Nonmalignant Adult Thoracic Lymphatic Disorders

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KEYWORDS

• Lymphangioma • Lymphangiomatosis • Lymphangiectasia • Generalize lymphatic anomaly (GLA) • Gorham-Stout disease (GSD) • Kaposiform lymphangiomatosis (KL) • Pulmonary lymphangiectasia • Yellow nail syndrome

KEY POINTS

- The thoracic lymphatic disorders typically present with symptoms of cough, shortness of breath, chyloptysis, or expectoration of branching casts.
- Typical pulmonary manifestations of the thoracic lymphatic disorders include chylous effusions, peribronchiolar interstitial infiltrates, and mediastinal masses.
- The emergence of sophisticated imaging techniques that characterize abnormal lymphatic flow promises to improve the classification and therapeutic approaches to the thoracic lymphatic disorders.

INTRODUCTION

Primary lymphatic anomalies comprise a bewildering array of congenital and acquired conditions that can affect every organ system containing lymphatic channels, generally considered to be all tissues except brain and bone marrow. Lymphatic anomalies usually come to medical attention during childhood or early adulthood, but can also present later in life. For the purpose of this article, the discussion focuses on the lymphatic disorders that involve thoracic structures, either primarily or as part of a more global lymphatic disease process, and which preferentially affect older children and adults.

The pulmonary lymphatics are a network of vessels that function to transport cells and fluids from the periphery of the lung to the central lymphatic conduits, in order to regulate tissue pressure and facilitate regional immune responses. The peripheral lymphatic vessels converge on the larger conduits coursing on the surface of major airways in the hila and mediastinum and ultimately drain into the right lymphatic duct and thoracic duct (TD). The right lymphatic duct inserts into the subclavian vein in the neck and drains the right upper lobe. The TD inserts into the left innominate vein at the junction with the internal jugular vein and drains the left lung, right middle and lower lobes of the right lung, as well as all structures below the diaphragm (Fig. 1). A broad discussion of lymphatic anatomy is beyond the scope of this article, but it is important to note that intestinal lymph (chyle) that contains chylomicrons (dietary fats) enters the TD at the level of cisterna chyli in the upper abdomen and is transported to the venous system in the neck. The primary route thorough which chylous fluid reaches the pleural space or other thoracic structures in subjects with chylous effusions, therefore, is either through (1) reflux from an obstructed or pressure-overloaded

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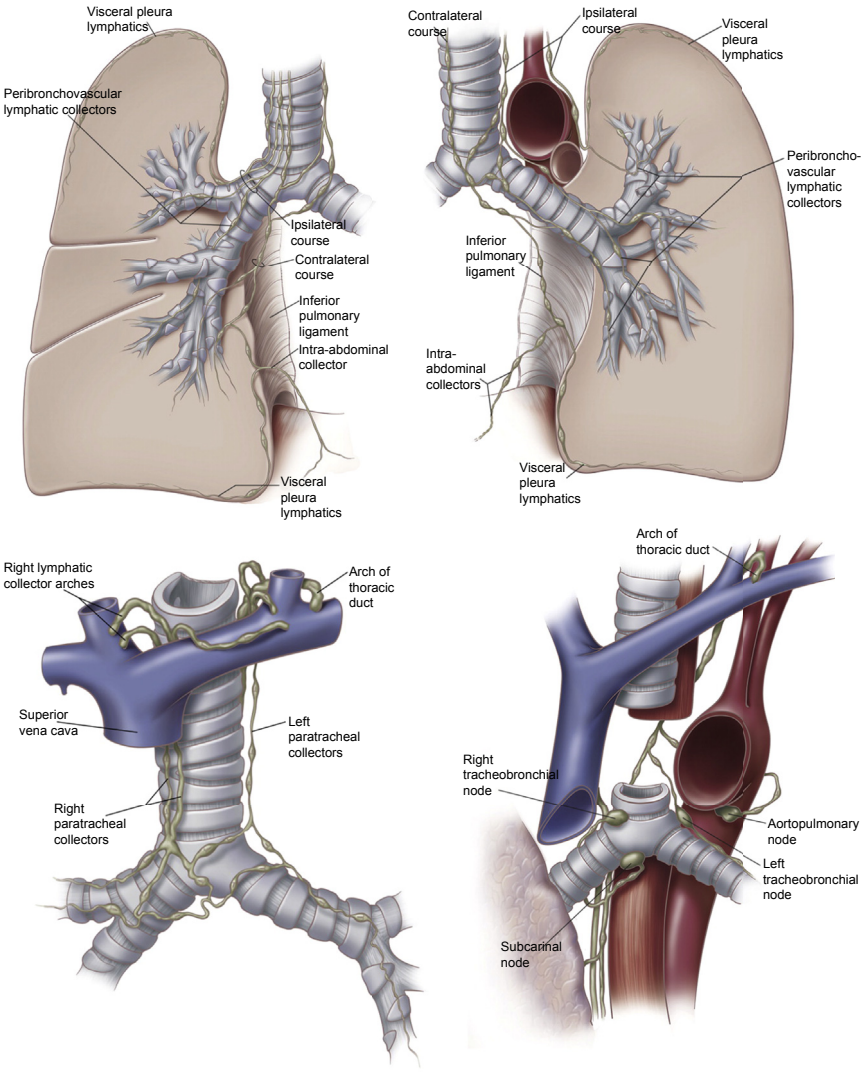

Fig. 1. Schematic representation of pulmonary lymphatic anatomy. (*Adapted from* Riquet M. Bronchial arteries and lymphatics of the lung. Thorac Surg Clin 2007;17:619–38; with permission.)

TD into the pulmonary lymphatic stream, or (2) through a pathologic connection between chylous lymphatics and the pleural space, airways, or lung parenchyma, as can occur following surgery or trauma or as part of a pathologic process. Other thoracic chylous complications that can result from these 2 processes include chylous congestion in the lung parenchyma,[1] plastic bronchitis (PB; expectoration of branching casts),[2] chylopericardium, and chyloptysis.

The thoracic lymphatic disorders (TLD) comprise a group of diseases that are variably associated with mediastinal or pulmonary masses, interstitial infiltrates, airway disorders including chyloptysis, PB, pleural effusions that are often chylous, and repeated pulmonary infections and bronchiectasis (eg, yellow nail syndrome, YNS). Extrapulmonary manifestations of the TLDs can include lymphatic leaks in various distributions (eg, chylous ascites), protein-losing enteropathy, recurrent fevers and prostration, lymphatic obstruction resulting in lymphedema of extremities, coagulopathy, and bony lesions. The TLDs often present in protean manner and can be congenital or acquired, localized or systemic. Attempts to classify these disorders have generally been based on defining commonalities of a limited number of cases or the consensus of experts.[3–6] Most of the published classification systems use inconsistent terminology and lack clear diagnostic, clinical, laboratory, or imaging standards. The TLDs are often grouped based on symptoms, age of presentation, histologic appearance, associated illnesses, or secondary imaging