

بِسْمِ اللّٰهِ الرَّحْمٰنِ الرَّحِیْمِ

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دانشگاه علوم پزشکی گیلان

LYMPHEDEMA:

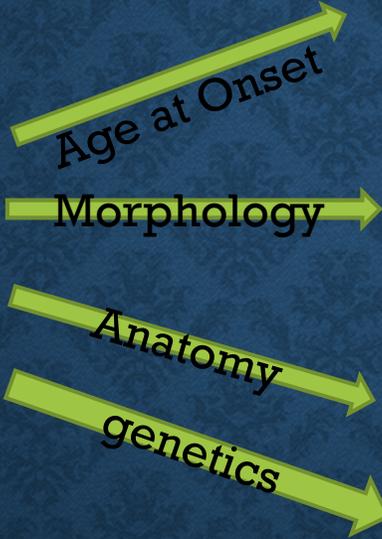
- **Lymphedema refers to tissue swelling caused by an accumulation of protein-rich fluid that's usually drained through the body's lymphatic system.**



Figure 168.2 Primary lymphedema of the right leg caused by hyperplasia of the lymphatics and valvular incompetence. Mid-calf skin vesicles contain a milky fluid because of lymphangiectasia and reflux of chyle.

CLASSIFICATION AND STAGING

- **Primary Lymphedema**



- **Secondary Lymphedema**

Lymphedema

Primary Lymphedema

- Occur in 1 of every 1000 individuals
- At birth, 1 in 6000 persons will develop lymphoedema with an overall prevalence of 0.13–2%^Q.
- Arise from congenital malformations such as lymphatics^Q

Secondary Lymphedema

- More common^Q
- Filariasis is MC cause worldwide^Q
- In Western countries: Result of neoplasms and their surgical treatments and radiotherapy^Q.

Congenital Lymphedema

- Onset before 1st year of life^Q
- Edema is typically present at birth^Q
- Can involve a single lower extremity, multiple limbs, genitalia or face^Q
- More likely to be bilateral and involve whole leg^Q
- Familial version of congenital lymphedema is

Lymphedema Precox

- Onset between 1-35 years^Q
- MC type, seen in 90% cases^Q
- More common in women^Q
- Mostly unilateral^Q
- Limited to the foot and calf^Q
- Familial version of lymphedema precox is known as Meig's disease^Q.

Lymphedema Tarda

- Onset after 35 years^Q
- Relatively rare

Primary Lymphoedema

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graph TD; A[Primary Lymphoedema] --> B[Congenital]; A --> C[Lymphoedema Praecox]; A --> D[Lymphoedema Tarda];
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Congenital

- Least common, approximately 2%
- More common to develop in male infants compared to female
- Symptoms displayed shortly after birth
- Commonly affects lower extremities

Lymphoedema Praecox

- Most prevalent type of primary lymphoedema
- Higher incidence rates in females during adolescent age
- Unilateral foot or calf usually affected

Lymphoedema Tarda

- Occurs in older adults that have a weak immune system



Figure 168.3 (A) Adult patient with congenital lymphedema. In addition to the bilateral arm lymphedema depicted, she has edema of both legs and the face. (B) Upper extremities of this patient's 18-year-old son, who has a similar distribution of lymphedema. This is an example of Milroy's disease.

