

# Diffuse pulmonary lymphangiomatosis: imaging findings

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## ABSTRACT

Diffuse pulmonary lymphangiomatosis is a rare pulmonary disorder affecting the lymphatic channels from the mediastinum to the pleura. The disease usually occurs in children and young adults and frequently ends with death due to progressive course. Imaging findings of the disease are based on lymphatic involvement which appear as mediastinal soft tissue infiltration and thickening of pulmonary peribronchovascular bundles and interlobular septae. In this report, spiral and high-resolution computed tomography, and ultrasonography findings of severe form of this rare disease are presented. Furthermore, some lymphatic disorders, which are called with similar name but different appearances on imaging, are discussed.

*Key words:* • lung • lymphangioma  
• tomography, X-ray computed • ultrasonography

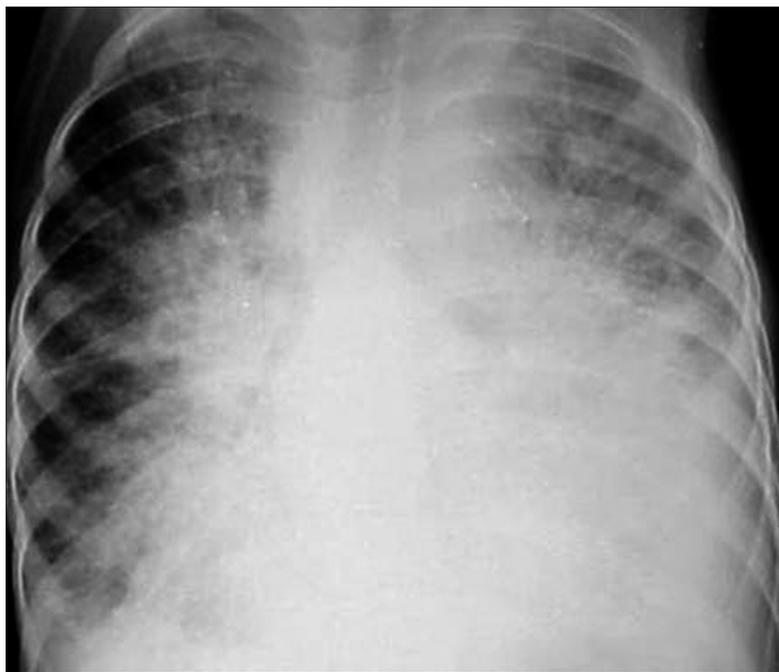
**D**iffuse pulmonary lymphangiomatosis (DPL) is a rare pulmonary lymphatic system disorder involving the entire lymphatic system from the mediastinum to the pleura. Although pathologically benign, it is a progressive and fatal disease. It is histologically observed that there is an increase in the diameter and number of involved lymphatics (1). The disease is generally seen among children and adolescents. Histologically, it is difficult to differentiate DPL from other lymphatic diseases like lymphangioma, lymphangiomyomatosis, and lymphangiectasia (1). Yet, such lymphatic diseases have some imaging characteristics which help in the diagnosis. DPL presents itself with diffuse mediastinal soft tissue infiltration, pulmonary interstitial parenchymal infiltration, and pleural effusion. Such diffuse parenchymal involvement in DPL can best be detected by high resolution computed tomography (HRCT). In the literature, there is a limited number of cases where interstitial involvement is shown by HRCT. In this case report, CT, HRCT, and ultrasonography (US) findings of a severe form of this uncommon disease are presented.

## Case report

An 8-year-old girl was admitted with non-productive cough, hemoptysis, and shortness of breath that became progressively worse for the last two months. Laboratory analysis yielded lymphocytosis (58%), decreased hemoglobin (9.5 g/dL) and hematocrite (30.9%) levels. Posteroanterior chest radiograph showed mediastinal enlargement, bilateral hilar and parahilar infiltrations which extended to the pleura and were more predominant on the left side, and pleural effusion (Figure 1). Contrast-enhanced CT of the chest showed diffuse soft tissue infiltration in the mediastinum without displacement of the vessels (Figure 2a). Bilateral peribronchovascular and interlobular septal thickening, ground glass opacities and multifocal air-trapping were observed in the lung parenchyma (Figure 2b). HRCT showed thickening of the interlobular septa displaying the anatomy of the secondary pulmonary lobules, especially in the upper lobes (Figure 2c). After thrombocyte replacement for thrombocytopenia, fine needle aspiration biopsy (FNAB) was performed from the mediastinal soft tissue. The biopsy result suggested a slowly progressing hematological tumor. A bone marrow aspiration biopsy was normal. Since the FNAB result was ambiguous, an open lung biopsy was performed with thoracotomy. Pathological results were compatible with DPL. Despite corticosteroid and interferon treatment which lasted eight months, the patient's shortness of breath and hemoptysis attacks progressed and a follow-up CT examination showed increase in the pulmonary infiltrations. Serial thoracic ultrasonography performed by a 12 MHz superficial probe showed progressively increased diffuse thickening in the parietal and visceral pleura as well as peripheral parenchymal in-

