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Symposium Highlight

LYMPHATIC IMAGING AND INTERVENTION IN CENTRAL LYMPHATIC DISORDERS

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ABSTRACT

Advances in lymphatic imaging for both diagnosis and intervention are reviewed, and specific examples given for protein-losing enteropathy, multi compartment lymphatic failure, congestive heart failure.

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Keywords: Lymphatic imaging, lymphatic intervention, central lymphatic disorders, thoracic duct, congenital heart disease

The lymphatic system plays a critical role in immune regulation, long-chain fatty acid absorption, and tissue fluid circulation. Lymphatic channels collect fluid from peripheral organs and tissues and transport it centrally toward the thoracic duct, which drains into the venous system at the junction of the internal jugular vein and subclavian vein. Approximately 2-3 liters flowing through the thoracic duct. The liver and intestine contribute significantly to this flow (1).

In patients with heart disease, central venous pressure (CVP) plays a crucial role in lymphatic dysfunction. Increased CVP leads to increased lymphatic production and impedes lymphatic drainage. When the capacity of the lymphatic system to drain the excess fluid is exceeded symptoms such as pleural effusions, ascites, edema, appear. In patients with congenital heart disease (CHD), especially single ventricle heart disease, conditions such as chylothorax, plastic bronchitis (PB), and protein losing enteropathy (PLE) can occur.

Lymphatic Imaging and Interventions

Recent advancements in lymphatic imaging and interventions have significantly improved the diagnosis and management of lymphatic disorders. Techniques such as T2 weighted MRI and dynamic contrast magnetic resonance lymphangiography (DCMRL) provide detailed visualization of lymphatic flow and anatomy (*Fig. 1a*) (2). These imaging modalities have been instrumental in understanding the pathophysiology of lymphatic disorders and guiding therapeutic interventions in patients with heart disease.

Techniques for lymphatic interventions have also advanced significantly over the past decade. Some of these techniques include lymphatic embolization, balloon dilation and stenting, and thoracic duct externalization

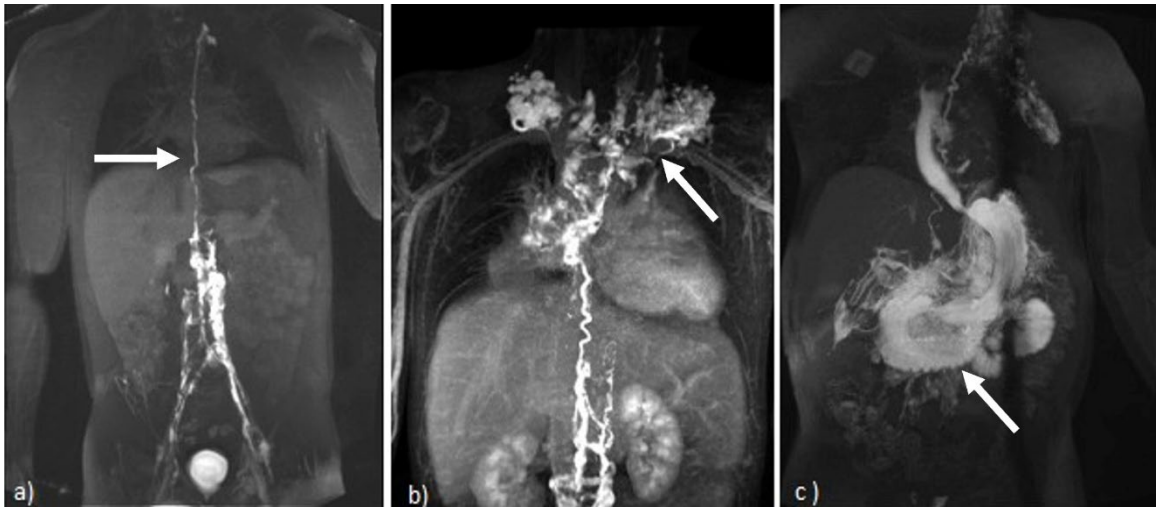

Fig. 1. *a) MIP coronal projection of intranodal DCMRL showing a normal central lymphatic system with normal TD (arrow). b) MIP coronal projection of intranodal DCMRL showing perfusion of the peribronchial networks (arrow). c) MIP coronal projection of intrahepatic DCMRL showing leak into the duodenum (arrow) with reflux into the stomach and esophagus.*

and drainage (3). Lymphatic embolization includes thoracic duct embolization (TDE) and selective lymphatic duct embolization (SLDE), and percutaneous needle embolization. SLDE, which leaves the TD intact, is preferred over TDE to minimize potential TDE complications. Percutaneous needle embolization, guided by fluoroscopy or ultrasound, involves direct injection of embolic agents through the needle directly into leaking channels. This technique is particularly effective for conditions like protein-losing enteropathy (PLE). Balloon dilation and stenting address TD obstructions by widening narrowed segments and maintaining duct patency with stents, though stent use requires caution due to occlusion risks. Thoracic duct externalization and drainage temporarily relieve symptoms in severe multi-compartment lymphatic disorders by draining lymphatic fluid externally, providing symptomatic improvement until definitive treatment is possible. This technique, described by Witte et al in 1969, demonstrated symptomatic improvement in patients with refractory heart failure through decreased thoracic duct afterload and brisk drainage (4). Further studies are necessary to understand the longevity and effectiveness of these interventions.