SECONDARY LYMPHEDEMA

- Cancer
- Filariasis
- Other Causes





DISEASE

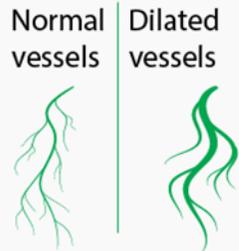
Infection

Filarial parasites spread by mosquitoes

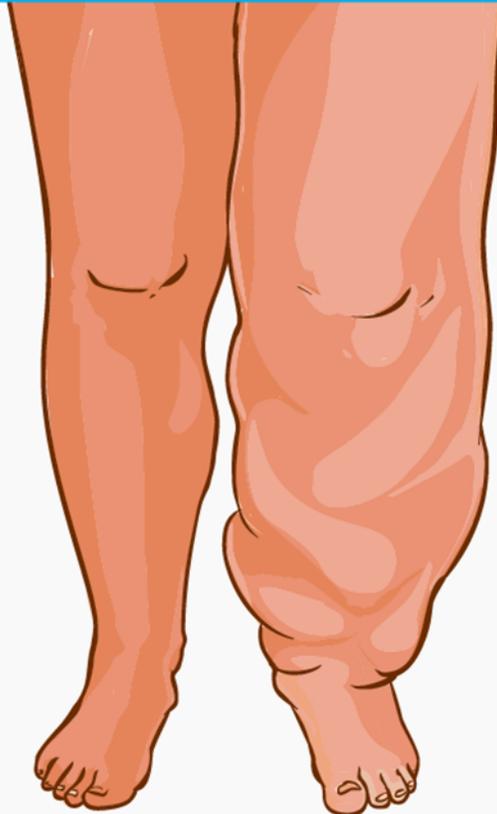


Disease

Impairs function of lymphatic vessels



856 Million people



ELIMINATION

Large-scale treatment of all at-risk populations can stop spread of infection

Vector control can supplement impact of large-scale treatment

Morbidity management & disability prevention to alleviate suffering due to disease

- **6.7 billion** treatments delivered (2000-2016)
- **499 million** people no longer require treatment
- Prevented or cured more than **97 million cases**
- **US\$ 100 billion** averted lifetime economic loss

Lymphatic Filariasis eliminated as a public health problem in 10 countries



Image credit: video.ink



CLINICAL STAGING

- Because none of the classification schemes addresses the clinical stage of the disease

Table 2. Staging of lymphedema adapted from The International Society of Lymphology

Stage	Description	Characteristics
0	Latent	Some damage to lymphatics; No visible edema yet
1	Spontaneously reversible, acute phase	Pitting edema; reversible with elevation of the arm. Usually, upon waking in the morning, the limb(s) or affected area is normal or almost normal size
2	Spontaneously irreversible, chronic phase	Spongy consistency and is "non-pitting," Fibrosis found in Stage 2 lymphedema marks the beginning of the hardening of the limbs and increasing size
3	Elephantiasis; irreversible, end-stage	Irreversible and usually the limb(s) is/are very large. The tissue is hard (fibrotic) and unresponsive; consider debulking surgery at this stage

CLINICAL PRESENTATION

- The clinical signs and symptoms of lymphedema largely depend on the **duration** and **severity** of the disease.
- **Edema**
- **Skin Changes**
- **Pain**
- **Infection**
- **Malnutrition and Immunodeficiency**
- **Malignant Tumors**

Stemmer's sign

- It reflects the **degree of distal fibrosis**, usually making it an especially accurate indicator for **primary lymphedema** of the lower extremity
- The sign is **positive** in **82% to 92% of primary** lower limb lymphedemas and in **56% of cases of secondary** lower limb lymphedema

