Lymphedema

Pathophysiology and clinical manifestations

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Learning objectives
After completing this learning activity, participants should be able to identity patients who are at risk for developing lymphedema; have an understanding of the etiology and pathophysiology of lymphedema; and describe the dermatologic manifestations of lymphedema.

Disclosures
Editors
The editors involved with this CME activity and all content validation/peer reviewers of the journal-based CME activity have reported no relevant financial relationships with commercial interest(s).

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Lymphedema is a localized form of tissue swelling resulting from excessive retention of lymphatic fluid in the interstitial compartment and caused by impaired lymphatic drainage. Lymphedema is classified as primary or secondary. Primary lymphedema is caused by developmental lymphatic vascular anomalies. Secondary lymphedema is acquired and arises as a result of an underlying systemic disease, trauma, or surgery. We performed PubMed and Google Scholar searches of the English-language literature (1966-2017) using the terms lymphedema, cancer-related lymphedema, and lymphatic complications. Relevant publications were manually reviewed for additional resources. This progressive chronic disease has serious implications on patients' quality of life. It is often misdiagnosed because it mimics other conditions of extremity swelling. There is no definitive cure for lymphedema. However, with proper diagnosis and management, its progression and potential complications may be limited. (J Am Acad Dermatol 2017;77:1009-20.)

Key words: lymphedema; pathophysiology; primary; secondary; skin.

PATHOPHYSIOLOGY

Key points

- **Lymphedema is caused by impaired lymphatic drainage in the presence of normal capillary function**
- **Lymphatic congestion induces chronic inflammation, which leads to fibrosis and further lymphatic damage**
- **Primary lymphedema is caused by genetic mutation or developmental abnormalities**

Abbreviations used:

APCD: advanced pneumatic compression device
BCRL: breast cancer–related lymphedema
ENV: elephantiasis nostras verrucosa

- **Filariasis is the most common cause of secondary (acquired) lymphedema worldwide**
- **Cancer-related treatment is the most common cause of secondary lymphedema in the United States**

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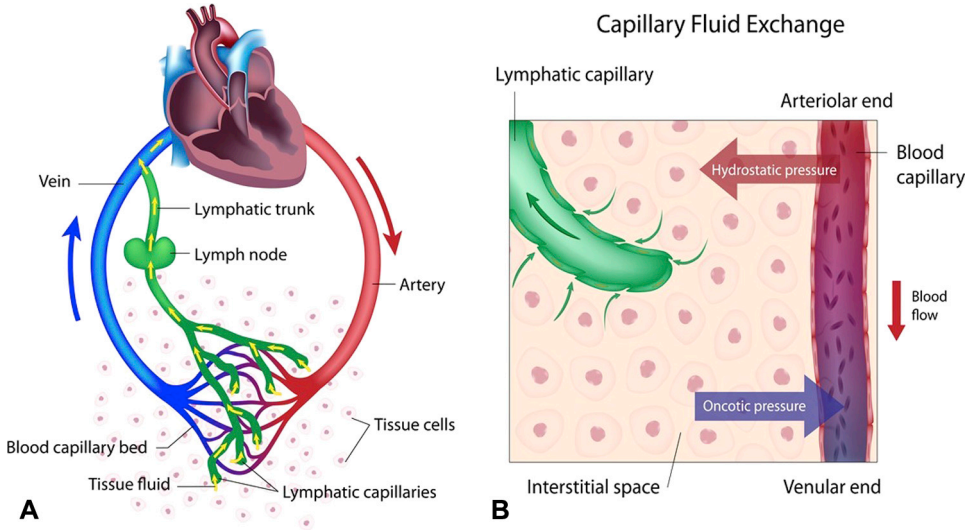

Fig 1. Normal lymphatic circulation. **A**, Lymphatic drainage returns tissue fluid to the bloodstream. **B**, Lymphatic capillaries collect excess fluid from the interstitial space. Around 90% of fluid filtered by blood capillaries will be reabsorbed and returned to the venous microcirculation. The remaining 10% is a protein-rich fluid "lymph" and will be drained by the lymphatic capillaries. Lymphedema develops when there is a malfunction in the lymphatic drainage system. (Copyright Alila Medical Media.)

The lymphatic system is composed of lymphatic organs, such as lymph nodes, tonsils, thymus, and the spleen, all of which are connected via a network of lymphatic vessels that run parallel to the venous circulation. The lymphatic system has 3 main functions: drainage of excess interstitial fluid, fat absorption, and immune surveillance. Interstitial fluid refers to the portion of fluid that leaks from blood capillaries into the tissue spaces. Most of the interstitial fluid (~90%) is reabsorbed via the venous microcirculation and returns to the bloodstream. The remainder (~10%) of the interstitial fluid has a relatively high protein concentration and is drained by blind-ended lymphatic capillaries (Fig 1). Once this protein-rich interstitial fluid enters the lymphatic capillaries, it is referred to as lymph. Lymph is then transported via the collecting lymphatic vessels, filtered through lymph nodes, and ultimately reenters the circulatory system near the point where the peripheral venous blood enters the right heart.[1] The normal lymphatic flow is 2 L to 3 L per day.[2] Unlike lymphatic capillaries, the collecting lymphatic vessels have smooth muscle walls and therefore the ability to contract and propel the lymphatic fluid forward. Impairment in the draining capacity caused by obstruction or lymphatic hypoplasia leads to an accumulation of interstitial fluid and tissue swelling known as lymphedema. The subsequent decrease in oxygen tension leads to chronic inflammation and reactive tissue fibrosis.

The lymphatic system has an important immune surveillance function. Circulating lymph transports various antigens and activated antigen-presenting cells into the lymph nodes to orchestrate the immune response.[3] Skin has an extensive presence of lymphatic capillaries. Patients with lymphedema are prone to recurrent skin infections because of the accumulation of peripheral tissue antigens. Chronic inflammation and the subsequent soft tissue fibrosis in lymphedema has been attributed to T_H2 immune response initiated because of lymphatic stasis.[4]

EPIDEMIOLOGY

Key points

- **Lymphedema is more common in females than males**
- **Lower extremity lymphedema is much more common than upper extremity lymphedema**

Incidence and prevalence

Lymphedema is a significant problem in the United States and throughout the world. It has been reported that lymphedema affects as many as 200 million people worldwide and approximately 3 million people in the United States.[5,6] Lymphedema affects females more often than males. Primary lymphedema is rare, with an estimated prevalence of 1 in 100,000 individuals, and usually occurs during childhood, but may present at any age.[7,8] Approximately 99% of individuals with lymphedema have a secondary disease.[5,9] The prevalence of

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