SPECIFIC CONDITIONS

Chylothorax and Plastic Bronchitis

Chylothorax is the accumulation of lymphatic fluid in the pleural space, often leading to respiratory distress and compromised lung function. Etiologically, it can be due to traumatic injury to the thoracic duct during surgery but more often is a result of abnormal lymphatic perfusion of the pulmonary interstitium and the mediastinum (*Fig. 1b*) (5). Imaging techniques such as DCMRL help in identifying the source and pathways of lymphatic leakage. Treatment strategies include dietary modifications, such as a low-fat diet or total parenteral nutrition to reduce lymph production, and pharmacological interventions like octreotide. Percutaneous interventions, such as selective lymphatic duct embolization (SLDE), target abnormal lymphatic branches to seal leaks and improve lymphatic drainage and are becoming the treatment modality of choice when conservative methods fail.

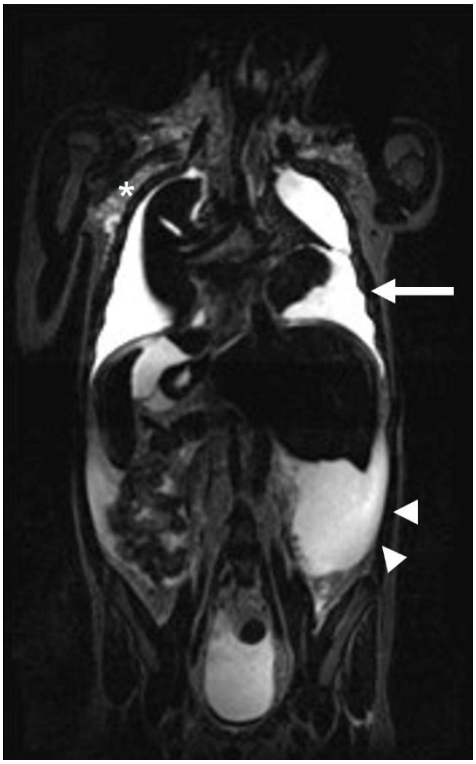

Fig. 2. *MIP coronal projection of T2 weighted MRI showing fluid accumulation in the chest (arrow) and abdomen (arrowheads) as well as skin edema (*) in a patient with multicompartment lymphatic dysfunction.*

Plastic bronchitis (PB) involves the formation of rubbery bronchial casts in the airways, which can lead to life-threatening respiratory obstruction. The etiology of PB involves retrograde lymphatic flow into the bronchial tree that leads to leak of protein into the airway (6). Imaging with DCMRL and T2-weighted MRI can visualize lymphatic flow abnormalities contributing to PB. Treatment includes bronchodilators, inhaled steroids, and pulmonary vasodilators to manage symptoms. For refractory cases, lymphatic interventions such as SLDE are used to occlude abnormal lymphatic channels and reduce lymphatic leakage into the airways.

Protein-Losing Enteropathy (PLE)

PLE is characterized by excessive protein loss into the intestinal tract, leading to hypoalbuminemia, diarrhea, edema, and ascites. It is primarily caused by abnormal lymphatic flow to the duodenum from the liver or mesentery, resulting in lymphangiectasia and increased mucosal permeability (*Fig. 1c*) (7-9). Imaging techniques like intrahepatic dynamic contrast MR lymphangiography (IH-DCMRL) are used to identify the lymphatic sources of PLE. Treatment includes dietary interventions, high-dose aldosterone antagonists, and sildenafil. For patients not responding to conservative measures, lymphatic embolization of hepatic and duodenal lymphatic networks and thoracic duct decompression (TDD) are considered (10-12). In severe cases, surgical interventions like Fontan takedown or orthotopic heart transplant (OHT) may be necessary.

Multicompartment Lymphatic Failure

This condition involves lymphatic dysfunction affecting multiple body compartments, such as the thorax, abdomen, and soft tissues (*Fig. 2*). Etiologically, it results from widespread lymphatic flow abnormalities and increased lymphatic pressure. Imaging with multicompartment DCMRL provides comprehensive visualization of lymphatic dysfunction across the different compartments. Treatment involves a combination of conservative management, including dietary modifications and pharmacotherapy, and interventional procedures like selective embolization and TDD. In severe cases, multi-compartment lymphatic failure may require advanced interventions such as OHT or ventricular assist devices (VAD) to manage symptoms and improve patient outcomes.

Congestive Heart Failure

Congestion is a primary contributor to the decompensation of heart failure (HF) (13). Most of the excess volume in patients with HF is located in the interstitial compartment. Inadequate decongestion, which leads to persistent interstitial congestion, is linked to worse

outcomes. Therapeutic interventions aimed at effective decongestion include traditional therapies such as diuretics, which enhance renal excretion of salt and water, and ultrafiltration. However, these methods often fall short due to diuretic resistance and renal dysfunction. Witte et al. demonstrated the resolution of interstitial congestion by external drainage of the TD. This resulted in short-term resolution of symptoms such as edema and effusions and improved renal function. However, the durability of this intervention is not clear and warrants further studies. Novel device therapies are currently being developed to enhance interstitial fluid excretion independently of renal function and diuretic efficacy.

CONFLICT OF INTEREST AND DISCLOSURE

The author declares no competing financial interests exist.

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