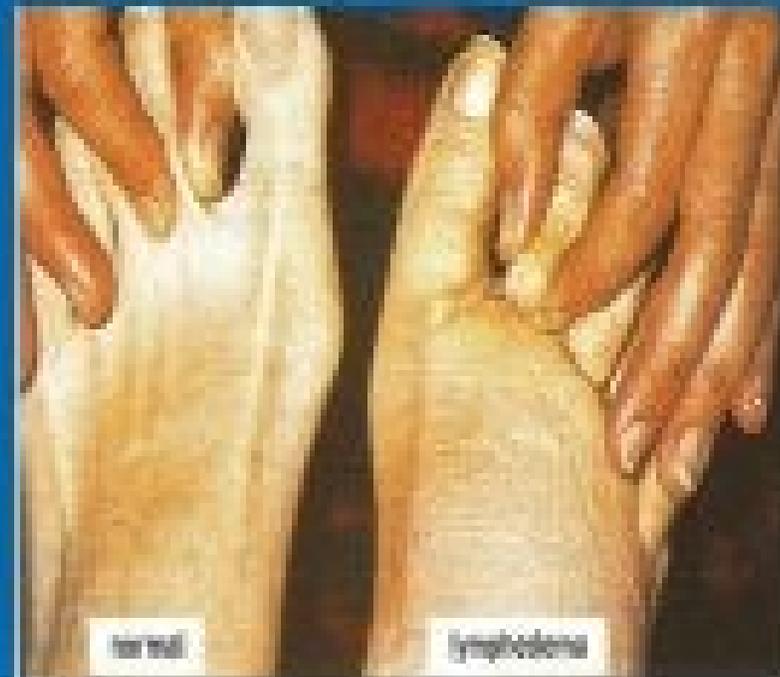


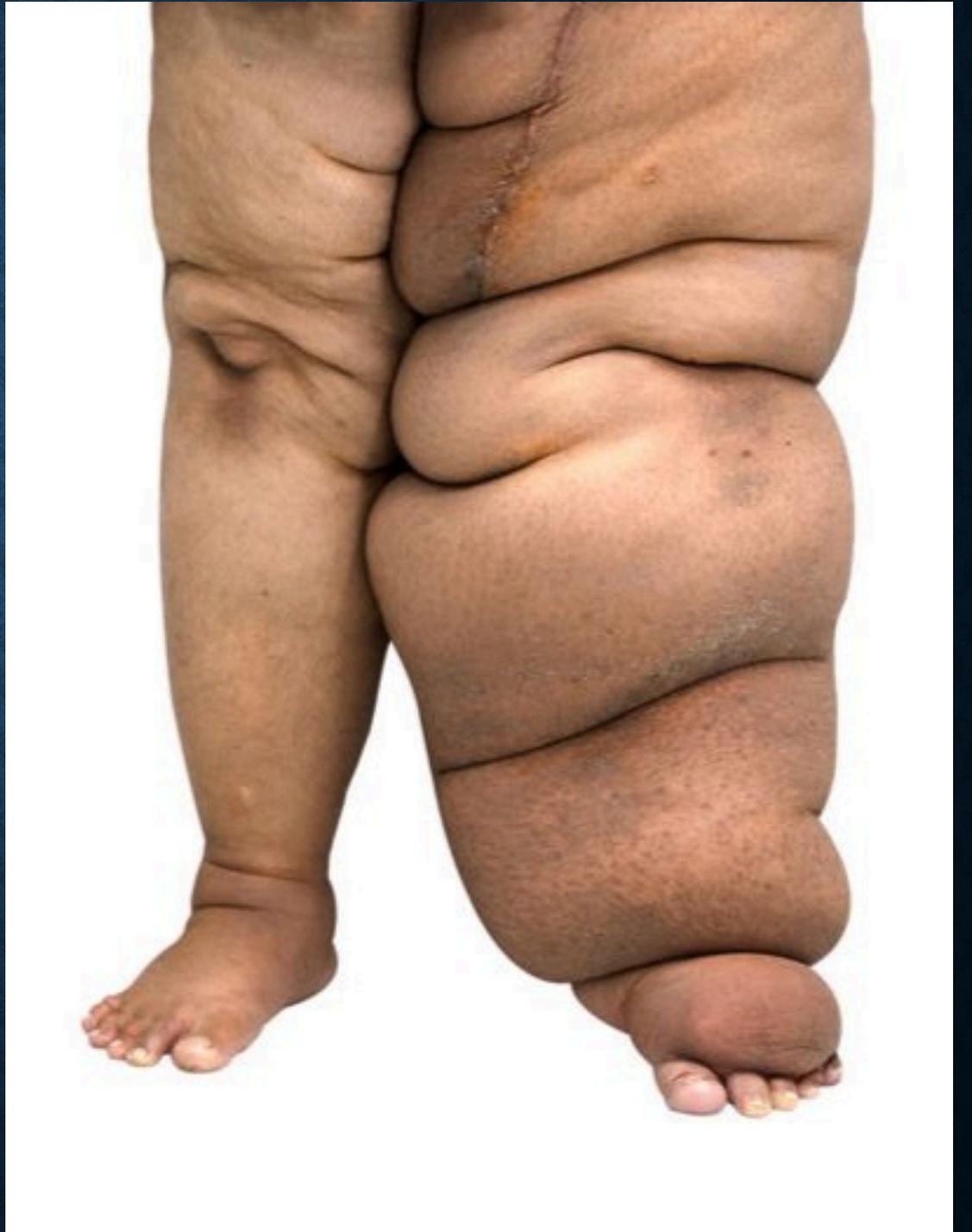
Figure 4.12)

