathogenesis and clinical phenotype, it can be classified as: primary or congenital pulmonary lymphangiectasia (CPL), or secondary. The diagnosis is made by clinical characteristic and imaging and histopathological exams features. Lung biopsy is considered the gold standard for the diagnosis of CPL and is also useful to distinguish between interstitial lung diseases or other forms of pulmonary lymphatic dilation. The definitive treatment of the isolated form can be curative with resection of the affected area. In diffuse form, the treatment is palliative and aims to limit the production and accumulation of lymph in the pulmonary serosa. Currently, the conservative treatment is recommended for chylothorax correction. This article aims to report the case of a patient with a diagnosis of congenital pulmonary lymphangiectasia with good evolution, emphasizing the importance of early diagnosis and therapeutic approach, and to compare with what has been described in the literature.

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Residência Pediátrica; 2023: Ahead of Print.
DOI: 10.25060/residpediatr-2022.v13n2-503

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INTRODUCTION

Pulmonary lymphangiectasia (PL) is a rare developmental disorder in which lymphatic vessel proliferation and dilatation is observed. Depending on pathogenesis and clinical phenotype, the condition may be categorized as primary or congenital pulmonary lymphangiectasia (CPL) or secondary PL[1].

PL was first described by Virchow in 1856, but the exact number of cases of the disease is unknown. Autopsy studies suggest that 0.5% to 1% of stillbirths and neonatal deaths are linked to PL. Two series including stillbirths described PL as the cause of death in 5 of 451 and 11 of 2,514 cases. Familial occurrence is rare[2,3].

The etiology of LP is unknown. Finegold et al. postulated that LP arises from a developmental error in which physiological regression of connective tissue does not occur until the twentieth week of life, causing a persistence of large lymphatic vessels[4].

Newborns with severe complications may require continuous positive airway pressure (CPAP), orotracheal intubation (OTI), and prompt drainage of excess fluid in the chest cavity.[3] Once the patient is clinically stable, curative treatment of the isolated form of the disease can be initiated with the resection of the affected area. Treatment for the diffuse form of the disease is palliative and designed to limit the production and accumulation of lymph in the serosa[5].

Initial treatment for chylothorax is conservative and includes thoracocentesis, water seal chest drainage, and ventilatory support. Enteral nutrition with medium-chain triglycerides and total parenteral nutrition have been used successfully, since absorption occurs through the portal system with decreased lymph formation. Treatment with octreotide and somatostatin may be attempted, while surgery may be reserved for patients unresponsive to these therapies[6].

The present article describes the case of a patient treated for CPL and underlines the importance of early diagnosis and treatment; it also offers an outline of what is prescribed in the literature.

CASE REPORT

D.L.O.P., a male infant, was born at the Toledo Maternity Hospital after 31 weeks and three days of gestation by cesarean delivery. His mother developed gestational hypertension and kidney failure, which promoted the interruption of pregnancy. The newborn had 1- and 5-minute APGAR scores of 8 and 9, respectively, a weight of 1,235 g, a height of 37 cm, and a head circumference of 28 cm. He required OTI for respiratory distress; surfactants were not used. Two days after birth, he was referred to a Neonatal Intensive Care Unit in Ivaiporã, and remained in hospital for 34 days on a mechanical ventilator; he could not be weaned.

Chest X-ray images showed multiple persistent cysts in his left hemithorax (Figure 1A). The patient was transferred to the Pequeno Príncipe Hospital in Curitiba 34 days after birth with a preliminary diagnosis of congenital cystic adenomatoid malformation (CCAM) for evaluation by a pediatric surgeon.

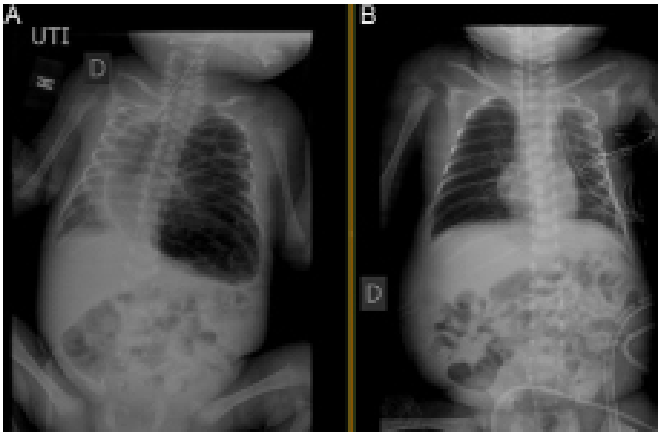


Figure 1. (A) Chest and abdominal X-ray images on admission (May 14, 2019) and (B) after resection of the affected lung lobe (May 16, 2019).

Physical examination on admission found the patient had a heart rate of 145 bpm, a respiratory rate 55 bpm, and an O_2 saturation of 84%; his general condition was regular; he was hypoactive, hydrated, and anicteric; he had diminished vesicular breath sounds in the apex of the left lung and presence of intercostal and subdiaphragmatic retractions. No other alterations were observed.

Other than showing low sodium levels (Na = 123 mmol/L) promptly corrected with 3% saline solution, the patient's laboratory test results were normal. X-ray images showed multiple cysts in the left hemithorax, while chest computed tomography (CT) scans showed multiple cystic malformations throughout the upper left lobe, deviation of mediastinal structures to the right, and atelectasis with air bronchograms in the lower left lobe, increased right parenchymal attenuation, and small isolated cysts in the middle lobe (Figure 2). The main diagnostic suspicion was still CCAM.