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**Figure 1.** Chest radiograph shows enlargement in the mediastinum, peribronchovascular infiltrations extending from hila to the periphery of the lung and pleural effusion. Note that the contours of the heart and the diaphragms are silhouetted by the consolidated lung.

filtrations (Figure 3). Radiation therapy could not prevent clinical deterioration, and the patient succumbed to respiratory failure.

### Discussion

Primary lymphatic disorders of the lung are rare, and their diagnosis and classification are difficult. At least four such diseases have been defined, which include lymphangioma, lymphangiomyomatosis, lymphangiectasis, and lymphangiomatosis. These have several shared pathological characteristics which lead to difficulties in differential diagnosis (1). Lymphangiomas are localized lymphatic malformations which have no connection with the normal lymphatic system (2). Fluid may accumulate in these lymphatic malformations and give them a cystic appearance. At this stage, they are called cystic hygroma and it is easier to differentiate them radiologically from other lymphatic disorders. Lymphangiomyomatosis generally affects women in reproductive age and is characterized by a proliferation of interstitial lymphatics. Air cysts develop in the lung parenchyma in the advanced stages of this disease (3). Common to both lymphangiectasis and lymphangiomatosis is that they might affect infants and children, they present chylous effusion and dyspnea, and cause a restrictive pattern in pulmonary function tests. While existing lymphatic channels

undergo dilatation with no new channels being formed in lymphangiectasis, new lymphatic channels appear in lymphangiomatosis and show abnormal dilatation (4, 5). Moreover, typical imaging characteristics of these diseases are quite helpful in diagnosis.

The radiological findings which lead to a diagnosis of DPL are diffuse mediastinal soft tissue infiltration, interlobular septal thickening, pleural effusion, and pleural thickening. Swensen et al. have identified in a HRCT study comprising eight patients with DPL smooth peribronchovascular and interstitial thickening in all patients (6). In the two cases reported by Higgins et al., nodular-type thickening was observed in addition to smooth interlobular septal thickening (7). In our case, smooth interlobular septal thickening was obvious, particularly in the upper lobes, demarcating the secondary pulmonary lobular anatomy (Figure 2c).

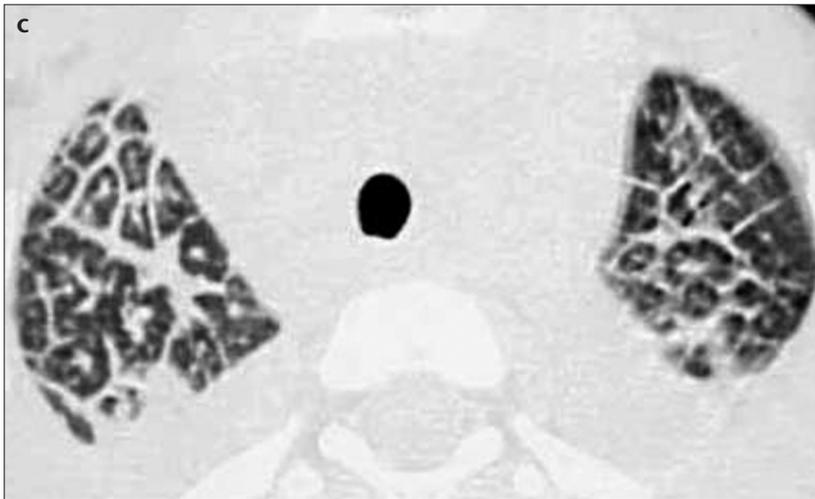
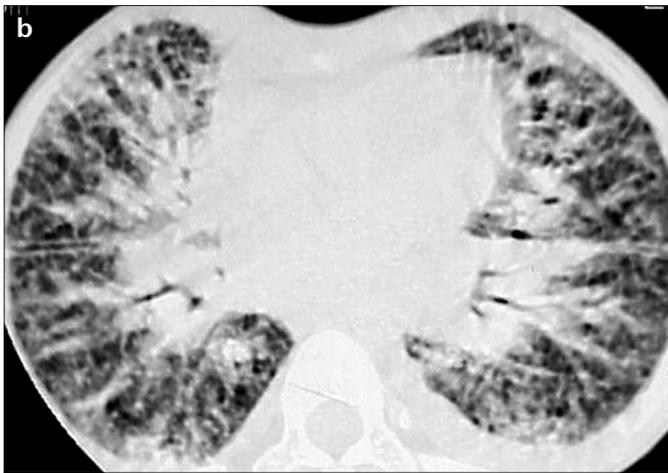
In the literature, there is no clear statement as to whether the mediastinal soft tissue infiltrations had mass effect on mediastinal vessels (6, 8). In the prospective study conducted by Swensen et al., all eight patients had diffuse mediastinal infiltrations on CT (6). Lynch et al. have observed mediastinal soft tissue mass in one of their two DPL cases (8). None of these studies, however, have quoted whether or not the mediastinal soft tissue infiltrations had