Stemmer skin fold sign. In contrast to the normal second toe, a distended skin fold is seen on the lymphedematous toe.



Figure 168.4 Chronic acquired lymphedema of the lower extremities. Note severe skin changes (A) and swelling of the foot (B) associated with squaring of the toes (Stemmer's sign) and the typical peau d'orange. (C) Severe lymphedema with subcutaneous lymph cysts and chronic verrucous superinfection.

(A)







DIAGNOSIS

- In most cases of advanced, sustained lymphedema, the characteristic clinical presentation, history, and physical findings establish the diagnosis with near certainty.
- Additional objective data may be required to confirm the presence of impaired lymphatic flow or the typical pattern of abnormal fluid distribution within tissues.
- The diagnosis is more difficult to establish in the early stages of disease, particularly when edema is mild or intermittent.

Testing

- Lymphoscintigraphy
- Direct Contrast Lymphangiography
- Computed Tomography and Magnetic Resonance Imaging

Patient's Initials _____

Clinic Number _____ Date _____

LYMPHOSCINTIGRAPHY DATA EVALUATION

Arms Legs

IMAGE	1 Hr		3 Hr		6 Hr		24 Hr	
	R	L	R	L	R	L	R	L
Lymph transport kinetics: 0 = no delay, 1 = rapid, 3 = low-grade delay, 5 = extreme delay, 9 = no transport								
Distribution pattern: 0 = normal, 2 = focal abnormal tracer, 3 = partial dermal, 5 = diffuse dermal, 9 = no transport								
Lymph node appearance time: Minutes								
Assessment of lymph nodes: 0 = clearly seen, 3 = faint, 5 = hardly seen, 9 = no visualization								
Assessment of lymph vessels: 0 = clearly seen, 3 = faint, 5 = hardly seen, 9 = no visualization								
Abnormal sites of tracer accumulation (describe)								

Figure 168.5 Evaluation form for calculation of the lymphatic transport index. (Modified from Kleinhans E, Baumeister RG, Hahn D, Siuda S, Büll U, Moser E. Evaluation of transport kinetics in lymphoscintigraphy: follow-up study in patients with transplanted lymphatic vessels. *Eur J Nucl Med.* 1985;10:349. Courtesy Springer-Verlag.)