After evaluation by a pediatric surgeon, a lobectomy was performed 36 days after birth. The patient was prescribed prophylactic antibiotic therapy with vancomycin and amikacin for 24 hours before surgery. He remained with a chest drain for six days.

Despite a preliminary diagnosis of CCAM, the surgical specimen sent for evaluation by a pathologist had findings consistent with CPL. Immunohistochemistry tests were positive for CD34, D2-40, ACTIN 1 A4 and CK AE1/AE3, consistent with lymphatic disease.

The patient was intubated after surgery with X-ray images showing good expansibility also in the left lung (Figure 1B). He was given dexamethasone for seven days, was extubated, and placed on non-invasive ventilation 11 days after surgery.

The patient started alternating between CPAP and oxygen therapy with an oxygen hood 27 days after surgery. Three days later, he was on a nasal catheter receiving oxygen at a rate of 1L/min. At this time, the patient showed a slight

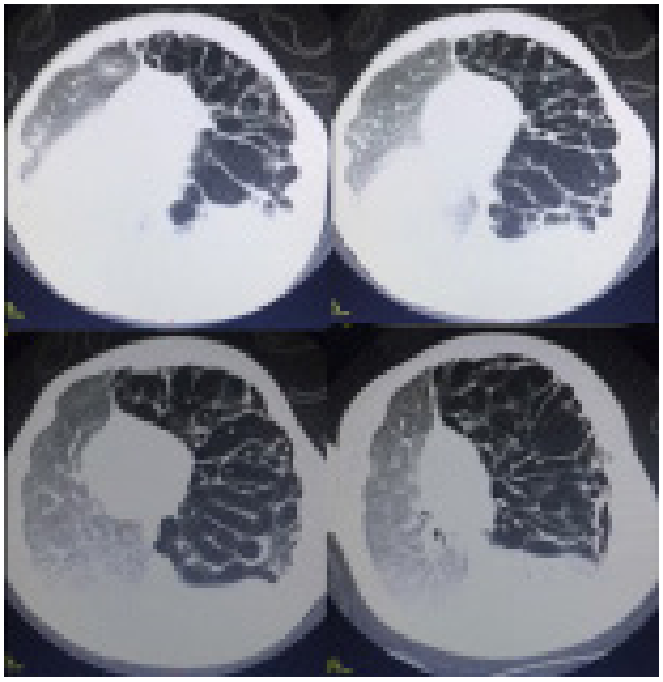


Figure 2. CT scans at admission (May 14, 2019).

decrease in vesicular breath sounds in the left apex of the lung and mild subcostal retractions, which remained until he was discharged.

The patient was transferred from the neonatal intensive care unit to the pediatric ward after 43 days of hospitalization, showing a good respiratory pattern and saturation above 90%. He no longer needed a nasal catheter after four days in the pediatric ward.

At three months and 23 days of age and gaining weight (3 kg), the patient was sent home breathing comfortably in room air and eating an orally-fed diet. A multidisciplinary follow-up appointment was scheduled with the pediatrics, pediatric surgery, neurology, ophthalmology, pulmonology, and nutritional support teams.

DISCUSSION

Pulmonary lymphangiectasia presents both early and late-onset symptoms. Early-onset symptoms, as in the case described, include respiratory distress, tachypnea, and cyanosis[2].

The classic imaging pattern for PL includes a diffuse nodular reticular infiltrate that does not improve antibiotics or diuretics, differently from cases of lung infection and congenital heart defects. In addition, diffuse interstitial opacity, Kerley B lines, pleural effusion, and lung hyperinflation may be found in neonates. CT scan findings usually include interlobular septal thickening, ground-glass opacity, and pleural effusion[1,7].

The x-ray images of the infant described in this case showed multiple persistent cysts in the left hemithorax. This finding, together with clinical signs and symptoms, were suggestive of CCAM, a condition in which a single or multiple cysts or a solid mass may be seen in x-ray images[7].

As the literature recommends in cases of symptomatic CCAM, surgical resection of the affected site was chosen after discussions with the pediatric surgery team[7].

The pathology report contained a diagnosis of congenital pulmonary lymphangiectasia (CPL). Despite the absence of the classic imaging signs of CPL, an open lung biopsy followed by immunohistochemistry testing is considered the gold standard diagnostic method. The report described dilatation of the subpleural, interlobar, perivascular, and peribronchovascular lymphatic vessels; microscopic findings included immunopositivity for CD31, CD34, and D2-40[2,5,8].

CPL is often disregarded in the differential diagnosis of neonatal respiratory failure, since it is a rare anomaly until recently associated with death in most cases, according to the literature. It must be differentiated from diseases such as congenital lobar emphysema, pulmonary interstitial emphysema, diffuse pulmonary lymphangiomatosis, and congestive heart disease[5,9].

Surgery for CPL is not commonly performed in newborns. Conservative management is usually preferred as initial treatment. The first case of neonatal surgery for CPL was described in 2014. The treatment was deemed successful and the patient developed normally until the age of two years. Currently, pleurectomy with chemical pleurodesis combined or not with thoracic duct ligation is used to treat chylothorax if conservative management fails[3,10].

In our case, although surgery was not performed for CPL, the patient benefited from the procedure, since surgery is a curative procedure in cases of lymphangiectasis restricted to the lungs, as seen in our patient.

Early PL is usually associated with death within hours of birth. However, advances in perinatal care, especially in respiratory management, have increased the survival of patients with the condition, as in the case reported. A condition once considered incompatible with life may offer a good prognosis, depending on disease severity and the care provided[1,2].

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