mass effect on the vascular structures. The absence of mass effect in our case is important in differentiating this disease from leukemia, lymphoma, sarcoidosis, and connective tissue diseases which are associated with lymphadenopathy.

Pleural thickening is one of the basic findings of DPL (6). However, to our knowledge, there is no detailed information about the nature of thickening in pleural leaves. In our case, high resolution US showed almost equal amounts of thickening in the parietal and visceral pleurae. Furthermore, US could clearly differentiate between pleural thickening and peripheral parenchymal infiltration. A chylous effusion is not characteristic for DPL, and serosanguineous pleural fluid might also be found (6), as was the case in our patient.

The chylothorax has been attributed to the leakage of the lymphatic fluid into the pleural space from the dilated lymphatics of pleura (9, 10). It has also been reported that mediastinal soft tissue infiltration may lead to lymphatic obstruction and consequently to chylothorax (9-11). However, although chylous pleural effusion was observed in all of the six DPL cases in the study of Aviv et al. (12), the presence of a mediastinal mass only in one case brings forward the need for further case series in order to clearly identify the pathogenesis of chylothorax seen in DPL.

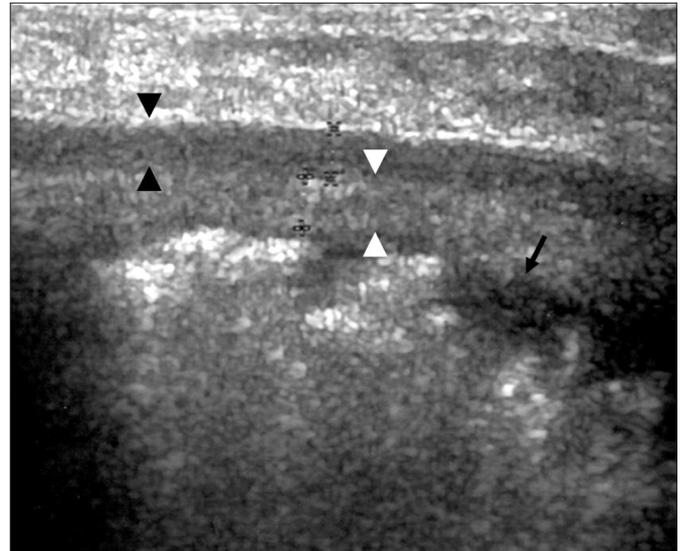
DPL is clinically and radiologically



**Figure 2.** a. Contrast-enhanced chest CT shows diffuse mediastinal soft tissue thickening, without mass effect on vessels. b. CT section at lung window shows bilateral peribronchovascular thickening, ground-glass opacities, and a mosaic attenuation pattern. c. HRCT section at the level of upper lobes demonstrates grossly thickened interlobular septae, revealing the polygonal anatomy of the secondary pulmonary lobules.

different from systemic lymphangiomas. Pulmonary lesions of lymphangiomas are radiologically different from DPL lesions. In systemic lymphangiomas, Laverdiere et al.

have identified cystic lesions accompanying linear opacities in both lungs (13). In a case with diffuse systemic lymphangiomas, massive chylothorax has been reported in the absence



**Figure 3.** Gray-scale ultrasonography shows prominent diffuse thickening of the parietal (black arrowheads) and visceral (white arrowheads) pleurae, and adjacent parenchymal infiltrations (black arrow).

of pulmonary parenchymal infiltrations (14).

In conclusion, DPL is a rare disease to be considered in the differential diagnosis of pulmonary lymphatic disorders, and is characterized by often diffuse interlobular septal thickening, pleural effusion, and mediastinal soft tissue infiltration that does not cause mass effect on vessels.

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