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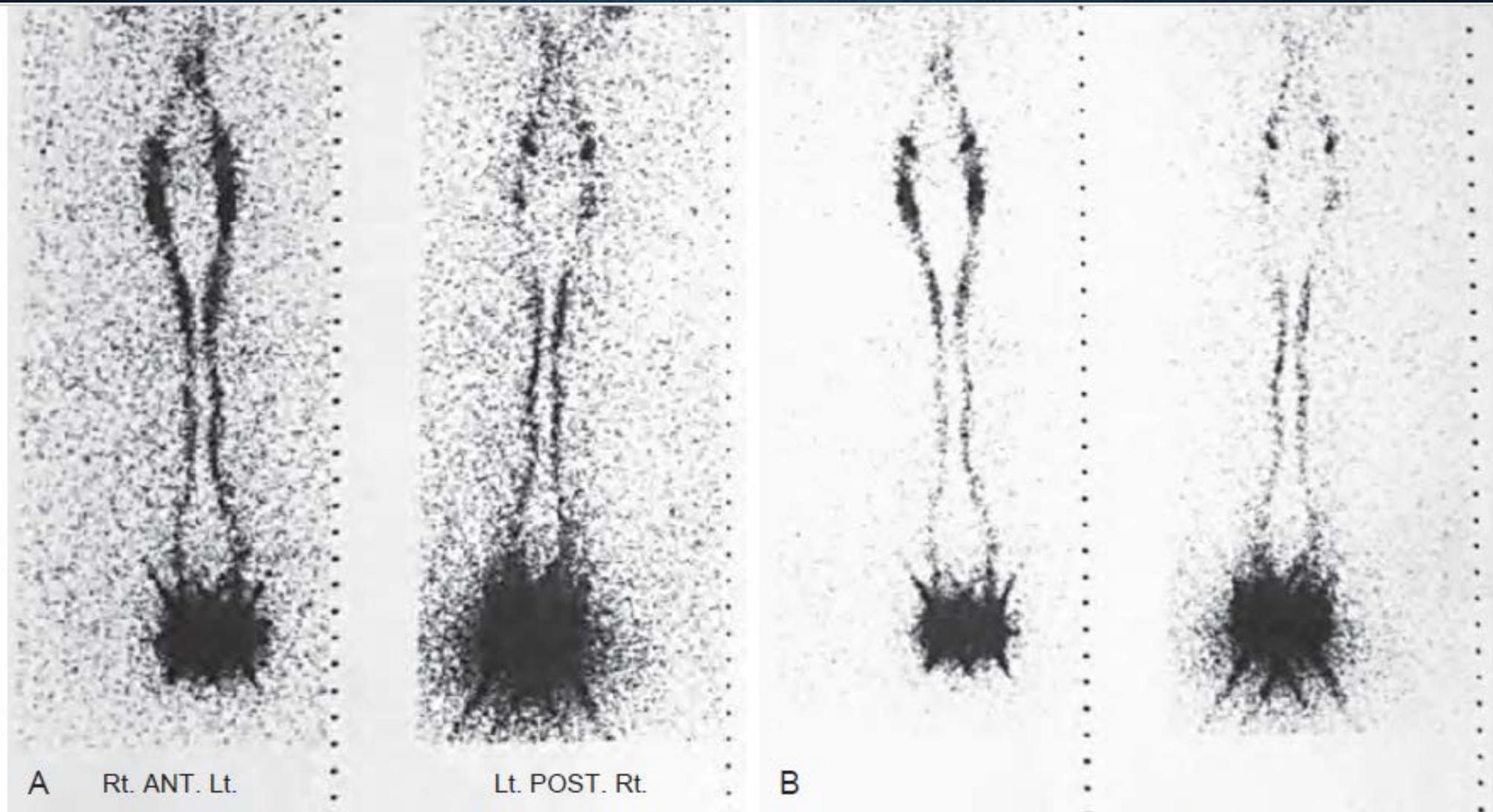


Figure 168.6 Anterior and posterior images in two intensity settings from a total-body scan with a dual-headed gamma camera. (A) Normal lymphoscintigram. (B) Higher intensity settings in the same patient. A large area of high activity and scatter is seen at the feet, where the injection was made. The single well-outlined band in each leg represents the main lymphatic channels. The lymph nodes in the groin and liver, the pelvic and para-aortic nodes, and an area at the site of the upper thoracic duct, are visualized.



Figure 168.7 Radionuclide lymphoscintigraphy in chronic bilateral lower limb lymphedema. The study demonstrates a dramatic degree of dermal backflow, suggesting the presence of lymphatic hypertension and valvular incompetence.

Lymphangiography

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graph TD; A[Lymphangiography] --> B[Direct Lymphangiography]; A --> C[Indirect Lymphangiography];
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Direct Lymphangiography

- Involves the injection of contrast medium (Isosulfan blue^Q) into a peripheral lymphatic vessel and radiographic visualization of the vessels and nodes^Q.
- Gold standard for showing structural abnormalities of larger lymphatics and nodes^Q
- Technically difficult, unpleasant for the patient
- May cause lymphatic injury
- Reserved for preoperative evaluation of megalymphatics being considered for bypass or fistula ligation^Q.

Indirect Lymphangiography

- Indirect lymphangiography involves the intradermal injection of water-soluble, non-ionic contrast into a web space, from where it is taken up by lymphatics and then followed radiographically^Q.
- It will show distal lymphatics but not normally proximal lymphatics and nodes^Q.

Computed Tomography and Magnetic Resonance Imaging

- Lymphedema is typically confined to the epifascial space of the skin and subcutaneous tissue, sparing the muscle.
- In venous edema, both the epifascial and subfascial compartments are affected/
- in lipedema, there is fat accumulation without fluid.

- The **greatest value** of CT and MRI in the evaluation of a patient with a swollen leg is to **exclude any obstructing mass in the pelvis** that may result in decreased transport capacity of the lymphatic system.

DIFFERENTIAL DIAGNOSIS

- Systemic Causes
- Venous Insufficiency
- Lipedema
- Vascular Malformation
- Other Causes

LYMPHEDEMA

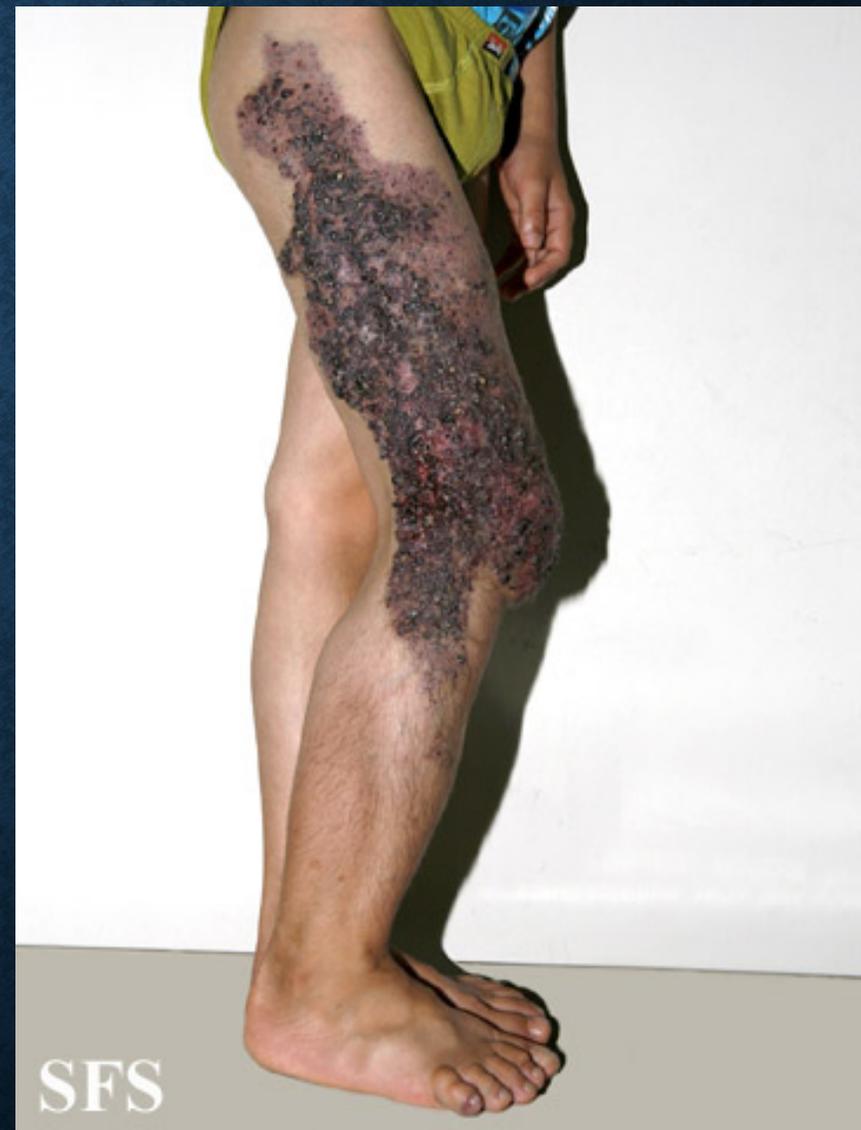
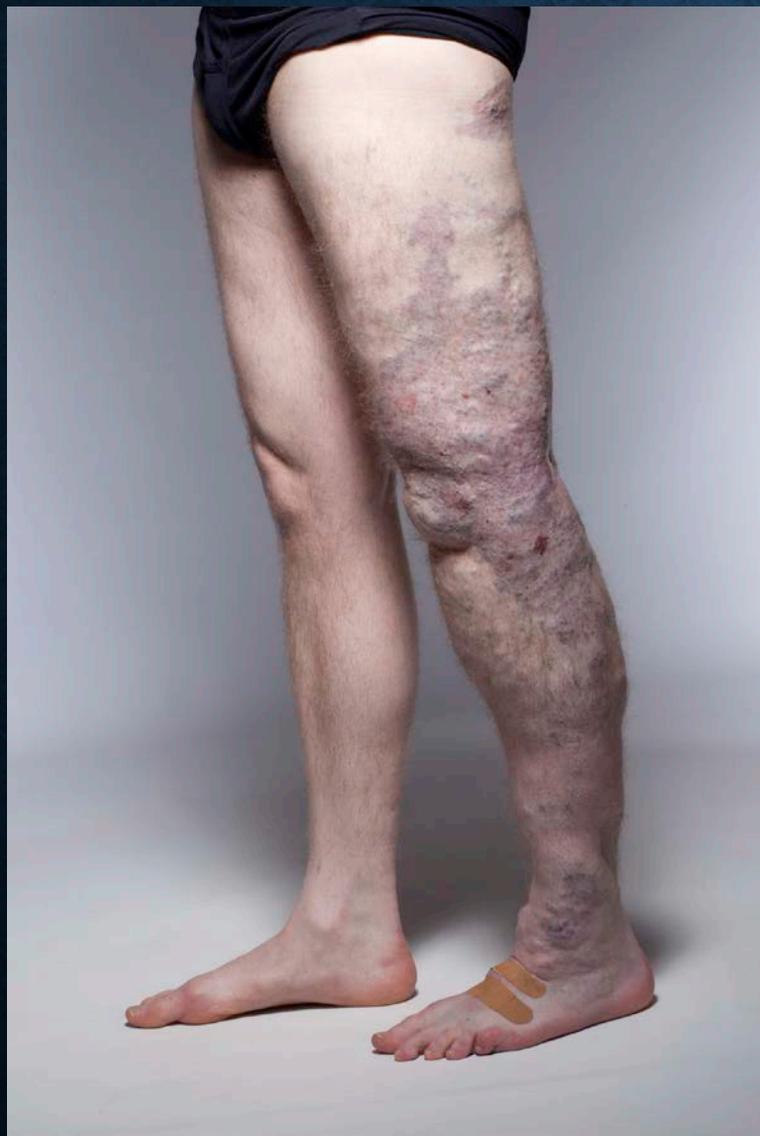


LIPPEDEMA





VASCULAR MALFORMATION



SFS



● ○ REDMI NOTE 9 PRO
∞ 64MP QUAD CAMERA